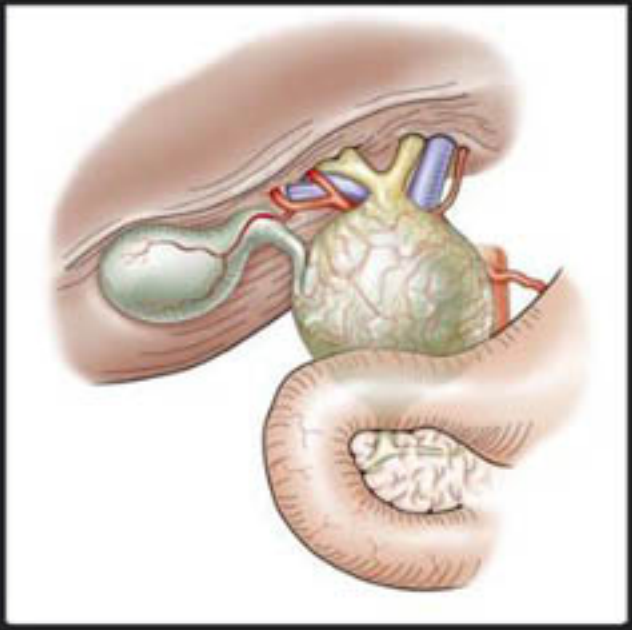
SIXTH EDITION

Operative Pediatric Surgery



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Operative Pediatric Surgery

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Preface

This is the sixth edition of *Operative Pediatric Surgery* and the second in which the two editors have collaborated. The new edition encompasses all the major advances in the specialty that have occurred in the past decade since the previous edition was published in 1995.

The most noteworthy advance in pediatric surgery has been the development of minimal invasive surgery, which has the advantages, in addition to obvious cosmetic superiority, of reduced postoperative pain, reduced metabolic response to operative trauma, and significant reduction of postoperative hospitalization. Numerous new authors have been recruited to contribute to this developing field of minimal invasive surgery.

Other new sections include congenital vascular malformations, the Nuss procedure for pectus excavatum, the Bianchi bowel-lengthening procedure, interventional radiology, and bariatric surgery. We have omitted sections that do not fall within the purview of the pediatric surgeon, such as cleft lip and palate, protruding ears, and congenital hand deformities.

We have retained the format of previous editions of initial principles and justification for the procedure, followed by preoperative investigations and preparation, the operative procedure, and postoperative management. All illustrations are simple black and white line drawings, which have maintained the consistently high standard demanded by Gillian Lee and her associates. A limited number of recommendations for further reading are included at the end of each chapter. These may not always appear to be up to date, but have been included as the operative technique may have remained unaltered over the years.

We wish to thank all authors for their efforts at achieving the highest standard of contribution. They were selected because of their established expertise in their fields and their international reputations, and we are grateful for their time and patience. We are confident that they will be pleased with and proud of the ultimate publication.

This textbook has succeeded in being generally accepted as the operative manual of choice for pediatric surgeons around the world and takes into account the varying practices of pediatric surgery in different countries. The previous edition completely sold out within a few years of publication, and we are confident that this expanded, up-to-date new volume will be as successful.

We trust that trainees as well as established pediatric surgeons will find this textbook useful as a guide when performing a wide range of procedures.

Lewis Spitz
Arnold G. Coran

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List of abbreviations used

ADH	Antidiuretic hormone
AFP	Alpha-fetoprotein
ATP	Adenosine triphosphate
BCG	Bacille Calmette–Guérin
BPD	Bronchopulmonary dysplasia
BXO	Balanitis xerotica obliterans
CAM	Cystic adenomatoid malformation
CFTR	Cystic fibrosis transmembrane (conductance) regulator
CGRP	Calcitonin gene-related peptide
CMV	Cytomegalovirus
CNS	Central nervous system
CPAP	Continuous positive airway pressure
CSF	Cerebrospinal fluid
CT	Computed tomography
CUSA	Cavitron Ultrasonic Surgical Aspirator
CVC	Central venous catheter
CVP	Central venous pressure
DMSA	Dimercaptosuccinnic acid
DTPA	^{99m}Tc-diethylenetriamine penta-acetic acid
EBV	Epstein–Barr virus
ECG	Electrocardiography
ECLS	Extracorporeal life support
ECMO	Extracorporeal membrane oxygenation
ECW	Extracellular water
ERCP	Endoscopic retrograde cholangiopancreatography
ESWL	Extracorporeal shock-wave lithotripsy
EXIT	Ex-utero intrapartum treatment
FAST	Focused abdominal sonography for trauma
FEV1	Forced expiratory volume in the first second
FRC	Functional residual capacity
FVC	Forced vital capacity
GFR	Glomerular filtration rate
GTN	Glyceryl trinitrate
Hb	Hemoglobin
hCG	Human chorionic gonadotropin
HD	Hirschsprung's disease
HIV	Human immunodeficiency virus
HMD	Hyaline membrane disease
ICP	Intracranial pressure
ICU	Intensive care unit
INR	International Normalized Ratio
Insl3	Insulin-like hormone 3
INSS	International Neuroblastoma Staging System
IR	Interventional radiology
IVC	Inferior vena cava
IVH	Intraventricular hemorrhage
IVU	Intravenous urography
LBW	Low birthweight
LDH	Lactate dehydrogenase
MACE	Malone procedure for antegrade continence enema
MAG-3	^{99m}Tc-mercaptoacetyltriglycine
MAGPI	Meatal advancement and glanuloplasty
MCT	Medium-chain triglyceride
MCUG	Micturating cystourethrography
MIBG	^{131}Meta-iodobenzylguanidine
MIS	Mullerian inhibiting substance
MRA	Magnetic resonance angiography
MRCP	Magnetic resonance cholangiopancreatography
MRI	Magnetic resonance imaging
NSAID	Non-steroidal anti-inflammatory drug
PDS	Polydioxanone
PEEP	Positive end-expiratory pressure
PEG	Percutaneous endoscopic gastrostomy
PET	Positron emission tomography
PICC	Percutaneously inserted central venous catheter
PSARVUP	Posterior sagittal anorectovaginourethroplasty
PTC	Percutaneous transhepatic cholangiography
PUJ	Pelviureteric junction
PVC	Polyvinyl chloride
PVT	Portal vein thrombosis
SCT	Sacrococcygeal teratoma
SGA	Small for gestational age
SMV	Superior mesenteric vein
STING	Subureteral [Teflon] injection
SVC	Superior vena cava
TBW	Total body water
TIPS	Transjugular intrahepatic portal systemic shunt
TPN	Total parenteral nutrition
US	Ultrasonography
UTI	Urinary tract infection
VA	Venoarterial
VATER (syndrome)	Vertebral defects, imperforate anus, tracheoesophageal fistula, radial dysplasia, and renal dysplasia
VUR	Vesicoureteral reflux
VV	Venovenous

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SECTION I

General

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1

Preoperative and postoperative management of the neonate

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INTRODUCTION

The neonate, infant, child, and adolescent differ significantly from each other and from the adult. The most distinctive and rapidly changing physiologic characteristics occur during the neonatal period. This is due to the newborn infant's adaptation from complete placental support to the extrauterine environment, differences in the physiologic maturity of individual neonates, the small size of these patients, and the demands of growth and development. Recent advances in neonatal care have resulted in the survival of increasing numbers of extremely low birth-weight (LBW) infants. Extreme prematurity magnifies the already dynamic and relatively fragile physiology of the newborn period, predisposing these tiny infants to physiologic derangements in temperature regulation, fluid and electrolyte homeostasis, glucose metabolism, hematologic regulation, and immune function. In addition, physiologic and anatomic organ system immaturity makes the preterm neonate vulnerable to specific problems such as intraventricular hemorrhage, hyaline membrane disease, and hyperbilirubinemia. From a surgical standpoint, these dynamic and fragile physiologic parameters are often the primary components that dictate the preoperative and postoperative management of the neonatal surgical patient. This chapter focuses on the physiology of the neonate undergoing surgery, highlighting the practical considerations of preoperative and postoperative management.

LOW BIRTH-WEIGHT INFANTS

Neonates may be classified according to their level of maturation (gestational age) and development (weight) (Tables 1.1 and 1.2). This classification is important because the physiology of neonates may vary significantly depending on these parameters.

Table 1.1 *Newborn classification by maturation (age)*

Classification	Age at birth
Preterm	Before 37 weeks' gestation
Term	Between 37 and 42 weeks' gestation
Post-term	After 42 weeks' gestation

Table 1.2 *Newborn classification by development (weight)*

Classification	Birth weight
Small for gestational age (SGA)	< 10th percentile
Appropriate for gestational age (AGA)	Between 10th and 98th percentile
Large for gestational age (LGA)	> 98th percentile

Under this classification system, a term, appropriate for gestational age infant is born between 37 and 42 weeks of gestation with a birth weight greater than 2500 g. However, approximately 7 percent of all babies do not meet these criteria. This may be due to prematurity or to intrauterine growth retardation. From a clinical standpoint, neonates born weighing less than 2500 g are broadly classified as *low birth-weight* (LBW) infants. Further sub-classification into moderately LBW, very LBW, and extremely LBW infants has been used for epidemiologic and prognostic purposes (Table 1.3). Using this terminology, LBW infants may be preterm and appropriate for gestational age, term but small for gestational age (SGA), or both. The following sections discuss several issues specific to the care of preterm and SGA infants.

Table 1.3 *Alternative newborn weight classification*

Classification	Birth weight (g)	Percent of premature births	Mortality rate vs. term infants
Low birth weight (LBW)	< 2500	-	-
Moderately low birth weight	Between 2500 and 1501	82	40 times higher
Very low birth weight	Between 1500 and 1001	12	200 times higher
Extremely low birth weight	< 1000	6	600 times higher

Preterm infant

By definition, *preterm infants* are born before 37 weeks' gestation. They generally have body weights appropriate for their age, although they may also be SGA. If the gestational age is not accurately known, the prematurity of an infant can be confirmed by physical examination. The principal features of preterm infants are a head circumference below the 50th percentile, thin, semi-transparent skin with an absence of plantar creases, soft and malleable ears with poorly developed cartilage, absence of breast buds, undescended testes (testicular descent begins around the 32nd week of gestation) with a flat scrotum in boys and relatively enlarged labia minora and small labia majora in girls.

In addition to these physical characteristics, several physiologic abnormalities exist in preterm infants. These abnormalities are often a result of unfinished fetal developmental tasks that normally enable an infant to transition successfully from intrauterine to extrauterine life. These tasks, which include renal, skin, pulmonary, and vascular maturation, are usually completed during the final weeks of gestation. The more premature the infant, the more fetal tasks are left unfinished and the more vulnerable the infant to adverse sequelae of an early birth.

This physiologic and anatomic vulnerability sets the preterm infant up for several specific and clinically significant problems:

- Central nervous system immaturity leading to episodes of apnea and bradycardia and a weak suck reflex.
- Pulmonary immaturity leading to surfactant deficiency, which can result in hyaline membrane disease (HMD).
- Cerebrovascular immaturity leading to fragile, unsupported cerebral vessels that lack the ability to autoregulate. This predisposes the preterm infant to intraventricular hemorrhage (IVH) – the most common acute brain injury of the neonate.
- Skin immaturity leading to an underdeveloped stratum corneum with significant transepithelial water loss (TEWL). This complicates the thermal regulation and fluid management of the infant.
- Gastrointestinal underdevelopment predisposing to necrotizing enterocolitis.
- Impaired bilirubin metabolism causing hyperbilirubinemia.
- Cardiovascular immaturity leading to a patent ductus arteriosus or patent foramen ovale. These retained elements of the fetal circulation can cause persistent left-to-right shunting, pulmonary hemorrhage and cardiac failure.

From a practical standpoint, the care of the preterm infant must be directed at preventing and/or treating these specific problems. Episodes of apnea and bradycardia are common and may occur spontaneously or as non-specific signs of problems such as sepsis or hypothermia. Prolonged apnea with significant hypoxemia leads to bradycardia and ultimately to cardiac arrest. All preterm infants should therefore undergo apnea monitoring and electrocardiographic pulse monitoring, with the alarm set at a minimum pulse rate of 90 beats per minute. In the neonate with respiratory difficulties, chest radiography will help to detect HMD and cardiac failure. The lungs and retinas of preterm infants are very susceptible to high oxygen levels, and even relatively brief exposures may result in various degrees of HMD and retinopathy of prematurity. Infants receiving oxygen therefore require continuous pulse oximetry monitoring, with the alarm set between 85 and 92 percent. Preterm infants may also be unable to tolerate oral feeding because they have a weak suck reflex, necessitating intragastric tube feeding or parenteral nutrition. Finally, impaired bilirubin metabolism may necessitate serum bilirubin monitoring and phototherapy for rising levels of unconjugated bilirubin that can lead to kernicterus.

Small for gestational age infant

Infants whose birth weight is below the 10th percentile are considered to be SGA. Newborns that are SGA are thought to be a product of restricted intrauterine growth due to placental, maternal or fetal abnormalities. Table 1.4 lists several conditions that may lead to intrauterine growth retardation in the neonate. However, it should be noted that not all infants in this group are truly growth retarded and therefore at higher risk. Some are simply born small as a result of a variety of factors, including race, ethnicity, sex, and geography. It is important to differentiate these infants from those whose relatively low birth weight is a result of a genetic or intrauterine abnormality.

The SGA infants can be divided into two broad categories: symmetric SGA infants and asymmetric SGA infants. This distinction is based primarily on when in the gestational period fetal growth was restricted. If fetal growth is restricted during the first half of pregnancy, when cellular hyperplasia

Table 1.4 *Common conditions associated with intrauterine growth retardation*

Age at delivery	Condition
Preterm	Placental insufficiency
	Discordant twin
	Chronic maternal hypertension
	Intrauterine infection
	Toxemia
Term	Congenital anomaly
	Microcephaly
Post-term	Placental insufficiency

and differentiation lead to tissue and organ formation, the neonate is generally a symmetric SGA infant. Fetal factors such as genetic dwarfism, chromosomal abnormalities, congenital abnormalities, inborn errors of metabolism, and fetal infection, as well as maternal factors such as genetics, toxin ingestion, and substance abuse are all causative etiologies. While only 30 percent of SGA infants fall into this group, they have the highest morbidity and mortality rates. In contrast, asymmetric SGA infants are those who experience restriction in intrauterine growth during the last half of gestation, often during the third trimester. This is usually due to an inadequate nutrient supply. An example of this is twin gestations. Though both infants may be full term at birth, they generally have a low birth weight because placental function is inadequate to meet the growth demands of both fetuses. Other causes of asymmetric growth retardation include maternal conditions that reduce uteroplacental blood flow, such as hypertension, toxemia, cardiac disorders, and renovascular disorders.

In general, SGA infants have a body weight that is low for their gestational age although their body length and head circumference are appropriate. The SGA infant is developmentally more mature than a preterm infant of equivalent weight, and therefore faces significantly different physiologic problems. Because of the longer gestational period and resultant well-developed organ systems, the metabolic rate of the SGA infant is much higher in proportion to body weight than that of a preterm infant of similar overall weight. Fluid and caloric requirements are therefore increased. Intrauterine malnutrition results in a relative lack of body fat and decreased glycogen stores. In fact, body fat levels in SGA infants are often below 1 percent of their total body weight. This, coupled with their relatively large surface area, greatly predisposes these infants to hypothermia and hypoglycemia. Close monitoring of blood sugar level is therefore essential. In addition, polycythemia is common in SGA infants due to increased red blood cell volumes. Occurring in 15–40 percent of asymmetric SGA babies, polycythemia may lead to hyperviscosity syndrome, characterized by respiratory distress, tachycardia, pleural effusions, and the risk of venous thrombosis. This may necessitate plasma exchange transfusions as well as frequent monitoring of the infant's hematocrit level. Finally, fetal asphyxia and distress due to inadequate placental support may lead to passage of meconium in utero. This results in an increased risk of meconium aspiration syndrome in SGA infants if the material is aspirated during labor and delivery. The perioperative management of these conditions is detailed in the sections below. While SGA infants are at significant risk for morbidity and mortality associated with these problems, their adequate length of gestation puts them at a relatively lower risk for many of the conditions that affect preterm infants such as retinopathy of prematurity, intraventricular hemorrhage, and HMD.

PHYSIOLOGIC CONSIDERATIONS IN THE PERIOPERATIVE CARE OF THE NEONATE

As stated above, the dynamic physiologic changes that occur during the neonatal period significantly influence the perioperative care of the newborn surgical patient. In particular, physiologic derangements in temperature regulation, glucose metabolism, hematologic regulation, immune function, and fluid and electrolyte homeostasis often dictate perioperative management strategies.

Thermoregulation

Neonates are susceptible to heat loss because of their large surface area, low body fat to body weight ratio, and limited heat sink capacity due to their small size. In addition, neonates have a relatively high thermoneutral temperature zone. The optimal thermal environment (thermoneutrality) is defined as a range of ambient temperatures in which an infant, at a minimal metabolic rate, can maintain a constant normal body temperature by vasomotor control. The environmental temperature must be maintained near the appropriate thermoneutral zone for each individual. In adults, this critical temperature range is 26–28 °C, whereas in the term infant it is 32–34 °C. In the LBW infant, this critical range is even higher, at 34–35 °C.

MECHANISMS OF HEAT LOSS

In the neonate, heat loss may occur by evaporation, conduction, convection, and radiation. Evaporative heat loss occurs as a result of TEWL and depends on the gestational age of the infant, the relative humidity, and other environmental conditions. In addition, the presence of liquid in contact with an infant's skin also contributes to evaporative heat loss. Conductive heat loss occurs when an infant's skin is in contact with a solid object of lower temperature, causing heat to flow from the infant to the object at a rate dependent on the temperature difference between the two as well as the insulating properties of the baby and the object. Similarly, convective heat loss occurs when the ambient air temperature is lower than the infant's skin temperature. Convective heat loss

depends on the temperature gradient between the infant's skin and the air as well as on the speed of the air current over the infant. Radiant heat loss occurs via the passage of infrared rays from the infant's skin to a cooler surface, such as the incubator or nursery wall. This type of heat loss is often the most difficult to control.

THERMOGENESIS IN THE NEONATE

Neonates generate heat by increasing metabolic activity. Unlike adults, they achieve this principally by non-shivering thermogenesis using brown fat. This has practical consequences because brown fat may be rendered inactive by pressors or anesthetic and neuromuscular blocking agents. Brown fat stores may also be depleted due to poor nutritional intake, such as in an SGA infant. When an infant is exposed to cold, metabolic work increases above basal levels and calories are consumed to maintain body temperature. If prolonged, this depletes the limited energy reserves of the neonate and predisposes to hypothermia and increased mortality.

PRACTICAL CONSIDERATIONS

The environmental temperature of the neonate is best controlled in an incubator by monitoring the ambient temperature and maintaining it at thermoneutrality. Inside the incubator, clothing on the infant can increase insulation, reducing radiant and convective heat loss. In particular, covering the head with an insulated hat can reduce heat loss and total metabolic activity during cold stress by up to 15 percent. Similarly, conductive heat loss is minimized by the use of insulating padding. The incubators themselves are plastic-walled containers that warm the infant by convection. The air in the incubator is heated by a heating element and then circulated by a fan. A servo system regulates incubator temperature according to the patient's skin temperature, which is monitored by a skin probe. In this manner, the infant's skin temperature is maintained at a relatively constant value. Double-walled incubators minimize radiant heat loss by maintaining the inner wall of the incubator at the same temperature as the air temperature inside the incubator. Finally, humidity can be provided to the incubator environment, thereby reducing evaporative heat loss.

Optimal air temperatures for individual infants vary with the gestational age and condition of the infant as well as with specific environmental factors such as humidity and airflow. Standard nomograms are available that aid in determining the appropriate incubator temperature necessary to achieve thermoneutrality. Term infants usually require the incubator air temperature to be 32–34 °C. Low birth-weight infants may require temperatures at or above 35 °C.

In contrast to incubators, radiant warmers provide open access to and visibility of the infants. They are often used for surgical patients for whom frequent access and tubes/lines are necessary. Radiant warmers generate heat by means of an overhead panel that produces heat in the infrared range. However, their side rails are only minimally protective against convective heat loss and often lead to higher evaporative water and heat losses. This evaporative heat loss may be reduced by plastic sheets.

The feedback mechanisms of both incubators and radiant warmers are used to maintain an infant's skin temperature in the normal range. The normal skin temperature for a term infant is 36.2 °C and for a LBW infant 36.5 °C. Increased metabolic activity can be detected by comparing skin and rectal temperatures, which normally differ by 1.5 °C. A decreasing skin temperature with a constant rectal temperature suggests that the metabolic rate has increased to maintain the core temperature.

In a cold environment, such as the operating room or radiology suite, heat loss may be reduced by wrapping the head, extremities, and as much of the trunk as possible in clothing, plastic sheets, or aluminum foil. A variety of 'warming blankets' are available. A plastic sheet placed beneath the infant decreases the humidity of the microenvironment between it and the sheet. After draping, the infant is covered by a large adhesive plastic sheet which diminishes evaporative heat and water loss and prevents the infant from becoming wet during the operation. Any exposed intestine (e.g., gastroschisis) should be wrapped in plastic. An overhead infrared heating lamp should be focused on the infant during induction of anesthesia, preparation for operation, and at the termination of the operation. Solutions used for skin cleansing and intracorporeal irrigation should be warmed.

Glucose homeostasis

The fetus receives glucose from its mother by facilitated placental diffusion; very little is derived from fetal gluconeogenesis. The limited liver glycogen stores accumulated during the later stages of gestation are rapidly depleted within 2 to 3 hours after birth. The blood glucose level of the infant then depends on the neonate's capacity for gluconeogenesis, the adequacy of substrate stores, and the energy requirements of the infant. It should be noted that the neonate's ability to synthesize glucose from fat or protein substrates is severely limited, necessitating the intake of exogenous carbohydrates to maintain adequate blood glucose levels.

HYPOGLYCEMIA

The risk of developing hypoglycemia is high in LBW infants (especially SGA infants), those born to toxemic or diabetic mothers, and those requiring surgery who are unable to take oral nutrition and who have the additional metabolic stresses of their disease and the surgical procedure. The clinical features of hypoglycemia are non-specific and include a weak or high-pitched cry, cyanosis, apnea, jitteriness or trembling, apathy, and seizures. The differential diagnosis includes other metabolic disturbances or sepsis. More than 50 percent of infants with symptomatic hypoglycemia suffer significant neurologic damage. Neonatal hypoglycemia is defined as a serum glucose level less than 1.66 mmol/L in the full-term

infant and less than 1.11 mmol/L in the LBW infant. However, neurologic abnormalities have been reported with higher blood glucose levels. Older children, particularly those with depleted stores and severe metabolic demands, are also at risk of hypoglycemia.

PRACTICAL CONSIDERATIONS

All pediatric surgical patients, particularly neonates, are monitored for hypoglycemia. To avoid delay, blood glucose levels can be rapidly determined in the neonatal unit using blood glucose reagent strips. These may be correlated at intervals with serum glucose determinations, the frequency depending on the stability of the patient. Any intravenous fluids administered should contain at least 10 percent dextrose. If non-dextrose-containing solutions such as blood or plasma are being administered, close monitoring of the blood glucose level is essential. Hypoglycemia should be treated urgently with intravenous 50 percent dextrose, 1–2 mL/kg, and maintenance intravenous dextrose, 10–15 percent, 80–100 mL/kg for each 24 hours.

HYPERGLYCEMIA

Hyperglycemia is commonly a problem of very LBW infants on parenteral nutritional support, because they have a low insulin response to glucose. Hyperglycemia may lead to IVH and renal water and electrolyte loss from glycosuria. Prevention of hyperglycemia is by small and gradual incremental changes in the glucose concentration and infusion rate.

Hematologic considerations

Total blood, plasma, and red cell volumes are higher during the first few hours after birth than at any other time in an individual's life. The levels may be further increased if a significant placental transfusion takes place at delivery (delayed umbilical cord clamping). Several hours after birth, plasma shifts out of the circulation and total blood and plasma volumes decrease. The high red blood cell volume persists, decreasing slowly to reach adult levels by the third postnatal month. Age-related estimations of blood volume are summarized in Table 1.5.

POLYCYTHEMIA

In addition to SGA infants, neonatal polycythemia occurs in infants of diabetic mothers and in infants of mothers with toxemia of pregnancy. In the neonate, polycythemia is defined as a central venous hematocrit greater than 65 percent or a hemoglobin level greater than 22 g/dL. Values at or above this threshold may be associated with high blood viscosity, which is further increased by a fall in body temperature. Partial exchange transfusion may be indicated because hyperviscosity may be an etiologic factor for several disorders, including central nervous system dysfunction and necrotizing enterocolitis.

Table 1.5 *Estimation of blood volume*

Age	Blood volume (mL/kg)
Preterm	85–100
Term	85
1–3 months	75
3 months to adult	70

ANEMIA

In the neonate, anemia is generally due to hemolysis, blood loss, or decreased erythrocyte production. Hemolytic anemia in the newborn is most often caused by placental transfer of maternal antibodies that destroy the infant's erythrocytes. Significant hemolytic anemia is most commonly due to Rhesus incompatibility producing jaundice, palor, hepatosplenomegaly, and, in severe cases, hydrops fetalis. In addition, congenital infections, inherited hemoglobinopathies, and thalassemias may all manifest as hemolytic anemia in the newborn period. In severe cases, these conditions may require exchange transfusions.

In addition to hemolysis, severe anemia in the neonate may be secondary to acute hemorrhage. This can occur as a result of placental abruption or in-utero bleeding into the intraventricular, intra-abdominal, subgaleal, or mediastinal spaces. Twin–twin transfusion syndrome may also result in severe anemia in the 'donor' co-twin. Finally, anemia of prematurity due to decreased red blood cell production is another cause of significant neonatal anemia. This occurs in preterm infants born before a gestational age of 30–34 weeks, before erythropoietin release by the kidneys has occurred.

HEMOGLOBIN

Infant erythopoiesis does not occur until approximately 2–3 months of age. Until that time, fetal hemoglobin represents the vast majority of circulating hemoglobin in the neonate. In fact, approximately 80 percent of an infant's circulating hemoglobin is fetal at birth. This is significant in that the high proportion of fetal to adult hemoglobin in neonates shifts their hemoglobin dissociation curve to the left. Since fetal hemoglobin has a higher affinity for retaining oxygen, lower peripheral oxygen levels are needed to release and deliver oxygen from fetal blood to the receiving end tissues. Thus, a high oxygen saturation percentage reading on a transcutaneous pulse oximeter may be associated with a relatively low blood pO_2 measurement. As fetal hemoglobin is broken down, a 'physiologic' anemia results with a nadir at about 2–3 months of age.

COAGULOPATHY

The routine administration of vitamin K to all neonates to prevent hypoprothrombinemia and hemorrhagic disease is established practice. This may be overlooked during the

activities attendant on major congenital anomalies or conditions requiring urgent surgical evaluation. When in doubt, 1.0 mg of vitamin K should be administered by intramuscular or intravenous injection.

JAUNDICE

Heme pigments, notably hemoglobin, are catabolized in the spleen and liver to produce bilirubin. The bilirubin is conjugated with glucoronic acid in the liver, forming a water-soluble substance that is excreted via the biliary system into the intestine. In the fetus the lipid-soluble, unconjugated (indirect) bilirubin is cleared across the placenta. In the fetal intestine beta-glucoronidase hydrolases conjugate bilirubin, which is then reabsorbed for transplacental clearance. Circulating unconjugated bilirubin is bound to albumin.

The neonate's capacity for conjugating bilirubin is not fully developed and may be exceeded by the bilirubin load, resulting in transient physiologic jaundice that reaches a maximum at the age of 4 days but returns to normal levels by the sixth day. Usually the maximum bilirubin level does not exceed 170 μmol/L. Physiologic jaundice is particularly likely to occur in SGA and preterm infants, in whom a higher and more prolonged hyperbilirubinemia may be encountered.

High serum levels of unconjugated bilirubin may cross the immature blood–brain barrier in the neonate and can act as a neural poison leading to kernicterus. This condition, in its most severe form, is characterized by athetoid cerebral palsy and sensorineural hearing loss. Predisposing factors are hypoalbuminemia, acidosis, cold stress, hypoglycemia, caloric deprivation, hypoxemia, and competition for bilirubin binding sites by drugs (e.g., furosemide, digoxin, and gentamicin) or free acids.

PRACTICAL CONSIDERATIONS

Clinical jaundice is apparent at serum bilirubin levels of 120–135 μmol/L. A rapid rise early in the neonatal period suggests hemolysis, secondary to inherited enzyme defects or to maternal–neonatal blood group incompatibilities. Prolonged (more than 2 weeks postnatally) hyperbilirubinemia is often associated with an increase in conjugated bilirubin due to biliary obstruction or hepatocellular dysfunction. Breast-milk jaundice commonly appears between 1 and 8 weeks of age. Mild indirect hyperbilirubinemia occurs with pyloric stenosis and quickly disappears after pyloromyotomy. Intestinal obstruction can intensify jaundice by increasing the enterohepatic circulation of bilirubin. Birth trauma with bleeding (e.g., caput medusae) can lead to jaundice as the blood is reabsorbed and hemolysed. Finally, jaundice is an early and important sign of septicemia.

If hemolysis is suspected, serial hematocrit estimations, reticulocyte counts, peripheral blood smears, and a Coomb's test are appropriate. Evaluation of neonatal sepsis includes hematocrit, white blood cell count and differential platelet count, chest radiography and cultures of blood, urine, and cerebrospinal fluid.

Phototherapy is widely used prophylactically in high-risk neonates to decrease the serum bilirubin levels by photodegradation of bilirubin in the skin to water-soluble products. It is continued until the total serum bilirubin level is less than 170 μmol/L and falling. The timing of phototherapy is based on the level of indirect bilirubin and the weight of the patient. Exchange transfusion is indicated if the indirect bilirubin level exceeds 340 μmol/L. The precise indications vary according to the individual patient, and in very LBW infants exchange transfusion is indicated at much lower serum bilirubin levels. Factors increasing the risk of kernicterus also influence the indications for exchange transfusion.

Immune function

As a group, neonates are particularly vulnerable to bacterial infections during the first 4 weeks of life. This may be due to maternal factors as well as to intrinsic deficiencies in their host defense system. Maternal factors independently associated with a higher incidence of neonatal sepsis include premature onset of labor, prolonged rupture of membranes (greater than 24 hours), chorioamnionitis, colonization of the genital tract with pathogenic bacteria such as group B Streptococci, and urinary tract infection. Neonatal factors include a diminished neutrophil storage pool, abnormal neutrophil and monocyte chemotaxis, decreased cytokine and complement production, and diminished levels of type-specific immunoglobulins including IgG, secretory IgA, and IgM. Overall, these factors lead to a significantly impaired host defense mechanism in the neonate with compromised anatomic barriers. These deficiencies are more severe in LBW infants.

PRACTICAL CONSIDERATIONS

The impaired immune function and compromised anatomical barriers of neonates may contribute to postoperative infection. Specifically, wound infections, as well as infections precipitated by indwelling catheters, may complicate the perioperative course of the neonate. For this reason, many surgeons advocate the use of prophylactic broad-spectrum antimicrobials in neonatal surgical patients. While this practice may be common, it should be noted that the specific antibiotics used as well as the duration of antibiotic therapy are very site-specific and surgeon-specific parameters. At this time, there are no conclusive studies supporting the use of any particular regimen. Therefore, the prophylactic use of antibiotics in these patients must be determined on a case-by-case and surgeon-by-surgeon basis.

Fluid and electrolyte homeostasis

FETAL TOTAL BODY WATER

In the fetus, total body water (TBW) constitutes 94 percent of the body weight during early gestation. As the fetus grows, this percentage progressively diminishes, to a value of 78 per-

cent at term. This then decreases further by approximately 3–5 percent during the first 5 days of life, eventually reaching adult levels by 9 months to 1 year of age. In addition to TBW, extracellular water (ECW) also declines until 1–3 years of age. In the term infant, ECW is often 40 percent of birth weight at 5 days. By 3 months of age, this value decreases to 33 percent, stabilizing at adult values of 20–25 percent by 1 to 3 years of age. Conversely, fetal intracellular water (ICW) slowly increases during gestation and the neonatal period. At 20 weeks' gestation, ICW is around 25 percent. This increases to 33 percent at the time of birth, finally reaching adult levels of around 44 percent by 3 months of age.

The neonate must complete these water redistribution tasks to transition effectively from the intrauterine to extrauterine environment. Under normal conditions, these changes in fetal body water progress in an orderly fashion in-utero and after birth. If this process is interrupted by premature birth or intrauterine growth retardation, specific tasks may be left uncompleted, predisposing the infant to an increased risk of developing serious complications such as patent ductus arteriosus and congestive heart failure.

RENAL FUNCTION

Compared to adults, the newborn infant has relatively low renal blood flow and plasma flow and high renovascular resistance. In fact, only 6 percent of the newborn's cardiac output is directed to the kidneys. This is in contrast to the 25 percent of cardiac output in adults. Overall, these factors lead to a relatively decreased glomerular filtration rate (GFR) in neonates. In term infants, the GFR rises rapidly during the first 3 months of life, nearing adult levels by 12–24 months of age. In premature infants, this process is delayed and GFR may lag behind that of the term infant.

In addition to GFR, the concentration capacity of the neonatal kidney is significantly lower than that of the adult kidney. Whereas the adult kidney can concentrate urine up to 1200 mOsm/kg, the neonatal kidney is only able to achieve 500–600 mOsm/kg. Furthermore, newborn renal tubules are relatively insensitive to the effects of antidiuretic hormone (ADH) and aldosterone compared to adults. These blunted responses are magnified in preterm infants. In addition, preterm infants are at a significant risk for salt wasting. This may lead to further growth retardation, as sodium appears to be a permissive factor for growth.

PRACTICAL CONSIDERATIONS FOR FLUID MANAGEMENT IN THE NEONATE

Effective fluid and electrolyte management involves:

- calculating the fluid and electrolyte requirements for maintaining metabolic functions;
- replacing fluid losses (third space, evaporative, insensible);
- considering pre-existing fluid deficits or excesses.

Taking these factors into consideration, a tentative program is devised for fluid and electrolytes administered for a finite period of time, usually 8 hours, although shorter intervals may be required in critically ill newborns. The response of the patient is then closely monitored and the treatment program adjusted accordingly.

CALCULATING MAINTENANCE NEEDS

The neonate's basic maintenance requirement for water is the volume required for growth, renal excretion (renal water), and replacing losses from the skin, lungs, and stool. Stool water loss has been estimated at 5–10 mL per 420 J expended, the lower figure applying to those patients not being fed. In the surgical patient with postoperative ileus, stool water loss is usually insignificant. Growth is inhibited during periods of severe stress and is also not a major factor under these conditions. The basal fluid maintenance requirement is therefore *renal water* plus *insensible loss*. Requirements during the first day of life are unique because of the greatly expanded extracellular fluid volume in the neonate, which decreases after 24 hours. In addition, neonates with intestinal obstruction are not hypovolemic as a result of intrauterine adjustments across the placenta. During these first 24 hours, basic maintenance fluid should not exceed 90 mL/kg in preterm infants weighing less than 1000 g or less than 32 weeks' gestational age. In larger infants, maintenance fluid rates should not exceed 75 mL/kg.

The basic electrolyte and energy requirements are provided by sodium chloride (2–5 mEq/kg per day) in 5 percent or 10 percent dextrose, with the addition of potassium (2–3 mEq/kg per day) once urine production has been established. Calcium gluconate (1–2 g/L fluid) may be added, especially in preterm infants.

RENAL WATER

The volume of water required for excretion by the kidney depends on the renal solute load and the child's renal concentrating ability. The solute load that the kidneys must excrete is derived from the endogenous tissue catabolism and exogenous protein and electrolyte intake. The osmolar load is thus reduced by growth and increased by tissue necrosis and high osmolar feeds/infusions. The volume of fluid administered should be sufficient to allow excretion of the solute load at an isotonic urine osmolality of 280 mOsm/dL. It is important to understand that there is no 'normal' urine output for neonates, due to the fact that the osmolar load is highly variable in newborns. The calculated ideal urine output, representing the renal water required to excrete an osmolar load, is also therefore variable.

INSENSIBLE LOSSES

Invisible continuing loss of water occurs from the lungs (respiratory water loss) and through the skin (TEWL), and constitutes the insensible water loss (IWL). Respiratory water loss (RWL) accounts for approximately one-third of IWL in infants older than 32 weeks' gestation and is approximately 5

mL/kg body weight per 24 hours at a relative humidity of 50 percent. The TEWL for a full-term infant in a thermoneutral environment is approximately 7 mL/kg body weight. The IWL for a full-term infant in the thermoneutral environment at 50 percent humidity is therefore 12 mL/kg per 24 hours.

The main factors that affect IWL are the gestational age of the infant and the relative humidity of the environment. For infants 25–27 weeks' gestation, TEWL has been estimated at 128 mL/kg per 24 hours at 50 percent relative humidity. The relative humidity has a marked inverse effect on TEWL, which decreases to almost zero as the relative humidity approaches 100 percent. Plastic sheets may be used to increase the relative humidity around the infant and reduce TEWL by 50–70 percent. Conversely, radiant warmers and phototherapy increase IWL. This loss is magnified in the preterm infant.

MANAGEMENT PROGRAM

The most commonly used method of calculating fluid requirements is based on body weight (Table 1.6). However, because of the many factors affecting maintenance requirements, there is no close or constant relationship between body weight and fluid and electrolyte needs. Thus, many surgeons advocate the use of a dynamic approach to fluid management. Such approaches generally begin with the administration of an initial fluid volume that is safe for the patient's status. This initial volume is essentially a 'best guess' volume. The effects of this volume on the patient's physiology are then monitored and appropriate changes are made.

Table 1.6 *Calculation of maintenance fluid requirements*

Body weight (kg)	Fluid volume per 24 hours
1–10	100 mL/kg
11–20	1000 mL + 50 mL for each kg over 10 kg
> 20	1500 mL + 20 mL for each kg over 20 kg

CALCULATION OF ADDITIONAL LOSSES

External losses from stomas, fistulae, and drainage tubes are directly measured and replaced volume for volume with an appropriate electrolyte solution. In neonates it is wise to measure the electrolytes in the fluid to guide replacement more accurately. Protein-rich losses (e.g., pleural fluid from chest tubes) are replaced with albumin solutions or fresh frozen plasma. Internal losses into body cavities or tissues (third space losses) cannot be measured, and adequate replacement of these losses depends on careful monitoring of the patient's response to fluid therapy.

CONSIDERING PRE-EXISTING FLUID DEFICITS OR EXCESSES

In addition to addressing maintenance requirements and additional losses, the fluid management of the neonate should include an assessment of any pre-existing fluid deficits or excesses. Pre-existing deficits may be due to in-utero or intrapartum hemorrhage as well as third space losses. Pre-existing excesses may be secondary to prematurity leading to a high TBW content. In all of these cases, the pre-existing condition should be considered when determining a fluid management plan.

MONITORING THE FLUID AND ELECTROLYTE PROGRAM

Once a fluid and electrolyte management program has been initiated, proper monitoring must occur to identify the newborn's response. In this manner, therapy may be adjusted dynamically to meet the specific needs of each neonate. The newborn's response to a fluid and electrolyte program may be monitored by clinical examination, body weight measurements, and urine volume and composition measurements.

CLINICAL FEATURES

Severe isotonic and hypovolemic dehydration results in poor capillary filling and collapse of peripheral veins. The skin is cool and mottled, with reduced turgor; the mucous membranes are dry and the anterior fontanelle is sunken. These findings occur with 10 percent body fluid losses in an infant of more than 28 days of age and with 15 percent losses in a neonate. Hypertonic dehydration is more difficult to detect clinically because the decrease in circulating blood volume is considerably less than the total loss of body fluids. Signs of shock occur late, and central nervous system signs such as lethargy, stupor, and seizures predominate.

BODY WEIGHT

Serial measurements of body weight are a useful guide to TBW in the neonate. Fluctuations over a 24-hour period are primarily related to loss or gain of fluid, 1 g body weight being approximately equal to 1 mL water. Errors will occur if changes in clothing, dressings, tubes, and standard intravenous boards are not accounted for, and if weighing scales are not regularly calibrated.

URINE VOLUME AND COMPOSITION

If the volume of fluid administered is inadequate, urine volume falls and its concentration increases. If excess fluid is administered, the opposite occurs. The authors aim to achieve a urine output that will maintain a urine osmolality of approximately 280 mOsmol/dL. In neonates, this usually results in a urine output of 2 mL/kg per hour. For infants and older children, hydration is adequate if the urine output is 1–2 mL/kg per hour with an osmolality of 280–300 mOsmol/kg. Serial hematocrit changes, in the absence of hemolysis or bleeding, also suggest a loss or gain of plasma water.

When the osmolar load is large, for example with extensive tissue destruction or with infusion of high osmolar solutions, urine flow may have to be increased to provide adequate renal clearance. Accurate measurements of urine flow and concen-

tration are fundamental to the management of critically ill infants and children. In this situation, the insertion of an indwelling urinary catheter is recommended.

The specific gravity of the urine is a reliable indicator of hypertonicity (> 1.012 specific gravity) and hypotonicity (< 1.008 specific gravity), but is unreliable if urine is in the isotonic range (1.009–1.011 specific gravity). When fluid monitoring is critical, urine osmolality estimations provide more precise information than specific gravity. An increase in osmolality suggests that too little water or too much electrolyte has been given. A fall in osmolality suggests that sodium replacement is inadequate or that too much water has been administered. An unexpected change in osmolality, particularly an increase, requires immediate determination of serum levels of electrolytes, blood urea nitrogen, and glucose values, and a calculation of the osmolality. Serum osmolality can be measured directly or calculated by the formula:

$$\text{osmalility} = \text{serum sodium} \times 1.86 + (\text{blood urea nitrogen}/2.8) + (\text{glucose}/18) + 5.$$

From this it is possible to determine whether the rise in osmolality is due to an increase in serum sodium, the development of hyperglycemia, or high blood urea nitrogen. Occasionally, the measured serum osmolality is higher than the calculated osmolality. This suggests that the increase in serum osmolality is due to some unidentified osmolar active substance such as a metabolic byproduct resulting from sepsis, shock, or radio-opaque contrast material.

A rising blood urea nitrogen level and falling urine output may be due to acute renal failure or prerenal oliguria with azotemia resulting from hypovolemia. The distinction between these two states is important for appropriate treatment. Initially, the response to a fluid challenge of 20 mL/kg 5 percent dextrose and sodium chloride over 1 hour is monitored. If oliguria persists, the sodium, creatinine, and osmolality levels in both the blood and urine are determined. The fractional excretion of sodium (Fe_{Na}) is calculated using the formula:

$$Fe_{Na} = (\text{urine Na}/\text{serum Na})/(\text{urine creatinine}/\text{serum creatinine}) \times 100.$$

A normal Fe_{Na} is 2–3 percent. A value below 2 percent implies prerenal azotemia, and a value above 3 percent implies renal failure.

Calcium and magnesium homeostasis

In addition to fluid and sodium management, calcium and magnesium homeostasis presents clinically significant challenges in the newborn surgical patient. The fetus receives calcium by active transport across the placenta, 75 percent of the total requirement being transferred after the 18th week of gestation. Hypocalcemia, defined as a serum level of ionized calcium below 1.0 mg/100 mL, is most likely to occur 24 to 48 hours after birth. Causes include decreased calcium stores, decreased renal phosphate excretion, and relative hypoparathyroidism secondary to suppression by high fetal calcium levels. Low birth-weight infants are at a great risk (particularly if they are preterm), as are those born of a complicated pregnancy or delivery (e.g., diabetic mother), or those receiving bicarbonate infusions. Exchange transfusions or the rapid administration of citrated blood may also lead to hypocalcemia. The symptoms of hypocalcemia are non-specific and include jitteriness, high-pitched crying, cyanosis, vomiting, twitching, and seizures. Diagnosis is confirmed by determining the serum calcium level. However, the ionized fraction of the serum calcium may be low, resulting in clinical hypocalcemia without a great reduction in total serum calcium. Therefore, evaluation of the serum ionized calcium level is often useful.

PRACTICAL CONSIDERATIONS

Hypocalcemia is prevented by adding calcium gluconate to daily maintenance therapy, 1–2 g/24 h intravenously or 2 g/24 h by mouth. Symptomatic hypocalcemia is treated by intravenous administration of 10 percent calcium gluconate in a dose of 1–2 mL/kg over 10 minutes; the rate should not exceed 1 mL/min.

Infants at high risk for hypocalcemia are also at risk for hypomagnesemia. In fact, the two conditions may coexist. If there is no response to attempted correction of a documented calcium deficiency, hypomagnesemia should be suspected and serum magnesium levels measured. Hypomagnesemia is corrected by administering 50 percent magnesium sulfate, 0.2 mEq/kg every 6 hours intravenously, followed by oral magnesium sulfate 30 mEq/day.

Although most seizures that occur in the neonatal period have a cerebral cause and are not secondary to hypoglycemia or hypocalcemia, hypocalcemia should be suspected in high-risk infants, particularly after surgery. Immediate blood glucose determination and serum glucose and calcium measurements should therefore be performed in a 'jittery' neonate. Treatment should be prompt, with intravenous glucose when hypoglycemia is suspected, followed by intravenous calcium if symptoms persist.

GENERAL CONSIDERATIONS IN THE PERIOPERATIVE CARE OF THE NEONATE

Monitoring

Due to the dynamic physiology of the neonatal period, newborn surgical patients should be monitored continuously in the neonatal unit. In addition to providing continuous oxygen saturation readings, transcutaneous pulse oximetry is useful for monitoring episodes of apnea and bradycardia, which can be common in preterm infants. Transcutaneous carbon dioxide monitoring is gaining popularity for those on mechanical ventilators – it provides accurate trending data.

Accurate monitoring of fluid status often requires an indwelling urinary catheter and frequent laboratory evaluations.

Invasive monitoring and access in the newborn can be achieved through the umbilical vessels as they are relatively accessible in the first 24 hours of life in this population. Specifically, umbilical venous catheters provide quick central venous access. A 3.5 French catheter is required for infants weighing less than 1500 g; infants weighing 1500–3500 g can accommodate a 5 French catheter. Umbilical artery catheters may be indicated in infants with significant respiratory distress or in any infant who may require frequent blood sampling. These catheters enter the aorta through the internal iliac arteries; 3.5 French catheters are used in infants weighing less than 1200 g, while 5 French catheters are used in those weighing more than 1200 g.

Nutrition

In neonatal surgical patients, proper nutrition must be delivered to meet their relatively large energy requirements. Specifically, neonates require a large energy intake because of their high basal metabolic rate, requirements for growth and development, energy needs to maintain body heat, and limited energy reserves. These requirements vary according to age and environmental factors and are significantly increased by cold stress, surgical procedures, infections, and injuries. Energy requirements are increased 10–25 percent by surgery, by more than 50 percent by infections, and by 150 percent by burns. Energy reserves are limited in the neonate, whose liver glycogen stores are usually consumed in the first 3 hours of life. These limited reserves are even more restricted in preterm and SGA infants.

The energy needs of individual newborns can be calculated according to the requirements for basal metabolism plus growth. Table 1.7 lists the energy requirements of children by age group. Consideration must also be given to the adequacy of energy reserves in the presence of stress factors such as cold, infection and trauma, and surgery.

Table 1.7 *Energy requirements by age*

Age	Energy required per 24 h (J)[a]
Basal metabolism: full-term infant	
Birth	134
2 weeks	202
1 year	168
Teen	97
Growth calories	
Birth	139
3 months	76
6 months	50
1 year	50
Teen	76
Total calories (maintenance and growth)	
Neonatal term (0–4 days)	462–504
Low birth-weight infant	504–546
3–4 months	420–445
5–12 months	420
1–7 years	378–315
7–12 years	315–252
12–18 years	252–126

[a]1 Kilocalorie = 4 Joules [J]

Pain management

Postoperative pain management in the newborn surgical patient may be challenging. In particular, the use of opioid analgesics in the neonate must be monitored carefully. As a group, neonates have a narrower therapeutic window for postoperative opioid analgesia than older age groups. In addition, neonates treated with opioids exhibit variable pharmacokinetics and are at a high risk for respiratory depression. Despite these challenges, postoperative opiate analgesia can be used effectively to control pain in neonates. However, this requires close monitoring and may necessitate consultation with a pain management service. In addition to opiate analgesics, acetaminophen and non-steroidal anti-inflammatory drugs (NSAIDs) may be used for pain control. In particular, the use of acetaminophen has had a long safety record in newborn patients.

Gastrointestinal decompression

The importance of gastric decompression in the neonate undergoing surgery cannot be overemphasized. The distended stomach carries the risk of aspiration and pneumonia, and may impair diaphragmatic excursion, resulting in respiratory distress. With congenital diaphragmatic hernia, ventilation is progressively impaired as the herniated intestine becomes distended with air and fluid. With gastroschisis, omphalocele and diaphragmatic hernia, the ability to reduce the prolapsed intestine into the abdominal cavity is impaired by intestinal distension. This may be alleviated by adequate orogastric or nasogastric decompression. A double-lumen sump tube, such as a Replogle® tube, is preferred, utilizing low continuous suction. If a single-lumen tube is used, intermittent aspiration is required. The correct position of the tube in the stomach is confirmed by carefully measuring the tube before insertion, by noting the nature of the aspirate, and by radiography. The tube should be carefully taped to avoid displacement. The use of gastrostomy tubes for postoperative gastric decompression is decreasing in popularity.

Diagnostic studies

Most laboratory tests pose an additional burden on the already stressed neonate. Therefore, diagnostic studies should be restricted to those essential for diagnosis and proper man-

agement. The volume of blood drawn for laboratory tests should be documented, as these small volumes cumulatively represent significant loss in a small infant.

When the patient is transferred to other departments for investigational procedures, monitoring and resuscitation equipment should be available, with a surgeon in attendance. All studies should be performed with minimal disturbance, taking steps to prevent heat loss. Before using hyperosmolar radio-opaque contrast materials, intravenous fluids must be administered and fluid deficits corrected, regardless of the route of administration. To counteract the osmotic effect of the contrast medium, an intravenous infusion of sodium chloride 34 mEq/L at twice the maintenance rate should be given during the radiographic study and for 2–4 hours afterwards. During this period the patient should be carefully monitored as described above.

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Pediatric anesthesia

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Pediatric anesthesia is recognized as a subspecialty. In different countries and institutions the upper age limit for pediatric patients can vary, usually between 14 and 18 years, but it is generally accepted that it is most important for infants and younger children to receive the specialist care of pediatric anesthetists for both physiological and psychological reasons, and to reduce the incidence of adverse events during even routine anesthesia.

The biochemical, physiological, and psychological needs peculiar to young children are best met in a children's environment, with all staff being trained and familiar with children and their needs. The recommended policy is to concentrate pediatric surgery and intensive care in children's departments and children's hospitals, so that each center for neonatal surgery serves a population of about 2 million. This is feasible because the transport of sick infants, even over very long distances, by surface or air, is now routine and safe, even for intubated and ventilated patients.

Many of the differences between adult and pediatric anesthesia are related to differences in anatomy and physiology – differences that are most marked in the very young. Surgical neonates with a gestational age as low as 24 weeks and a weight of 450 g are now surviving, so that the implications of the traditional neonatal period of 28 days of life in terms of development have become meaningless: the neonatal period is defined as up to 44 weeks after conception (Fig. 2.1).

The weight of the infant and an assessment of the function of its various bodily systems are more important guides than the age from birth. For a given gestational age, the morbidity and mortality are greater, the lower the birth weight. Therefore all children should be accurately weighed on admission to hospital. Many surgical procedures in children aged up to 3 years are for the correction of congenital defects, and it is important to remember that such defects are often multiple. For example, a cardiac abnormality may be present in children with a cleft palate or esophageal atresia.

PHYSIOLOGIC DIFFERENCES BETWEEN NEONATES AND OLDER CHILDREN

Infants have poor respiratory reserves, and respiratory failure is a common sequel to pathology in any other system. Total pulmonary resistance, at 25 cmH_2O/L per second, is five times that of the adult.

Lung compliance is very low (6 mL/cmH_2O compared with 160 mL/cmH_2O in the adult), but the infant chest wall is a very compliant structure, so that small infants have great difficulty in maintaining a normal functional residual capacity (FRC) in states in which pulmonary compliance is further

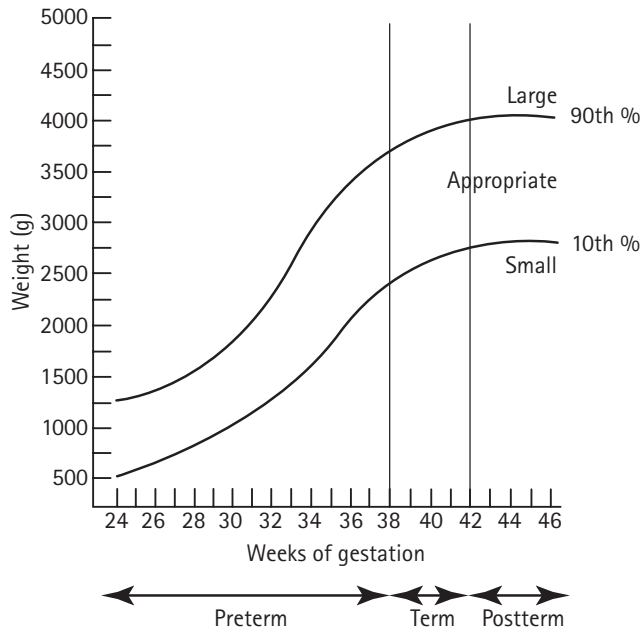

Fig. 2.1 Percentile chart showing appropriate weight for gestational age.

reduced. The chest wall provides no counter-resistance to the collapsing forces of the lungs, as it will do later in life. Hence, the response to constant distending pressure in the form of positive end-expiratory pressure (PEEP) or continuous positive airway pressure (CPAP) is strikingly beneficial in this age group.

After birth, an eight-fold increase in the number of alveoli occurs and the adult number is reached by the age of 6 years. The resistance of the airways (and thus the work of breathing) remains high, until finally, the airways begin to enlarge; this occurs at the same time as the full complement of alveoli is present. Closing volume occurs within tidal breathing until 6 years of life, so there is an increase in physiologic right-to-left shunt during this period, with an even greater effect on oxygenation should the FRC fall, as it does with pulmonary disease or during anesthesia.

The resistance of the nasal passages in neonates is relatively great (45 percent of the total). Neonates are obligatory nose breathers, so respiratory obstruction may occur if the nares are blocked, for example by choanal atresia or by a large nasogastric tube. This dependence on nose breathing can be exploited with great effect in the use of nasal prongs or masks to apply distending pressure in the form of CPAP.

Alveolar ventilation and oxygen consumption per unit body weight are twice those of the adult, as manifest by the alarming rate at which cyanosis appears if ventilatory problems arise.

Respiration during the early months of life is purely diaphragmatic (the bucket-handle effect of ribs becomes operational toward the end of the first year of life), so that respiratory failure may ensue if diaphragm movement is restricted, for example by abdominal distension or with phrenic nerve palsy. Attempts to increase alveolar ventilation can be made only by increasing the respiratory rate, which explains why a rising respiratory rate is a diagnostic sign of increased respiratory distress in infants. Phrenic nerve palsy may occur as part of a birth injury, but is more commonly associated with damage to the phrenic nerve during thoracic or cardiac surgery.

The circulation of the neonate is labile and may revert to the fetal pattern, with blood flowing from right to left through a patent ductus arteriosus and/or through the foramen ovale, if subject to conditions that promote pulmonary vasoconstriction: a state known as transitional circulation. This state, previously known as persistent fetal circulation, is a transitional state between the fetal circulation, including the placenta, and that of the adult, when the right and left sides are quite separate (Fig. 2.2).

The duct may reopen with exposure to hypoxia or fluid overload, until it is firmly closed by fibrosis after 3–4 weeks. Attempts can be made to close the duct pharmacologically, using small doses of a prostaglandin synthetase inhibitor such as salicylate or, more commonly, indomethacin, with a fair chance of success. Conversely, prostaglandin E_2 may be infused to maintain ductal patency where this is essential, for example in severely cyanosed infants with pulmonary artery atresia, until a systemic–pulmonary shunt of the Blalock type is created surgically.

The liability of the pulmonary vasculature is caused by abundant arteriolar smooth muscle, extending more peripherally than in later life (due to a failure of normal regression of the muscle in the first few hours of life). These arterioles constrict in response to hypoxia, hypercapnia, or acidosis via an adrenergic mechanism (this response is abolished after sympathectomy).

Some infants develop this state of persistent pulmonary hypertension of the neonate following a rise in pulmonary vascular resistance and a right-to-left shunt through the ductus arteriosus or patent foramen ovale.

Such a condition, with a shunt of 80 percent and critical hypoxemia, occurs commonly in respiratory distress of the premature infant, congenital diaphragmatic hernia, meconium aspiration, β-hemolytic streptococcal infections and post-maturity.

If left untreated, these infants will die in a vicious cycle of cyanosis, acidosis, and falling cardiac output. Steps must be taken to reverse the high pulmonary vascular resistance: a high inspiratory oxygen concentration (FiO_2), hyperventilation, if possible, and pH > 7.4. In the past, vasodilating drugs such as tolazoline, prostacyclin or glyceryl trinitrate have been used, but nowadays inhaled nitric oxide is the treatment

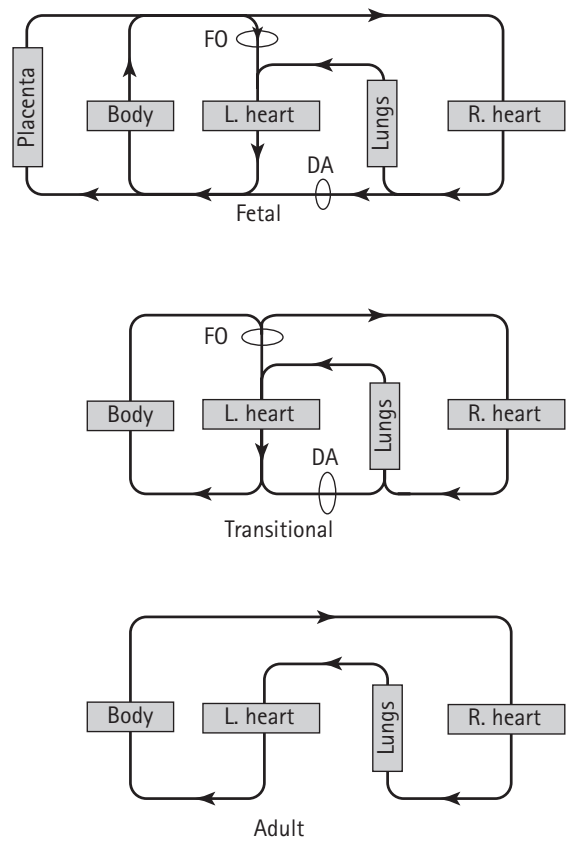

Fig. 2.2 Transitional circulation: FO, foramen ovale; DA, ductus arteriosus.

of choice. Extracorporeal membrane oxygenation (ECMO) may also be used for this condition at some specialist centers.

Neonates do attempt to maintain core temperature at 37 °C, but may not succeed because of an initial low basic metabolic rate, a large surface-to-weight ratio, immature sweat function, and an inability to adapt to adverse conditions. Superficial thermoreceptors exist in the trigeminal area of the face; hence a cold stimulus causes an increase in metabolic heat production from hydrolysis of triglycerides in brown fat, causing a great increase in oxygen consumption, which may make existing hypoxia worse. Brown fat is distributed over the back and provides thermal lagging for the major intrathoracic vessels. The metabolic response to cold is inhibited by general anesthesia, hypoxia, hypoglycemia and prematurity. Neonates are nursed in the neutral thermal environment at which their oxygen demands are minimal, as low as 31 °C for a 3-kg term baby and up to 36 °C for those with low birth weight (Fig. 2.3). If preterm infants are allowed to cool, there is increased mortality and morbidity. They are more likely to develop respiratory disease, acidosis, hypoxia, coagulopathy, intraventricular hemorrhage, and a subsequent slower rate of brain growth.

The mean cord hemoglobin concentration is approximately 18 g/dL at birth and rises by 1–2 g/dL in the first days of life because of low fluid intake and a decrease in extracellular fluid volume. After that, the level declines (see Figure 2.4) and causes the physiologic anemia of infancy. Premature babies have a greater fall because of lower red cell production and survival. At birth, 70 percent of the hemoglobin is HbF, which has a greater affinity for oxygen, possibly because of a relative insensitivity to 2,3-diphosphoglycerate, which itself lowers the oxygen affinity of the hemoglobin molecule. The HbF is replaced by HbA by 3 months of age, at which time sickle tests become positive in children with sickle-cell disease, although most will have been diagnosed by electrophoresis in the newborn period. A hemoglobin concentration < 10 g/dL is always abnormal and should be investigated. Non-emergency surgery is usually delayed pending the investigation of severe anemias.

The blood volume of an infant with normal hemoglobin is estimated to be 80–85 mL/kg. In very premature infants it is greater, perhaps as much as 100 mL/kg. The blood volume of the neonate is more variable than that of the older infant and depends on the magnitude of the placental transfusion. Difficulties may arise if blood replacement is based on percentage of the estimated blood volume.

Carbohydrate reserves of the normal neonate are relatively low and, as most glycogen is synthesized after 36 weeks' gestation, those of preterm infants may be very low. Blood sugar levels should average 2.7–3.3 mmol/L (50–60 mg/dL), and hypoglycemia of less than 1.6 mmol/L (30 mg/dL) is treated by infusion of 10 percent dextrose, or a bolus of 1 mL/kg 25 percent dextrose if urgent correction of hypoglycemia is indicated. Frequent testing for blood sugar using point of care equipment gives improved control. There is no agreement as to the hypoglycemia effect of preoperative fasting, but 4 hours between the last clear drink and induction of anesthesia should be the very maximum, and small children should receive a drink of clear fluid containing sugar up to 2 hours preoperatively. The usual regimen for preoperative starvation at the major children's hospitals worldwide is 6 hours for food and formula feeds and 4 hours for breast-milk, with 2 hours for clear fluids. Premature and very small babies receiving more frequent feeds may have shorter periods of starvation if this is discussed with the anesthetist. Children below 15 kg in weight are at greatest risk from perioperative hypoglycemia.

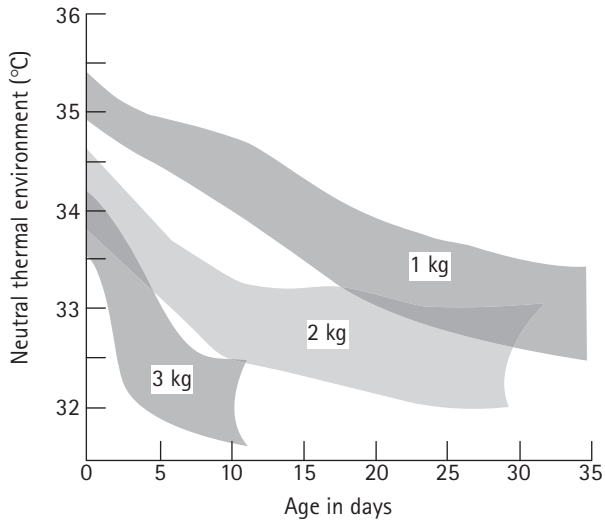

Fig. 2.3 Neutral thermal environment for three groups of infants of differing birth weight.

Maturity of liver enzyme systems is complete by 2 months of age. The synthesis of vitamin-K-dependent clotting factors II, VII, IX, and X is suboptimal until then. Minimal levels of clotting factors occur on the second or third day of life, and this is partially prevented by routine oral administration of

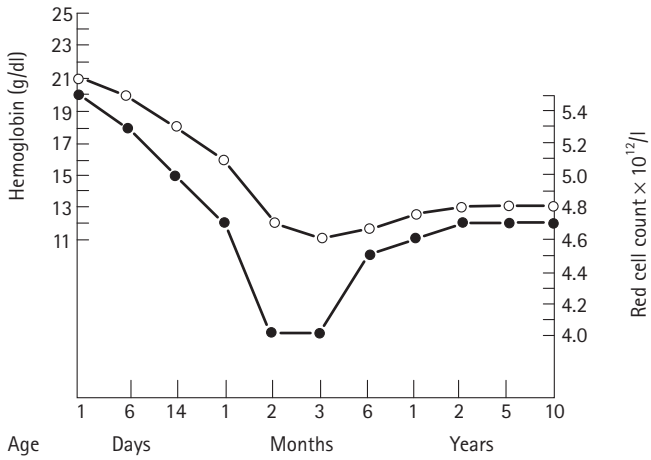

Fig. 2.4 Changes in hemoglobin concentration and red cell count in the first 10 years of life; ○, hemoglobin; ●, red cell count.

vitamin K to all neonates or by intramuscular injection if the oral route is not available.

Hepatic immaturity also means that drugs metabolized in the liver, such as barbiturates and opiates, should be used with extreme caution. The conjugation of bilirubin is very inefficient, and uncoupling of at least one of the two molecules occurs at times of stress such as during hypoxia or acidosis. After liver maturity is reached, most drugs are well tolerated because of the high metabolic rate of the young child.

The neonate has no diuretic response to a water load for the first 48 hours of life. By the end of the first week, dilute urine can be produced, but the output falls before the full load has been excreted.

Fluid maintenance requirements for full-term infants start at 20–40 mL/kg per 24 hours on day 1, increasing by 20 mL/kg each day until the levels shown in Table 2.1 are reached by the end of the first week of life.

Fluid retention associated with the surgery is usually severe, and restriction to the requirements suggested for the neonatal period is necessary during and after operation. All intravenous fluids for maintenance therapy should contain 4 percent or 10 percent glucose in newborns (depending on blood sugar), with abnormal losses being replaced with an isotonic replacement fluid separate to the maintenance solution. Added potassium chloride may be required. Hyperglycemia may occur even in very low birth-weight neonates undergoing surgery if glucose solutions are continued during the operation.

Table 2.1 *Basic fluid requirements of neonates 7 days after birth*

Birth weight (g)	Volume/24 h (mL/kg)
< 1000	180
1000–2500	150
> 2500	120

ASSESSMENT OF THE PATIENT

Fitness for general anesthesia and surgery must be assessed in relation to the urgency of the surgery. Assessment often involves weighing up the risks related to an associated medical problem against the benefits of surgery. This requires cooperation between the anesthetist and the surgeon. Many centers run preoperative clinics in which medical problems can be identified, appropriate investigations performed, and treatment instituted in order to optimize the child's condition prior to surgery. The parents and patients can also be given the necessary instructions for admission to hospital, which is especially important for day-cases.

Elective surgery should not take place when the patient has an acute intercurrent illness. The operation should be deferred about 1 month after the last symptoms of respiratory tract infection, croup, or the acute exanthems have subsided, as related adverse events can occur for up to 6 weeks. However, some children requiring surgery, especially for ear, nose and throat procedures, may suffer repeat upper respiratory infections, and the next episode may occur if the operation is postponed for too long a period.

After bronchiolitis, pulmonary abnormalities of increased resistance and reduced compliance may persist for as long as

Table 2.2 *Prophylactic regimens for dental, oral, respiratory tract, or esophageal procedures*

Situation	Agent	Regimen
Standard general prophylaxis	Amoxicillin	Adults: 2.0 g Children: 50 mg/kg orally 1 h before procedure
Unable to take oral medications	Ampicillin	Adults: 2.0 g i.m. or i.v. Children: 50 mg/kg i.m. or i.v. within 30 min before procedure
Allergic to penicillin	Clindamycin	Adults: 600 mg Children: 20 mg/kg orally 1 h before procedure
	Cephalexin[a] or cefadroxil[a]	Adults: 2.0 g Children: 50 mg/kg orally 1 h before procedure
	Azithromycin or clarithromycin	Adults: 500 mg Children: 15 mg/kg orally 1 h before procedure
Allergic to penicillin and unable to take oral medications	Clindamycin or cefazolin[a]	Adults: 600 mg Children: 20 mg/kg i.v. within 30 min before procedure Adults: 1.0 g Children: 25 mg/kg i.m. or i.v. within 30 min before procedure

i.m., intramuscularly; i.v., intravenously.

[a]Cephalosporins should not be used in individuals with immediate-type hypersensitivity reaction (urticaria, angioedema, or anaphylaxis) to penicillins.

1 year. In patients with chronic respiratory disease, lung function is assessed by measuring airway resistance, compliance, and lung volumes, and by ventilation/perfusion scans. Baseline blood gas estimations may show metabolic alkalosis compensating for respiratory acidosis, or a raised $PaCO_2$ if there is incipient respiratory failure. Patients with values that are 50 percent of the predicted normal may be expected to develop respiratory problems after anesthesia and surgery, and in those with only 30 percent of predicted values with a resting $PaCO_2$ above 40 mmHg (5.3 kPa), postoperative respiratory support should be anticipated after major surgery and possibly even after apparently trivial procedures.

Preoperative antibiotic prophylaxis against subacute bacterial endocarditis is essential for patients with corrected or uncorrected congenital heart disease undergoing surgery associated with bacteremia. Antibiotics appropriate to the surgery and anesthesia are required. (See Table 2.2 for dental surgery prophylaxis.)

The history, including exercise tolerance and the physical examination of the child, should reveal any potential problems such as respiratory obstruction or respiratory failure. The anesthetist is also alerted to possible problems, such as a difficult intubation due to a small jaw or limited mouth opening and neck movement, for example in patients with Pierre–Robin sequence or Still's disease.

DAY-CASE SURGERY

Most minor surgery of all specialties is performed on a day-case basis; even in specialist centers this may approach 40 percent, and in many other centers almost all cases are admitted and discharged on the day of surgery. This arrangement is more cost effective, more convenient for the parents, and has obvious psychological advantages for the child. Anesthetic techniques of premedication, intubation, inhalation or intravenous anesthesia, local blocks, and postoperative analgesia need not differ significantly from those for hospitalized patients and are geared to a rapid and pain-free return to normal function. However, facilities must be available to admit a child overnight if the anesthesia or surgery has not been straightforward or if the parents feel they cannot manage at home.

In general, babies of less than 46 weeks since conception should not be treated on a day-stay basis, even for minor surgery. If they have a history of previous apneic attacks, or are ex-premature babies, there is a risk of apnea occurring up to 24 hours after the operation.

Infants and small children are vulnerable to the psychological stress of being in hospital and undergoing surgery. They are totally dependent on their parents, and prolonged separation in the early months of life may cause problems with maternal bonding. Children between 2 and 4 years of age are especially vulnerable, as they may have unreasonable fears about hospitals and surgery but may not yet have developed the intellectual mechanism to deal with these fears. Full preparation with a kindly and sympathetic approach is therefore required. Some children, despite this, will develop behavioral changes, which may last days or occasionally weeks.

PREMEDICATION

There is no ideal agent for premedication. The aim is to achieve mild sedation for most children, since a dose required to produce sleep in most, will cause over-sedation in a few. In recent years, preoperative medication has become less important, with parents always being present at induction and the universal use of topical anesthetic creams to allow painless intravenous induction of anesthesia. Opioid premedication is rarely used, as intramuscular injections are so disliked, and intraoperative and postoperative analgesia is usually managed by specific measures involving regional analgesia or intravenous opioid infusions.

Predication drugs include midazolam 0.5 mg/kg, temazepam, 0.50–1 mg/kg, and chloral hydrate 30–50 mg/kg – all administered orally. Other routes of administration, such as nasal or rectal, have been used, but no one method or agent has been shown to be superior.

The use of atropine as an anti-sialogogue used to be widespread, but since the development of non-irritant inhalational agents, it is now largely confined to specialist areas of practice such as upper airway endoscopy.

EQUIPMENT

Specialized apparatus with low resistance to breathing (less than 30 cmH_2O/L per second during quiet breathing) and minimal dead space is necessary as infants already have a high airway resistance and a rather higher ratio of dead space to tidal volume than adults.

Jackson Rees' modification of Ayre's T-piece has almost universal approval for small infant anesthesia. The T-piece has been extensively studied and no rebreathing with spontaneous or controlled ventilation occurs unless the fresh gas flow is reduced below 220 mL/kg per minute. No circuit should be used at the limit of its function, so in practice at least 4 L fresh gas flow is used. Commercially available scavenging devices are widely available for the T-piece.

With the advent of newer and more expensive volatile agents such as isoflurane and sevoflurane, circle systems are widely used for both spontaneous and intermittent positive pressure ventilation.

Clear plastic, cuffed facemasks provide a better fit to the face, and a firm fit also enables distending pressure to be applied to spontaneously breathing patients, which prevents stridor, promotes gas exchange, and prevents reduction in FRC. The apparently larger dead space of some such masks is usually unimportant, as the fresh gas flow streamlines within the mask.

The infant larynx lies higher in the neck (opposite the fourth cervical vertebra) and more anterior than in the adult and, as the epiglottis is relatively large, laryngoscopy is best performed with a small, straight- bladed laryngoscope, the tip of which picks up the epiglottis. Perfect sizing, positioning, and fixation of the tracheal tube are central to pediatric anesthesia and intensive care.

The correct size of tube is that which allows a small air leak between it and the mucosa of the cricoid at a peak inspiratory pressure of 25 cmH_2O. The cricoid ring is the narrowest part of the upper airway in a child and is easily damaged by too large a tracheal tube, resulting in postoperative stridor or even subglottic stenosis (1 mm of mucosal edema in the infant cricoid will reduce the airway by 60 percent). Uncuffed tracheal tubes with a small leak have been routinely used in both anesthesia and intensive care; however, the use of circle systems with low gas flow, and also the potential problems with a leak around the tube with non-compliant lungs, have reintroduced the debate on the use of cuffed tubes in infants and children.

A small airway inserted alongside an oral tube can splint it and prevent lateral movement and kinking. An oral tube is usually fixed to the face by two pieces of adhesive strapping to prevent dislodgement. Nasal intubation is preferable for some head and intraoral surgery and is also a more secure route for long-term ventilation in infants and children, apart from very premature infants in whom the nasal cartilage is too soft and can erode due to the continuous pressure from the tube. Cuffed tracheal tubes are used routinely in older children, usually over the age of 8 years.

The laryngeal mask airway (LMA) designed by Sir Archie Brain, is routinely used in pediatric anesthesia and is available in sizes 1, 1½, 2, and 2½, which are suitable for all sizes of children. There is an increase in the complication rate with diminishing LMA size. Intermittent positive pressure ventilation can be performed easily with the laryngeal mask in children, although many pediatric anesthetists would avoid this technique in smaller infants, as the potential to inflate the stomach and develop decreased compliance from diaphragmatic splinting may be significant. The newly introduced disposable laryngeal masks will remove the theoretical risks of prion and other infective agent transfer.

WARMING DEVICES

There are numerous heating devices available to help maintain normothermia in small children and infants during anesthesia. Many are either under-patient or over-patient hot air or water heaters with thermostatic controls. The efficacy of most of these devices makes it mandatory that temperature measurement of the patient is closely monitored. In children receiving large volumes of intravenous fluids, especially blood, fluid warmers are essential. The most effective of these warm the fluid right up to the point where it enters the patient, such as the coaxial inline water heater.

GENERAL PRINCIPLES OF ANESTHESIA

The principles of anesthesia in infants and children are similar to those for adults; however, infants weighing less than 5 kg are usually intubated for anesthesia, however minor the surgery, to allow controlled ventilation and avoid hypoxemia and hypercapnia. Metabolic studies clearly show that even neonates mount harmful stress responses to surgery, and that these can be obtunded by opioid or regional anesthesia. Adverse responses may contribute to morbidity and mortality or the prolongation of recovery.

Induction and maintenance agents

Induction of anesthesia is usually achieved either by inhalation or intravenous administration. Halothane continues to be a popular agent worldwide as it is readily accepted by children, but in many developed countries it has been replaced by the newer agent sevoflurane, which provides rapid and well-tolerated induction of anesthesia, with improved maintenance of cardiac output.

Intravenous induction is usually with Propofol (3 mg/kg), the non-barbiturate induction agent, as it is very short acting and may therefore be of some advantage for day-case patients or where rapid recovery is desirable. Thiopentone sodium in doses of 4–5 mg/kg may be used as an alternative. Ketamine (1–2 mg/kg intravenously) is a useful agent for children with cardiovascular instability as it may enhance cardiac output. Halothane and isoflurane are commonly used for maintenance of anesthesia. The metabolites of halothane may cause sensitization and severe liver failure in postpubertal patients, but although this is only very rare in children, its use has declined considerably. Halothane may preserve blood flow to the liver better than other agents and is thus not contraindicated in children with liver disease. The higher cost of isoflurane has been offset by the widespread use of circle systems with low fresh gas flow rates.

Muscle relaxants

Sensitivity of the neuromuscular junction to non-depolarizing muscle relaxants exists during the first 2–3 weeks of life. This, together with wide individual variation, makes careful titration of dose with effect mandatory.

The progress of action of the relaxants is monitored with a peripheral nerve stimulator using the 'train of four' response, although the post-tetanic count is more sensitive. Atracurium besylate is the relaxant of choice for neonates as its metabolism is independent of hepatic and renal function, and it may be given by bolus (0.5 mg/kg) or continuous infusion (9 mcg/kg per minute). The short-acting non-depolarizing relaxant mivacurium, inactivated by plasma cholinesterase, may also be given by infusion. Pancuronium is often preferred for cardiac surgery because of its long duration of action. In this setting, its tendency to produce tachycardia

may be offset by the administration of very large doses of opiate.

The reported resistance to succinylcholine (suxamethonium) in the neonate is caused by the dilution of a given dose in the relatively large extracellular fluid volume. The indications for the use of this agent are significantly reduced with the introduction of newer agents; however, the rapid and optimal conditions produced for intubation are useful in rapid sequence inductions and critical airway management.

Prevention of aspiration

Cricoid pressure to prevent the aspiration of regurgitated gastric contents is as effective in infants as it is in adults if correctly applied and used when indicated, for example in patients with intestinal obstruction.

ANALGESIA

Analgesia is balanced with anesthesia to provide stress-free conditions for surgery with improved outcomes. The technique used can either involve intravenous opioids such as fentanyl (1–10 mcg/kg) or morphine (0.05–0.2 mg/kg) or be regional, or a combination of the two. Great care must be taken when opioids are given to neonates unless postoperative mechanical ventilation is planned. Older infants and children tolerate up to 10 μ/kg fentanyl without the need for postoperative ventilation, and this provides excellent analgesia.

Regional anesthesia

Central or peripheral nerve blocks usually, but not necessarily, associated with light general anesthesia or sedation are routine in pediatric anesthesia. They obviate the need for opioid analgesia in high-risk groups such as ex-premature infants. In day care, they result in good postoperative analgesia with a reduced incidence of side effects. Techniques such as spinal or extradural blocks with catheters are used even in neonates and have the advantage that they can be continued into the postoperative period. Sacral lumbar and thoracic roots up to T10 may be blocked by caudal analgesia using 0.25 percent plain bupivacaine. Newer local anesthetic agents such as ropivacaine and levo-bupivacaine appear to be less cardiotoxic than the older agents.

Most pediatric surgery is suitable for the use of local anesthetic techniques of some type. Caudal epidural blocks are widely used, very safe, and easily performed in most children and are suitable for perineal, lower abdominal, and lower limb surgery in small children. In older ambulant children, the numb legs postoperatively may be a disadvantage, and ileo-inguinal nerve block may be preferable for inguinal herniotomy or orchidopexy. Penile blocks can be used for circumcisions and minor hypospadias repair, although caudal blocks are preferable for more extensive repairs. Axillary brachial plexus blocks are also easily and safely performed in children and are suitable for most upper limb surgery. Simple infiltration combined with either opiate intravenous analgesia or simple analgesics is also very effective, both for short procedures and for immediate postoperative analgesia.

Postoperative pain management

Since it was discovered that postoperative pain in children was being seriously under-treated, a great deal of attention has been given to the subject of acute pain relief in this patient group. Many children's hospitals and large centers have established acute pain services with physicians and nurses to treat, audit, and research this problem. Neonates present a unique problem of assessment and of treatment due to their sensitivity to the respiratory depressant effects of opioid analgesia. Regional techniques as already described, including simple wound infiltration, are used whenever possible. Paracetamol is safe and effective in neonates in doses not exceeding 60 mg/kg per 24 hours. Non-steroidal anti-inflammatory drugs (NSAIDs) can be given to children over the age of 6 months, and possibly younger, provided they have normal renal function and are not wheezy, and are excellent in combination with paracetamol, especially for ambulatory care. Analgesia for neonates, infants, and children after major surgery is based on morphine intravenous infusions unless there is an epidural infusion. They obviate the need for painful intramuscular injections and avoid the peaks and troughs of bolus administration. A regimen appropriate to the patient, surgery, location of nursing (intensive care, high dependency, or ward), and the institutional protocols is essential, but most are based on infusions delivering 20–40 mcg/kg per hour for older children and infants, with neonates receiving lower doses.

Patient-controlled microprocessor pumps can be used by children as young as 5 years, whilst younger children usually benefit from a higher background delivery supplemented by nurse-controlled boluses. Continuous epidural infusions of local anesthetics with or without opioids are widely used for pain relief after major abdominal and thoracic surgery in pediatric practice.

MONITORING

Suitable adaptations of standard techniques of monitoring used in adult practice are acceptable for all children, including neonates. Minimal standards of monitoring include electrocardiography (ECG), pulse oximetry, non-invasive blood pressure measurement, inspired and expired gas analysis, with CO_2 and O_2, and anesthetic agents. In addition, many anesthetists find the precordial or esophageal stethoscope a useful adjunct in pediatric practice. For all but the very briefest procedures, and for any in which a heating device is

used, central temperature monitoring – usually nasopharyngeal – is mandatory. Peripheral temperature monitoring is useful in prolonged procedures and can help inform volume replacement. Direct measurement of arterial and central venous pressure is used routinely for much major pediatric surgery. The easy access to sampling blood, and the use of near-patient blood testing, allow close control of fluid therapy, blood replacement, and blood sugar monitoring.

FLUID MAINTENANCE

Care is required with clear maintenance fluids during neonatal surgery, as fluids to flush drugs may be sufficient for requirements, particularly if the neonate has been receiving preoperative intravenous fluids. Neonates who have been on preoperative glucose infusions are prone to hypoglycemia if the glucose infusion is not continued. Frequent blood glucose monitoring is essential.

Older children may be given intraoperative fluids (as balanced electrolyte solution) at 6–10 mL/kg per hour. The routine use of the traditional 4 percent dextrose and 0.18 percent saline solution is no longer encouraged, as it is increasingly clear that hypoglycemia is rare outside the neonatal period and the injudicious use of functionally hypotonic solutions may be associated with severe hyponatremia.

Abnormal losses should be replaced with appropriate isotonic solutions such as 0.9 percent saline, human albumen solution, or artificial colloid solutions. The use of red cells is avoided unless absolutely necessary and should be guided by bedside testing of hematocrit. Much lower hemoglobin levels are now tolerated than in the past, and most anesthetists would not transfuse a fit older child without ongoing losses unless the hemoglobin fell below 7.0 g/dL. Techniques such as cell salvage and acute normovolemic hemodilution are increasingly used to avoid transfusion in major surgery.

RESPIRATORY SUPPORT AND POSITIVE PRESSURE VENTILATION

Many machines exist for the intraoperative mechanical ventilation of children. T-piece occluding machines such as the Penlon 200 series with the Newton valve are satisfactory for simple cases in children with normal lungs, but most centers now use dedicated pediatric ventilators. Whatever ventilator is used, it is essential that is has a reliable alarm system.

Hand ventilation is still the 'gold standard' in situations of rapidly changing pulmonary compliance or if there is tracheal compression. Controlled ventilation should be used for all neonates because the respiratory depressant effect of inhalational anesthesia is so great at this age. Older infants may tolerate short periods of spontaneous ventilation via a tracheal tube, laryngeal mask, or facemask.

At the end of surgery, infants are extubated when fully awake once spontaneous respiration is judged to be adequate. Because of the low respiratory reserve at this age, however, respiratory failure may ensue. Acute respiratory failure is a clinical diagnosis based on a rising respiratory rate (> 60/min), pulse rate, cardiac output, and oxygen dependence and on an assessment of the work of breathing as shown by intercostal recession, tracheal tug, nasal flaring, restlessness, and grunting. An inability to clear secretions or apneic attacks are further pointers. Blood gas levels may confirm the clinical impression and may be measured to determine baseline values.

Distending pressure in the form of CPAP can be useful when an infant cannot maintain adequate saturations (> 90 percent) in 60 percent oxygen. The CPAP may be administered via a tight-fitting facemask or nasal prong in an attempt to avoid tracheal intubation.

Prolonged intubation requires nasal plain polyvinyl chloride (PVC) tubes. All the complications of blockage, dislodgement, and subglottic stenosis can be avoided by meticulous care. The tube must be of a size to allow effective ventilation but also some leakage of air around it, or damage to the mucosa of the cricoid will result. Intubation may be continued if necessary for many weeks without resorting to tracheostomy. As mentioned earlier, there is a resurgence of interest in the use of cuffed tubes, and some centers are using them without any apparent increase in complications.

The art of ventilating babies with modern infant ventilators consists of using the facilities of the machines to minimize the factors such as high inspired oxygen concentrations and barotrauma known to be associated with bronchopulmonary dysplasia (BPD).

Infant lungs are particularly liable to be damaged by intubation and mechanical ventilation, with factors such as high inspired oxygen concentrations and high peak airway pressures being incriminated in the production of BPD. The lung architecture is progressively deranged, with fibrosis and the formation of cysts, which in turn demands higher oxygen and ventilator pressures to maintain adequate gas exchange. Unless the factors known to produce BPD are minimized, the condition will progress until ventilation becomes impossible. Machines must have the facility to allow high respiratory rates, variable inspiration:expiration ratio, control of peak pressures, and full humidification. They should also have an alarm system for disconnection and power failure, a facility for constant distending pressure, and the ability to wean the patient from the ventilator by using intermittent mandatory ventilation or similar.

Most infant ventilators also have the facility for patient-triggered ventilation, which may be useful when there are difficult weaning problems.

Constant distending pressure, whether used with intermittent positive pressure ventilation (PEEP) or spontaneous breathing (CPAP), will improve the relation between FRC and closing volume in the lungs, and reduce right-to-left intrapulmonary shunting. By keeping the small airways distended, it will also cause a fall in pulmonary resistance and in the work of breathing.

Positive end-expiratory pressure of up to 10 cmH_2O is used routinely in patients with increased pulmonary water. The distending pressure may preserve surfactant, but, as PEEP is increased, pulmonary vascular resistance rises and there is an increased incidence of pneumothorax.

Formulae should not be relied upon for setting up patients for ventilation because of the internal compliance of the machine, its tubing, and other variables: 10 mL/kg tidal volume or 20–25 cmH_2O peak inflation pressure is a reasonable initial setting, but adequate chest expansion should be confirmed clinically. Inspired oxygen concentration should be set at the level the child needs before mechanical ventilation is instituted and monitored initially by means of the pulse oximeter. The lowest FiO_2 necessary to maintain satisfactory saturations should be used.

All children ventilated in an intensive care unit require analgesia such as morphine infusion, at least at the start, but later they may be ventilated with simple sedation such as midazolam 0.1–0.2 mg/kg intravenously, or chloral hydrate 30 mg/kg by nasogastric tube. However, it may be necessary to paralyze patients with severe pulmonary disease with infusion of atracurium or vecuronium in order to limit peak airway pressures. Ventilation in such children should not aim to achieve normal carbon dioxide levels at the expense of excessive airway pressure. Newer ventilatory modes such as pressure regulated volume control (PRVC) may also help to minimize barotrauma.

Patients are extubated when coping on low levels of CPAP. Those with stiff lungs who are CPAP dependent may continue on CPAP after extubation, using a nasal prong. No pediatric patient should be left to breathe through a tracheal tube without distending pressure, since, without the physiologic levels of CPAP, the FRC will fall with increased resistance to gas flow, increased work of breathing, and increased right-to-left intrapulmonary shunting.

Invasive and non-invasive blood gas analysis is essential for setting up and maintaining patients on mechanical ventilation, but plays a less important part in monitoring them during the weaning process, when clinical observation of respiratory rate and effort is more important.

High-frequency jet ventilation or oscillation, negative pressure ventilation and ECMO are used in various centers for difficult patients or cases of extreme respiratory failure.

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3

Vascular access

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HISTORY

William Harvey's description of the heart and circulation in 1628 provided the anatomical basis for subsequent vascular interventions. By the early twentieth century, intravenous therapy was becoming established, replacing proctoclysis and cutaneoclysis as a means of delivering fluids and drugs to patients. Effective antimicrobials, chemotherapeutic agents, total parenteral nutrition (TPN), and the evolution of a culture of intensive care are among the medical advances that have encouraged the development of innovative strategies for vascular access in children over the last 50 years. This group of procedures is now one of the most common performed by pediatric surgeons.

PRINCIPLES AND JUSTIFICATION

Venous and arterial access is a key part of the management of many children who require investigations, monitoring, and specific interventions. Access procedures may facilitate the monitoring of physiologic, hematologic and biochemical indices. Therapeutic indications include the delivery of fluids, blood products, nutrition and drugs, hemodialysis, as well as miscellaneous interventions such as endovascular surgery and cellular transplantation.

Some children put forward for vascular access may be suitably managed by simpler, less invasive, and equally effective means. Despite improvements in devices, insertion techniques, and postoperative care, distress and complications related to catheter insertion are not infrequent and may be severe. In consultation with other members of the multidisciplinary team, it is the responsibility of the surgeon to ensure that the vascular access procedure proposed for each child can be justified.

OVERVIEW OF DEVICES AND TECHNIQUES

In most children, short-term venous access for sampling and the delivery of non-irritant infusions is achieved by the use of a short 14–26 gauge cannula into a superficial vein in an upper or lower limb. In addition to easily visible veins, useful sites include the long saphenous vein anterior to the medial malleolus at the ankle, the cephalic vein at the wrist, the interdigital vein between the fourth and fifth metacarpals on the dorsum of the hand, the external jugular vein (EJV), the superficial temporal vein of the scalp in small infants, and, occasionally, superficial veins on the trunk.

For children who require intravenous access for several days, the insertion of a 'short long line' early in the course of treatment, while superficial veins are well preserved, may reduce the frequency of recannulation. A number of suitable devices are available. Unless coincident with a general anesthetic for another reason, these procedures are generally performed in the awake child with the aid of topical anesthetic creams or sprays. Familiarity with venepuncture in children and maintenance of competence are recommended for pediatric surgeons.

The large number of central venous access devices fall into four broad categories: percutaneously inserted central venous catheters (PICCs), non-tunneled central venous catheters (CVCs), tunneled CVCs (such as Hickman or hemodialysis catheters), and venous port devices (Table 3.1). The type of central venous access device used will depend on the requirements of the individual child. The ideal position of the CVC tip is contentious. The options include the superior vena cava (SVC), the right atrium, and at the junction between the two. Individual patient requirements, government guidelines, manufacturer's recommendations, and institutional and personal experience and practice should be taken into consideration when deciding the final position. In general, our preference is for placement in the proximal right atrium. Stiff catheters inserted percutaneously are placed low in the SVC.

Table 3.1 *Types of central venous access device*

Device	Characteristics	Advantages	Disadvantages	Typical indications
Non-tunneled central venous catheter	Short, relatively stiff, usually multiple-lumen catheter	Ease of insertion	High infection rate	Short-term intravenous therapy or pressure monitoring
Single-lumen Hickman catheter	Relatively soft catheter	Relatively low infection rate	No possibility of co-administration of incompatible infusions	Total parenteral nutrition, low-intensity chemotherapy (e.g., nephroblastoma)
Multiple-lumen Hickman catheter	Relatively soft catheter with two or three lumens	Co-administration of blood products, parenteral nutrition and drugs	Higher infection rate	Intensive chemotherapy protocols, bone marrow transplantation
Non-cuffed tunneled central venous catheter	Small-caliber soft catheter with one or two lumens	Ease of removal	Higher rate of inadvertent removal and infection	Short- to medium-term access with reliable blood sampling
Non-tunneled hemodialysis catheter	Large-diameter catheter with offset lumens	Ease of insertion	Short lifespan, higher infection rate	Short-term hemodialysis, plasmapheresis, stem cell harvest
Tunneled (permanent) hemodialysis catheter	Large-diameter catheter with offset lumens	Long lifespan, low infection rate	Higher incidence of damage to vein	Long-term hemodialysis
Central venous port device	Subcutaneous port with attached venous catheter	Even lower infection rate than Hickman catheter	Requires needle access, longer scar than Hickman catheter	Chemotherapy, conditions requiring regular transfusions of blood products (e.g., hemophilia) or antibiotics (e.g., cystic fibrosis)
Peripherally inserted central venous catheter	Small-caliber soft catheter with one or two lumens	Safe insertion without general anesthetic, ease of removal	Higher rate of inadvertent removal and occlusion	Short- to medium-term access (e.g., for antibiotic therapy)

Emergency intraosseous access and arterial cannulation for monitoring are described below, but other vascular procedures such as arterial access for diagnostic and therapeutic purposes, endovascular surgery, and extracorporeal membrane oxygenation are beyond the scope of this chapter.

PREOPERATIVE ASSESSMENT AND PLANNING

A specific history and clinical examination are imperative, especially if there has been previous central venous cannulation. These should include the proposed site of insertion and exit site of the device and a search for stigmata that suggest potential venous access difficulty, such as multiple scars and dilated anterior chest wall veins. If such stigmata exist, preoperative vascular imaging may be indicated.

For the purposes of central venous access, the central veins may be defined as the SVC, the right atrium, and the suprahepatic inferior vena cava (IVC). The choice of vessels in order of preference should be determined in advance, taking into account factors such as previous cannulation, the presence of a tracheostomy, and the child's skin condition. In general, the right internal jugular vein (RIJV) is the best site for central venous access. Other potential veins, in the usual order of preference, are the left internal jugular, external jugular, axillary, common femoral, and subclavian veins. If ultrasound-guided puncture is an option, this order may change, and other veins such as the brachiocephalics may be considered. Less conventional methods, such as recanalization of occluded veins, the use of small collateral veins, and transhepatic or translumbar access to the IVC, are occasionally required. These should be undertaken by personnel skilled in image-guided techniques and are outside the remit of this text.

Consent should be obtained by the operating surgeon or a colleague with a clear understanding of the procedures and devices involved. Baseline blood tests should be performed and hemoglobin, platelet levels, and coagulation parameters optimized prior to surgery. With proper planning, most of these procedures can be carried out on elective lists, reducing the risk to patients by a reduction of out-of-hours operating.

Anesthesia

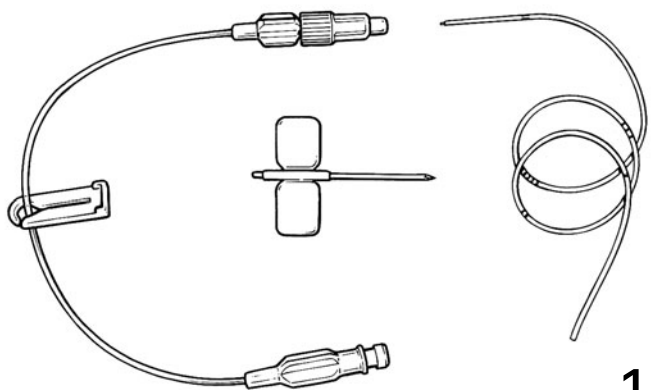
Anesthetic experience should be appropriate to the complexity of the child undergoing surgery. Postoperative management may require intensive care facilities, which should be arranged in advance.

Anesthetic strategies for vascular access procedures in children differ widely amongst centers. Intravenous sedation may be used with good results. Our institutional preference is for general anesthesia, but selected children will tolerate CVC insertion under local anesthetic with or without an inhalational agent.

In the majority of cases, the airway is best secured by muscle paralysis and an endotracheal tube. A laryngeal mask may be appropriate for some patients. Standard monitoring includes oxygen saturation, end-tidal carbon dioxide, temperature, blood pressure, and electrocardiography. The airway must be monitored closely, especially each time the position of the head is altered. Injection of local anesthetic at the site of all incisions is recommended. This should be done before incisions are made, as it allows for a lighter general anesthetic and removes the risk of damaging the device following implantation.

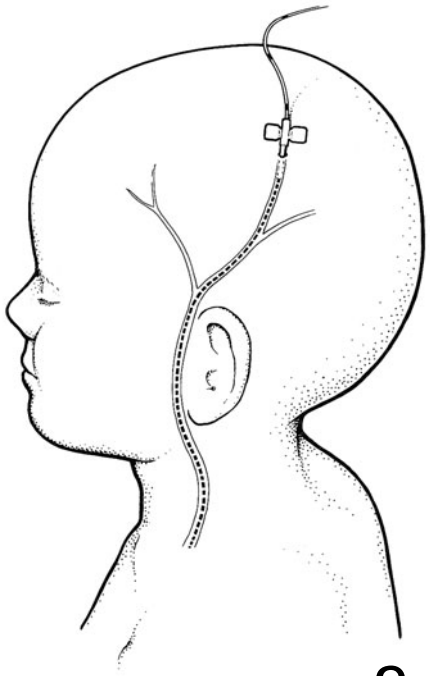
PERCUTANEOUS CENTRAL VENOUS CATHETER INSERTION IN THE INFANT VIA A PERIPHERAL VEIN

1 In the small infant, percutaneous insertion of a fine Silastic CVC can be performed awake or with sedation. These are small, single-lumen catheters (outer diameter 0.6 mm, maximum crystalloid flow rate about 6 mL/min), and are particularly useful in neonates where the need for venous access is anticipated for a period of weeks. Repeated sampling shortens the lifespan of these catheters.



1

2a,b Popular insertion sites include the median cubital vein at the elbow, the long saphenous vein anterior to the medial malleolus, and the superficial temporal vein. For supracardiac veins, the distance from the chosen insertion site to the right nipple is measured as a guide to the length of the catheter that should be inserted. Good nursing assistance is essential. After antiseptic preparation of the skin, venepuncture is performed with a 19-gauge butterfly needle and the fine Silastic feeding line is inserted into the needle and threaded up the vein using fine, non-toothed forceps. The progress of the catheter may be interrupted at venous junctions, but manipulating the limb will usually allow it to be advanced further. Once inserted to the desired distance, the butterfly needle is withdrawn and the line is connected to the infusion system via an inner blunt metal cannula and flushed with heparinized saline. Gentle suction on a 2-mL syringe should allow blood to be aspirated if the catheter tip lies in a large vein. The external catheter is firmly secured with a small piece of gauze covered by a transparent adhesive dressing. The position of the catheter tip should be confirmed radiologically. It is usually visible on a plain film with magnification, but it is easier to see if 0.5 mL of intravenous contrast material is injected prior to the chest radiograph. Line breakage is usually the result of perforation of the Silastic tube by the blunt metal cannula. This is easily repaired by removal of the damaged part of the line, retrieval of the metal cannula, and then re-insertion into the new distal end of the catheter. This is then re-connected to the infusion system.



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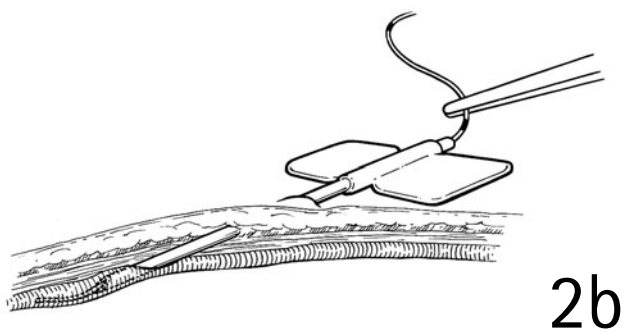
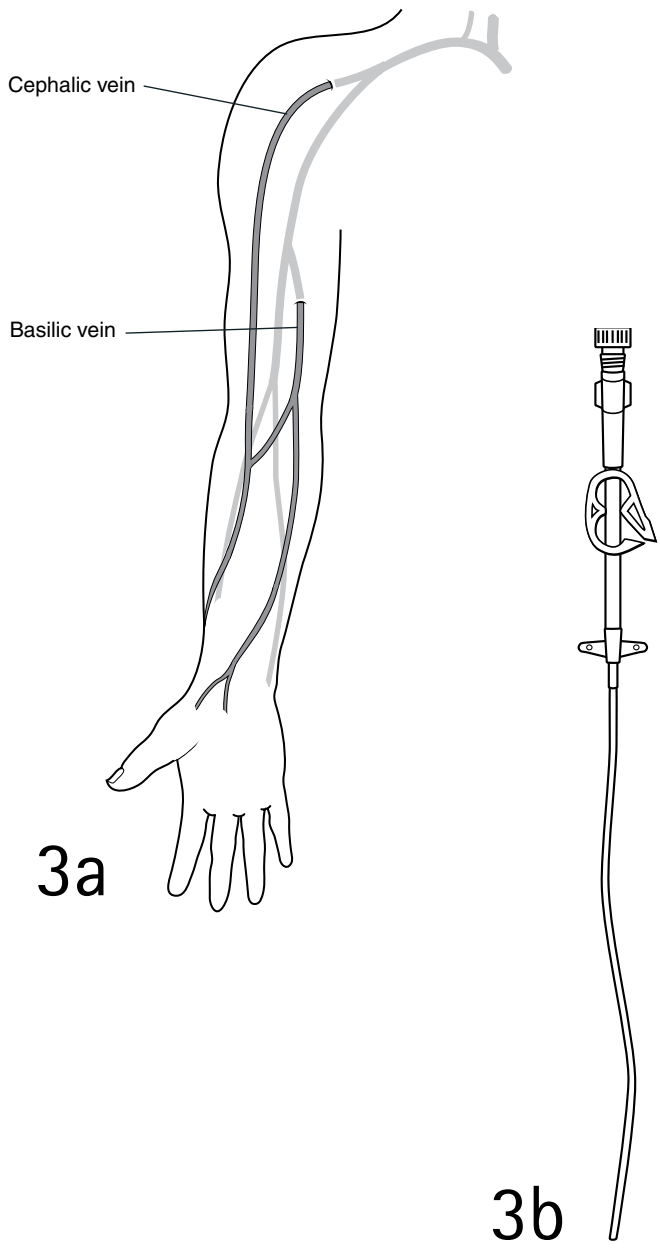


2b

Peripherally inserted central venous catheters

3a,b These are single-lumen or dual-lumen catheters typically made of silicone elastomer or polyurethane. They range in size from 2 Fr (0.67 mm) to 6 Fr (2.0 mm). Some PICCs have valves at the tip or hub, to prevent reflux of blood into the catheter. Although they are intended for short- to medium-term use (from 1 week to a few months), they are occasionally left in for much longer. Upper limb veins are generally used for PICCs, and many are inserted without image guidance. Imaging with ultrasound may, however, be required to gain access to a suitable vein, and the use of fluoroscopy significantly improves the chance of achieving a suitable final catheter tip position. The procedure is easier to perform in older children with larger cephalic and basilic veins. In certain circumstances, when a short-duration catheter is required, a device of this type may be used as a tunneled CVC. The advantage over cuffed catheters is that it is easy to remove without sedation or anesthesia. For peripheral access, the arm is stabilized on a support board. Standard, sterile skin preparation is carried out and a local anesthetic is injected after selection of an appropriate entry site to the basilic or cephalic vein, usually above the elbow. The vein is punctured with a 21-gauge needle or 22-gauge cannula. Aspiration of blood confirms successful puncture. A 0.018-in (0.46-mm) guidewire is then advanced into the vein. If resistance is felt at this point, the needle or cannula should be repositioned (it is usually too far in). An appropriately sized PICC is selected. The needle or cannula is removed, and a peel-away sheath of diameter just sufficient to accept the PICC is advanced over the guidewire. The guidewire should be fixed relative to the patient, and pressure applied over the puncture site as this is done. The guidewire and the dilator of the peel-away sheath are then removed, and the PICC is inserted into the sheath. It is usually easier to advance the PICC to a central position if its stiffening wire is left in. In certain places, especially near the termination of the cephalic vein in the deltopectoral groove, it may be easier to fix the stiffening wire and advance the PICC over it, unsupported. When the tip lies in the low SVC or upper right atrium, the peel-away sheath is split and removed, aspiration of blood is confirmed, and the catheter is flushed with normal saline. It is then sutured to the skin, and a transparent occlusive dressing applied. When no suitable superficial vein is available, ultrasound-guided puncture of a brachial vein (vena comitans of the brachial artery) is usually successful.

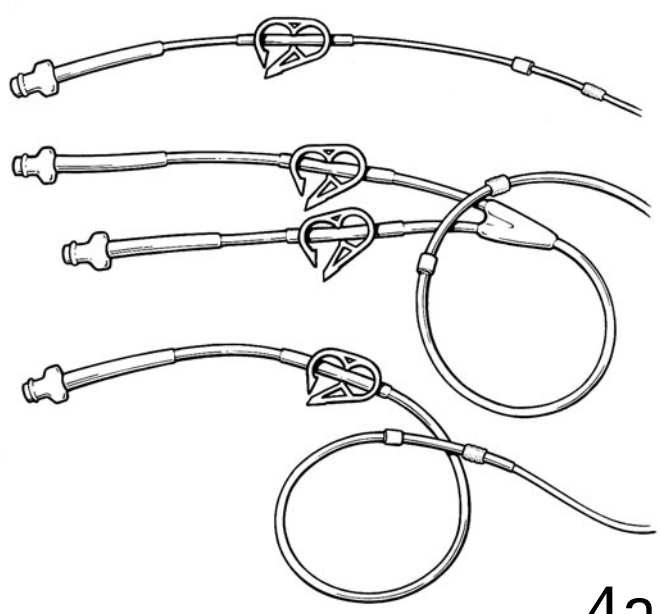


3a

3b

Non-tunneled (percutaneous) central venous catheter insertion

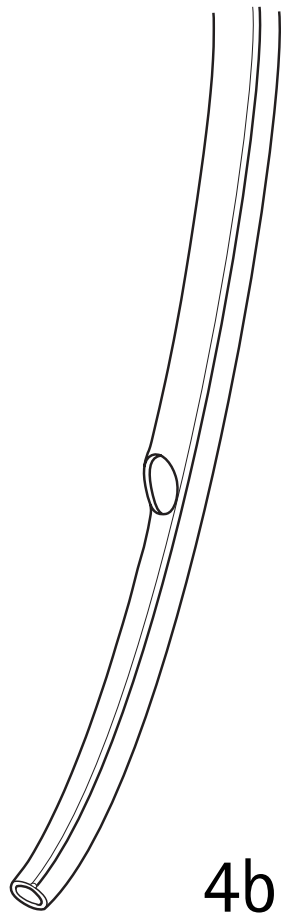
These short catheters, designed for insertion directly over a guidewire, include temporary hemodialysis catheters. Non-tunneled catheters are usually intended for short-term use (less than 10 days), because of the high rate of infection when they are left in for longer than this. Most of these lines are inserted using a percutaneous technique based on anatomical landmarks. Ultrasound guidance is recommended and increasingly being used for these and other CVC insertions.



4a

Tunneled central venous catheters

4a,b These are intended for medium- to long-term venous access. There are two main types. *Hickman* and similar catheters have single or multiple lumens, a size range of 2.7–12 Fr, and a tissue in-growth cuff made of Dacron, which lies in the subcutaneous tunnel. The cuff is intended to reduce the risk of ascending infection and inadvertent removal. *Hemodialysis* catheters are similar, but have two lumens with offset openings at the tip, to prevent re-circulation of blood during hemodialysis. The techniques for the insertion of these catheters are described below.

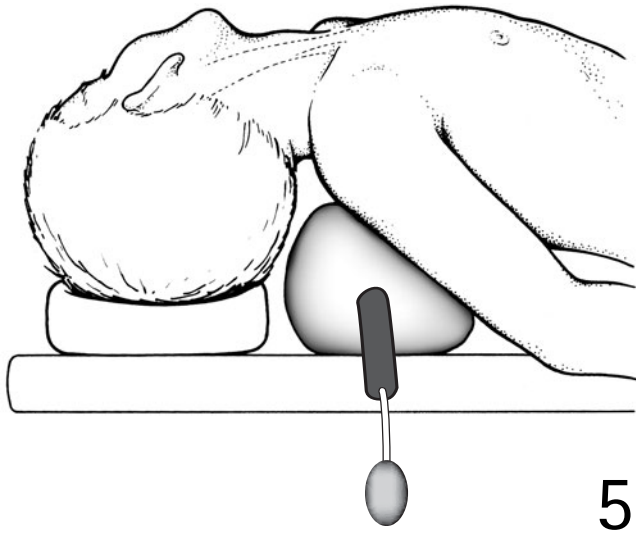


4b

OPEN INSERTION OF TUNNELED CENTRAL VENOUS CATHETER

INTERNAL JUGULAR VEIN (IJV)

5 The patient is positioned supine. A radiolucent pad (preferably inflatable) is placed under the scapulae and the head is turned slightly to the contralateral side. Alternatively, a soft roll may be utilized. Loupe magnification is an asset, particularly in infants and small children. After thorough skin preparation of the operative field, including the planned catheter exit site, the ipsilateral nipple, and the neck, the drapes are secured with adhesive plastic or sutures.



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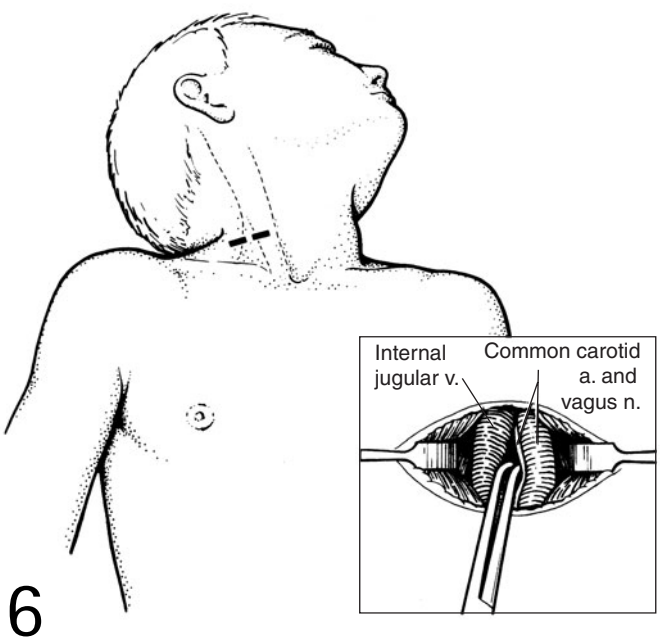
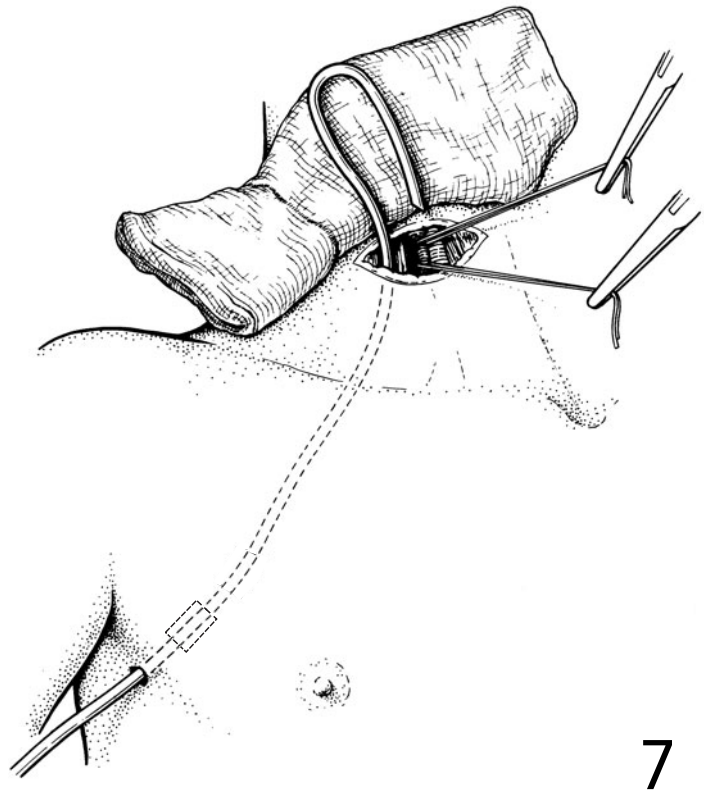


6

6 A short skin crease incision is made 1–2 cm above the clavicle, overlying the diverging clavicular and sternal heads of the sternomastoid. The incision is deepened through the platysma and the cervical fascia. The two heads of the sternomastoid are separated by blunt dissection. Small retractors are inserted to facilitate exposure of the IJV. Picking up and incising the fascia investing the IJV makes subsequent dissection easier. Using a Mixter, blunt right-angled forceps, a plane is developed on either side of the vein and the instrument is passed around the vein once a clear window has been established. A thin Silastic vessel loop is used to sling the vein. With this as a gentle retractor, a second vessel loop can be passed and a 1–2 cm length of vessel exposed between the slings. The retractors can be removed and the slings relaxed while the catheter tunnel is created.

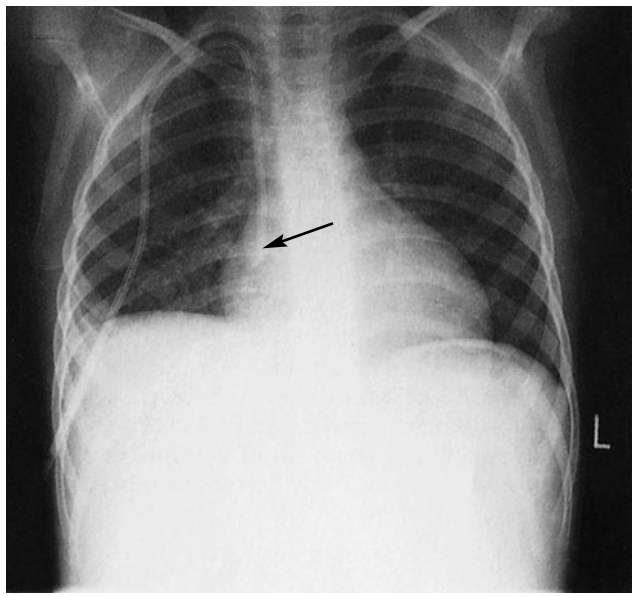
7 Various exit sites over the chest wall are possible, but the developing breast and strap lines should be avoided in girls. A small skin incision is made and a track of sufficient size to accommodate the catheter is developed using a hemostat. Either a blunt tunneling rod to which the catheter is attached or a hollow tunneler is used to pass the catheter to the cervical wound. Soaking the Dacron cuff with aqueous antiseptic may be useful. The Dacron cuff is then positioned about 2 cm from the exit site in order to facilitate future line removal. The distal catheter is cut to length with the tip beveled. On the right, the distance to the mid right atrium is estimated by a point just above the right nipple line, and on the left just below the right nipple line.



7

The prepared section of vein is elevated between the two slings by the assistant. Using fine, non-toothed forceps and microvascular scissors, a short venotomy is made. This commences with the decisive use of the scissors to make a very small initial incision. This is followed by insertion of the closed scissor tips into the venotomy to widen it until it is equal to the external diameter of the catheter. The beveled catheter tip is now introduced into the vein with the aid of two non-toothed forceps. The assistant gently relaxes the lower sling to allow distal passage of the catheter and then tightens it to prevent back bleeding. The catheter should pass freely and, once inserted, free bidirectional flow should be confirmed. If required, the venotomy is closed around the catheter with 6/0 polypropylene vascular sutures, care being taken to avoid narrowing the vein. Hemostasis is checked with the slings relaxed. A purse-string suture around the catheter should be avoided, as this may result in shearing of the vein upon removal.

8 The position of the catheter is checked using fluoroscopy. Suboptimal positions resulting from anatomic variation or wrong catheter length should be corrected (e.g. left-sided SVC). The sternomastoid muscle is loosely approximated with an absorbable suture and the cervical wound is closed in two layers using a fine absorbable subcuticular suture. A topical biological skin glue and an adhesive dressing are an acceptable alternative. The catheter exit site incision is approximated around the catheter with a 4/0 monofilament suture and firmly tied without compression of the catheter to aid fixation. The catheter is flushed with heparinized saline (10 unit/mL heparin). The exit site is dressed with gauze and a transparent adhesive plastic dressing. Adhesive tape is also used to secure the external part of the catheter, which should be looped to ensure that any pull on it does not result in direct pressure to the line at the exit site. Unless soiled, the dressing is changed after 1 week.



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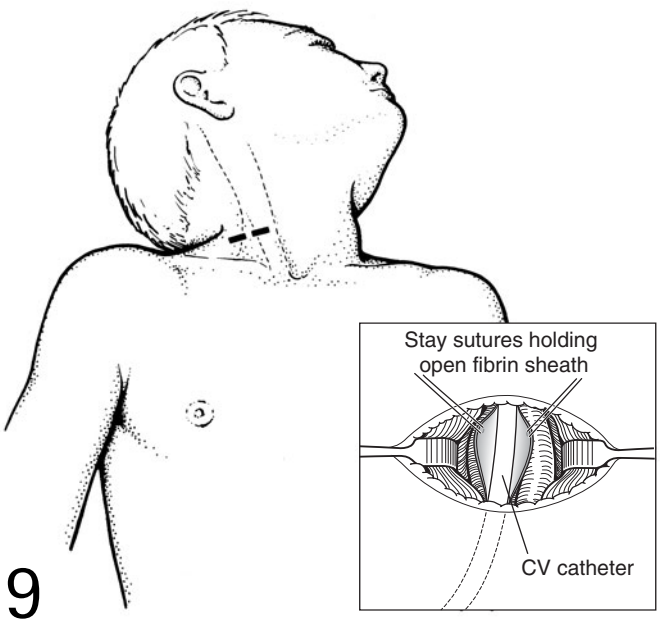


9

If the external portion of a Hickman line is damaged, it may be suitable for repair. This is carried out using a kit supplied by the manufacturer. The line is divided at the line of fracture with or without excising the perforation. A cannula is interposed between the two ends and secured with adhesive. The line is not disturbed for 24 hours.

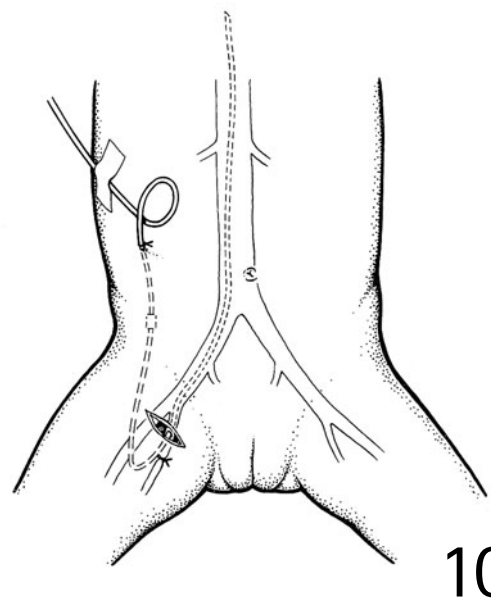
VEIN RE-USE AND LINE REPAIR

9 When indicated for mechanical failure in a tunneled central line that has been in place for weeks or more, the open exchange of a CVC provides very satisfactory results. The technique involves palpation of the old CVC just below the level of insertion into the right internal jugular vein (RIJV), dissection onto the catheter, careful incision of the pericatheter fibrin sheath and control with fine stay sutures. At this stage, a new CVC (same size or smaller than the old) is tunneled via a fresh site beside the old catheter in the neck and cut to length. With an assistant keeping the sheath open with gentle traction on the stay sutures, the surgeon removes the old line and immediately replaces it with the new. The final position is confirmed on screening and adjusted if required. The technique is simple and reliable. Its main drawback is that it cannot be used to increase the size of a catheter.

ALTERNATIVE SITES

Usually, the external jugular vein (EJV) is easily visible and requires minimal dissection. It may provide a very useful alternative to the IJV; however, its use is limited by the caliber of the vein and the occasional difficulty of negotiating the junction with the subclavian vein.

10 The long saphenous vein is approached by a short transverse incision 1 cm below the groin skin crease, medial to the femoral artery. The vein is controlled with fine vessel loops. The catheter tip is positioned in the right atrium and the exit site on the lateral abdominal wall.



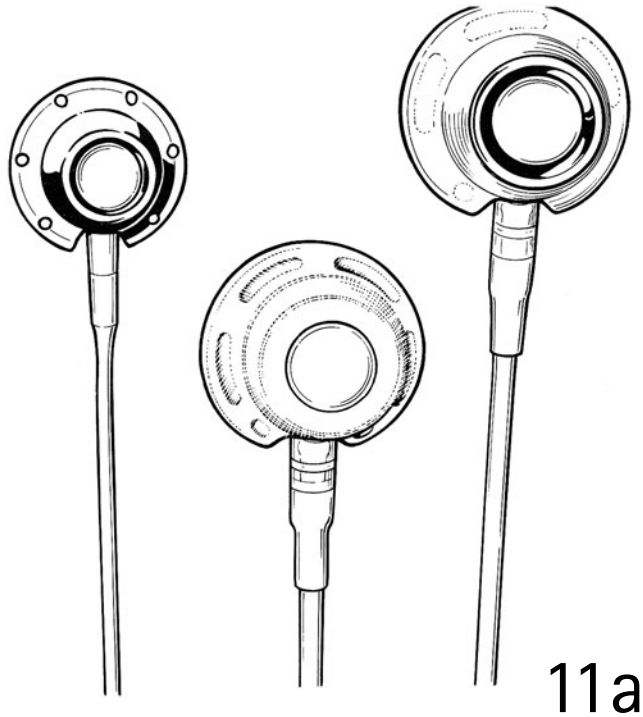
10

The femoral vein is especially useful for short-term access, but may be used as a route for long-term catheters. The axillary vein is easily approached by an axillary incision and dissection, but size can be restricting. The common facial vein is a large anterior tributary of the IJV in infants that may be entered midway between the angle of the mandible and the clavicular head.

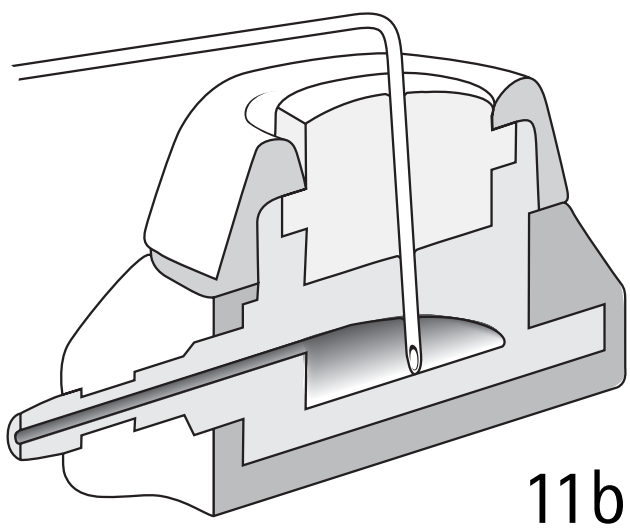
The cephalic vein is accessed in the deltopectoral groove but tends to be small in young children. In some children who have required repeated, chronic venous access complicated by central vein thrombosis, other routes that can be used include the azygos, epigastric, iliac and renal veins, the IVC, and the right atrium. In these difficult cases, preoperative and intraoperative image guidance is advisable. Percutaneous techniques may avoid major dissection.

Ports

11a,b Totally implantable vascular access devices, or 'ports', have a catheter connected to a small reservoir, which is implanted subcutaneously. A thick silicone membrane forming the roof of the port can be repeatedly injected percutaneously using a 22-gauge side-fenestrated, non-coring (Huber) needle. The ports are made from stainless steel, titanium, or hard plastic and are available in different shapes and sizes. Those with a preconnected catheter are easier to insert. One variety is designed to be implanted in the arm, with central venous access through a peripherally inserted catheter. Because they have no external catheter, port devices have certain advantages over tunneled CVCs. In particular, they are less likely to require removal for infection, they cannot be accidentally removed, and they allow for activities such as swimming. They are therefore preferable for most children who require only intermittent (e.g., weekly) access, including those with hematological diseases and cystic fibrosis. They are less appropriate for children who cannot tolerate regular needle access, or who require continuous access, for example those who will need intensive chemotherapy or parenteral nutrition.

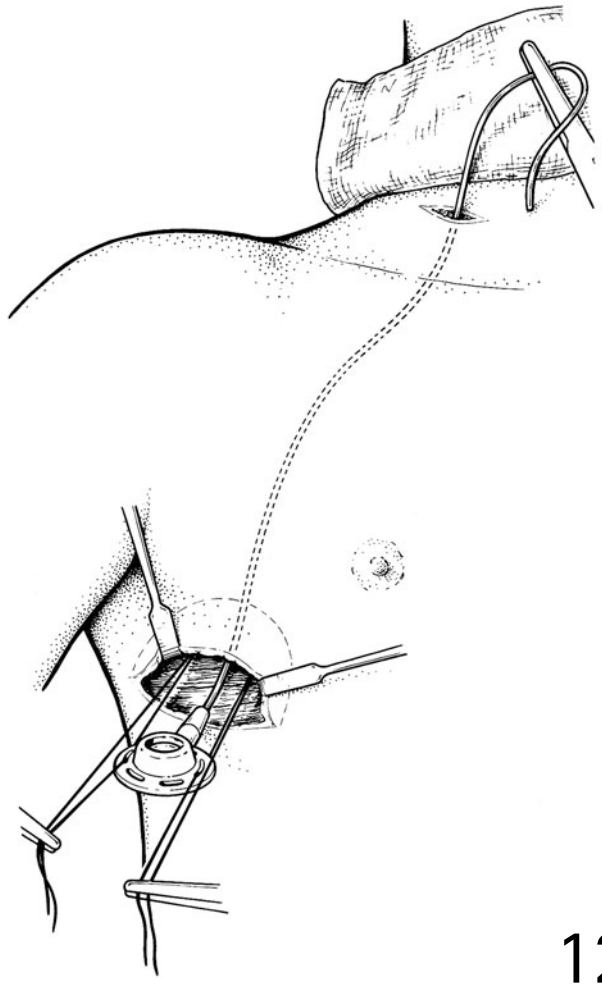


11a



11b

12 At the predetermined reservoir site, which must be easily accessible and rest on a firm surface such as the anterolateral chest wall, the skin incision is deepened with diathermy. Hemostasis must be meticulous. A subcutaneous pocket is developed beneath the superficial fascia in such a way as to avoid placing the port directly under the skin incision. Placement of the port above rather than below the incision may reduce the impact of wound-related problems on port and line function. Before implanting the port, it is helpful to place non-absorbable sutures through the muscular fascia and the circumference of the port; when tied, these provide three-point fixation of the device. The catheter must be tunneled from the reservoir pocket to the site of venous access, such as the IJV in the neck. The port is flushed with saline, ensuring there are no kinks in the catheter. The distal catheter is cut to length (see above) and inserted by a cut-down or percutaneous technique. After confirming the catheter tip position by fluoroscopy, the port is flushed with heparinized saline and the skin incision is closed in two layers with an absorbable subcuticular skin suture.



12

Each injection must access the port vertically through the center of the silicone diaphragm such that the needle touches the base plate. As the needle is withdrawn, the port should be held in place and positive injection pressure applied to prevent reflux of blood into the catheter. A careful aseptic injection technique must always be used and the system flushed periodically.

Removal

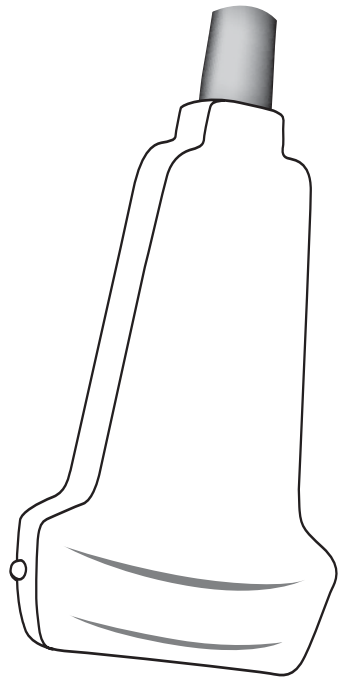
Most cuffed external catheters and all ports require a short general anesthetic for removal. With the former, the Dacron cuff can usually be dissected free with a hemostat and fine scissors via the exit site incision, which is then closed with absorbable sutures or skin tapes.

Ultrasound-guided insertion of central venous catheters

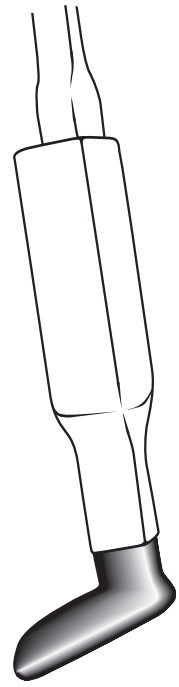
The use of real-time ultrasound guidance makes central venous access easy, quick, and safe in all but the most difficult cases. Potential advantages over surgical placement of central lines include a very high success rate at the first site attempted, a good cosmetic result because of the short puncture site incision, a short procedure time, and virtually no need for preoperative imaging in children who have had multiple central veins accessed in the past. It is recommended over 'blind' insertion and may eventually supersede the open technique.

EQUIPMENT

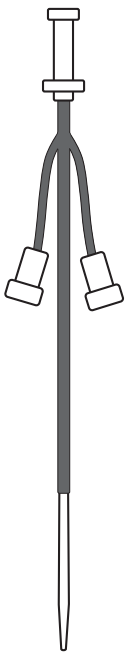
13a,b A high-frequency (≥ 7 MHz) linear-array transducer is appropriate for the majority of punctures. In very small patients, a small 'hockey-stick' transducer is a useful tool. The transducer is placed in a sterile probe cover for operative use.



13a



13b



14a

14b

14c

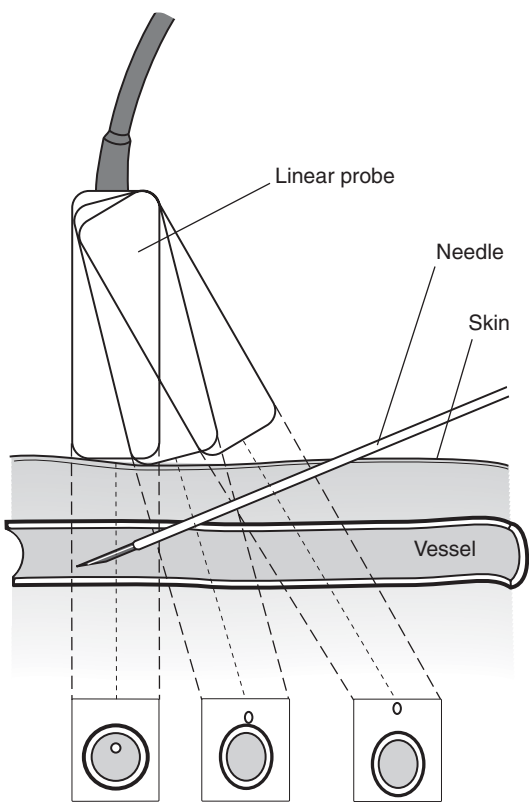
14a–c In children weighing less than 10 kg, a 21-gauge one-part needle or 22-gauge cannula should be used for central venous puncture. These devices accept a 0.018-inch (0.46-mm) guidewire. The best guidewires have a stiff shaft and a short floppy tip. The stiff part of the wire is necessary to allow insertion of a peel-away sheath. In larger children, a bigger (19-gauge or 18-gauge) needle permits the use of a thicker guidewire, making insertion easier. The percutaneous insertion of tunneled catheters requires the use of a peel-away sheath. These are available in a wide range of sizes. Although the stated size of a sheath is equal to the diameter of a catheter that can be introduced through it, this should be checked in advance, as it is sometimes necessary to use a sheath 0.5 Fr larger than the catheter.

INSERTION TECHNIQUE

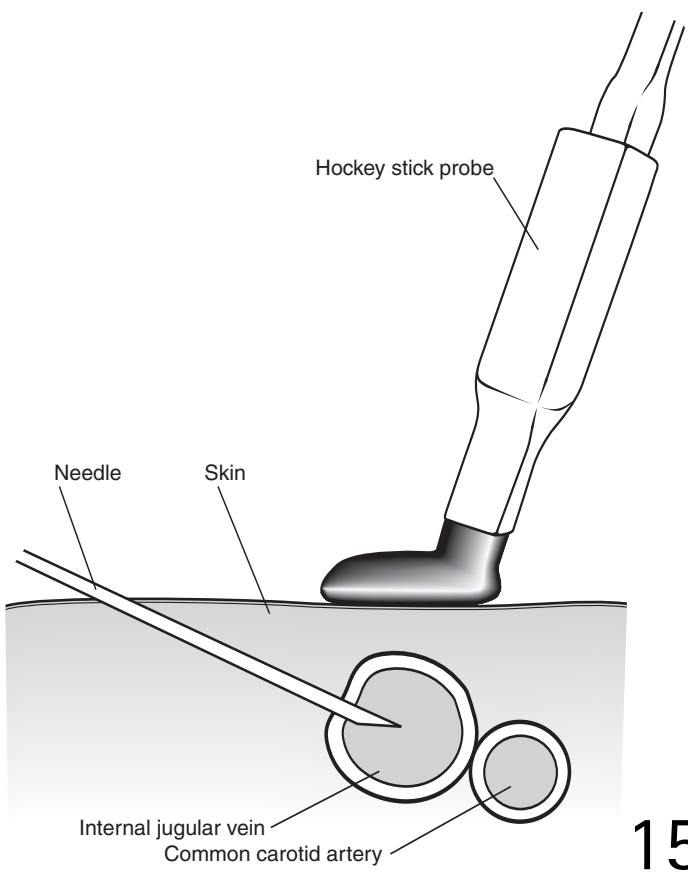
15a–c Ultrasound is used to assess the available vessels and select one for access. Here again, the site of first choice is the RIJV. Skin preparation and draping are as described for open insertion. A trajectory for puncture is established using a 23-gauge needle attached to a syringe. The needle stops short of the vein wall. A small amount of local anesthetic is infiltrated as the needle is withdrawn and a skin crease stab incision made at the point of skin entry of the trajectory needle. This is widened slightly with a hemostat. We recommend tunneling the line (and, when indicated, the creation of a subcutaneous port pocket) at this stage. The line is brought through the stab incision in the neck and wrapped in antiseptic-soaked gauze. The needle for vein puncture is attached to a syringe and inserted at the medial end of the cervical incision, taking care not to damage the catheter.

There are two methods of puncturing the vein. In the first, the needle is advanced along the line of the vein, puncturing its anterior surface, with the probe held perpendicular to the needle and vein. The second method may be better for tunneled catheters and is our preferred technique: the anterolateral surface of the vein is punctured with the probe held in the same plane as the needle. In either case, it is important to puncture the vein with a sharp, stabbing motion to ensure that the tip of the needle enters the lumen of the vein with the bevel pointing downwards. The needle should be seen to move freely in the lumen, without a 'tent' of intima over the tip, and venous blood should aspirate freely. It is easy to create a subintimal hematoma if care is not taken at this stage. Inadvertent puncture of the opposite wall of the vein is usually not a problem, as the needle can be withdrawn into the lumen with ultrasound guidance. Once in the center of the vein, the angle of entry of the needle may be altered slightly so that it is pointing centrally. The guidewire is advanced into the vein, and its position confirmed by fluoroscopy. If it is easy to pass the guidewire through the right atrium and down the IVC, this should be done, as it makes insertion of the peel-away sheath easier and safer.

Following removal of the needle, the peel-away sheath is advanced over the guidewire under fluoroscopic control. *It is crucial to fix the guidewire (relative to the patient) at this stage.* If this is not done, the dilator of the peel-away sheath may cause serious damage to the SVC or heart. Catheter length can be determined as previously described by measurement against the nipple. Alternatively, fluoroscopy can be performed with the catheter on the anterior chest wall, projected over the peel-away sheath. If the catheter is cut at the T7 level, its tip will lie in the upper right atrium. The guidewire and the dilator of the peel-away sheath are removed. Mechanical ventilation with positive end-expiratory pressure effectively pre-

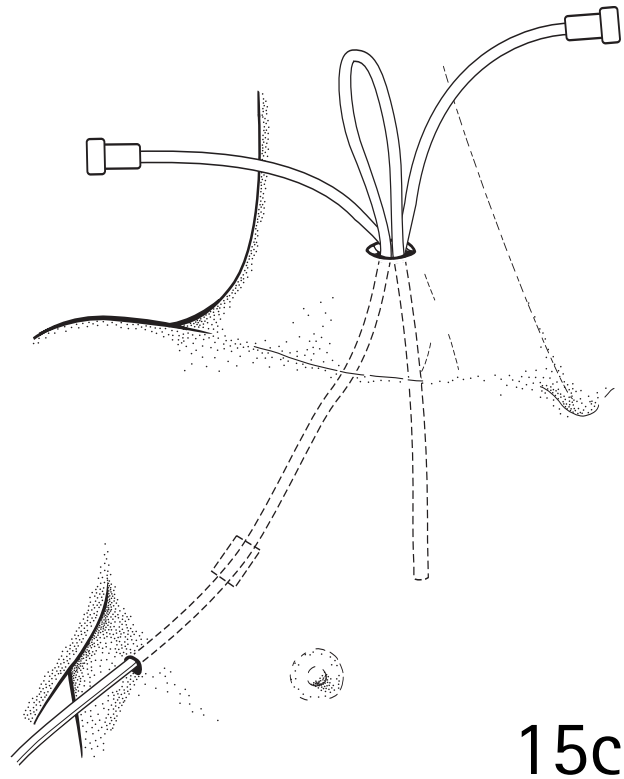


15a



15b

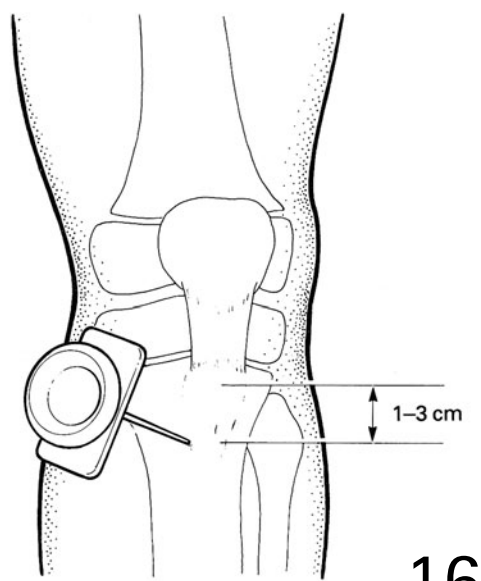
vents air entering the peel-away sheath at this stage, but great care should be taken to avoid air embolism if the patient is breathing spontaneously. The catheter is advanced through the sheath, which is then partially split. The position of the catheter tip is confirmed with fluoroscopy, and adjusted if necessary. The sheath is completely split and removed. The catheter is flushed with heparin (10 unit/mL) and sutured to the skin at the exit site. The cervical puncture can be closed with a subcuticular suture or tissue glue and adhesive tape. With minor modifications, this technique can be used at other sites or with other systems such as venous port devices.



15c

EMERGENCY INTRAOSSEOUS VENOUS ACCESS

16 This route provides immediate vascular access during life-threatening emergencies in young children when rapid venous return cannot be achieved (cardiac arrest, shock, burns, and trauma). Contraindications include fracture or infection near the insertion site. After antiseptic preparation, the skin is punctured with a scalpel blade. The intraosseous needle is inserted into the medullary cavity of the proximal tibia through the middle of its flat anteromedial surface, 1–3 cm below the medial tuberosity (depending on the size of the child). The infusion needles (14–18 gauge) have an inner occluding stylet designed to facilitate bone penetration and should be inserted almost perpendicularly to the bone but angled slightly away from the growth plate. Upon entering the marrow cavity, the resistance suddenly decreases. The needle should then stand firmly in the bone. It should be possible to aspirate bone marrow or flush the needle easily without extravasation. The needle flange is adjusted to skin level and taped in position. The patient's leg should be restrained with a support behind the knee. Crystalloids, blood products, and drugs can be infused, but blood sampling may occlude the needle.

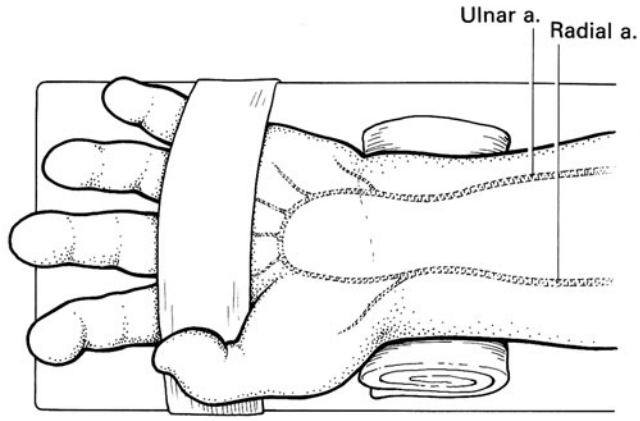


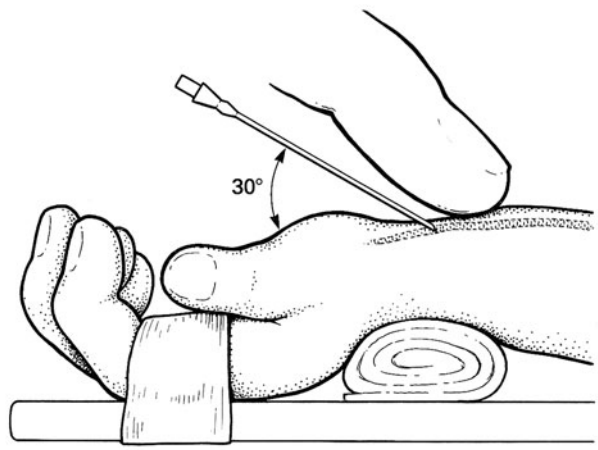
16

The infusion needle should be removed once suitable conventional access has been obtained if potential complications (extravasation, compartment syndrome, fractures, osteomyelitis, fat embolism) are to be avoided. The distal femur and distal tibia are alternative sites.

ARTERIAL PUNCTURE AND CANNULATION

Intra-arterial access is used to provide continuous monitoring of systemic arterial blood pressure and to enable repeated arterial sampling for blood gas measurements.





17 The radial artery is the preferred site for both percutaneous and cutdown cannulation. The presence of adequate collateral flow must first be checked by the Allen test: both arteries are occluded at the wrist and after releasing the ulnar artery alone, the hand should flush pink (most hands have an ulnar dominant palmar arch). A small roll is placed under the supinated, extended wrist and the palm is taped to a padded surface, keeping the fingers exposed in order to assess the distal circulation. The skin is cleaned with antiseptic and a small quantity of local anesthetic is injected subcutaneously over the radial artery just proximal to the transverse crease at the wrist. The skin is punctured with a No. 11 scalpel blade, and a 22-gauge or 24-gauge Teflon cannula with a needle stylet is selected according to the size of the child. The artery position is verified by palpation.

Two techniques are used. In the first, the needle and Teflon cannula are advanced at about 30° to the skin until a flashback of blood is seen. In the transfixion method, the artery is transfixed by the needle and cannula. The needle is then removed and the cannula is gently withdrawn until arterial blood appears, when it is advanced up the artery lumen.

In the cutdown technique, a small transverse incision over the artery allows the vessel to be punctured and cannulated under direct vision, with the option of proximal and distal vessel control. The catheter hub is sutured in place and the skin is sutured around the cannula.

Arterial cannulas require continuous perfusion with 0.5–1.0 mL/h heparinized saline. Because of the risk of serious complications (ischemia, embolism, hemorrhage, sepsis), arterial access requires an even higher level of vigilance and should be used for the shortest possible time. The cannula should be removed if signs of digital ischemia develop. Alternative sites for arterial access include pedal, umbilical, femoral, brachial, and axillary arteries, but complications are more frequent than with radial artery cannulas.

POSTOPERATIVE CARE

The postoperative care of CVCs is crucial to longevity and optimal function. Tunneled CVCs should be looped and then secured with occlusive, see-through dressing. This ensures that distal traction is not directly transmitted to the line at the exit site. The development of dedicated teams and carefully defined protocols that cover aspects of management such as frequency of flushing, line-handling techniques for sampling, and infusions has also helped to reduce complications. If well cared for, the majority of catheters last the duration of the planned course of treatment. However, complications which may shorten this duration are well recognized and are summarized in Table 3.2.

Table 3.2 *Complications of central venous access*

Timeline	Complication	Prevention	Management
Immediate	Pneumothorax	Avoid blind procedures	Chest drain
	Hemorrhage	Meticulous technique	Digital pressure, vein repair
	Air embolism	Positive pressure ventilation	Patient positioning, resuscitation
	Arterial puncture	Avoid blind procedures	Digital pressure
	Cardiac arrhythmia	Fix guidewire	Remove irritation, carotid massage, adenosine
	Malposition	Intraoperative fluoroscopy	Line revision
Early	Accidental removal	Secure exit site suture, loop catheter	Line replacement
	Catheter-related sepsis	Meticulous aseptic technique for insertion and subsequent line access	Antibiotic therapy, catheter removal
	Chest wall hematoma	Check and normalize coagulation parameters preoperatively	Pressure dressing
	Cardiac tamponade	Site stiff percutaneous catheters in low superior vena cava	Pericardiocentesis, pericardiotomy
Delayed	Catheter-related thrombosis	Site catheter tip in upper right atrium, flush regularly	Conservative management, anticoagulation, thrombolysis
	Extravasation		
	Port–catheter separation	Use of preconnected catheter	Urgent exchange
	Catheter migration	Site catheter tip in upper right atrium	Catheter revision
	Catheter fracture	Gentle handling and careful removal	Fluoroscopic catheter retrieval of embolized fragments

FURTHER READING

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SECTION II

Head and Neck

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4

Ranula

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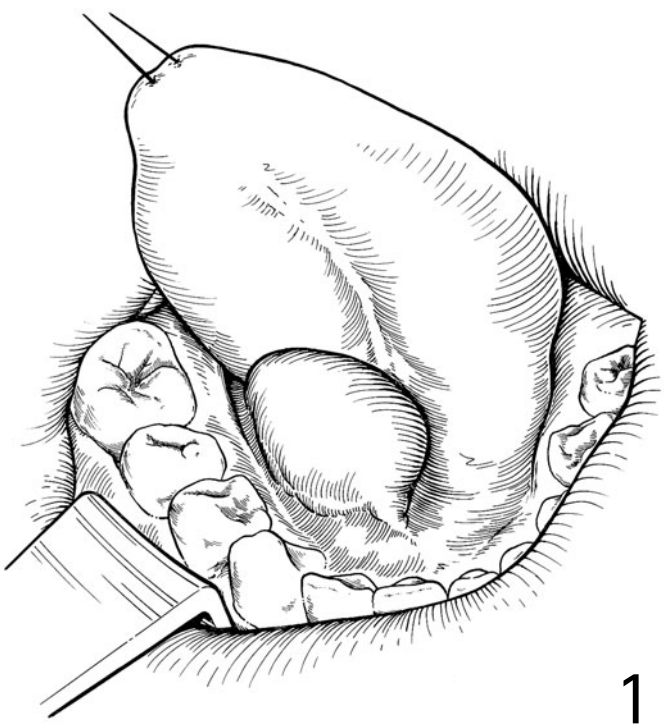
HISTORY

Ranulas are histologically benign lesions that present in the floor of the mouth. They arise from the sublingual salivary glands and have also been known as mucoceles, mucous extravasation cysts, and simple mucous retention cysts. Ranulas may be 'simple' or 'plunging', the latter being much more difficult to manage. Until the 1960s, little was written in medical literature about this entity. Early statements suggested that ranulas were found mainly in African tribes or dogs. The lack of knowledge of the pathophysiology led to varied recommendations for treatment (radiation, grommet insertion, marsupialization, and total excision), which resulted in a high incidence of recurrence and high morbidity. Since the sublingual gland has been recognized as the site of origin, treatment has been much more successful.

PRINCIPLES AND JUSTIFICATION

The sublingual gland has been identified as the site of origin of ranulas because of its high protein fluid content and the observation that excision of the sublingual glands markedly reduces recurrence.

1 The simple ranula (a simple cyst) probably represents obstruction of the duct and has an epithelial lining. These ranulas are confined to the sublingual space. However, the more common variety (the 'plunging' ranula) probably represents mucous extravasation and is thus a pseudocyst without an epithelial lining but with a lining of connective tissue or granulation tissue. The 'plunging' ranula therefore dissects deeply into the soft tissue and fascial planes of the neck.



1

Trauma to the sublingual gland may be an important etiologic factor in ranulas that develop later in life. Crysdale reported that persistent ranulas occurred in 5 percent of patients undergoing submandibular duct relocation surgery. Ligation of the sublingual gland duct in some laboratory animals has also led to lesions that are identical to naturally occurring ranulas.

The modalities for treatment have varied because of the initial misunderstanding about the cause of ranulas. The methods of therapy have included simple incision, marsupialization or fistulization with or without packing of the intraoral wound, destruction of the 'lining', grommet insertion, radiation therapy, excision of the sublingual gland with or without drainage or excision of the pseudocyst, and total surgical excision (via an intraoral or cervical approach). In the rare cases of well-encapsulated simple cysts of less than 1 cm diameter with an epithelial lining and no history of trauma, intraoral excision of the cyst will often be successful.

In recent years, the use of OK-432 (Picibanil) has been recommended by some authors. OK-432 is a streptococcal preparation that was originally developed as an immunotherapy agent to treat malignancies. It has been very effective in the reduction of ascites and pleural effusions and has more recently been used by Ogita and others to treat cystic hygromas. For cystic lesions, Woo et al. would prepare OK-432 by diluting 0.1 mg of OK-432 in 10 mL of normal saline with contrast. Under fluoroscopic guidance, the surgeons then puncture the mass and aspirate as much of the contents as possible. Then they inject the same amount of OK-432 solution as was aspirated. There may be side effects, such as pyrexia, which can be controlled with antipyretics. If necessary, a second injection can be given 3 weeks later.

There is a wide variety of opinions regarding whether to excise or marsupialize a ranula as well as how to approach excision (if this is chosen), and finally whether to remove or leave the ipsilateral sublingual gland. Guerrissi and Taborda suggest that an endoscopic intraoral approach for excision of the submandibular gland can be safely performed (thus eliminating the cervical incision) in 'plunging' ranula derived from the submandibular gland.

Takagi et al. have described an innovative approach in four patients with 'plunging' ranulas. Their technique involves making a 1 cm incision into the swelling in the floor of the mouth and draining the fluid. They then place a rubber Penrose drain into the incision (beyond the mylohyoid muscle into the submandibular space) and suture the drain into place. Rolled gauze and tape are then used as a pressure dressing in the submandibular area. The drain and pressure dressing are continued for 3 weeks (the patients receive antibiotics for 3 days). This technique has yielded no recurrences and does not entail external cutaneous incisions.

Baurmash cautions against the 'blanket' removal of the sublingual gland as a routine treatment for every ranula. He points out that there are ranulas that arise from the ducts of Rivini, as retention phenomena involving the orifice of Wharton's duct, and as a result of extravasation of saliva from a perforated submandibular duct. These ranulas are superficial and small to medium in size, so he recommends marsupialization as the treatment of choice.

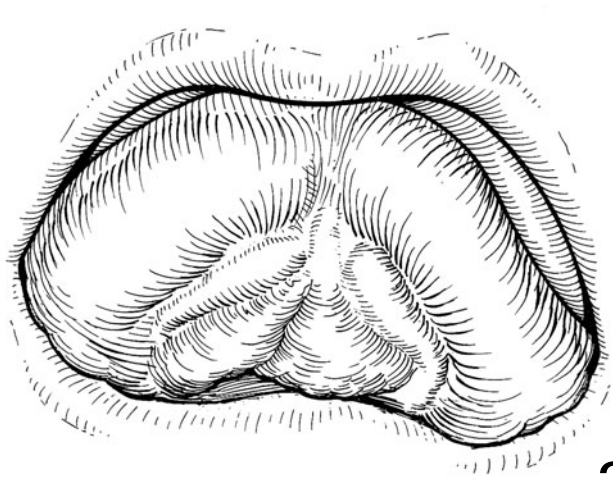
Pandit and Park recommend initial observation when a ranula occurs, as they have seen spontaneous resolution over a 4–6-month period. However, if the ranula does not resolve, they recommend a transoral approach for excision of the pseudocyst as well as the ipsilateral sublingual gland, along with relocation of the submandibular duct if necessary to obtain optimal exposure. It should be noted that Haberal et al. found that regardless of whether one chose an intraoral or external cervical incision, rupture of the ranula does not increase the risk of recurrence. They also found that there was not a significant difference in recurrence based upon marsupialization versus excision, so their recommendation in cases of oral ranula is that marsupialization with packing should be chosen as a conservative initial approach, with total excision of the sublingual gland reserved for recurrences or 'plunging' lesions.

This author has developed the following paradigm for the treatment of ranulas. If the lesion is a pseudocyst without an epithelial lining, if the ranula is recurrent, if the cyst is greater than 1 cm in diameter, and if there is a history of antecedent trauma, excision (usually intraoral) with removal of the ipsilateral sublingual gland is preferred. In extensive 'plunging' ranulas extending inferiorly beyond the mylohyoid muscle, a cervical neck incision may be used to give good access to both the sublingual and the submandibular glands. However, whether an intraoral or cervical approach is used, extensive dissection of the cyst in an attempt to remove all of the cyst lining is unnecessary when the sublingual gland is removed.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of ranulas (especially 'plunging' types) must include cystic hygroma, thyroglossal duct cyst, second branchial cleft cyst, enteric cyst, dermoid or epidermoid cyst, and inflammatory lesions. Malignant neoplasms such as squamous cell carcinoma are very unlikely in children.

2 Cystic hygromas arise from aberrant development of fetal lymphatic tissue in the neck and may extend into the floor of the mouth, tongue, or submandibular space. These lesions are usually easily compressible, transilluminate well, and are multilocular. They grow progressively, especially during upper respiratory infections, and may present with dangerous airway compromise. The differentiation between a cystic hygroma and ranula is difficult when the lesion is located in the superior anterior neck with extension into the sublingual or submandibular spaces.



2

Thyroglossal duct cyst tracts usually have well-defined borders, may elevate when the tongue is protruded, are close to the midline (20 percent are lateral), may enlarge during upper respiratory infections, are usually at the level of the hyoid bone, and are associated with the strap muscles.

Second branchial cleft cysts are usually located along the anterior border of the sternocleidomastoid muscle at the level of the angle of the mandible and may enlarge with upper respiratory infections.

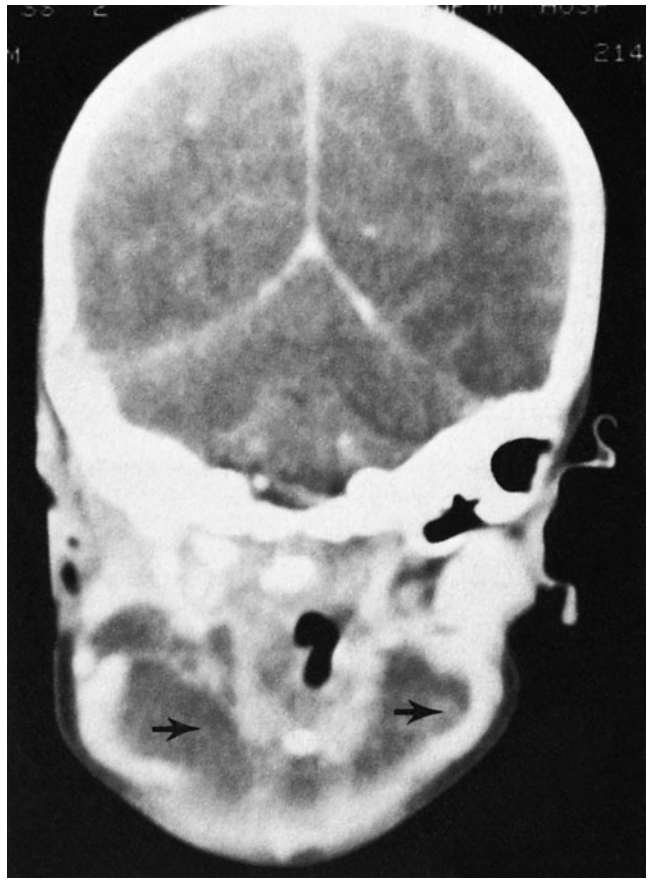
Epidermoid inclusion cysts are usually superficial, adherent to the skin, may be in the sublingual space, and may be difficult to differentiate from a ranula radiographically.

Lipomas grow slowly, are not fluctuant, and do not grow in response to inflammation.

Dermoid cysts may be similar to epidermoid cysts but have different signals radiographically on magnetic resonance imaging (MRI).

Benign cervical lymph nodes are rarely found in the floor of the mouth.

Inflammatory lesions secondary to sialadenitis or oral inflammatory processes will usually involve several spaces in the neck. Computed tomographic (CT) findings are usually somewhat different from those of a ranula.



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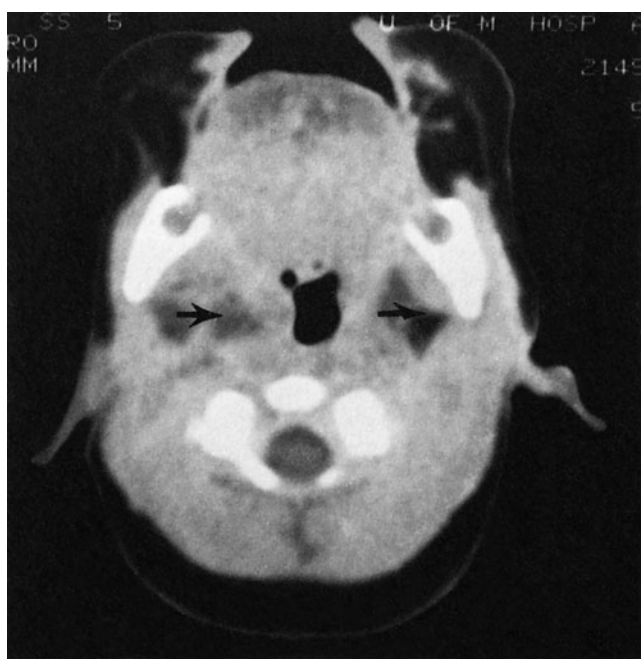


3b

3a,b Antenatal diagnosis may be with fetal ultrasound. If a ranula is diagnosed antenatally, some authors have suggested that obtaining fetal karyotype (by amniocentesis) and chorionic villous sampling is warranted. Garcia et al. have recommended that ultrasonography should be the initial imaging study used for the examination of pediatric salivary gland lesions, as most are benign and are well visualized with this modality. They feel that ultrasonography may differentiate intraglandular and extraglandular lesions, but admit that additional studies such as color Doppler (for vascular lesions), CT scans, or MRI may be needed. Unruptured ranulas may appear as simple cystic lesions, while those where there is ductal obstruction and rupture will appear as the extravasation type.

Computed tomographic scans are extremely useful in differentiating ranulas from the lesion previously described. A 'plunging' ranula has a water content on CT scan, while dermoid and epidermoid cysts have high protein and fat contents, lipomas have a low attenuation, and the thyroglossal and branchial cleft cysts have different locations in the neck. An anterior cervical cystic hygroma will generally contain septae, while a 'plunging' ranula is usually a single cavity. The demonstration of a unilocular, cystic mass in the sublingual space will usually be either a ranula or an epidermoid cyst. Since all 'plunging' ranulas arise from the sublingual glands, the lesions must involve or adjoin the sublingual space in every case. If no sublingual space extension or abutment occurs, the diagnosis of 'plunging' ranula is very unlikely.

Magnetic resonance imaging may be useful in verifying the cystic nature of the lesion, in providing information about the thickness and vascularization of the cyst wall, and for determining the relationship between the cyst and the surrounding tissues so that surgical planning may be more precise. Kurabayashi et al. found that, in 20 patients with ranulas, the lesions were well-defined, homogeneous masses yielding a low signal on T1-weighted images and a very high signal on T2-weighted images. 'Plunging' ranulas were found to extend into the sublingual space anteriorly (producing a so-called 'tail sign') and into the parapharyngeal space superiorly. Other types of cystic masses in the Kurabayashi study had one or more MRI findings that were different from those of ranulas and could be easily differentiated.

PREOPERATIVE

Anesthesia

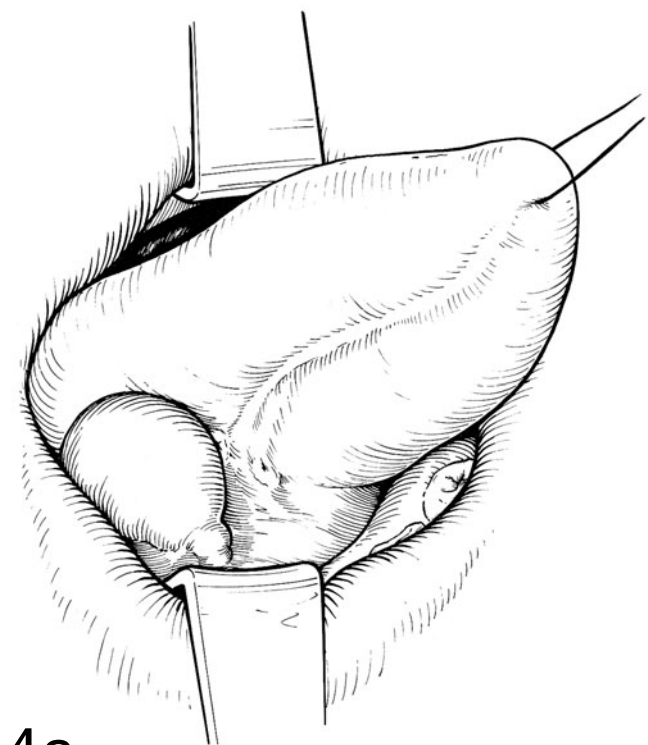
These patients must have general anesthesia with either oral or nasal endotracheal intubation. The author prefers nasotracheal intubation, which gives complete access to the oral cavity and the floor of the mouth. The patient should not be given a long-acting muscle relaxant (again, the author prefers that *no* relaxants be administered), as it is desirable to be able to monitor tongue 'twitches' as an indication of the close proximity of the motor nerves to the tongue. The hypopharynx should be packed with a gauze pack.

OPERATION

4a–d After suitable general endotracheal anesthesia is performed, a mouth gag (usually a Jennings type) is utilized to hold the mouth open. An alternative in cases where the mouth does not accommodate the gag is to place a small dental bite block posteriorly, or an assistant can retract the mouth with a Weider retractor or a small Army–Navy retractor. The tongue can be retracted by placing a heavy silk suture through the tongue tip.

Both Wharton's (submandibular) duct orifices are identified and the ipsilateral orifice is usually cannulated with a lacrimal probe to lessen the chance of injury. Three or four stay sutures (3/0 silk) can be placed around the planned mucosal incision. The mucosa is incised with coagulation unipolar or bipolar cautery. Avoidance of injury to the lingual and hypoglossal nerves is also paramount, and the surgeon can use one of two approaches. The first is to make an incision in the floor of the mouth directly over the gland with the duct cannulated with a rigid lacrimal probe. The second is to make an incision in the lingual gingival sulcus at the first molar on the ipsilateral side, extending across the midline as far as is necessary for adequate exposure. A full-thickness flap of mucoperiosteum is elevated from the lingual surface of the mandible to the floor of the mouth. This will allow the sublingual gland to bulge against the periosteum that is incised, causing the gland to herniate through the incision and make its excision much easier.

If the lesion is well encapsulated and appears to have a distinct wall, the intraoral excision without removal of the ipsilateral sublingual gland is warranted. If the lesion is not well encapsulated but has several loculations that are ruptured, the ipsilateral sublingual gland should be removed also via the intraoral approach. After removal of the cyst and sublingual gland, the intraoral incision is closed with interrupted 3/0 or 4/0 polyglactin (Vicryl) sutures.



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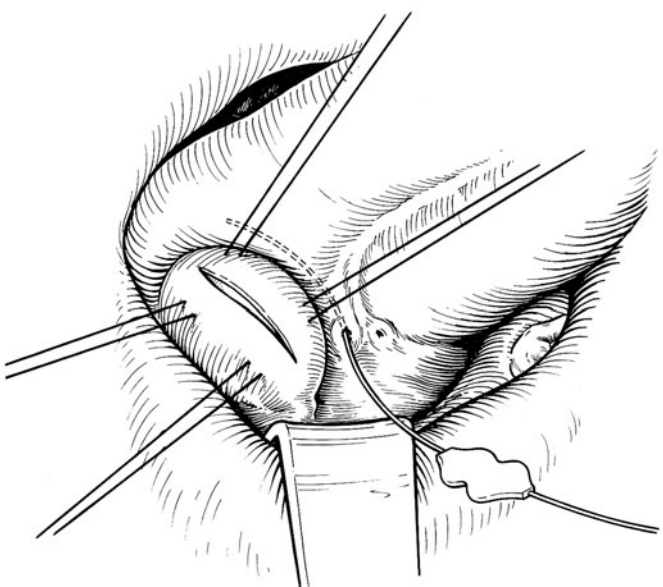

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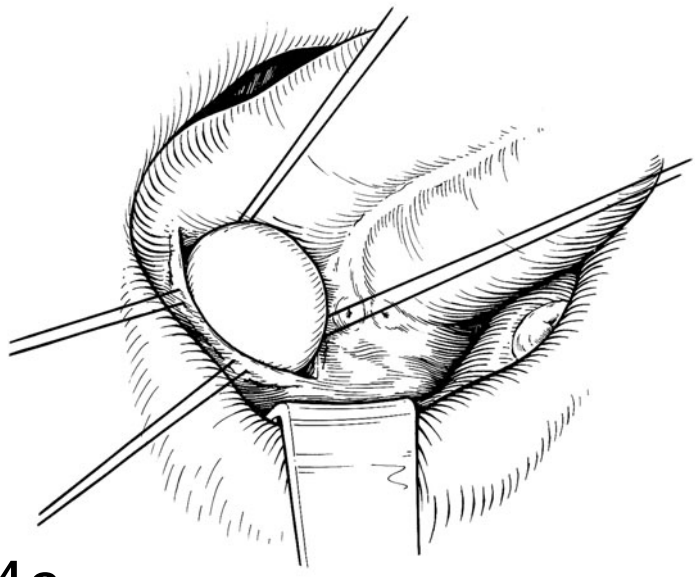

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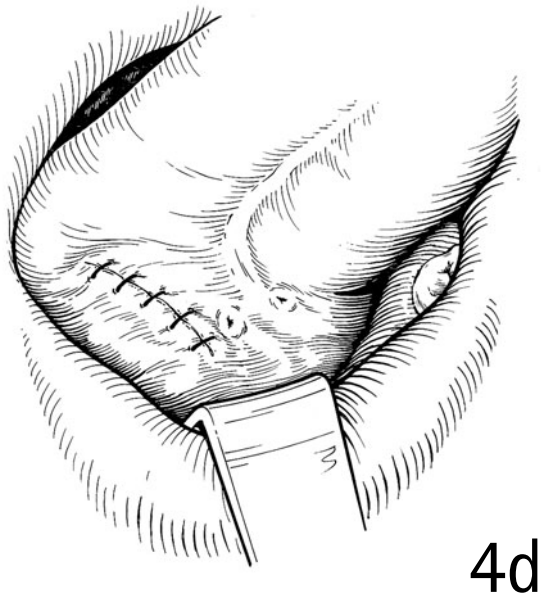

4d

If the ranula is 'plunging' and very large, a cervical incision may be considered. This approach allows the surgeon to have a wider access to the cyst and sublingual gland because the excision of the submandibular gland makes exposure of these structures easier and also makes identification of the lingual nerve more obvious. Again, even in the external approach, extensive dissection of the cervical cyst is not usually necessary. The neck incision is drained with a suction drain, such as a Jackson–Pratt drain system. The subcutaneous tissue is closed with a 3/0 polyglactin or chromic suture and the skin is closed with a running 4/0 subcuticular suture.

POSTOPERATIVE CARE

The infant should refrain from having a nipple or pacifier, as it may traumatize the incision site. Oral feedings are usually initiated 8–12 hours after surgery, using a red rubber catheter for gavage feedings for 24–48 h. If an external approach is employed, the Jackson–Pratt drain is hooked to wall suction for 24 hours and then placed to bulb (or grenade) suction for 24 hours before being removed.

OUTCOME

Both this author and others, as reported in the literature, have found the above approaches to be very successful.

FURTHER READING

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Garcia CJ, Flores PA, Arce JD, Chuaqui B, Schwartz DS. Ultrasonography in the study of salivary gland lesions in children. *Pediatric Radiology* 1998; **28**: 418–25.

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5

Thyroglossal cyst and fistula

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HISTORY

The thyroglossal duct was first described in 1723 by Vater, who called it the 'lingual duct'. It was later referred to as the canal of His following his descriptions in 1855 and 1891.

PRINCIPLES AND JUSTIFICATION

Development of thyroid gland and thyroglossal tract

The thyroid gland develops as a median thickening of the floor of the pharynx at the level of the second branchial arch (tuberculum impar), during the fourth week of gestation, and descends to its final position in the neck, leaving the thyroglossal duct extending caudally from the foramen cecum of the tongue to the pyramidal lobe of the gland, passing anterior, through, or posterior to the hyoid bone. The lateral lobes of the gland receive contributions from the fourth branchial clefts, which form the medullary C cells. Early in the fifth week of gestation, the attenuated duct loses its lumen and shortly afterwards breaks into fragments. Thyroid remnants may be found along the course of the thyroglossal duct.

Thyroglossal cysts

Thyroglossal cysts, the most common anterior cervical swelling in children, most frequently arise just inferior to the level of the hyoid bone. Occasionally the duct deviates anterosuperiorly once it has passed the hyoid bone, giving rise to a thyroglossal cyst in the submental triangle, where it may be mistaken for a dermoid cyst. Although dermoid cysts may occur below the hyoid bone, they are more common in the submental triangle and can be distinguished from thyroglossal cysts by their softer, 'putty-like' consistency. Very occasionally, aberrant thyroid glandular tissue is found along the course of the thyroglossal duct.

Thyroglossal fistulas

A thyroglossal fistula usually results from rupture or incision of an inflamed thyroglossal cyst. The fistulous opening is usually at the level of the original cyst, but may appear lower down in the neck.

PREOPERATIVE

Assessment and preparation

In thyroid hypoplasia, a small central area of aberrant ectopic thyroid tissue may be mistaken for a thyroglossal cyst, and it is recommended that the precise location of the thyroid gland is determined, using isotope scanning or ultrasound examination, before undertaking surgery, as removal of the aberrant tissue may result in permanent hypothyroidism. The incidence of such aberrant tissue is, however, low (about 1 percent of all thyroglossal abnormalities) and it is easily recognizable when the lesion is exposed.

Occult staphylococcal infection is common in these cysts, and perioperative antibiotic cover using a penicillinase-resistant agent such as flucloxacillin or fusidic acid is usually indicated.

Anesthesia

General anesthesia using an orotracheal or nasotracheal tube is recommended.

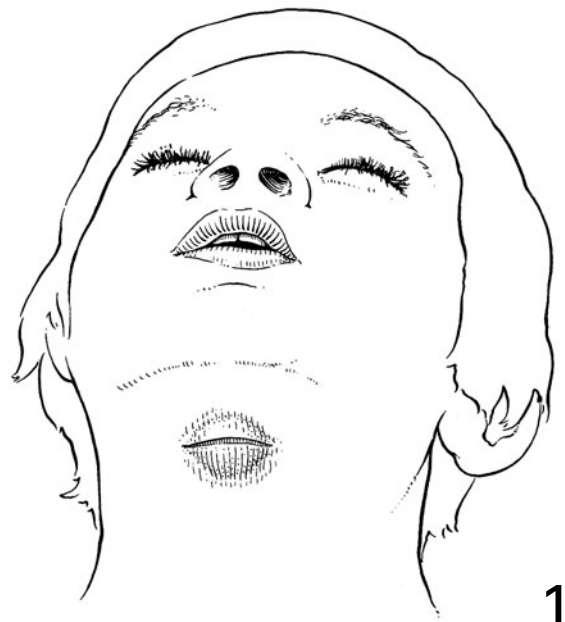
OPERATIONS

Excision of thyroglossal cyst

The aim of surgery is to remove the entire duct, including the central part of the body of the hyoid bone, to the level of the foramen cecum.. Because side branches may arise from the duct within the muscles of the tongue, the intraglossal part of the duct should be removed with a surrounding cuff of muscle approximately 0.5 cm in diameter. Complete excision is essential to prevent recurrence and eliminate the risk of malignant degeneration. All thyroglossal cysts, however small, should be excised to avoid the risk of infection, which makes subsequent surgery more difficult, morbidity and recurrence rates higher, and cosmetic results less satisfactory. The operation may be performed on a day-case basis provided meticulous hemostasis has been achieved.

POSITION OF PATIENT

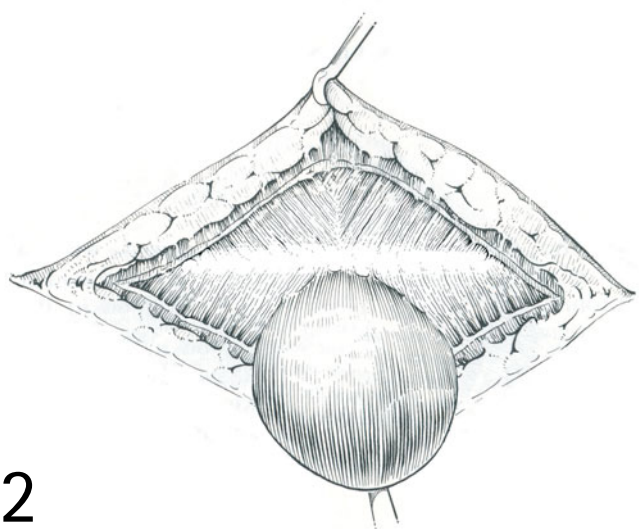
The patient is placed supine with the head extended and the shoulders elevated on a small sandbag.



1

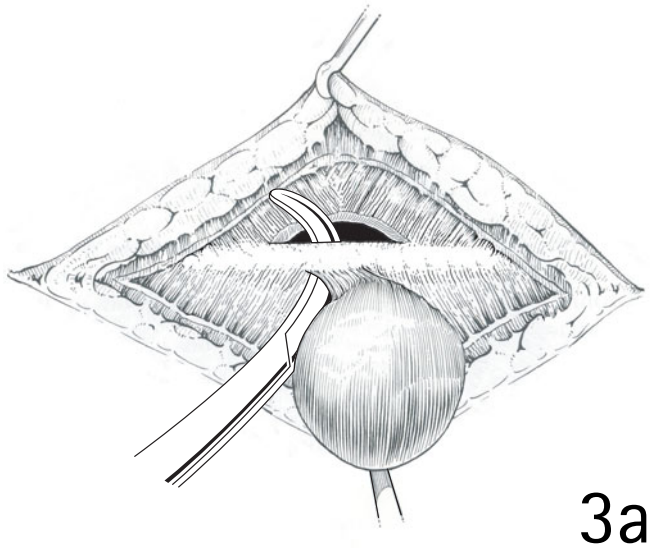
INCISION

1 A short (usually less than 3 cm) transverse incision is made in a skin crease over the main prominence of the cyst. Some authors recommend infiltration of the skin with epinephrine (adrenaline) to reduce bleeding.



2

2 The subcutaneous fat, platysma, and deep cervical fascia are incised in the line of the incision with a diathermy needle and the cyst is freed from its superficial attachments by a combination of sharp and blunt dissection. Meticulous hemostasis is essential so that the operative field is not obscured.



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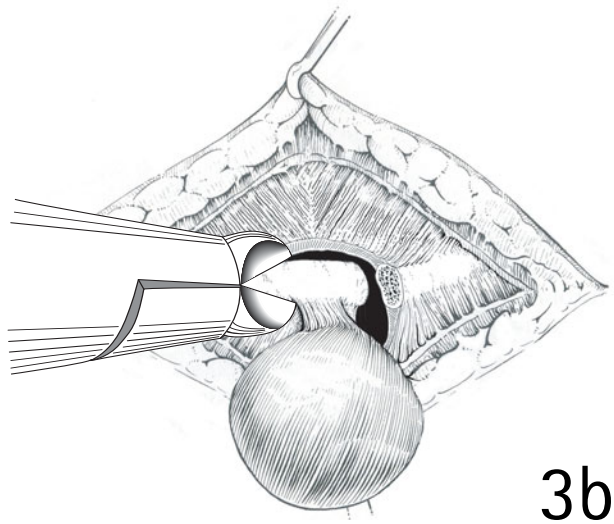
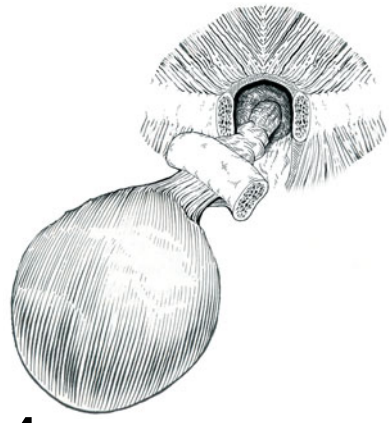


3b

DISSECTION

3 The thyroglossal tract is identified at its deep attachment to the cyst and followed between the sternohyoid muscles to the hyoid bone. The centrum of the hyoid bone is freed from the sternohyoid muscles below and the mylohyoid and geniohyoid muscles above with a diathermy needle. The thyrohyoid membrane is separated from the posterior aspect of the centrum using artery forceps, a closed pair of scissors, or a McDonald dissector. Small bone-cutting forceps or strong Mayo scissors are then used to divide the body of the hyoid 5 mm to either side of the midline. This maneuver is facilitated by grasping and steadying the bone with Kocher artery forceps.

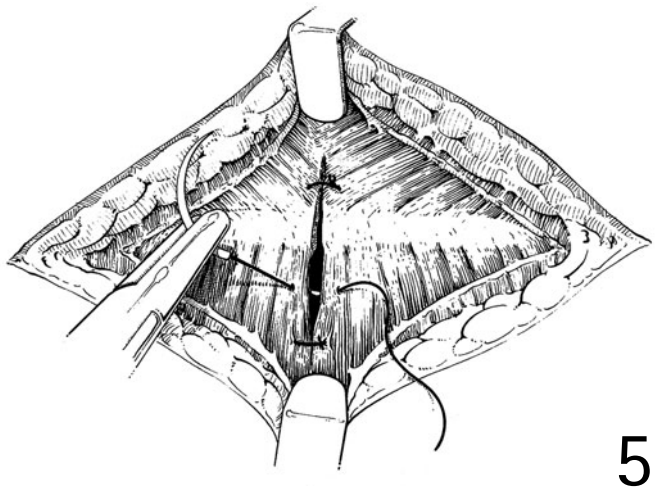


4

4 A cylinder of geniohyoid and genioglossus muscles 0.5 cm wide including the duct is excised to the foramen cecum; this is best performed using needle diathermy. It has been suggested that the dissection is made easier if the anesthetist uses a finger to depress the foramen cecum into the wound, but this is seldom of practical value and is a potential danger to the anesthetist. Meticulous hemostasis using diathermy will prevent postoperative respiratory obstruction due to hematoma formation.

WOUND CLOSURE

5 The muscles are approximated in the midline using sutures of 3/0 polyglycolic acid or chromic catgut to aid hemostasis. It is not necessary to reconstitute the hyoid bone because its cut ends tend to be approximated by the muscle sutures.



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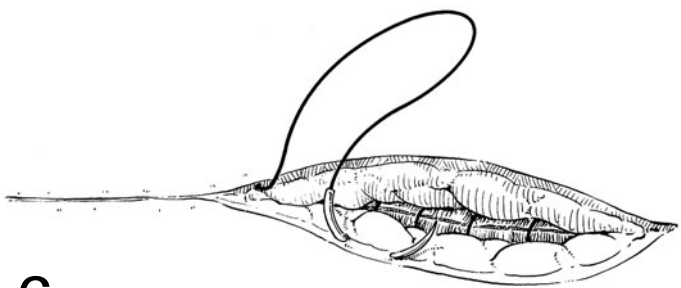


6

6 The fascia and platysma are closed with a continuous suture of 3/0 or 4/0 polyglycolic acid and the same suture is used in the subcutaneous layer to appose the skin edges. Alternatively, the skin edges may be approximated with self-adhesive wound tapes. The use of non-absorbable skin sutures or clips is not recommended, as their removal causes anxiety and discomfort. If adequate hemostasis has been secured, no drains or dressings are required.

Excision of thyroglossal fistula

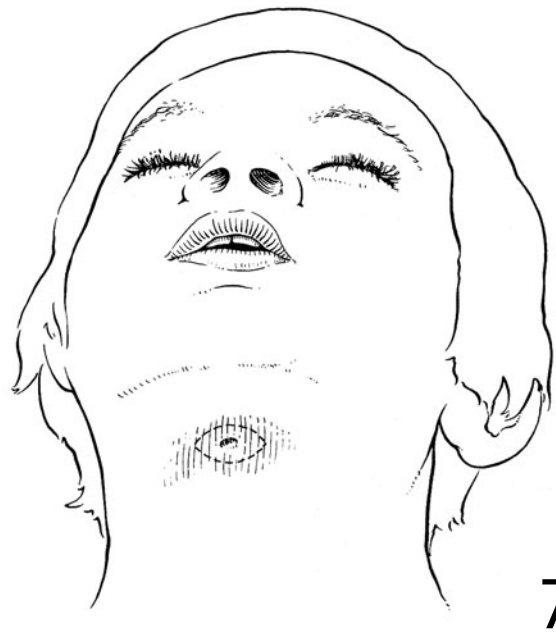
Treatment is similar to that for an uncomplicated thyroglossal cyst. All traces of the fistula must be excised to the foramen cecum.

POSITION OF PATIENT

The patient should be positioned as for excision of a thyroglossal cyst (see p. 54).

INCISION

7 A small elliptical incision is made around the orifice of the fistula (sinus) in the neck.



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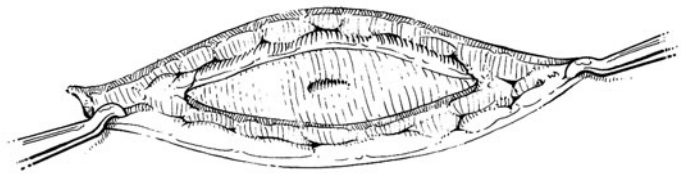

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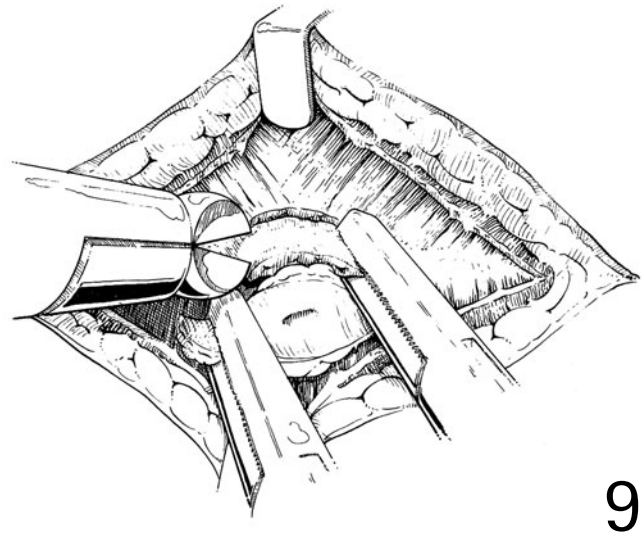
8 The ellipse of skin (including the sinus or fistulous tract) is traced through the subcutaneous tissues towards the hyoid bone.

RESECTION

9 The centrum of the hyoid bone is resected with the fistulous tract.



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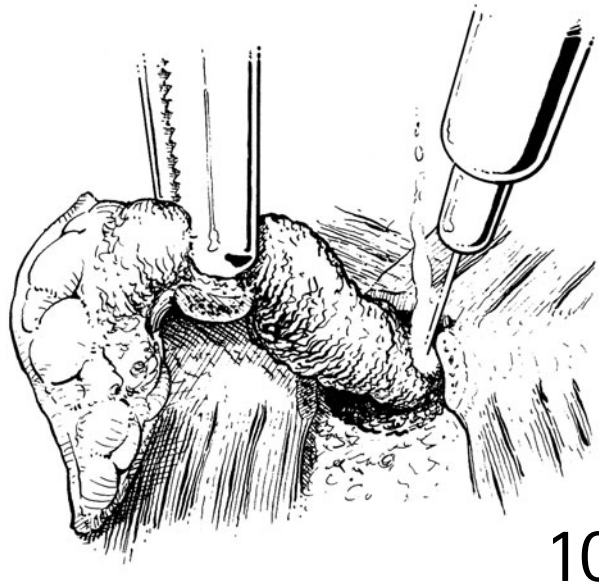
10 A core of geniohyoid and genioglossus muscles, including the tract, is excised up to the level of the foramen cecum.



10

WOUND CLOSURE

The wound is closed as for a thyroglossal cyst. It may be necessary to insert a drain if perfect hemostasis cannot be guaranteed or if there is florid inflammatory edema.

POSTOPERATIVE CARE

It is important to ensure that the airway does not become obstructed by reactionary hemorrhage. Infection is common, and spreading cellulitis may cause airway compression, so oral antibiotics should be continued for 2–3 days after the procedure. The child may be allowed home on the day after surgery.

OUTCOME

If the thyroglossal duct is excised with the cyst, recurrence is unlikely. Recurrence rates of 5–7 percent are reported. Cysts may recur, however, sometimes as much as 10 years later, in more than 20 percent of patients treated by local excision only. This rate is reduced to 5 percent by removing the central part of the hyoid bone with the cyst. The recurrence rate of a thyroglossal fistula is higher than that for the uncomplicated cyst.

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6

Branchial cysts, sinuses, and fistulas

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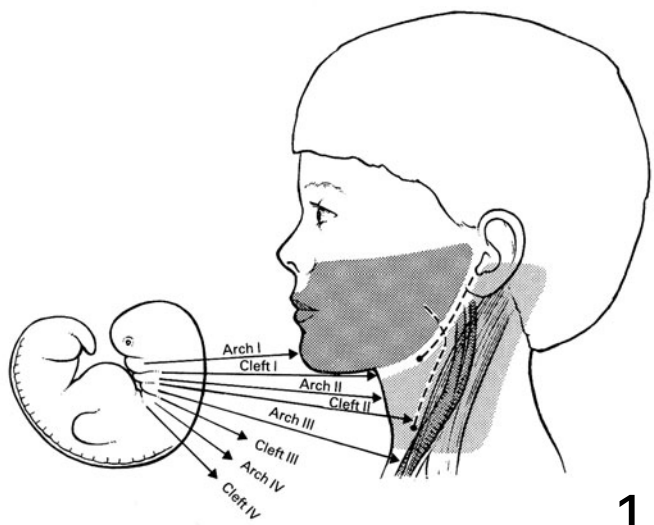
PRINCIPLES AND JUSTIFICATION

Cysts, sinuses, and fistulas of the neck derived from branchial cleft remnants are common in the pediatric age group. Sinuses and fistulas are encountered more commonly in infants and children, while branchial cysts occur more often in older children and young adults. Remnants of the first and second branchial apparatus are most common, with abnormalities of the second cleft outnumbering those of the first by 6:1. Abnormalities of the third and fourth branchial apparatus are rare, but recent case reports and reviews indicate that they may be more common than previously supposed.

A simple knowledge of head and neck embryology is helpful in understanding these abnormalities. The branchial arches appear by the 15th day of fetal life and present as bar-like ridges separated by grooves or clefts. Five paired ectodermal clefts and five endodermal pouches separate the six branchial arches. A closing membrane lies at the interface of the pouches and clefts. The four clinically significant arches and clefts are shown.

The pathogenesis of branchial cleft anomalies is controversial, and may occur as any combination of sinus, fistula, and cyst. Incomplete obliteration of the branchial apparatus, primarily the cleft, is accepted as the most likely etiology. Most branchial anomalies arise from the second branchial apparatus as the second branchial arch overgrows the second, third, and fourth branchial clefts, and finally fuses with the lateral branchial wall. As the arches coalesce during the growth of the embryo, part of the first branchial cleft remains open as the eustachian tube and auditory canal. The second branchial cleft normally closes completely; however, either branchial cleft may form a sinus tract or cyst as it coalesces.



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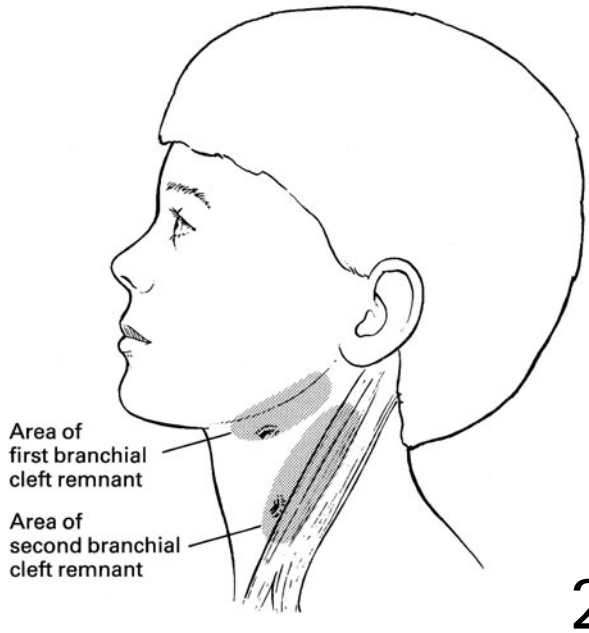
2 Remnants of the first branchial cleft occur along an imaginary line extending from the auditory canal behind and below the angle of the mandible to its midpoint. Second branchial cleft remnants are found anywhere along an imaginary line extending from the tonsillar fossa down to a point on the lower one-third of the anterior border of the sternocleidomastoid muscle.



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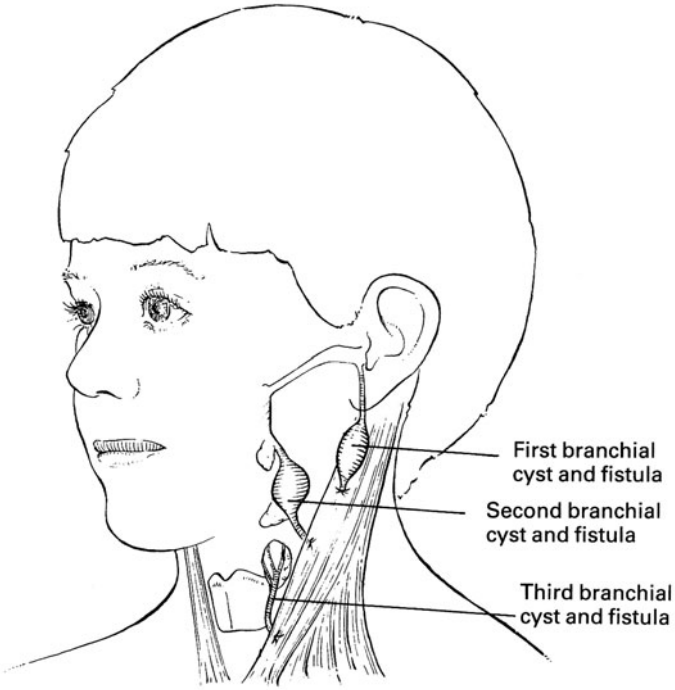
Although branchial apparatus anomalies may present at any age, most branchial sinuses present clinically soon after birth or before the age of 10 years.



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3 The more common second branchial cleft sinus presents as a pinpoint opening on the anterior border of the sternocleidomastoid muscle, one-quarter to one-third of its length cephalad from the sternal end. The defect is usually characterized by the appearance of small drops of clear fluid at the opening or by the occurrence of infection in the tract itself. The anomaly may be either unilateral or bilateral, and may be familial. Tracts that have an exterior opening occasionally become infected, although infection is a more common problem in sinuses and cysts in the older age group. Cysts of the first branchial cleft usually present as enlarging masses near the lower pole of the parotid gland and are more commonly seen in older children and young adults. Cysts of the second branchial cleft usually present in children and young adults as a mass at the mandibular angle along the anterior border of the sternocleidomastoid muscle, often associated with upper respiratory infection.

Case reports and recent reviews suggest that cysts or acute infections arising from a third or fourth branchial pouch sinus are rare but well-defined entities that offer diagnostic and therapeutic challenges not encountered with the anomalies of the first and second branchial remnants. These lesions present as an air-containing inflammatory lateral neck mass in the neonate or as acute suppurative thyroiditis in the infant or child. The etiology for both presentations is a fistulous track from the piriform sinus, most commonly on the left side, occurring as a result of a persistent remnant from the third or fourth branchial pouch. This condition should always be suspected in a neonate presenting with an inflammatory lesion containing air in the left side of the neck. Similarly, acute suppurative thyroiditis is rare and its presence should prompt consideration of a piriform sinus fistula as the etiology. Treatment of the acute infection should be followed by surgical extirpation in all cases.

PREOPERATIVE

Assessment and preparation

Cysts and sinuses of the first and second branchial clefts are diagnosed by their clinical appearance on careful physical examination. No special diagnostic imaging is indicated. The operation may be performed at any age, usually at the time of diagnosis, the main consideration in neonates being the availability of sophisticated pediatric anesthesia. The use of sclerosing solutions is contraindicated and may be dangerous. If infection is present, a course of antibiotics should be administered first. With respect to cysts or sinuses suspected to be of third or fourth branchial pouch origin, barium studies may demonstrate the presence of a piriform sinus fistula, particularly after a course of antibiotics and resolution of the surrounding inflammation. Computed axial tomography has also proved useful in diagnosing lesions of a third or fourth branchial pouch origin. If imaging techniques are not successful after resolution of the inflammation, then the next time the inflammation recurs, compression of the pus-filled cyst during endoscopy may reveal the origin of the fistula as pus exudes from the piriform sinus.

Anesthesia

Endotracheal inhalation general anesthesia, preferably by a pediatric anesthetist, is preferred.

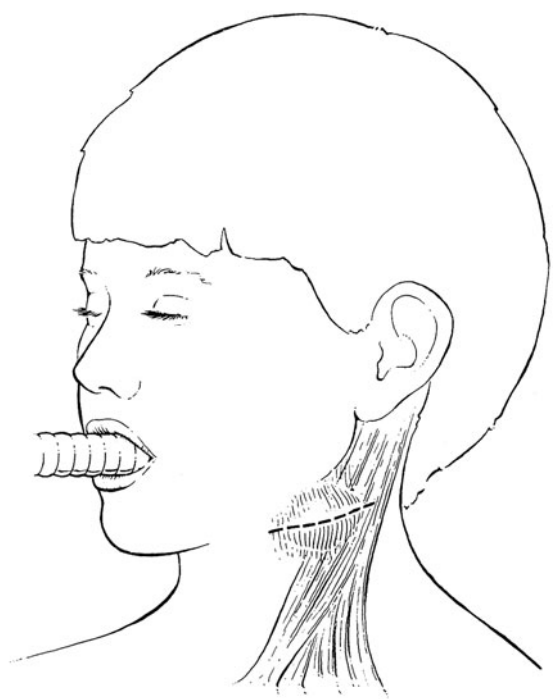
OPERATIONS

Position of patient and preparation

The patient is placed in the supine position with a sandbag beneath the shoulders and a soft ring headrest beneath the cranium so that the neck is extended. The head of the table is raised slightly to diminish venous blood pressure in the head and neck. The chin is turned away from the side of the lesion, and the skin is prepared with 10 percent povidone-iodine solution. The field is draped with four towels held to the contours of the neck with plastic barrier drapes with one adhesive edge (Steridrape 1010). An electrocautery grounding plate is applied to the thigh.

Exposure of cyst

4 The most common branchial cyst is derived from the second branchial cleft. The skin incision is made over the cyst along Langer's lines or in a natural skin crease in order to obtain the best cosmetic result. The length of the incision will vary with the size of the cyst.



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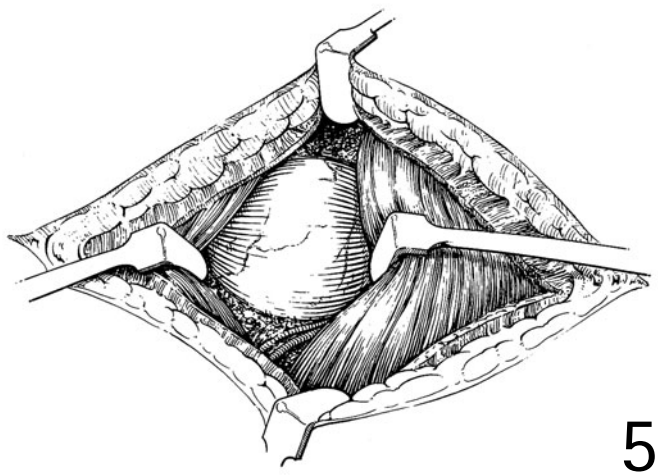
Infiltration of the overlying skin and adjacent tissues with dilute norepinephrine (noradrenaline) (1:1000 in isotonic saline) is optional, but generally unnecessary. A scalpel is used to incise the skin only, and subsequent dissection is accomplished by lifting the tissues off the underlying structures with Adson's tissue forceps and dissecting with a fine hemostat and electrocautery. The incision is carried through the subcutaneous tissues and platysma to the level of the cyst. The cyst is exposed by retracting the skin and muscle flaps, which is best accomplished with a self-retaining or ring retractor.

5 The deep cervical fascia is divided next to the anterior border of the sternocleidomastoid muscle, allowing the belly of the muscle to be retracted away from the cyst. Exposure is extended anteriorly and medially by retraction of the sternohyoid muscle. The fascia and soft tissue overlying the cyst are lifted and incised carefully to expose the superficial aspect of the cyst.



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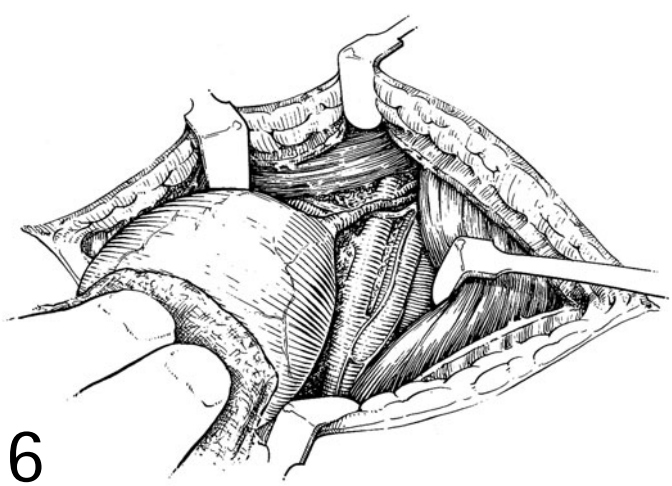

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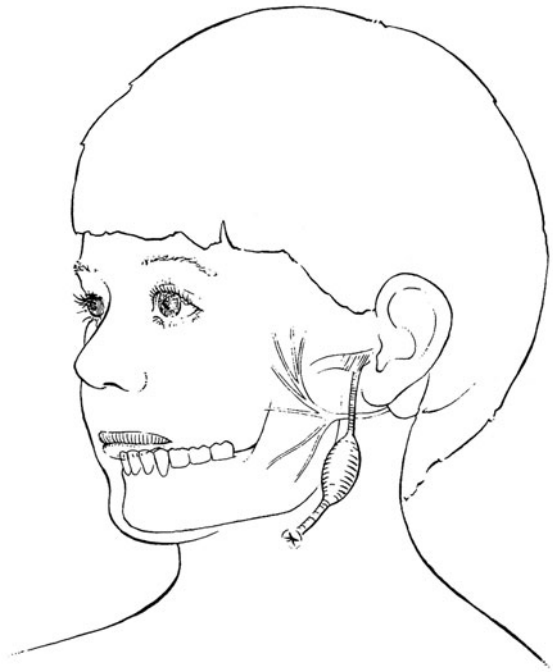
Dissection and removal of a second branchial cleft cyst

6 Great care is taken to avoid rupture of the cyst, as a tense cyst wall is easier to define and dissect than a collapsed cyst. Adjacent structures are separated from the cyst by blunt and electrocautery dissection along the cyst wall, special care being taken along the deep aspect of the cyst where the jugular vein and carotid arteries are in intimate relation. The pedicle of the cyst generally lies posterior to the jugular vein, usually coursing between the carotid artery bifurcation. It is then dissected cephalad towards the tonsillar pillar, where it is clamped and suture ligated with fine non-absorbable suture. Meticulous hemostasis is obtained and the wound irrigated with 1 percent povidone-iodine solution before closure.

Dissection and removal of a first branchial cleft cyst

7 During dissection of the less common first branchial cleft cyst, care must be taken to avoid damage to the adjacent facial nerve in cases where there is a tract leading up to or into the external auditory meatus. Not uncommonly, the deep or superficial lobes of the parotid gland must be mobilized. A neurosurgical nerve stimulator is often helpful during the dissection.



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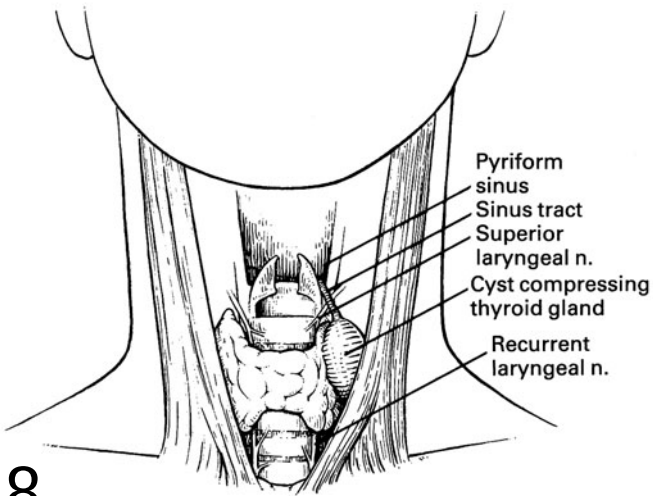
Dissection and removal of a third or fourth branchial pouch cyst and sinus

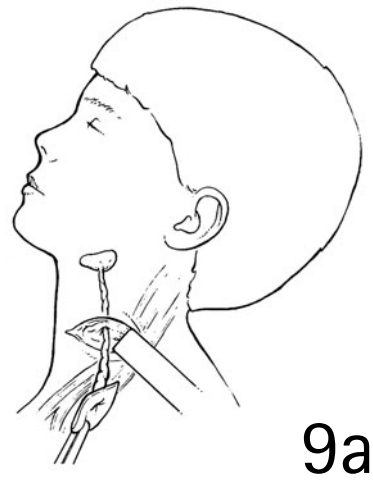
Cysts and sinuses of the third and fourth branchial pouch are clinically similar because of their common origin in the piriform fossa and presentation as a neck or thyroid abscess. Common presentations include an air-containing inflammatory lateral neck mass requiring repeated incision and drainage. Preoperative resolution with antibiotics should be followed by a barium swallow or contrast computed tomography to allow visualization of the piriform sinus tract common to each of these anomalies. Exploration of the neck with excision of the entire tract to the level of the piriform sinus is necessary to prevent recurrence. Operative endoscopy at the start of the operation may enable cannulation of the tract from above, which greatly facilitates localization of the tract during resection. Once the cyst and tract are resected, the histological finding of squamous cell epithelial lining confirms the diagnosis of a branchial anomaly.



8

8 The thyroid gland is exposed through a standard collar incision, and the left lobe is mobilized. The recurrent and superior laryngeal nerves and parathyroid glands should be identified and protected. If no discrete cyst or tract is found, the fistula may be located at the laryngeal level near the cricothyroid membrane. The fibers of the inferior constrictor muscle are bluntly spread to expose the piriform recess. Extreme caution should be exercised in this region to preserve the external branch of the superior laryngeal nerve. The tract usually passes inferiorly, external to the recurrent laryngeal nerve along the trachea to the superior pole of the thyroid. It may end blindly near the gland or actually penetrate the capsule to terminate in the parenchyma of the left thyroid lobe. Thyroid lobectomy or resection of the superior pole is carried out as indicated by the extent of the cyst.

Excision of a second branchial cleft sinus



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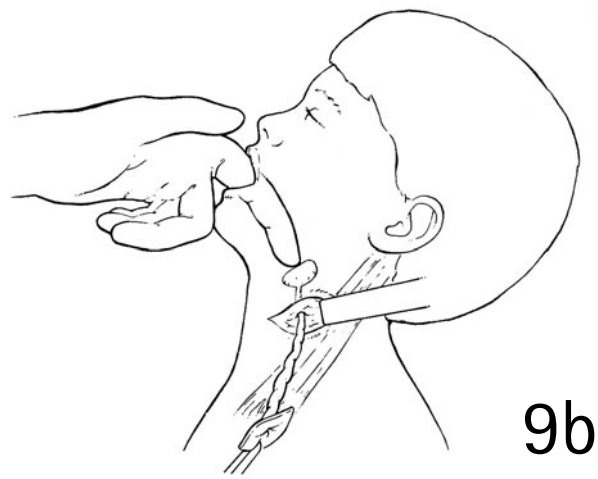


9b

9a,b The operation to excise a second branchial cleft sinus begins with an elliptical transverse incision at the sinus opening, and cephalad dissection of the tract to its furthest extent, generally at the level of the tonsillar pillar. The dissection is kept directly on the tract to avoid injury to contiguous structures, e.g., the internal jugular vein, the bifurcation of the carotid artery and the hypoglossal nerve. The operation can almost always be carried out through a single elliptical incision if the tract is kept under gentle traction and the anesthetist places a gloved finger in the tonsillar fossa and exerts downward pressure towards the field of dissection. In addition, the anesthetist's finger helps to localize the tonsillar fossa at the end point of dissection, where the sinus tract is suture ligated and divided. Dissection of the sinus tract may be facilitated by passing a fine silver probe or piece of heavy nylon suture the length of the tract and clamping this in position as the dissection progresses.

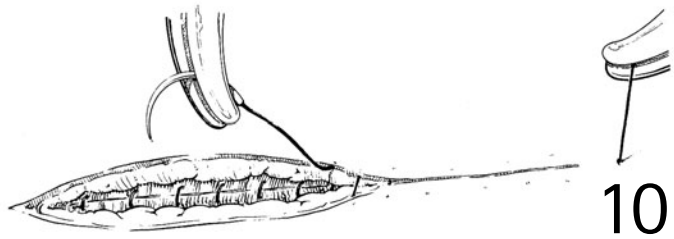
Minor branchial remnants

A branchial arch remnant may also occur along the lower anterior border of the sternocleidomastoid muscle near the sternoclavicular joint, and typically consists of a small cartilaginous mass presenting in the subcutaneous tissue. The lesion is usually visible and palpable, and bilateral occurrences are common. An accompanying sinus or cyst is seldom present, and infection is uncommon. Excision may be carried out for cosmetic reasons or may be delayed indefinitely.

Preauricular sinuses or pits are common and have been attributed to vestiges of the first branchial cleft. These lesions probably relate to the infolding and fusion associated with formation of the ear. Asymptomatic lesions require no treatment; draining sinuses and infected cysts require antibiotic treatment, incision, and drainage if they fail to resolve, and later excision to prevent recurrence.

Wound closure

10 The cervical fascia and platysma are closed with interrupted sutures of 4/0 polyglycolic acid. When the dissection is carefully performed and the cyst or sinus is not infected, the incision may be closed without using a drain. The skin is closed with a running 5/0 subcuticular absorbable suture or pull-out nylon suture. Steristrips and a clear plastic dressing (Opsite) are applied as a wound dressing.



POSTOPERATIVE CARE

The operations described are carried out as outpatient procedures unless unusual difficulties are encountered or unless a drain is placed. Antibiotics (penicillin or a cephalosporin) are continued for 1–5 days after operation, depending on the presence of infection or degree of contamination. An occlusive dry sterile dressing is kept in place for 48 hours, after which, the patient is permitted to bathe and shower normally. If a drain is placed, it is removed after 24 hours or whenever drainage has ceased. The patient is seen 7 days after operation for a wound check.

OUTCOME

The outcome for the procedures described is usually excellent, both functionally and cosmetically. Operative damage to related anatomical structures is rare and should not occur provided the surgeon has adequate knowledge of the anatomy and that meticulous hemostasis is obtained during dissection to ensure a clear field. Failure to excise the cyst or sinus completely may lead to its recurrence, in which case the patient should be treated with antibiotics and a thorough diagnostic re-evaluation initiated.

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7

External angular dermoid cyst

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PRINCIPLES AND JUSTIFICATION

Dermoid cysts are congenital cysts that result from sequestration of ectodermal and mesodermal elements. They occur along lines of embryologic closure, and the external angle of the supraorbital ridge is the most common site. External angular dermoid cysts are situated beneath the muscle and lie in a shallow depression in the outer table of the bone of the skull. A 'pit' within the bone is invariably present in the base of the bony depression through which the cyst receives its blood supply. Occasionally these cysts have an intracranial extension in a 'dumb-bell' fashion, although this is far less common than with dermoid cysts situated at the internal angle of the eye or over the bridge of the nose.

External angular dermoids present as rounded, soft, semi-mobile swellings in the lateral part of the eyebrow. They are usually asymptomatic, although with time can enlarge and rupture or become infected. There are few conditions that cause diagnostic confusion in the infant or child. Treatment is by surgical excision of the cyst, which must be complete to prevent recurrence.

PREOPERATIVE ASSESSMENT AND PREPARATION

Radiology

The diagnosis is made on clinical grounds. If there is any suspicion of intracranial extension, magnetic resonance imaging should be performed preoperatively. However, unlike for dermoid cysts situated medially on the face, these are not indicated routinely. Skull X-rays are not helpful for excluding an intracranial extension as they may be misleading.

Consent

Informed consent must be obtained from one of the child's parents or the child's legal guardian. The complications of bleeding, infection, recurrence, and keloid scarring must be explained as part of this process.

Anesthesia

General anesthesia is administered via an endotracheal tube and the patient is placed supine on the operating table. The patient's head is positioned on a head ring.

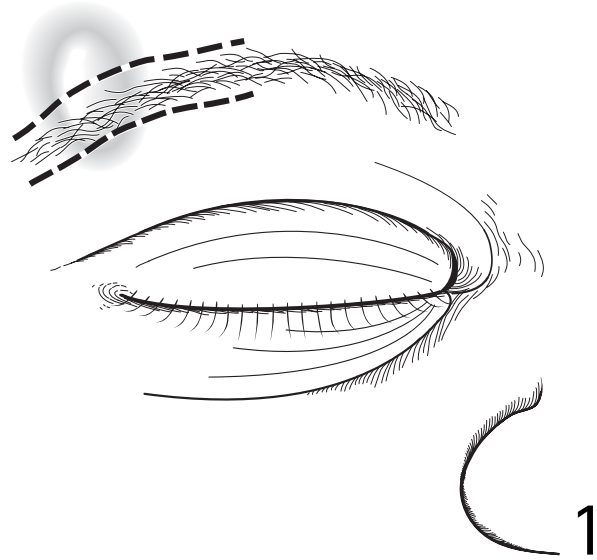
OPERATION

Skin preparation and draping

There is no need to shave the eyebrow for this procedure. The eyelids are taped closed to prevent soiling of the eye. The skin is prepared with either aqueous betadine or aqueous chlorhexadine. A head towel is used to maintain the sterile field.

Incision

1 A 1–1.5 cm incision is made over the cyst in line with the upper or lower margin of the eyebrow. Care is taken not to cut the hair follicles of the eyebrow. Although this incision gives a good cosmetic result, an alternative approach is to make the incision in the palpebral crease itself.



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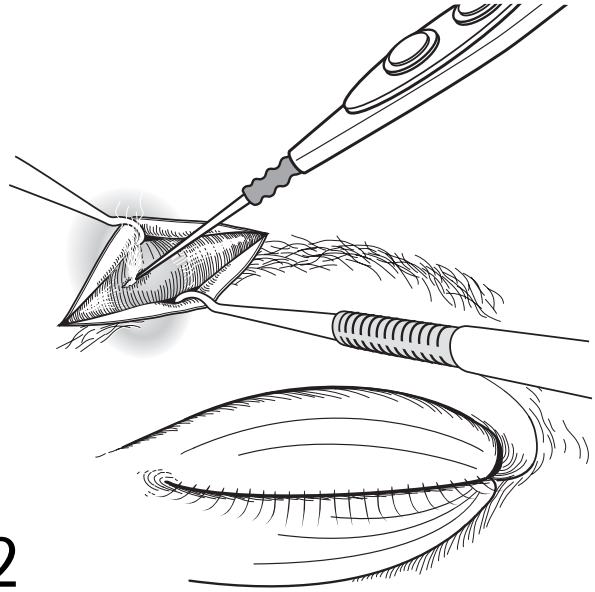

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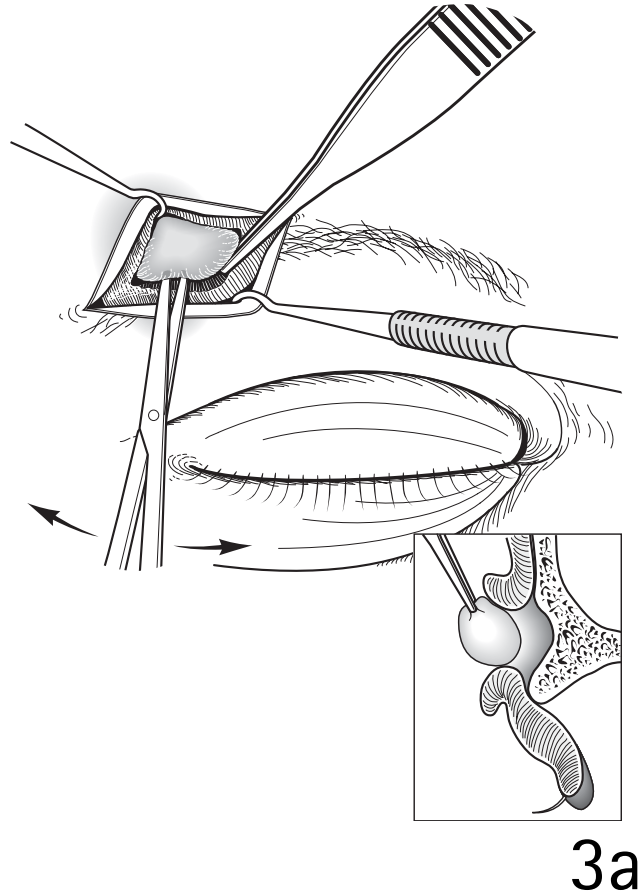
Dissection and excision

2 The incision is deepened using a fine monopolar diathermy point until the cyst becomes visible. Care is taken to avoid diathermy contact with the skin edges.



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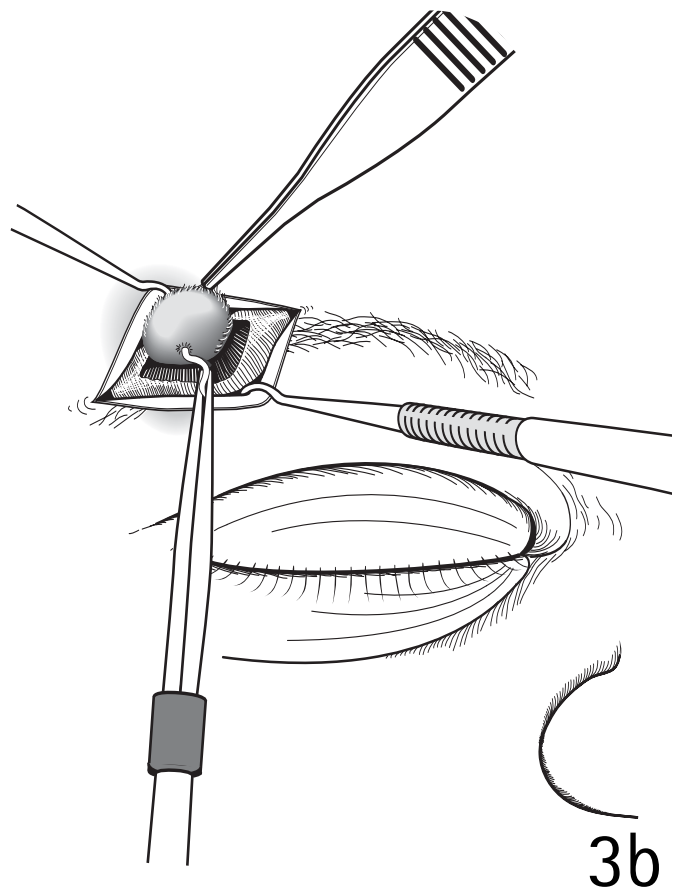


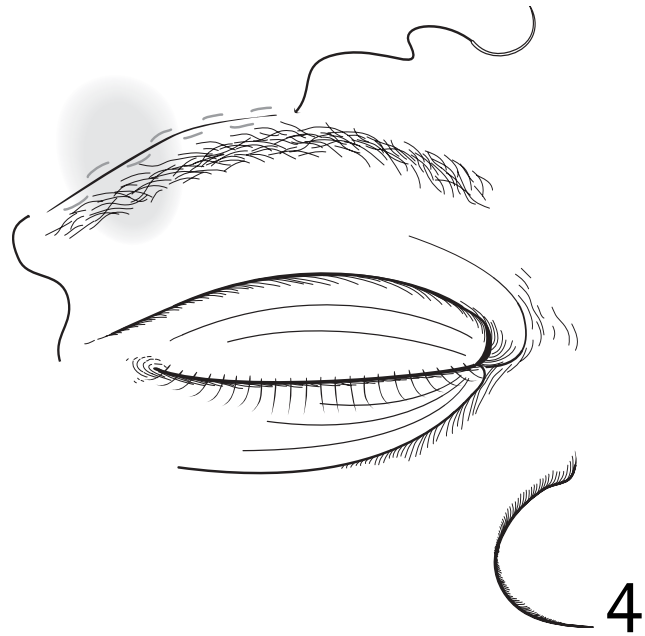
3b

3a,b The cyst is then mobilized using a mixture of blunt-scissor dissection and dissection with bipolar diathermy forceps. Meticulous hemostasis is ensured throughout. Instruments should not be applied directly to the cyst as this usually results in its rupture. The cyst is freed from its deep attachments, which often requires excising a circumferential rim of periosteum in continuity with the cyst. The feeding vessels passing through the pit of the bony depression are coagulated. Occasionally this pit needs to be packed with bone wax to secure hemostasis. The cyst is removed and sent for routine histopathology to confirm the diagnosis. If rupture does occur during mobilization, the wound should be irrigated with saline and the contents and lining of the cyst completely removed to prevent recurrence.

Closure

4 Once hemostasis has been confirmed, interrupted 4/0 polyglycolic sutures are placed in the muscle and fat layers, ensuring that the knots are buried. The skin is either closed using a subcuticular 5/0 undyed polyglycolic suture or a more predictable cosmetic result is achieved using interrupted 6/0 nylon sutures, although the latter can only be used in those infants in whom removal of sutures is practical. A Steristrip dressing is applied along the wound, followed by a small gauze/Mefix pressure dressing to prevent hematoma formation and thereby optimize the cosmetic result.

In the rare event of an intracranial extension, a formal craniotomy is required because the intracranial portion may be the larger of the two elements.



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POSTOPERATIVE CARE

The majority of these procedures are performed as day cases. The pressure dressing is maintained for 48 hours after the operation and the wound kept dry during this time. If interrupted non-absorbable sutures have been used for skin closure, these should be removed 5 days postoperatively to prevent a tissue reaction to them.

OUTCOME

Complications are rare following this procedure. Postoperative hematoma and infection can occur, but are minimized by the techniques described above. Incomplete excision may result in recurrence of the cyst. The cosmetic result is usually good, although a keloid scar may result, especially if the wound becomes infected or the child is of Afro-Caribbean descent.

8

Sternocleidomastoid torticollis

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HISTORY

Sternocleidomastoid torticollis is the term used to describe the presence of a shortened, fibrosed sternocleidomastoid muscle (SCM) which results in traction of the mastoid process toward the sternoclavicular joint. The head, therefore, is rotated away from and tilted toward the involved SCM.

The etiology of torticollis remains undefined. Light microscopic evaluation of the involved SCM demonstrates replacement of muscle bundles by dense fibrous tissue. Approximately 25–30 percent of patients presenting with SCM torticollis have a history of breech presentation at birth, with 62 percent involved in a complicated birth. Recent evidence suggests that SCM torticollis may be the manifestation of an in-utero positional disorder with development of an SCM compartment syndrome. Bilateral SCM fibrosis is present in 2–3 percent of cases.

PRINCIPLES AND JUSTIFICATION

Approximately 30–40 percent of patients will present at age 1 to 8 weeks with a hard 1–3 cm, painless, *discrete mass*, or pseudotumor, located within the substance of the middle or inferior portions of the SCM. This is often, but not always, accompanied by torticollis, with the face turned away from and the head tilted toward the side with the tumor. Such pseudotumors consist of fibrous tissue and mesenchyme-like cells, fibroblasts, and myoblasts at various stages of differentiation and degeneration. The remainder of patients will present with muscular torticollis (30 percent), in which there may be *diffuse fibrosis* of the SCM, or postural torticollis (22 percent). Passive neck range of motion exercises that emphasize rotation toward and side flexing away from the affected SCM should be performed by the parents under the guidance of a physical therapist. The parents should be encouraged to place toys and other desirable objects on the ipsilateral side in order to encourage turning toward that side. With physical therapy, resolution of the SCM pseudotumor and/or fibrosis is usually observed over the following 3–4 months. Therefore operative intervention is not necessary in the majority (> 90 percent) of newborns and young infants, but is required in 25 percent of the 3–6-month-old infants, 70 percent of the 6–18-month-old children, and 100 percent of those children older than 18 months. Torticollis will persist in approximately 7 percent of newborns, but this occurs usually in those with severe rotational limitations. In fact, large series have demonstrated that with the use of manual stretching neck exercises, less than 8 percent of those with an SCM pseudotumor and 3 percent of those with muscular torticollis will require operative intervention. In general, operation is indicated in patients who are unresponsive after more than 6 months of manual stretching. Acquired plagiocephaly with flattening of both the frontal area on the ipsilateral side and the occipital region on the contralateral side may be noted in the first few months of life, but usually resolves spontaneously by 1 year of age after the infant begins to sit up.

Presentation in the child older than 1–2 years of age is more often refractory to conservative management, and operation is indicated if failure of physical therapy has been determined. Facial hemihypoplasia, which involves flattening and underdevelopment of the malar eminence and downward displacement of the eye, ear, and mouth on the affected side, may appear as early as 6 months of age, but most often appears after age 3 to 5 years. The development of facial hemihypoplasia is an indication for operative intervention, as potential resolution of skeletal abnormalities and subsequent normal growth and development of the facial skeleton will occur only after prompt operative correction of the torticollis.

PREOPERATIVE ASSESSMENT AND PREPARATION

Preoperative evaluation includes assessment to exclude other causes of abnormal head posture. Non-SCM causes for torticollis exist in 18 percent of patients and include neurologic disorders in 9 percent, such that the patient holds the head in a compensatory position most commonly due to the presence of ocular problems such as nystagmus or strabismus, or vestibular disorders. Ocular examination is indicated in patients without a clear musculoskeletal cause of torticollis. Benign paroxysmal torticollis is an episodic functional disorder of unknown etiology that occurs in the early months of life in healthy individuals and resolves by the age of 5 years. It is thought that this disorder occurs as a migraine equivalent or due to a dystonic SCM. The child's head tilts to one side for a few hours or days, often without any associated symptoms, although vomiting, pallor, and ataxia may occur. Posterior fossa tumors may also present with torticollis, frequently accompanied by headache, nausea, and vomiting. Cervical spine radiologic evaluation is necessary to exclude congenital osseous deformity of the cervical spine and the Klippel–Feil syndrome or C1-C2 cervical spine subluxation, which may occur in 6 percent of patients presenting with torticollis. Usually the SCM is normal in these cases, but occasionally SCM fibrosis may coexist with other causes of torticollis. Atlantoaxial rotatory subluxation may be the cause of acute torticollis in children and also may be noted following head and neck procedures or infection. Imaging with computed tomography (CT) or magnetic resonance imaging (MRI) may be diagnostic and is indicated in children with torticollis.

The diagnosis of an SCM pseudotumor and torticollis in the newborn or infant presenting with an SCM mass can almost always be made based on physical examination, history, and clinical progression. Occasionally, ultrasound evaluation may help to differentiate such an SCM pseudotumor from a mass secondary to lymphadenopathy or neoplasm.

Photographic and/or CT evaluation for hemihypoplasia helps with the diagnosis and should be performed in any patient in whom torticollis persists beyond 6 months of age and as baseline before operative intervention. Radiologic evaluation of the hips and lower extremities should be performed because of the relatively high incidence (28 percent) of congenital dysplasia of the hip and abnormalities of the lower extremities in patients with torticollis. Hip dysplasia is observed in 7 percent of infants with a pseudotumor and is more commonly noted in infants with greater limitations in neck rotation. Preoperative open-mouth radiography of the odontoid process should be performed to look for tilt of the odontoid process to the side of the torticollis.

Anesthesia

General anesthesia with endotracheal intubation is required. The endotracheal tube should be secured sufficiently and placed appropriately to allow sterile preparation of the entire neck and full rotation of the neck in both directions.

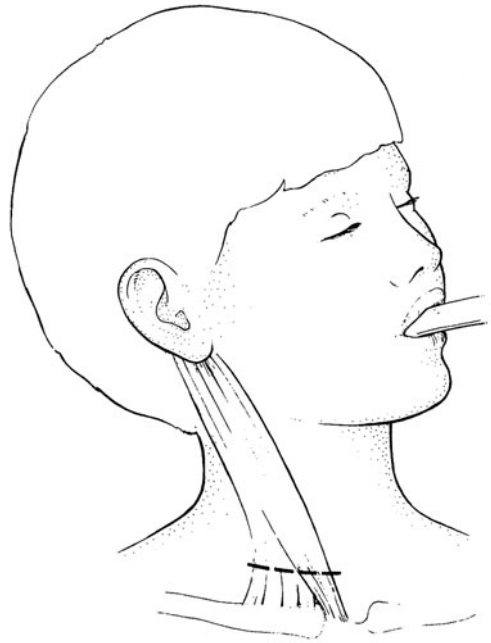
OPERATION

The standard operation involves division of the SCM at the point where the sternal and clavicular heads converge. However, reports suggest that botulinum toxin injection into the SCM or lengthening of the SCM at its mastoid insertion may be effective. Selective peripheral denervation of the cervical muscles, including the spinal accessory branches to the SCM and C1-6 posterior nerve branches on the involved side, may also be effective, especially in those with an initial response to botulinum toxin injection.

Position of patient

The patient is placed in the supine position with a roll placed transversely under the shoulders in order to provide neck extension. The bed is flexed such that the head of the patient remains in a 30° upright position in order to decrease venous bleeding. The head is turned slightly to the side opposite the affected SCM. A sponge 'doughnut' is placed under the head to maintain head position. The prepared field should include the entire neck extending up to just above the mandible, down to the areas of both shoulders, and to the infraclavicular regions bilaterally. Crumpled drapes are placed in the posterior aspect of the junction of the neck and shoulders bilaterally. Drapes are then placed along the inferior border of the mandible, at the level of the clavicle, and bilaterally along the posterolateral aspect of the neck.

1 An approximately 3 cm, transverse, curvilinear incision is made in a skin crease one to two fingerbreadths above the clavicle.



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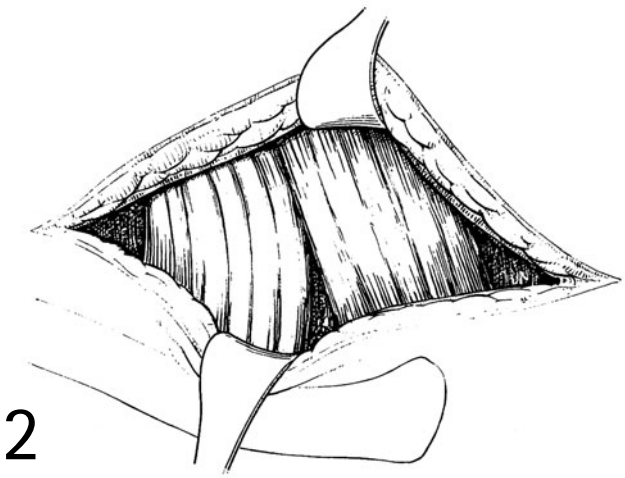

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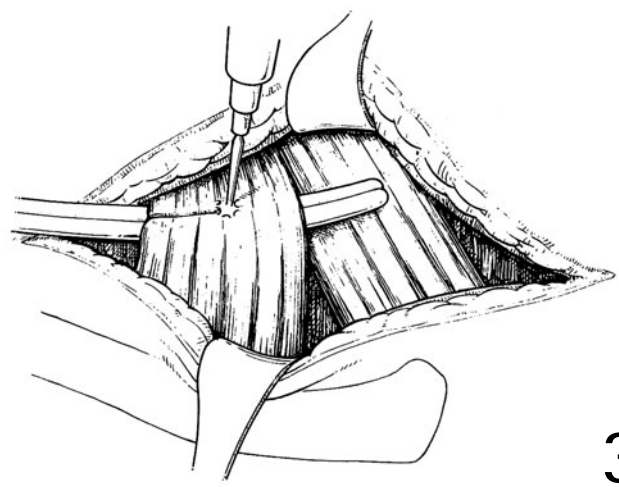
2 The platysma is divided. Subplatysmal flaps are developed along the SCM for 1–2 cm superiorly and to the level of the clavicle inferiorly. Division of the external jugular vein may be necessary. The posterior aspect of the SCM is then dissected free from the underlying carotid sheath structures at the point where the sternal and clavicular heads converge.

3 The sternal and clavicular heads, including the underlying investing cervical fascia, are divided at that level using electrocautery, with care taken to ensure hemostasis. The spinal accessory nerve is usually located superior to this region.



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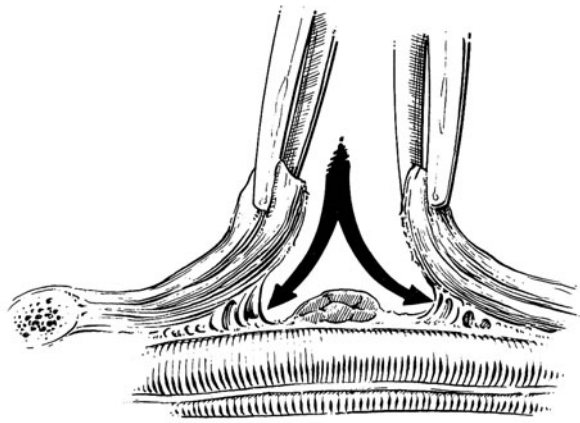
4 The ends of the transected SCM are then dissected free from the underlying carotid sheath structures. Limited dissection is performed superiorly while more extensive dissection is performed inferiorly down to the level of the sternum and clavicle.



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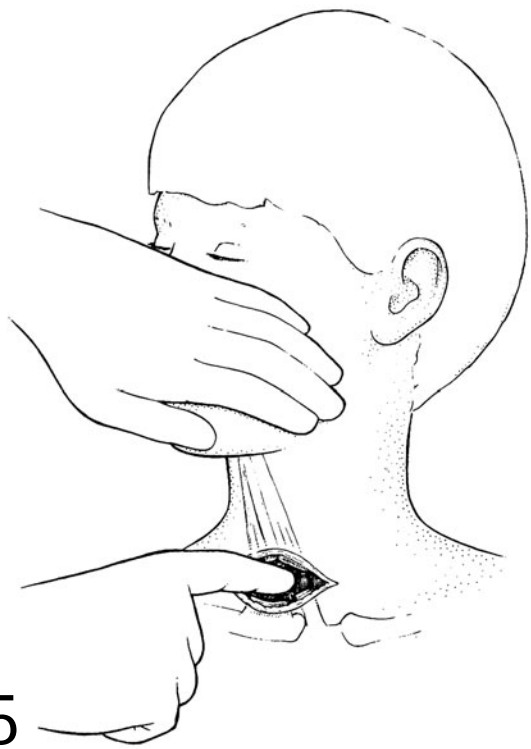

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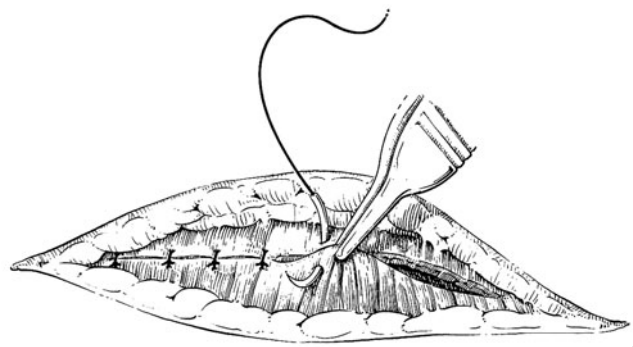
5 The head is then turned from side to side and the depths of the wound palpated to ensure that all contracted tissue is released. If tight lateral structures, such as the deep cervical fascia lateral to the SCM, are identified, then these should be divided under direct vision. Although unusual, fascial tissue involved with the omohyoid muscle and the carotid sheath may occasionally require division.

6 The platysma muscle is then re-approximated with interrupted 3/0 vicryl sutures.



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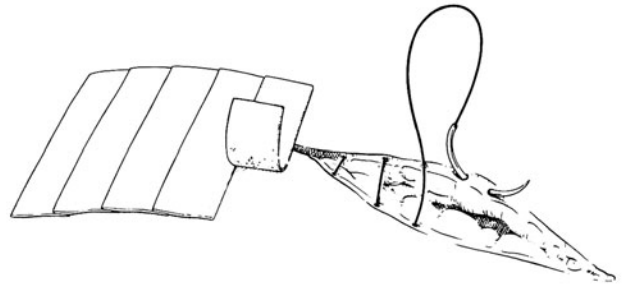
7 The skin is approximated with a running 5/0 vicryl subcuticular suture. Steristrips are placed perpendicular to the incision in overlying fashion such that a dressing is formed. The patient is maintained in a 30° head-up position during and after recovery.



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POSTOPERATIVE CARE

For the following 48 hours the patient remains supine in the 30° head-up position without the use of a pillow. Careful assumption of the erect posture with initial assistance is then allowed. Some clinicians advise the use of a neck collar in the postoperative period. Physical therapy with resumption of passive neck range of motion exercises is then reinstituted and continued for the following 6 months. Full neck range of motion should be achieved by 7–10 days postoperatively. Older children must undergo retraining of the neck-righting reflexes in front of a mirror over the following months. Occasionally, a patient who is too young or not willing to cooperate with postoperative physical therapy will require placement of a Minerva cast for 6 weeks in order to maintain the head rotated toward the operatively treated side and tilted laterally toward the opposite side.

The most common complication is that of postoperative bleeding. Hematoma formation over the hours to days after the procedure may require evacuation. Specific attention to hemostasis during division of the muscular fascial tissues and to potential bleeding from tributaries of the internal jugular vein should prevent this complication. The risk of airway compromise secondary to postoperative bleeding is low. Injury to the spinal accessory or facial nerve is unusual.

OUTCOME

In the more than 90 percent of patients who are managed without operative intervention, follow-up should include monitoring to ensure resolution of the SCM pseudotumor, residual SCM fibrosis, and torticollis. Specifically, close follow-up should ensure the early detection of facial hemihypoplasia in those patients with persistent torticollis after 6 months of age.

After operation, recurrent torticollis is observed in less than 3 percent of patients. Normal neck movement is observed with long-term follow-up in 88 percent of patients. Follow-up should include evaluation for facial hemihypoplasia and plagiocephaly, which, if present, will resolve. In addition, because of the high incidence of residual cervical and thoracolumbar scoliosis in patients with torticollis, radiologic spine evaluation is indicated to ensure identification and appropriate follow-up of any spinal deformities.

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9

Tracheostomy

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PRINCIPLES AND JUSTIFICATION

Tracheostomy should be performed as an elective procedure using the conventional dissection technique. Percutaneous dilatational tracheostomy, now popular for adults, is not suitable for children because the airway is small and often precarious; the trachea is soft, flexible, and very mobile; landmarks can be difficult to palpate; and a displaced tube cannot be rapidly replaced.

INDICATIONS

- Airway obstruction.
  - Congenital anomalies, e.g., laryngeal web, subglottic stenosis.
  - Failed extubation with increasing subglottic edema and evidence of mucosal damage.
  - External trauma, e.g., 'hanging'-type injuries.
  - Acute infection, e.g., acute epiglottitis or laryngotracheobronchitis, if endotracheal intubation is not available.
  - Tumors, e.g., hemangioma, lymphangioma, recurrent respiratory papillomatosis.
  - Functional obstruction, e.g., bilateral vocal cord palsy, cricoarytenoid joint fixation.
  - Tracheomalacia or extrinsic compression of the trachea.
- Long-term respiratory support, for example, for pulmonary pathology.
- Clearance of secretions, for example, in neurological injury or disease.
- As a covering procedure to secure the airway for subsequent surgery on the larynx, pharynx, or temporomandibular joints.

PREOPERATIVE

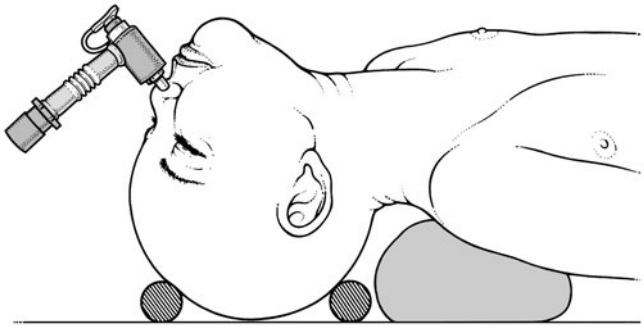
Anesthesia

The operation is performed under general anesthesia. Children are usually intubated, but in cases where intubation is deemed impossible a laryngeal mask airway (LMA) may be used, or even a facemask. Exceptionally it might be necessary to undertake the procedure under local anesthesia, using lignocaine (lidocaine) 1 percent with epinephrine (adrenaline) 1:200 000.

OPERATION

Position of patient

1 The neck must be hyperextended. A jellyroll or sandbag is placed under the shoulders. The occiput is supported by a head ring. The patient is tipped into a slightly head-up position. A fenestrated 'circumcision' drape is convenient and allows an anesthetic connector to be passed easily beneath it into the surgical field when the tracheostomy tube has been inserted. Bipolar diathermy should be available.



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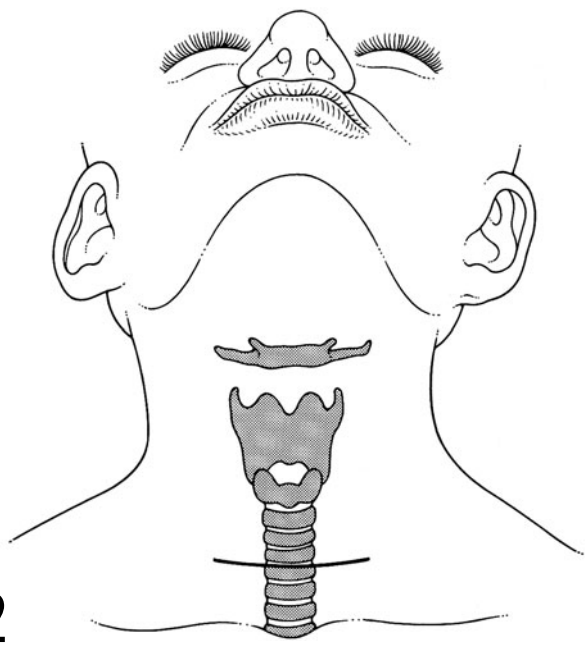

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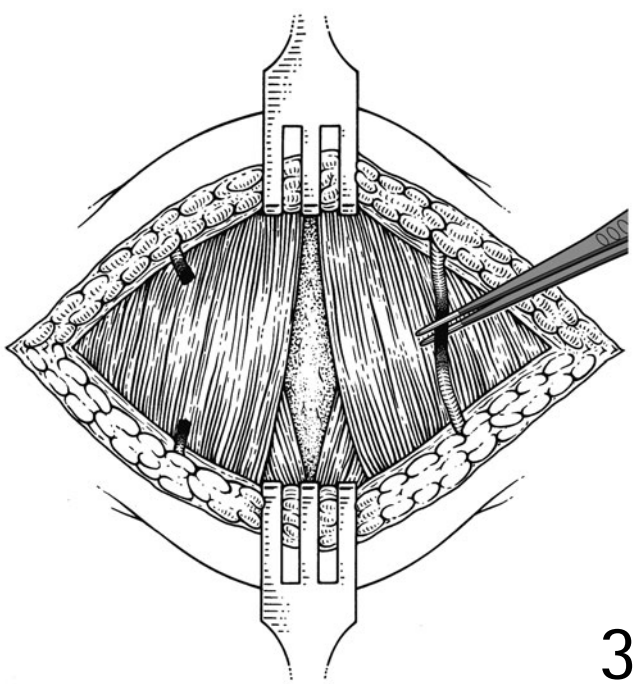
Incision

2 The neck is palpated carefully so that the hyoid bone, thyroid notch, and cricoid cartilage can be felt and marked. In neonates, the cricoid cartilage is not easily palpable. The skin midway between the cricoid and the suprasternal notch is marked and infiltrated using 1 percent lignocaine with 1:200 000 epinephrine. A horizontal skin incision approximately 2 cm in length is made through the infiltrated area, cutting through fat and platysma. In infants and young children it is advantageous to de-fat the skin along the incision margins, for which purpose microbipolar diathermy forceps are useful.

3 The incision is deepened in a horizontal plane until the deep cervical fascia investing the sternohyoid and sternothyroid strap muscles is encountered. Branches of the anterior jugular vein will be seen during this dissection; these should be coagulated and divided with the bipolar diathermy.



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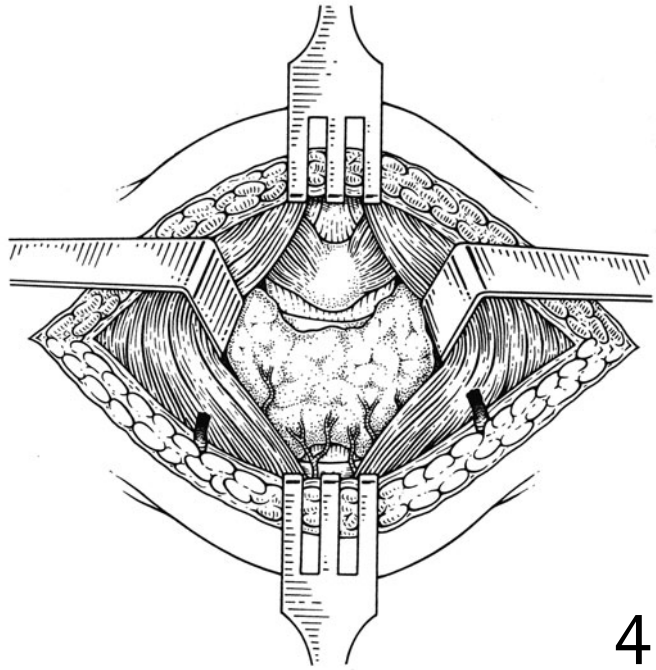
4 Having identified the strap muscles, a condensation of the investing layer of the deep cervical fascia will be seen running vertically between them in the midline. This interval is entered by blunt-scissor dissection and the strap muscles are separated and retracted laterally. Sprung self-retaining retractors of the Aberdeen pattern are very convenient for this purpose.



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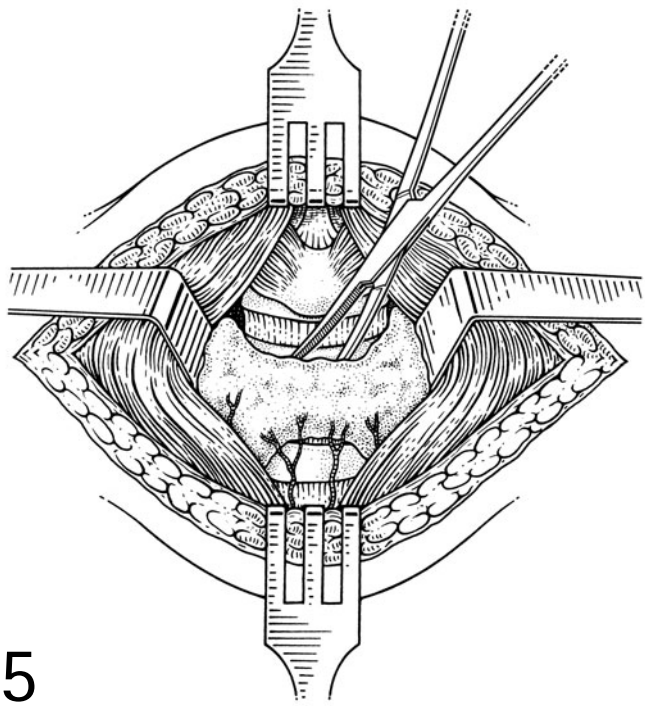
Exposure of the thyroid gland

Retraction of the strap muscles reveals the thyroid gland with the thyroid isthmus joining the two lobes of the gland. Above the isthmus the cricoid is seen, although in the neonate it is easier to identify by palpation, feeling for its prominence with fine curved 'mosquito' artery forceps.



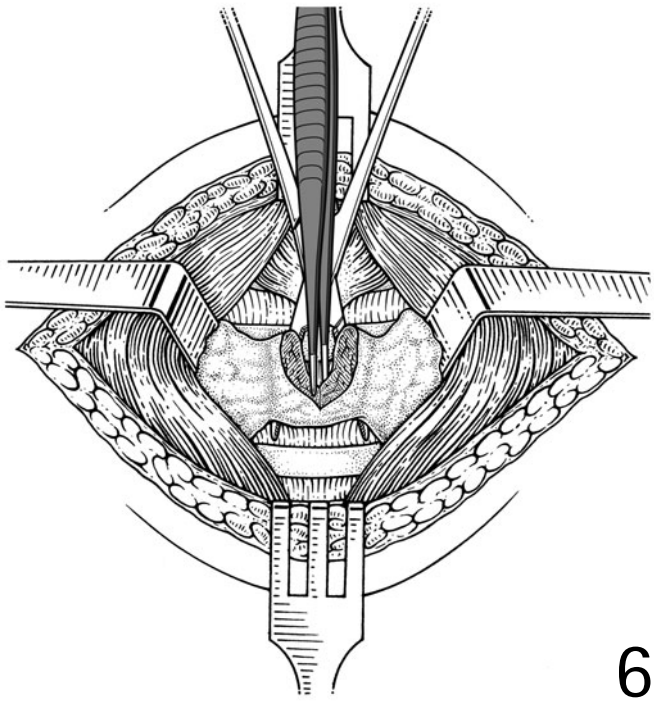
5

Dissection of the thyroid isthmus

5 The size and relationship of the thyroid isthmus to the trachea are variable, but in all cases the isthmus should be divided. A small incision is made through the condensation of the pretracheal fascia at the upper border of the isthmus, and the isthmus is then separated from the underlying trachea by blunt dissection using fine curved artery forceps. Branches of the inferior thyroid vein will be encountered at the lower border of the isthmus, and these should be coagulated with diathermy.

Division of the thyroid isthmus

6 The artery forceps are spread beneath the isthmus, and bipolar diathermy is used to divide it bloodlessly. In adolescents with a bulky, vascular isthmus it may be preferable to divide the isthmus between hemostats and secure the ends by suture transfixion and ligation.



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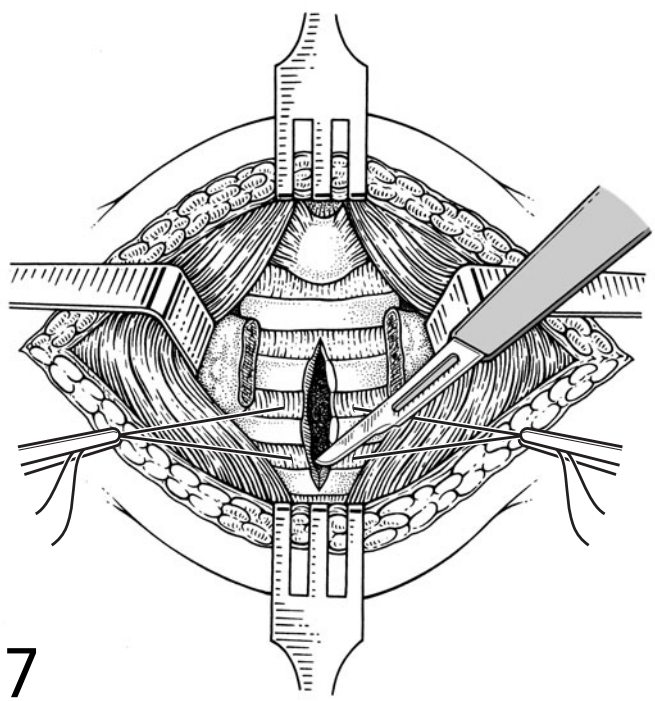
Opening the trachea

Careful hemostasis is achieved and the tracheal rings counted. The first tracheal ring must *not* be included in the tracheostomy, and in neonates and infants, where distances are small, it is preferable to preserve the second ring as well.



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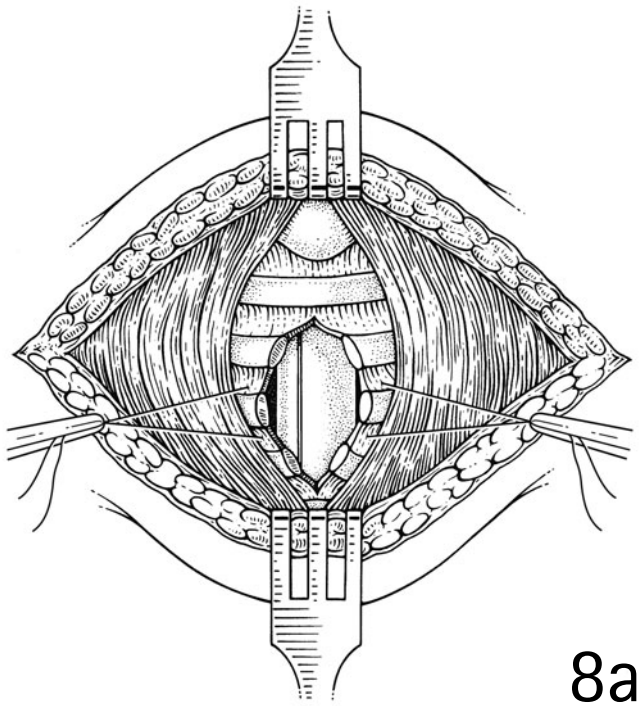
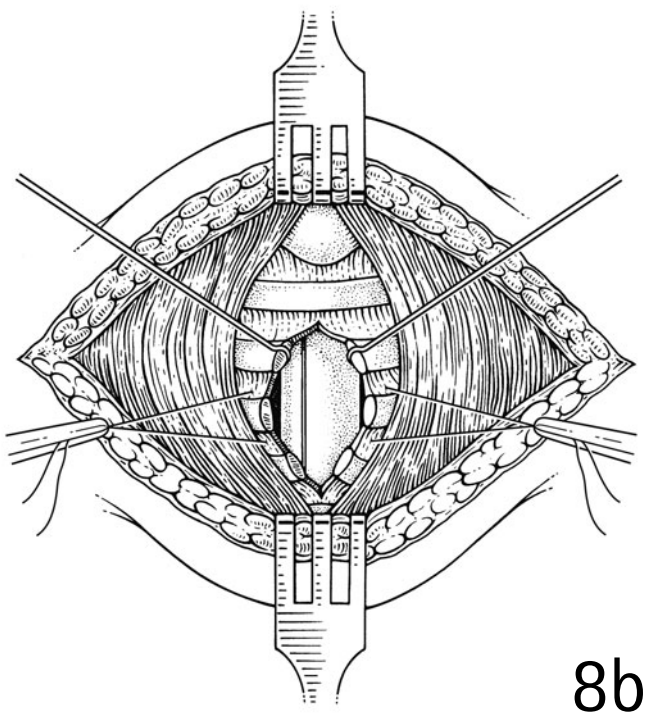
7 Before making the incision, Prolene stay sutures are placed to help distract the tracheal opening: these are taped to the skin and left in place until the first tracheostomy tube change. For routine tracheostomy in children, a vertical incision is made in the trachea through the third, fourth and fifth tracheal rings. If a subsequent reconstructive operation on the larynx is planned, the incision in the trachea may be made through the fourth, fifth, and sixth tracheal rings. Care should be taken with a low tracheostomy that the end of the tracheostomy tube is clear of the carina, and in infants it is important to use a shorter 'neonatal'-length tube. A small sucker should be used to prevent blood from entering the trachea.



8a

8a,b The stay sutures are now used to distract the cut edges of the trachea, if necessary, aided by fine skin hooks. The endotracheal tube will then be clearly seen.



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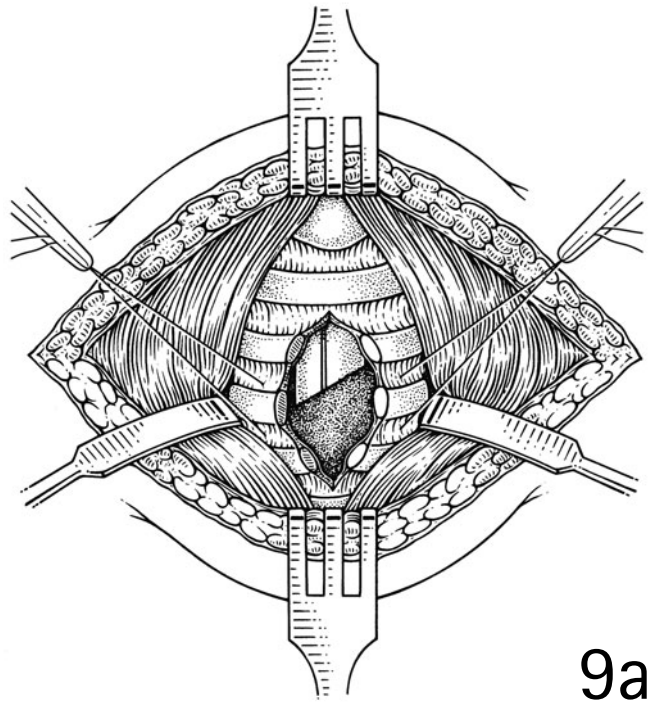

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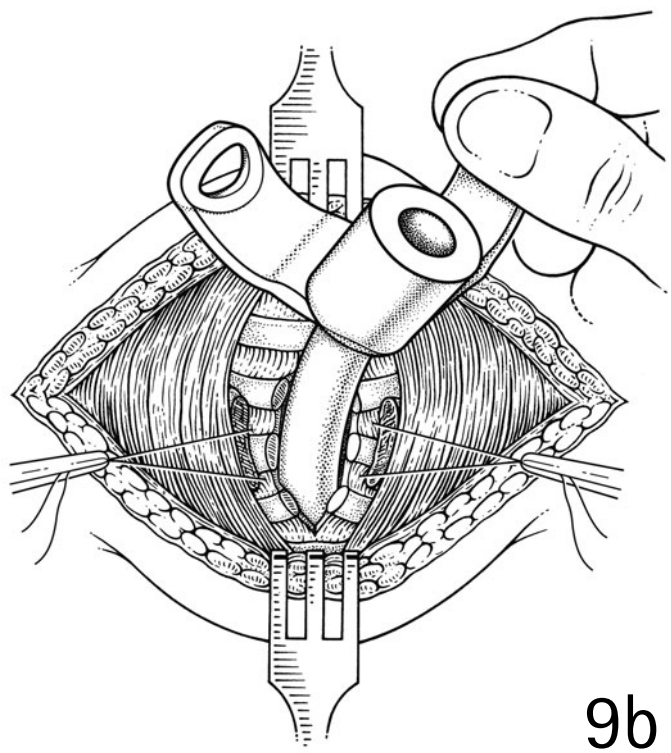


9b

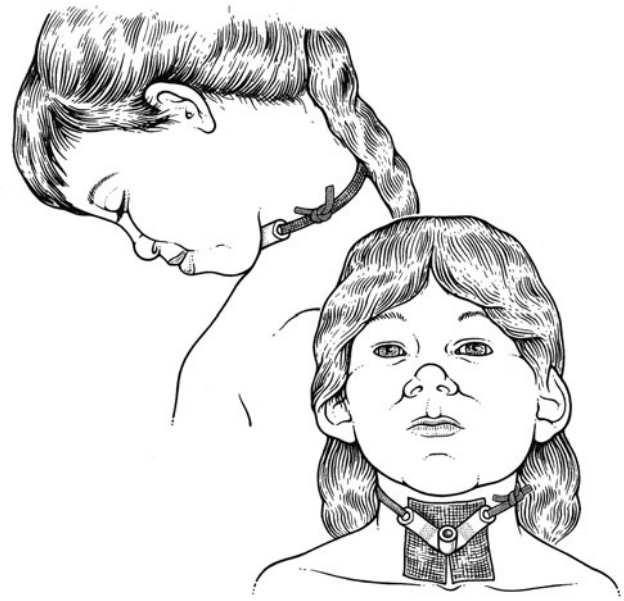
Insertion of tracheostomy tube

9a,b The anesthetist is asked to withdraw the endotracheal tube gently until its tip is just above the tracheostome. A plastic tracheostomy tube with a 15 mm termination is then inserted into the tracheal opening, a sterile anesthetic connector is fitted to it, and the end of the connector is passed out of the surgical field beneath the drape to the anesthetist.

Fixation of tracheostomy tube

When the tracheostomy tube is fully inserted it must be held by the assistant until it is properly secured. The skin edges are partly approximated with a suture on each side of the tube. It is essential to leave a gap around the tube to avoid postoperative surgical emphysema.

10 The jellyroll is then removed from underneath the patient's shoulders and the tracheostomy tube is secured by tying the tapes with a reef knot *with the neck flexed*. It is extremely important to remember this neck flexion and to tie the tapes fairly tightly; failure to do this may result in the tube becoming dislodged. Dislodgement of the tracheostomy tube must be avoided during the immediate postoperative period, as its subsequent re-insertion can be difficult in the first few postoperative days. A non-adherent dressing is applied to the wound, with a keyhole to accommodate the tracheostomy tube. This dressing should be kept clean and changed when necessary. At the end of the procedure the surgeon should listen to the chest with a stethoscope to ensure that air entry is symmetrical; if it is reduced on the left, this suggests that the tracheostomy tube is too long and has intubated the right main bronchus.



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POSTOPERATIVE CARE

The following must be at the bedside.

- A spare tracheostomy tube of the same pattern and size as that in the patient, and a tube one size smaller.
- Suction apparatus and sterile suction catheters with holes in the side of their tips.
- A properly trained nurse.
- Humidification.

On return to the ward, a chest radiograph should be arranged to check the length of the tracheostomy tube with regard to its proximity to the carina, and to ensure that there is no pneumothorax. The tracheostomy tube must be kept clear of secretions by the regular application of suction, initially at half-hourly intervals. Suction is facilitated by the instillation of 0.5 mL of sterile normal saline into the tracheostomy tube first. A sterile catheter is then inserted into the tracheal lumen gently but swiftly, and suction is applied only as the catheter is withdrawn.

If the tracheostomy tube is properly positioned and functioning satisfactorily, respiration should be virtually inaudible. If bubbling noises are heard, further suction is required.

Adequate humidification should be provided, using a head-box for neonates and a tracheostomy mask for older children. This will reduce the tenacity of the secretions and allow the tracheostomy tube to remain patent with less frequent suction.

The tracheostomy tube is changed routinely after 7 days. However, if any difficulty is encountered during suction, if breathing becomes noisy despite suction, or if the child begins to phonate, the tube must be changed immediately. This should be done with adequate help: the child's neck must be extended with a rolled towel under the shoulders, good illumination is essential and a headlight is desirable. As the tracheostomy tube is withdrawn, the stay sutures are pulled to distract the edges of the tracheal opening and bring them up to the surface of the neck. The tracheostome should thus be clearly visible, especially if the subcutaneous fat was reduced at the time of surgery, and a new tube is inserted under direct vision. If difficulty is encountered, a tracheal dilator is too large to be useful in small children; it is better to insert a suction catheter into the tracheal lumen and 'railroad' the new tube over it into the trachea.

The most common cause of difficulty in suction is displacement of the tube in front of the trachea. This is always accompanied by audible respiration and by the child being able to phonate.

COMPLICATIONS

Intraoperative

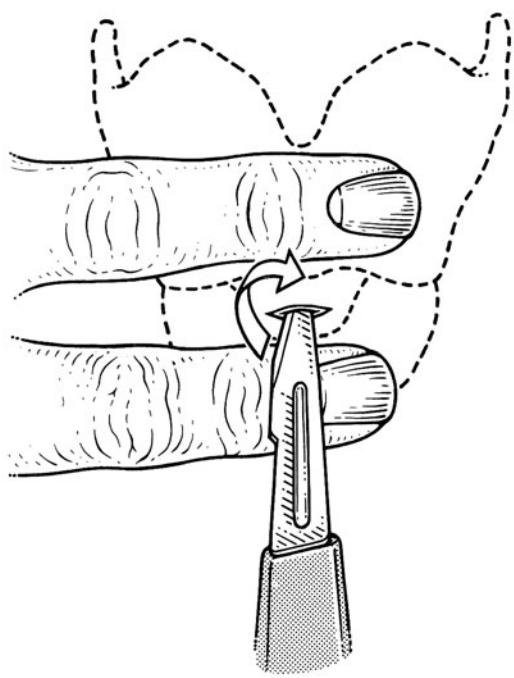
- Bleeding may be encountered from an abnormally high innominate artery. Careful controlled dissection of the structures in the lower part of the neck should avoid this.
- Damage to the cervical pleura may cause a pneumothorax; dissection lateral to the trachea should be avoided.
- Injury to the esophagus should not occur when an elective tracheostomy is performed.

Postoperative

- *Blocking and displacement of the tracheostomy tube.* These complications are preventable by careful nursing and attention to detail when tying the tracheostomy tube tapes.
- *Postoperative pneumonia.* This risk is small if a sterile technique is used for tracheal toilet and effective humidification is maintained. Adequate physiotherapy to encourage coughing is also desirable.
- *Surgical emphysema.* If this occurs, the wound should be opened fully. It is caused by suturing the wound too tightly around the tracheostomy tube.
- *Hemorrhage.* Reactionary hemorrhage from the wound may occur if hemostasis has been inadequate, a risk minimized by the use of bipolar diathermy dissection. Erosion of the anterior wall of the trachea may well occur if a metal tracheostomy tube of incorrect curvature is used. The overlying innominate artery may also be damaged, in which event a frequently fatal hemorrhage will result.
- *Stenosis.* This may occur at the tracheostome if cartilage is removed during the tracheostomy, or at the tip of the tube if suction is too vigorous or incorrectly applied. If the tracheostomy is placed too high and the cricoid cartilage is damaged by the tube, a subglottic stenosis may ensue.
- *Granuloma formation.* A suprastomal granuloma may form in the tracheal lumen at the upper margin of the tracheostome where it is irritated by the tube. If it becomes large enough to interfere with tube changes, phonation, or decannulation, it should be removed either with punch forceps via the stoma or endoscopically using the KTP laser through a bronchoscope. A tube-tip granuloma may arise as a result of irritation of the tracheal wall by either the tip of an ill-fitting tracheostomy tube or unskilled and overvigorous suction.
- *Suprastomal collapse.* Some degree of suprastomal collapse of the anterior tracheal wall at the upper margin of the tracheostome is almost inevitable in long-term pediatric tracheostomies. Usually it is mild, but occasionally it is sufficiently severe to hinder eventual decannulation. When the suprastomal flap occludes up to 50 percent of the airway, it can be successfully vaporized using the KTP laser through a bronchoscope. If more than 50 percent of the airway is occluded, a formal surgical decannulation may be needed, with anterosuspension of the suprastomal flap or even stomal reconstruction using a cartilage graft.
- *Difficult decannulation.* In the absence of persisting pathology, decannulation is most safely undertaken by progressively downsizing the tube to one of 3.0 mm inner diameter, blocking it for 24 hours, and then removing it if the child can manage comfortably both awake and asleep with the blocked tube in situ.
- *Persistent tracheocutaneous fistula.* After prolonged tracheostomy, removal of the tube may result in a persistent fistula at the site of the tracheostomy in up to 50 percent of patients. If it fails to close spontaneously and continues to leak mucus, the tract may be excised and a formal closure performed.

CRICOTHYROTOMY

In an emergency, for example, in the case of acute epiglottitis when sudden airway obstruction may occur and peroral endotracheal intubation may not be possible, a cricothyrotomy should be considered in preference to a crash tracheostomy, which can be extremely difficult in the child whose trachea is soft and may not be easy to identify.



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11 The child is held with the neck extended and the left hand is placed with the first finger over the cricoid cartilage and the second over the thyroid cartilage. A transverse stab incision is made through the cricothyroid membrane. The blade of the knife is then turned vertically, establishing the airway so that a tube of suitable size may be inserted to preserve the airway and to enable general anesthesia to be undertaken safely. Elective tracheostomy is then performed.

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10

Cervical lymphatic malformation

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HISTORY

Cervical lymphatic malformations have traditionally been called cystic hygromas or lymphangiomas. Most commonly, those with large cysts have been called hygromas and those with more tissue parenchyma have been called lymphangiomas. Both terms are antiquated and should be abandoned. In modern medical parlance, the suffix 'oma' implies a tumor with active cellular proliferation. The International Society for the Study of Vascular Anomalies (ISSVA) has accepted the terminology proposed by Mulliken in 1982 more appropriately describing these lesions as lymphatic malformations. They are subdivided into macrocystic, microcystic, and combined varieties.

PRINCIPLES AND JUSTIFICATION

Lymphatic malformations represent morphogenic errors in the development of the lymphatic vessels. They are most commonly located in regions of confluence of major lymphatic channels, including the neck (75 percent), axilla (20 percent), mediastinum, retroperitoneum, pelvis, and groin. Cervical lesions may extend into the mediastinum and/or axilla. Malformations may be detected antenatally and are usually visible at birth. Occasionally, a mass may not become apparent for months or, rarely, years.

These vascular anomalies may become infected or sustain intralesional hemorrhage in addition to causing disfigurement. Very extensive cervical lesions may threaten the airway immediately at delivery. On occasion, ex-utero intrapartum treatment (EXIT) procedures may be indicated to prevent neonatal asphyxiation.

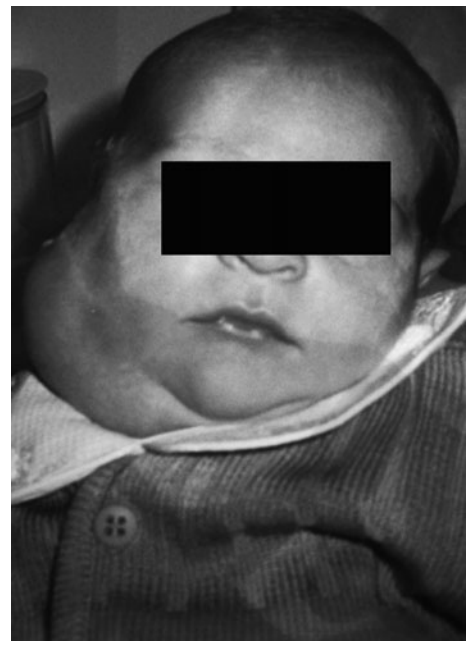
Predominantly macrocystic lymphatic malformations may often be successfully treated with cautious intralesional sclerotherapy. Sclerosants employed include ethanol, doxycycline, sodium tetradecyl sulfate, and OK-432 (an investigational agent). Sclerotherapy is best performed under fluoroscopic guidance. It is not effective for microcystic components. Thus surgical excision is warranted for many microcystic and mixed lesions.

Excision may result in a number of potential complications. Prolonged drainage is virtually assured, and families should be informed that surgical drains will remain for weeks to months. Re-enlargement of residual lesion is not uncommon if an anatomic region is not adequately resected. Vesicle formation in and around surgical scars may occur if the malformation involves the dermis of the remaining lesion. Malformations tend to invest normal structures. Thus injury to the thyroid, parathyroids, main and accessory thoracic ducts may occur. In particular, the vagus, phrenic, recurrent laryngeal, accessory, transverse cervical, and marginal mandibular nerves are at risk. A Horner's syndrome may result from stellate ganglion injury. Great care must be taken to avoid injury to the brachial plexus. Bleeding should be minimal, with blood transfusion rarely necessary. Mortality is not expected.

PREOPERATIVE ASSESSMENT AND PREPARATION

Physical examination

1 Physical examination is usually sufficient to make a diagnosis. The posterior triangle of the neck is the most common location. The lesion is usually ballotable and may transilluminate.



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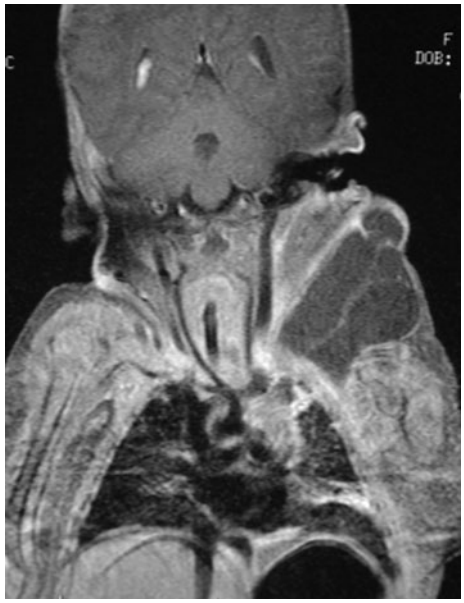

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Imaging

2 Ultrasound can differentiate macrocystic from microcystic components. Magnetic resonance imaging (MRI) is most useful in establishing the anatomic extent of a lesion and its relationship to vital structures.

Timing and extent

Unless there is actual or threatened airway impairment, resection can be scheduled at 3–6 months of age. On rare occasions, a lesion may deflate to become undetectable during this interval. Very extensive lesions often require staged procedures. An anatomic region with specific defined borders should be chosen and dissected completely in each procedure to minimize the necessity to return to a scarred, incompletely resected area. For example, a cervical lesion extending into the axilla should be resected down to the brachial plexus, with the intent to complete the infra-plexus component on a separate occasion. Facial extension is often best left until children are older, as they will often 'grow into' their lesion to achieve a better contour than might be achieved surgically. Bilateral lesions are generally best handled one side at a time. Since these lesions are histologically benign, the goal of resection is to be '99 percent' complete within the chosen region, while preserving all vital neurovascular structures. In order to achieve this degree of extirpation while preserving all vital

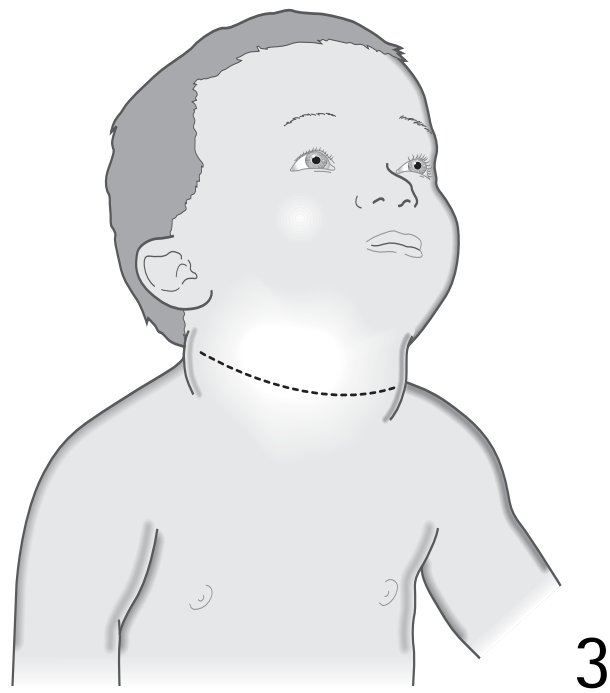
structures, a lengthy, meticulous dissection may be required for extensive lesions. It is crucial to avoid letting the clock determine the pace or endpoints of dissection. An inadequate resection will undoubtedly lead to a much more difficult secondary operation, with greater risk of morbidity.

Laboratory studies

Hematologic and chemistry studies are not crucial. A specimen for blood crossmatch should be sent to the blood bank, although transfusion should not often be necessary.

Anesthesia

General anesthesia with endotracheal intubation is mandatory. Pharmacologic paralysis should be avoided to permit nerve stimulation during dissection. Direct laryngoscopy prior to incision may be elected to confirm preoperative vocal cord function.



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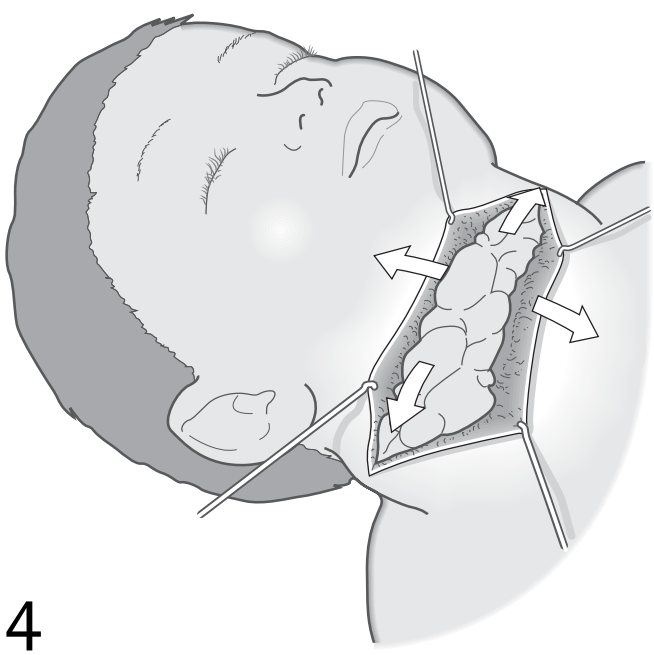
OPERATION

Position and incision

3 The patient is supine with a roll under the shoulders to extend the neck. The head is turned away from the lesion. The exposure is through a supraclavicular incision parallel to the clavicle.



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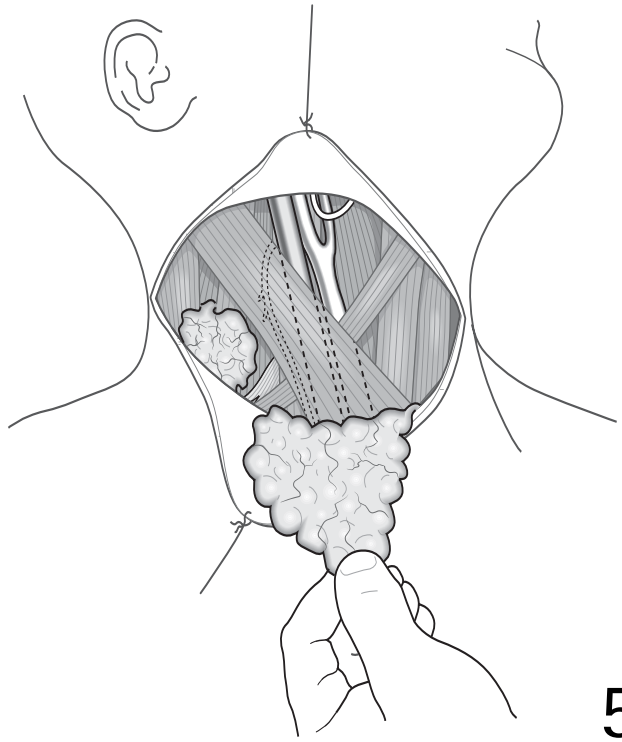
Exposure

4 Superior and inferior flaps are created deep to the platysma, extending superiorly, medially, and laterally to the extent of the superficial aspect of the malformation.

Deep posterior dissection

5 The malformation is meticulously dissected from the underlying and adjacent strap muscles, neurovascular structures, thryroid, trachea, and sometimes esophagus. The vagus, recurrent laryngeal, and phrenic nerves should be identified and preserved, even if the malformation tissue must be split to free these structures. Successful dissection is facilitated by full cysts, so an attempt should be made to avoid deflating macrocystic components for as long as possible.



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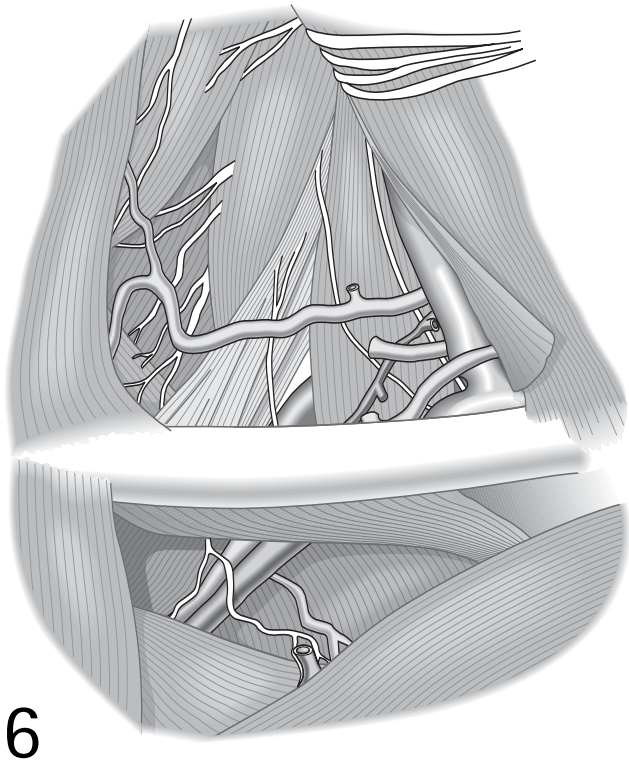

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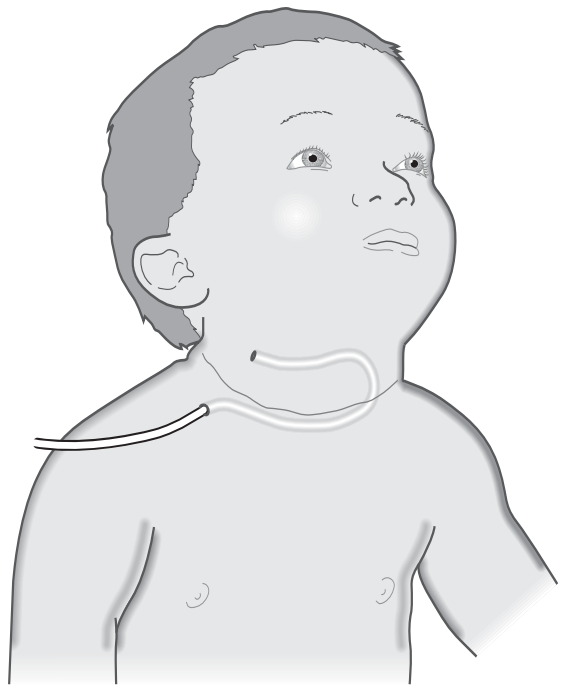
Root of neck

6 After deep neck structures have been identified, the malformation is dissected off the underlying brachial plexus. Care must be taken to avoid injury to the subclavian vessels and the thoracic duct (left) or accessory thoracic duct (right). If the lesion extends into the mediastinum, the malformation may be lifted superiorly out of the thoracic inlet, facilitating at least partial separation from the thymus and removal. Sternotomy need not be performed in the absence of symptoms referable to the intrathoracic component.

Closure

7 Excess skin is resected only if prominently redundant after removal of the underlying malformation tissue. A closed suction drain is placed in the wound and brought out through a reasonably long subcutaneous tunnel to avoid leakage around the drain. The platysma and subcutaneous tissues are closed with running braided, absorbable suture. This skin is closed with a running subcuticular closure.



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POSTOPERATIVE CARE

Long procedures with extensive dissections may require a period of postoperative intubation. Pain management is generally straightforward, as muscular disruption is minimal. An oral diet may be resumed as soon as tolerated. Closed suction drains may be required for weeks after hospital discharge. Parents should be taught to empty and record drain output. Postoperative infection is not uncommon and generally requires intravenous antibiotics.

OUTCOME

Although re-enlargement occasionally occurs, satisfaction is generally achieved. Contour is not always completely symmetric with the uninvolved side. Neurovascular injury can occur, but should be uncommon.

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11

Preauricular sinus

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PRINCIPLES AND JUSTIFICATION

The preauricular sinus is a congenital sinus whose opening is situated on the anterior aspect of the helix of the pinna. It results from ectodermal inclusions that arise during fusion of the six fetal tubercles that form the pinna. The course of the sinus is unpredictable and often consists of multiple branches. It passes anteriorly and inferiorly through the subcutaneous tissue to end in a racemose group of preauricular cysts and, contrary to some reports, frequently extends deep to the subcutaneous tissue to be attached to the cartilage of the pinna. The sinus is often familial, and may be unilateral or bilateral. In many cases the preauricular sinus is completely asymptomatic and requires no treatment. However, it may present with an intermittent discharge or become infected, or as a preauricular abscess. Total excision of the sinus and the underlying group of preauricular cysts is essential for cure of the symptomatic sinus and this may also require excision of a small amount of underlying cartilage at the point of attachment. Incomplete excision results in a significant recurrence rate.

PREOPERATIVE ASSESSMENT AND PREPARATION

Definitive surgery should not be undertaken in the presence of active infection, which should be treated with anti-staphylococcal antibiotics. In the case of abscess formation, this requires either aspiration or excision and drainage. At the time of formal excision of the sinus, prophylactic antibiotics should be administered in those cases that have had previous episodes of infections.

Investigations

The decision about treatment is made on clinical grounds. No further investigations are routinely required preoperatively.

Consent

Informed consent must be obtained from one of the child's parents or the child's legal guardian. The complications of bleeding, infection, recurrence, and hypertrophic scarring must be outlined as part of this process.

Anesthesia

General anesthesia is administered by an endotracheal tube. Infiltration of the wound with local anesthetic agents often distorts the dissection plane if done preoperatively, and this is therefore best reserved for the end of the procedure. The patient is placed supine on the operating table and the patient's head positioned on a head ring.

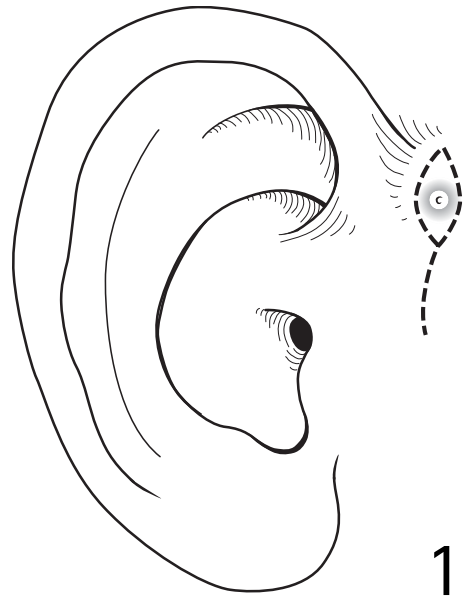
OPERATION

Skin preparation and draping

Any hair anterior to the pinna should be shaved. The skin is prepared with either aqueous betadine or aqueous chlorhexidine. If bilateral sinuses are present, a head towel is used to maintain a sterile field. In the case of a unilateral sinus, a standard four-drape technique can be used. Although injection of methylene blue has been advocated as a method for identifying the ramifications of the sinus, in practice this does not prove helpful.

Incision

1 An elliptical incision is made around the punctum. The ellipse should also include any scarring that has resulted from previous infection. The incision is extended inferiorly in a vertical plane immediately in front of the pinna. An alternative incision is an inverted L-shape with an associated skin flap.



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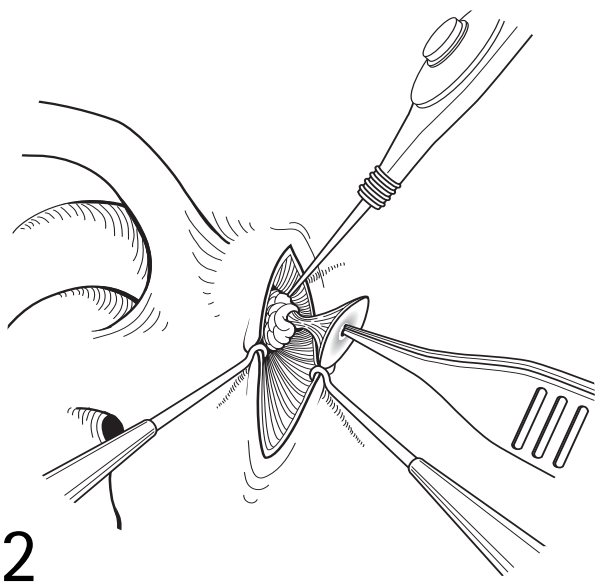

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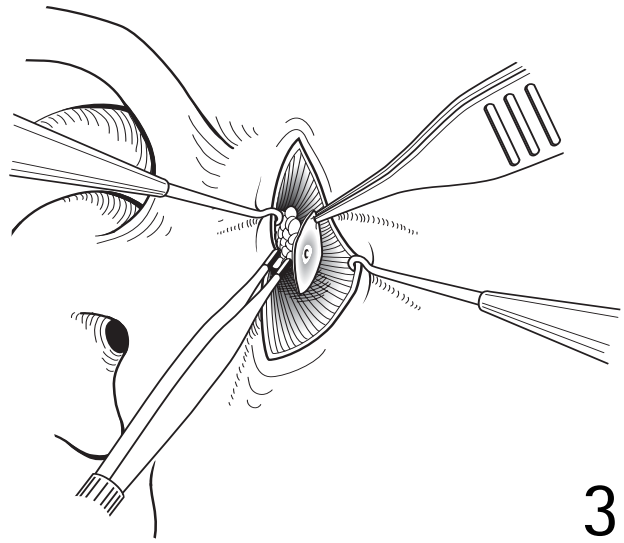
Dissection and excision

2 The incision is deepened using a fine monopolar diathermy point. The ellipse of skin containing the punctum and underlying sinus is then held using either toothed Adson forceps or a mosquito hemostat. Dissection is continued and the underlying group of preauricular cysts identified. Sometimes insertion of a lacrimal probe is helpful to identify the course of the sinus.

3 Further dissection is performed using bipolar diathermy forceps, ensuring that the skin ellipse, sinus, and cyst are dissected en bloc. As stated above, the deep attachment of the sinus is often lying on the cartilage, and excision of a small amount of cartilage at the point of attachment is often required to prevent recurrence. During deep dissection, particular care is taken to preserve the superficial temporal artery and the preauricular nerve. The sinus together with the collection of cysts is then removed and sent for routine histopathology. Meticulous hemostasis is ensured throughout.



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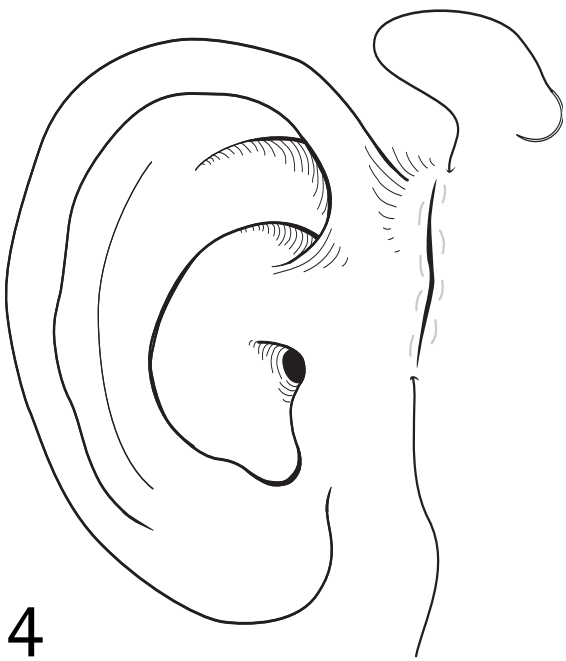
Closure

Once hemostasis has been confirmed, interrupted 5/0 or 4/0 polyglycolic sutures are used to close any defect in the cartilage. Drainage of the wound is not routinely required. Bupivacaine 0.25 percent is infiltrated into the wound to ensure postoperative analgesia. Interrupted 4/0 polyglycolic sutures are then placed in the subcutaneous tissue, ensuring that the knots are buried.



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4 The skin is closed using either a subcuticular 5/0 undyed polyglycolic suture or, for a more predictable cosmetic result, interrupted 6/0 nylon sutures, although the latter can only be used in those infants for whom removal of sutures is practical. A Steristrip dressing is applied along the wound together with a small gauze/Mefix pressure dressing to prevent hematoma formation, and thereby optimize the cosmetic result.

POSTOPERATIVE CARE

Excision of a unilateral sinus is usually performed as a day case. The pressure dressing is maintained for 48 hours after the operation and the wound kept dry during this time. If interrupted non-absorbable sutures have been used for skin closure, these should be removed 5 days postoperatively to prevent a tissue reaction to them.

OUTCOME

Postoperative hematoma and infection can occur, but are minimized by the techniques described above. Incomplete excision may result in recurrence of the sinus. This risk is increased in those cases in which recurrent infection has preceded excision, as the dissection planes are often less clearly defined. Using the elliptical incision described above, the cosmetic result is usually good, although hypertrophic scarring may result, especially if the wound becomes infected or the child is of Afro-Caribbean descent.

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SECTION III

Thoracic

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12

Thoracic surgery: general principles of access

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PRINCIPLES AND JUSTIFICATION

The chest of the infant is relatively short in its longitudinal axis, so a posterolateral incision entering the chest through the fifth intercostal space will provide adequate exposure for most procedures. The compliant ribs can be widely retracted and it is not necessary to remove or transect a rib. Care must be taken to avoid fracturing the ribs with the rib spreader by ensuring that they are adequately mobilized anteriorly and posteriorly. The length of the incision and the choice of rib space to be entered will depend on the procedure to be performed. For esophageal atresia, a higher approach through the fourth or third space is usually best, while an approach below the fifth space is required for access to the diaphragm (bearing in mind that in the infant the liver is relatively large and elevates the right hemidiaphragm) or to the distal esophagus on the left. The axillary approach through a short cosmetic incision in an axillary skin crease provides access to the upper thorax without dividing major muscles. The posterior incision, which curves upwards around the scapula, is rarely required.

A median sternotomy is used for access to the mediastinum or to explore both lungs simultaneously.

OPERATIONS

Anesthesia

General anesthesia with endotracheal intubation is adequate for most procedures. For certain operations on the lung or airway, selective intubation of the opposite lung may be required.

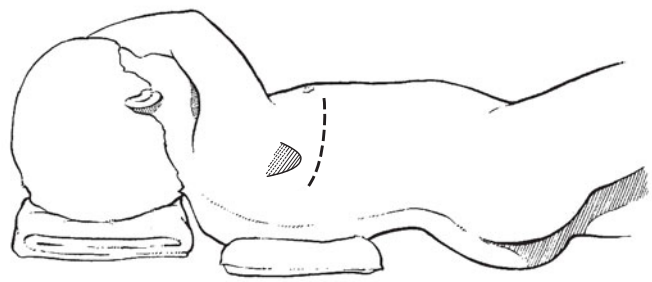
Posterolateral thoracotomy

POSITION OF PATIENT AND INCISION

1 The child lies in the lateral position. The arm on the side of the incision lies forwards and upwards over the face on an armrest or padding. A bolster placed under the chest approximately in the line of the nipples may aid the exposure. The child is firmly stabilized in position with sandbags or pads at the front and back of the chest and pelvis and adhesive strapping attached across the hips to the operating table and similarly across the uppermost arm and shoulder.

The skin is widely cleansed with antiseptic solution. Sterile towels are placed to keep the nipple, lower scapula, spine, and costal margin visible as landmarks. A large adhesive plastic sheet is applied to stabilize the towels and reduce heat loss. The lower chest wall is included in the operative field for later chest drain placement if required.

The incision starts from the anterior axillary fold on a line inferior to the nipple, and extends posteriorly below the inferior tip of the scapula, the position of which is marked on the skin before the arm is elevated. From here the incision curves gently cranially as far as the erector spinae muscle.



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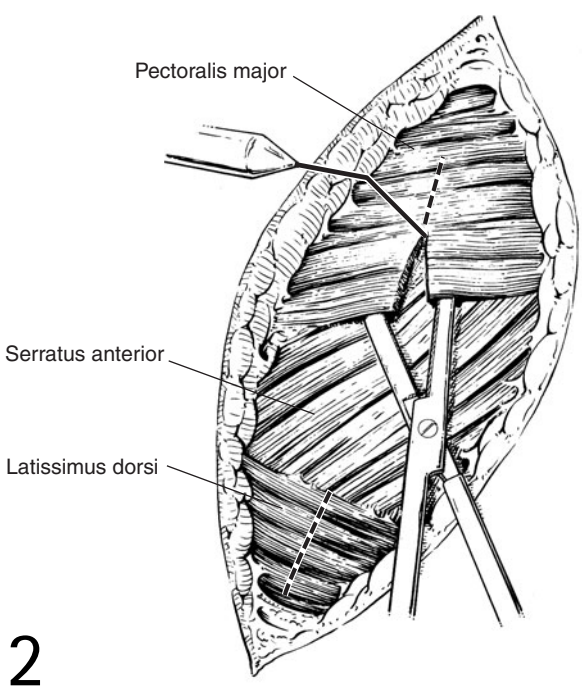

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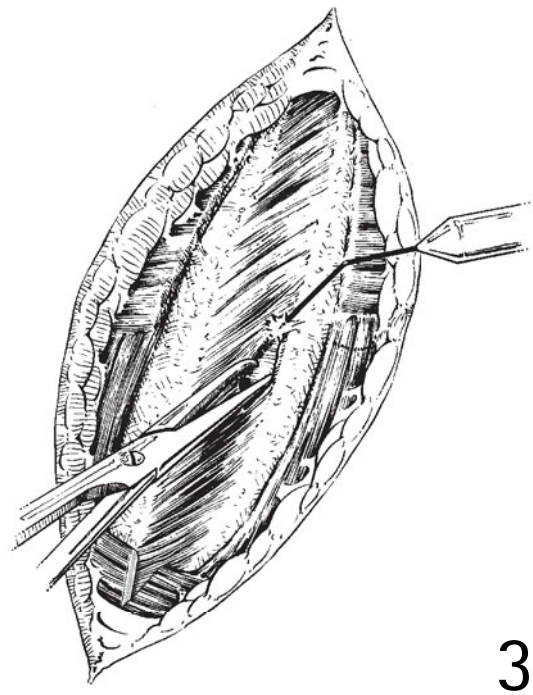
2 The subcutaneous tissues are divided, followed by the pectoralis major and latissimus dorsi muscles, and, on a deeper plane, the serratus anterior muscle. Artery forceps are used to elevate each muscle while it is being cut with the diathermy. Bleeding vessels are accurately coagulated using fine-toothed forceps. The long thoracic nerve runs near the anterior border of the serratus anterior muscle and must be protected; should it be necessary to divide the nerve, this should be done as far caudally as possible to minimize the effect on the function of the muscle. The scapula is then elevated with a retractor and the ribs are palpated and counted; in an infant, the highest that can be palpated usually is the second rib. The ribs above and below the target rib space are marked with the diathermy.

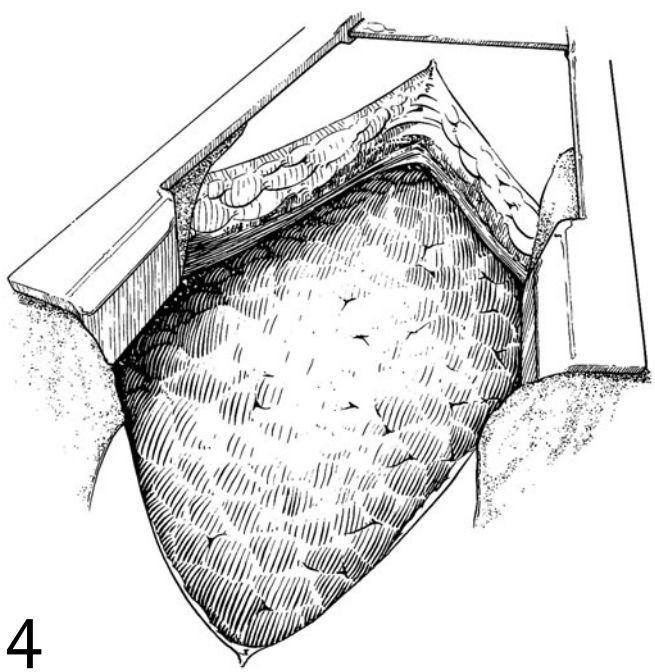
OPENING THE CHEST: INTERCOSTAL TRANSPLEURAL APPROACH

3 The intercostal muscles are divided with the diathermy, keeping to the upper border of the lower rib to avoid the neurovascular bundle. A short incision is made initially, using artery forceps to spread the muscle, and, with the lungs deflated by the anesthetist, the pleural cavity is entered.



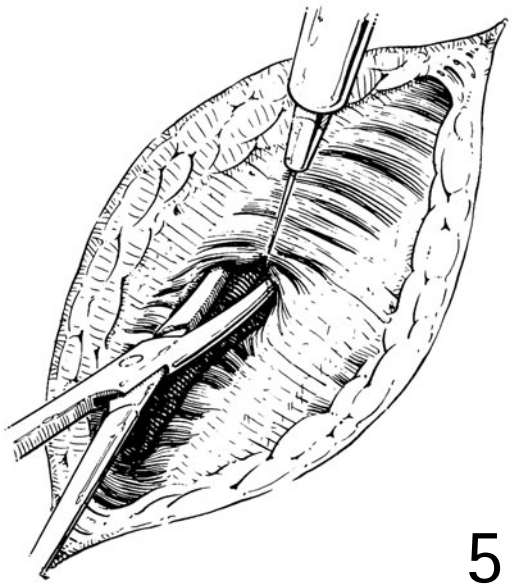
3

The remaining intercostal muscles are divided with the diathermy, using a peanut swab or retractor to protect the lung.



4

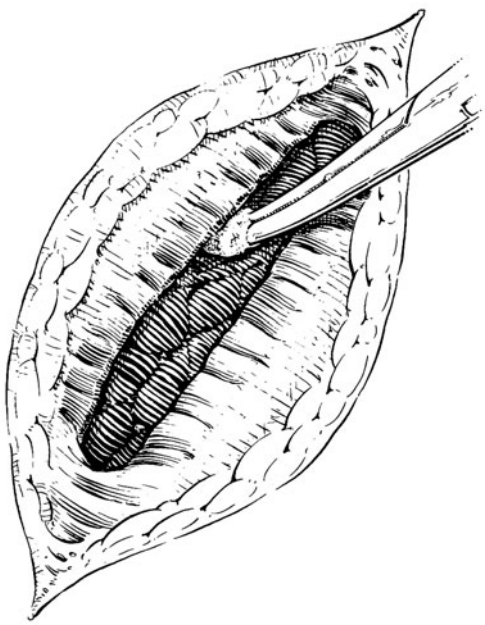
4 Rib retractor (such as the Finochietto retractor for an infant) is inserted and opened slowly while dividing the intercostal muscles anteriorly and posteriorly. Care must be taken to avoid excessive pressure from the retractor as this may cause a rib fracture, particularly posteriorly. The chest wall muscles and subcutaneous tissues may also need to be incised further to ease the tension on the rib.



5

OPENING THE CHEST: INTERCOSTAL EXTRAPLEURAL APPROACH

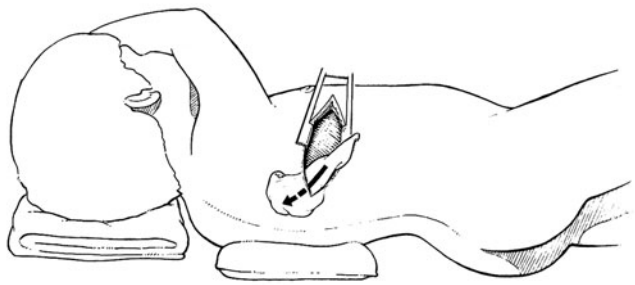
5 Division of the intercostal muscles begins as shown in Figure 3, using artery forceps to enter the extrapleural space in the posterior half of the incision and diathermy to incise the muscle. Great care must be taken not to open the parietal pleura.



6

6 A moist peanut swab is used carefully to extend the dissection in the extrapleural plane. Anteriorly, the parietal pleura are adherent to the ribs and easily torn, and the dissection must proceed in a posterior direction.

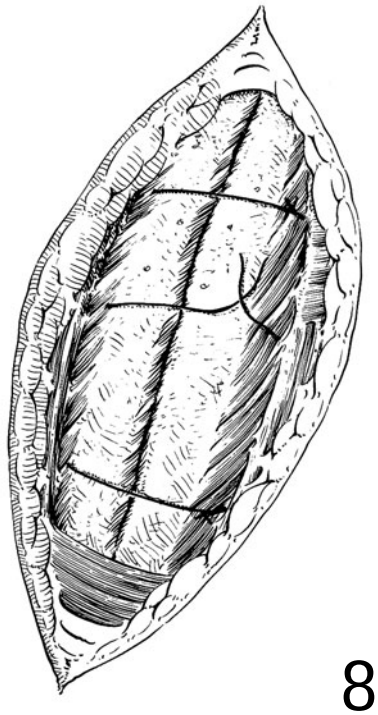
7 A moist gauze swab is unfolded and gently packed into the extrapleural space posteriorly and cranially in order to further strip the pleura from the chest wall. The swab is removed, a small rib retractor is inserted, and the extrapleural dissection to the posterior mediastinum is completed under direct vision.



7

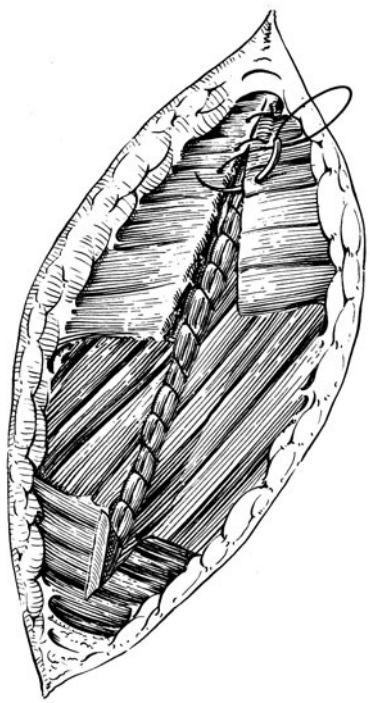
WOUND CLOSURE

Before closing the chest, a long-acting local anesthetic is infiltrated around the intercostal nerves as they lie near the neck of the ribs above and below the level of the thoracotomy to enhance postoperative analgesia. The margins of the skin incision are infiltrated prior to skin closure, as is the site of exit of the chest drain. If a chest drain is required it is inserted before closing the chest through a short transverse incision in the mid-axillary line, two intercostal spaces below the thoracotomy incision. It should not be placed posteriorly where the patient will lie on it. The drain is sutured to the skin and connected to an underwater seal.



8

8 Three or four pericostal sutures are passed around the ribs above and below the incision, taking care to avoid the intercostal vessels, and firmly tied to approximate the ribs. In the neonate, an absorbable suture is used and the ribs must be approximated gently. Tight apposition of these pliable ribs may result in them fusing together; this may be avoided by placing an instrument (e.g., artery forceps) between the ribs while the suture is tied. If possible, the intercostal muscles are repaired, but this is not essential.

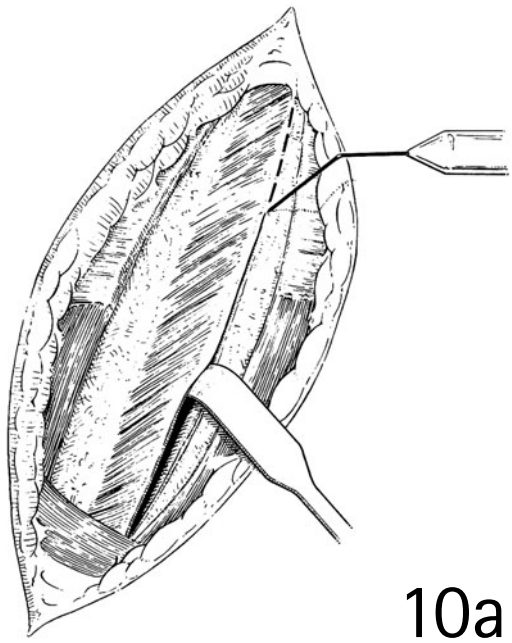


9

9 The chest wall muscles and subcutaneous tissues are repaired in anatomic layers using continuous absorbable sutures; accurate closure enhances the functional and cosmetic outcomes. In particular, the edges of the divided muscles must be aligned accurately. The skin is closed with an absorbable subcuticular suture or adhesive strips.

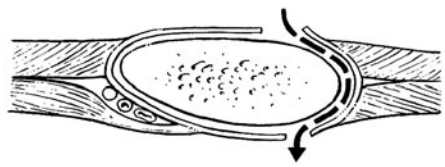
OPENING THE CHEST: SUBPERIOSTEAL APPROACH

This approach, which may be used even in infants, preserves the intercostal muscles.

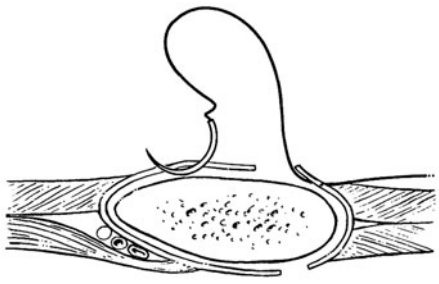


10a

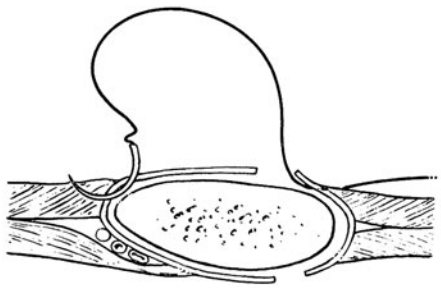
10a,b On the rib below the intercostal space to be entered, the periosteum is incised longitudinally using the diathermy. The periosteum is elevated in an anterior direction off the upper border of the rib using a curved raspatory. This exposes the posterior periosteum, which is incised to enter the extrapleural space (see Figures 6 and 7) or the pleural cavity.



10b



11a



11b

WOUND CLOSURE

11a,b To close the chest, pericostal sutures are inserted and the ribs approximated, avoiding undue tension in the neonate (see Figure 8). The cranial edge of the incised periosteum and intercostal muscle is sutured using an absorbable suture to the periosteum on the rib or to the periosteum and intercostal muscles at the lower edge of the rib, avoiding the neurovascular bundle.

POSTOPERATIVE CARE

Effective analgesia is essential for the patient's comfort and to facilitate early postoperative physiotherapy and mobilization, which will reduce the risk of atelectasis and pulmonary infection. The chest drain is painful and should be removed as soon as any pneumothorax has resolved and fluid drainage is minimal.

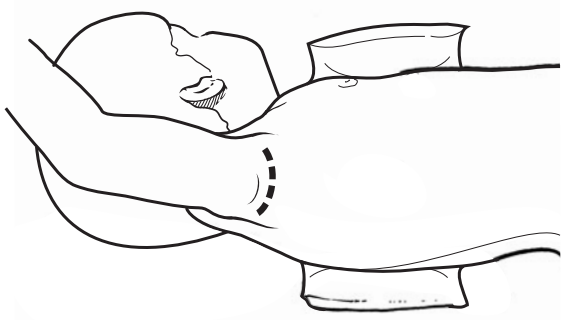
COMPLICATIONS

Early complications, which include bleeding from damaged intercostal vessels and wound infection, are not common. Chest tube displacement will occur if the tube is not securely fixed to the chest wall by strong adhesive tape. Damage to the nerve supply to the serratus anterior muscle results in winging of the scapula. If the incision crosses the inferior angle of the scapula, adhesions to the scapula may develop, leading to restricted shoulder movement and an unsightly scar. In the infant there may be abnormal development of the chest wall; in some patients this is associated with fusion of adjacent ribs at the site of the thoracotomy. Scoliosis of the thoracic spine may develop as the infant grows, and follow-up should include examination of the spine.

Lateral thoracotomy

AXILLARY (MUSCLE-SPARING) APPROACH

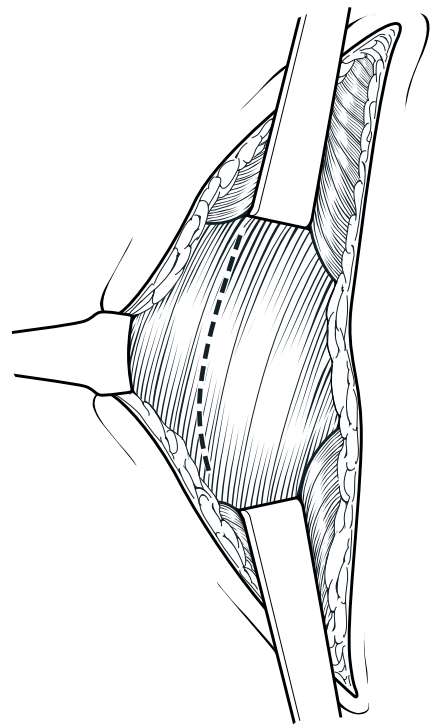
This approach provides access to the upper thoracic cavity and posterior mediastinum using a cosmetic, muscle-sparing incision.



12

12 With the infant in the lateral position, the incision is made in a high axillary skin crease, extending from the anterior axillary fold formed by the pectoralis muscles to the posterior axillary fold formed by the latissimus dorsi muscle. A vertical incision in the mid-axillary line has also been described; although this retracts up into the axilla with time, in some patients the scar is irregular and unsightly.

13 The axillary fatty tissue and lymphatics are retracted cranially and the pectoralis and latissimus dorsi muscles are retracted to expose the serratus anterior muscle, which is split along the line of its fibers, preserving the long thoracic nerve and adjacent vessels lying along its anterior border. Alternatively, the serratus anterior may be mobilized posteriorly and retracted anteriorly to preserve the long thoracic nerve. The thoracic cavity is entered through the third or fourth intercostal space and extrapleural or transpleural dissection proceeds as described above.

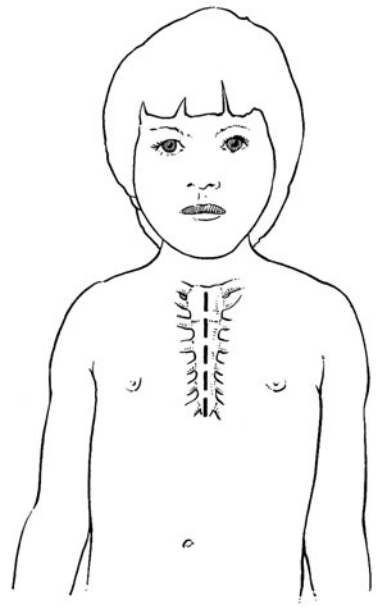


13

Median sternotomy

POSITION OF THE PATIENT AND INCISION

14 The child lies supine, with a bolster placed under the upper thorax to hyperextend the thoracic spine and bring the sternum forward. Slight elevation of the head of the operating table will reduce venous congestion. The skin is incised from just below the suprasternal notch to the xiphoid cartilage. To minimize blood loss, diathermy is used to incise the subcutaneous tissues and periosteum.



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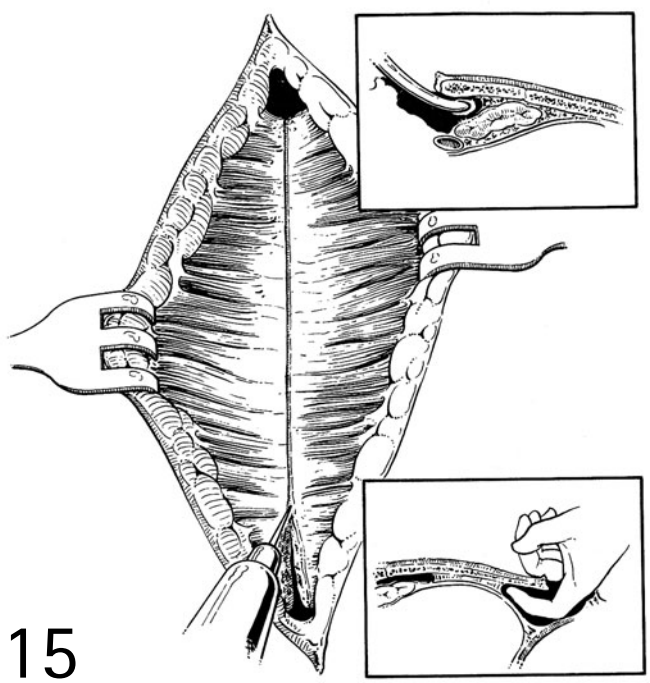


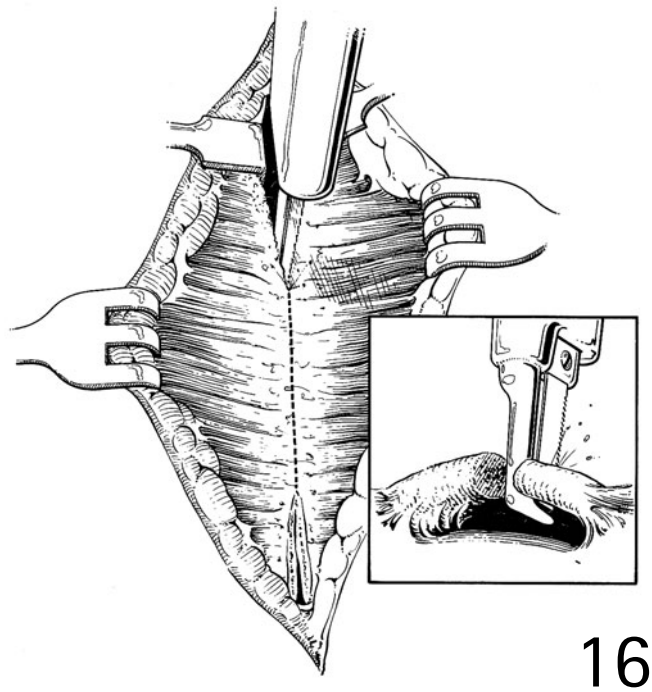
15

15 The subcutaneous tissues over the sternum are divided with diathermy. In the suprasternal notch the soft tissues are dissected off the superior and posterior aspects of the manubrium with sharp and blunt dissection. Inferiorly, the xiphoid is mobilized and divided vertically with heavy scissors or diathermy. It is possible now to enter the retrosternal plane cranially and caudally, using a peanut swab or finger to free the mediastinal structures from the back of the sternum.

DIVIDING THE STERNUM

16 The sternum is divided vertically in the midline using a suitable bone saw. In the infant, shears or sturdy scissors are adequate. Ventilation should be suspended while this is done to avoid opening the pleura. Bleeding, which is usually from the periosteum or bone marrow, is controlled with diathermy and bone wax.

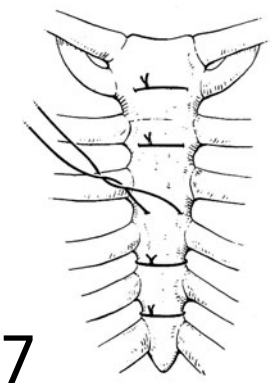
A self-retaining retractor is inserted and opened slowly while dissecting the soft tissues off the inner aspect of the ribs on either side, taking care not to tear the pleura. In the infant it may be necessary to mobilize and retract or partially excise the thymus gland; this is done by sharp and blunt dissection from below cranially, ligating larger vessels. Laterally, care must be taken not to damage the phrenic nerves.



16

WOUND CLOSURE

Whether or not the pleural cavity has been opened, one or two suction drainage catheters should be placed in the retrosternal space and brought out through stab incisions below and lateral to the xiphoid process, and sutured to the skin.



17

17 The two halves of the sternum are apposed with several strong sutures (polydioxanone in infants, nylon or wire in the older child) placed around or through the sternum and firmly tied. The subcutaneous tissues are closed with a continuous absorbable suture. The skin is approximated with a fine absorbable subcuticular suture. Prior to skin closure, the margins of the incision are infiltrated with local anesthetic.

POSTOPERATIVE CARE

Systemic analgesia is essential to ensure early mobilization. The drains are removed after 24 hours if there is no drainage or clinically significant pneumomediastinum.

FURTHER READING

Bianchi A, Sowande O, Alizai NK, Rampersad B. Aesthetics and lateral thoractomy in the neonate. *Journal of Pediatric Surgery* 1998; **33**: 1798–800.

Singh SJ, Kapila L. Denis Browne's thoracotomy revised. *Pediatric Surgery International* 2002; **18**: 90–2.

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13

Esophageal atresia with and without tracheoesophageal fistula

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HISTORY

The first description of esophageal atresia is credited to Durston who, in 1670, described esophageal atresia in one of a pair of conjoined twins. Thomas Gibson in 1697 accurately described the clinical features of esophageal atresia. In 1913, Richter proposed a plan of management, which comprised dividing the tracheoesophageal fistula (TEF) and feeding the infant by gastrostomy until the 'technical difficulties of an esophageal anastomosis' had been overcome. Ladd and Leven were independently the first to achieve long-term survival in 1939, but only by a staged approach. Haight in 1941 is credited with the first successful primary anastomosis.

PRINCIPLES AND JUSTIFICATION

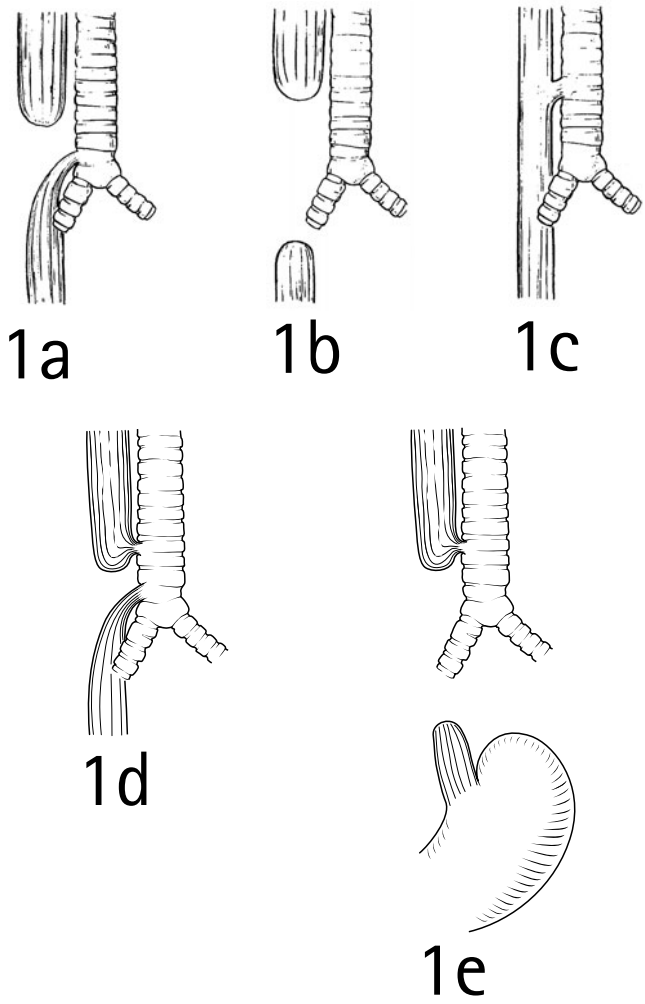
Embryology

The respiratory primordium first appears as a ventral diverticulum of the foregut at the 22nd to 23rd day of gestation. This is followed by a period of rapid growth when the ventrally placed trachea separates from the dorsally placed esophagus. One theory postulates that the trachea becomes a separate organ as a result of rapid longitudinal growth of the respiratory primordium away from the foregut. An alternative theory is that the trachea initially grows as part of an undivided foregut and then becomes a separate structure as a result of a separation process that starts at the level of the lung buds and proceeds in a cranial direction. This process is associated with a precise temporospatial pattern of expression of the developmental gene sonic hedgehog (*Shh*). The primary defect resulting in esophageal atresia is persistence of an undivided foregut either as a result of failure of tracheal growth or of failure of the already specified trachea to separate physically from the esophagus.

Classification of esophageal atresia

1a–e The various types of esophageal atresia are:

a esophageal atresia with distal fistula (85 percent);
b esophageal atresia without fistula (7 percent);
c H-type TEF (4 percent);
d esophageal atresia with proximal and distal fistula (3 percent);
e esophageal atresia with proximal fistula (1 percent).



Associated anomalies

At least 50 percent of infants with esophageal atresia have one or more associated anomalies (Box 13.1). Most common are cardiac malformations, particularly ventricular septal defects and tetralogy of Fallot, and these are responsible for the majority of deaths. Next most common are gastrointestinal and anorectal anomalies, followed by genitourinary tract abnormalities. The VACTERL association (V = vertebral, A = anorectal, C = cardiac, T = tracheo, E = esophageal, R = renal, L = limb) is a well-known combination of defects.

A full clinical examination should be made at the outset for associated anomalies, and special investigations, including echocardiography and renal ultrasonography, are carried out as indicated.

Box 13.1 *Anomalies associated with esophageal atresia*

Cardiovascular	29%
Anorectal	14%
Genitourinary	14%
Gastrointestinal	13%
Vertebral/skeletal	10%
Respiratory	6%
Genetic	4%
Other	11%

Prognosis

In 1962, Waterston proposed a risk classification for infants with esophageal atresia based on his experience in the management of 218 infants as follows.

- *Group A* (95 percent survival): birth weight over 5.5 lb and well.
- *Group B* (68 percent survival):
  - birth weight 4-5.5 lb and well,
  - higher birth weight, moderate pneumonia, and congenital anomaly.
- *Group C* (6 percent survival):
  - birth weight under 4 lb,
  - higher birth weight, severe pneumonia and severe congenital anomaly.

An amended classification more relevant to modern pediatric surgical practice based on the experience of treating 372 infants between 1980–1992 is shown in Table 13.1.

Table 13.1 *Prognosis based on 372 patients treated for esophageal atresia 1980–1992*

		Survival (%)
Group I	Birth weight > 1500 g and no major cardiac anomaly	97
Group II	Birth weight < 1500 g *or* major cardiac anomaly	59
Group III	Birth weight < 1500 g *plus* major cardiac anomaly	22

Diagnosis

ANTENATAL

Polyhydramnios is present in 95 percent of patients with isolated atresia without fistula and in 35 percent of cases with a distal fistula.

With expert fetal ultrasonography it is possible to visualize the blind upper esophageal pouch filling and emptying, and an inability to detect the stomach would suggest atresia without fistula.

POSTNATAL

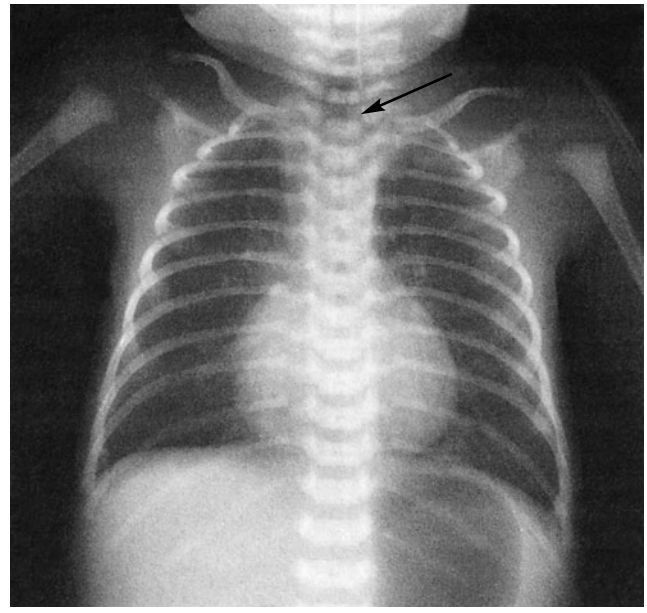
A nasogastric tube (8–10 gauge) should be passed at birth in all cases where polyhydramnios was present during pregnancy. Failure to advance the tube beyond 10 cm from the nose or mouth indicates esophageal atresia.

Symptoms that develop in the neonatal period include: inability to clear secretions from the mouth, cyanotic episodes with or without attempting to feed the infant, inability to swallow, and respiratory distress.

PREOPERATIVE

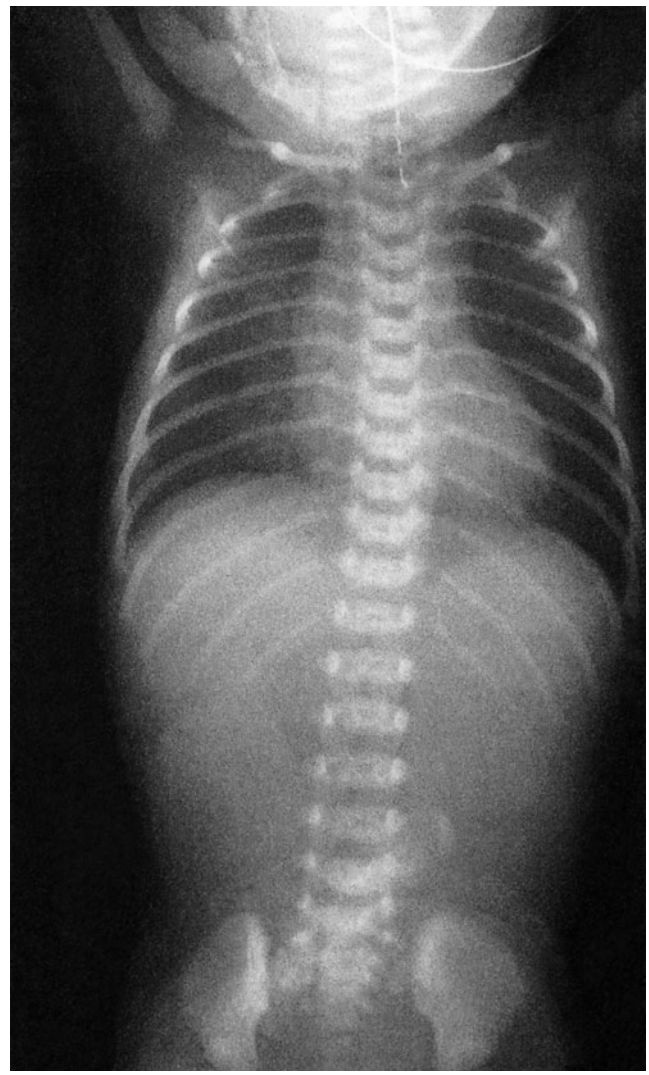
Investigations

2 Plain radiography of chest and abdomen demonstrates a radio-opaque nasogastric tube (Replogle type) arrested in the upper mediastinum, and air in the stomach and intestine confirms the presence of a distal TEF.



2

3 The absence of intestinal air suggests the diagnosis of pure esophageal atresia. Contrast studies of the upper pouch are seldom indicated. Endoscopic examination of the upper esophagus and/or bronchoscopy immediately before surgery will detect an upper pouch fistula in most cases. If contrast medium is used, the examination should be performed with extreme care by an experienced radiologist.



3

Initial management

Immediate surgery for esophageal atresia is seldom necessary. Scheduling the operation 12–24 hours after admission allows for full assessment for associated anomalies and resolution of pulmonary atelectasis if aspiration has occurred prior to referral. Neonates with respiratory distress requiring assisted ventilation, particularly if associated with gastric distension, should undergo emergency transpleural ligation of the distal fistula. This will immediately improve the respiratory status, and gas exchange in the lungs will improve as the escape of gas through the fistula is halted. In some infants the improvement is so dramatic as to allow primary repair of the atresia to proceed. In others, the procedure is terminated pending improvement in the infant's condition. The repair can be safely postponed for up to 7 days; further delay increases the risk of the fistula reopening. Gastrostomy as a primary procedure in these high-risk neonates is to be avoided.

While awaiting surgery, the upper pouch is continuously aspirated using a Replogle tube attached to low-pressure suction. Intravenous fluids (10 percent dextrose in 0.18 percent saline) will maintain fluid and electrolyte balance and prevent hypoglycemia. Preoperatively, a vitamin K analog should be routinely administered intramuscularly.

Choice of operation

In most neonates with esophageal atresia and a distal TEF, division of the fistula and primary anastomosis of the esophagus are possible. The anastomosis should be attempted, even if performed under extreme tension. It is important to recognize the presence of a right-sided aortic arch, best identified on an echocardiogram, as in these cases a left-sided thoracotomy may provide easier access to the mediastinum.

A short upper pouch on the preliminary plain radiograph

may also indicate that a primary anastomosis may be difficult. The presence of a distal TEF requiring division dictates the necessity for right thoracotomy, and the possibility of obtaining a satisfactory primary anastomosis should not be ruled out until the anatomy has been inspected at the time of the thoracotomy.

The absence of a lower pouch fistula is usually associated with a long gap between the upper and lower esophagus. This situation is usually managed by a preliminary feeding gastrostomy. If esophageal replacement is the procedure of choice, a cervical esophagostomy is necessary. The alternative is a delayed primary anastomosis after several weeks of gastrostomy feeding and upper pouch suction. The decision about when to attempt the delayed primary anastomosis is based on radiological assessment of the intervening gap. With a Replogle tube in the proximal esophagus and a urethral dilator introduced into the distal esophagus through the gastrostomy stoma under fluoroscopic control, the size of the gap between the upper and lower esophagus is measured. If the gap measures less than the width of two vertebral body, primary anastomosis should be attempted. A gap greater than six vertebra indicates the need for an esophageal replacement. In general, it is not profitable to wait longer than 8–12 weeks before deciding how to proceed.

PREOPERATIVE

The Replogle tube or a similar large-bore tube should be in position in the upper pouch. Careful attention is paid to maintaining body temperature with a heating blanket, and to preventing heat loss by covering the infant with foil. Broad-spectrum antibiotics should be administered either preoperatively or at the time of induction.

Anesthesia

Premedication is with atropine alone. The endotracheal tube requires careful positioning to permit adequate ventilation with minimal gas flow through the fistula. The majority of pediatric anesthetists will control ventilation from an early stage following intubation. An intravenous infusion is sited in a limb other than the right upper limb.

Preliminary endoscopy

Many surgeons advocate preliminary bronchoscopy to define the site of entry of the distal TEF, to exclude the presence of a proximal fistula, and to assess the degree of tracheomalacia. A distal TEF entering the trachea at the level of the carina may indicate a 'wide-gap' atresia. Esophagoscopy will define the length of the upper pouch and exclude an upper pouch fistula (more common in isolated atresia) that enters the side of the proximal pouch at some distance from its distal end.

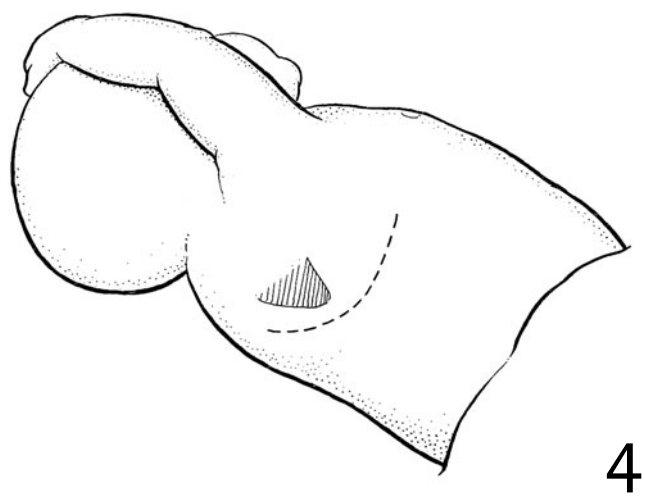
OPERATION

Classic repair

INCISION

4 The infant is positioned on the left side and stabilized with strapping or sandbags. The right arm is extended above the head and fixed. Care must be taken to ensure that the neck is flexed. A curved incision is made 1 cm below the inferior angle of the scapula extending from the mid-axillary line to the paravertebral region posteriorly.

Division of the subcutaneous tissues and muscles is carried out with diathermy to minimize blood loss. The latissimus dorsi muscle is divided along the length of the incision. Serratus anterior is retracted forwards, but if additional exposure is required, the lowermost fibers only are divided in order to preserve its nerve supply. Alternatively, access to the intercostal spaces may be via the angle of auscultation whose borders are trapezius above, latissimus dorsi below, and the medial border of the scapula laterally. Following division of the muscles, the scapula is elevated and the rib spaces are counted by palpation.



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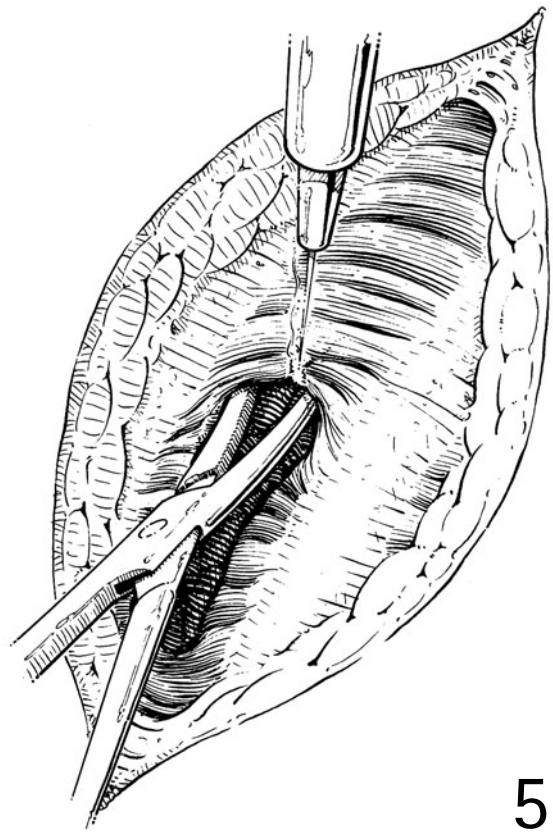
5 The thorax is entered through the fourth or fifth intercostal space by carefully dividing the external and then the internal intercostal muscles. The muscle fibers are gently elevated from the underlying parietal pleura and divided using diathermy.



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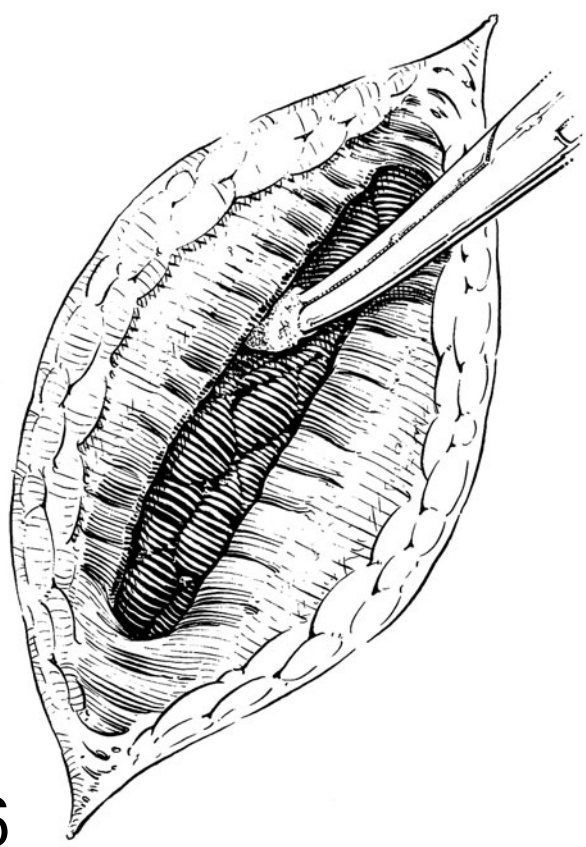
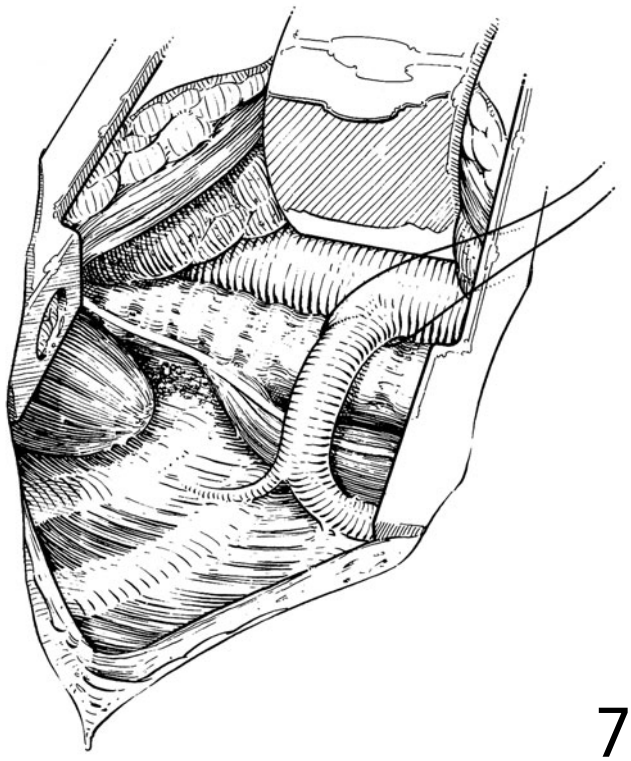


6

6 Having exposed the pleura through the intercostal space, stripping of the pleura from the chest wall is best carried out by the gentle insertion of a gauze swab into the extrapleural space. On withdrawing the swab, an extensive area of dissection will have resulted; a rib spreader can then be inserted and the ribs gently separated. Further posterior dissection of the pleura is achieved by using moist pledgets; a pair of pledgets used simultaneously is most satisfactory.

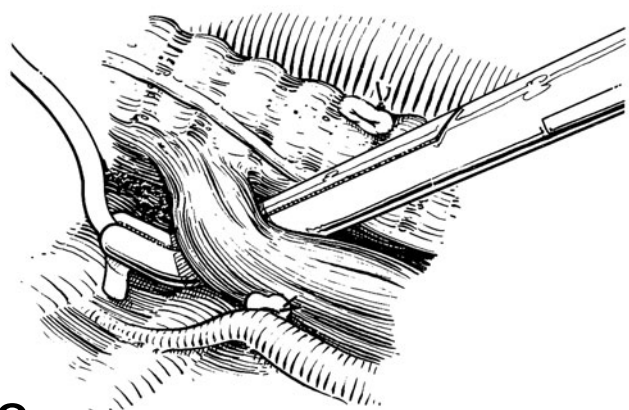
7 The azygos vein and the posterior mediastinum should be exposed, enabling the lower pouch, upper pouch, and fistula to be seen. Anterior dissection of the pleura should be sufficient only to allow the ribs to be adequately spread. Very occasionally, the size or position of the fistula may make it impossible for the anesthetist to ventilate the lungs adequately. In that situation the more rapid transpleural approach to the fistula may be necessary. In order to expose the posterior mediastinum effectively, lung retraction is essential, but care must be taken to ensure that the retractor does not compress the mediastinal structures.



7

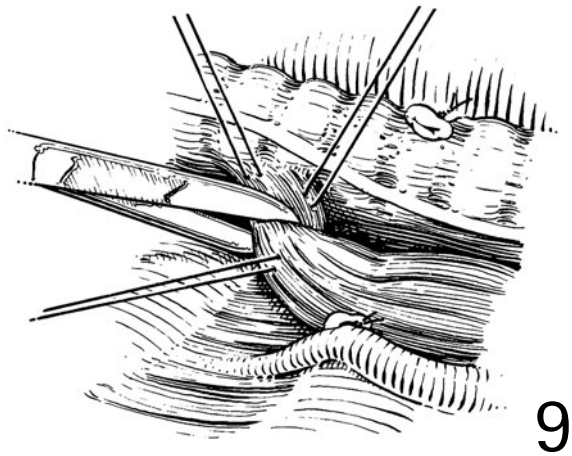
MOBILIZATION OF LOWER ESOPHAGUS AND DIVISION OF FISTULA

The azygos vein is ligated and divided. The lower esophagus may be obvious, distending with each inspiration as it lies in the lower posterior mediastinum. The close proximity of the vagus nerve to the lower esophagus aids in its identification.

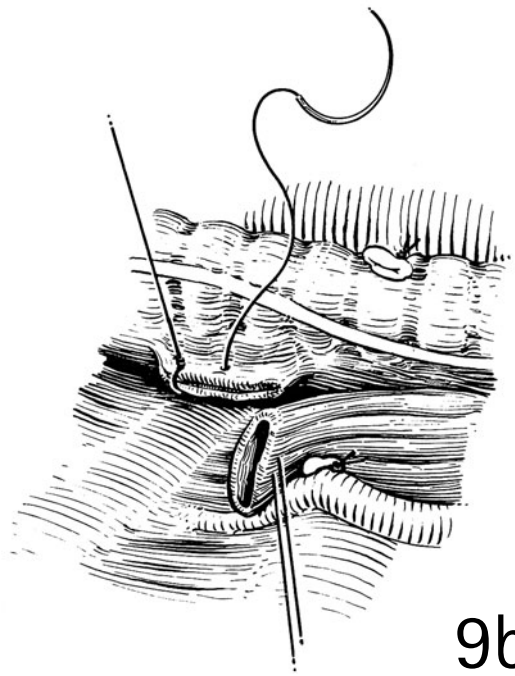


8

8 Every attempt must be made during dissection to preserve the fibers of the vagus nerve supplying the lower esophagus. The lower esophagus is dissected circumferentially to just distal to the fistula and a tape is placed around it. Traction on this tape controls the fistula and enables the junction of the lower esophagus and trachea to be accurately defined and dissected.



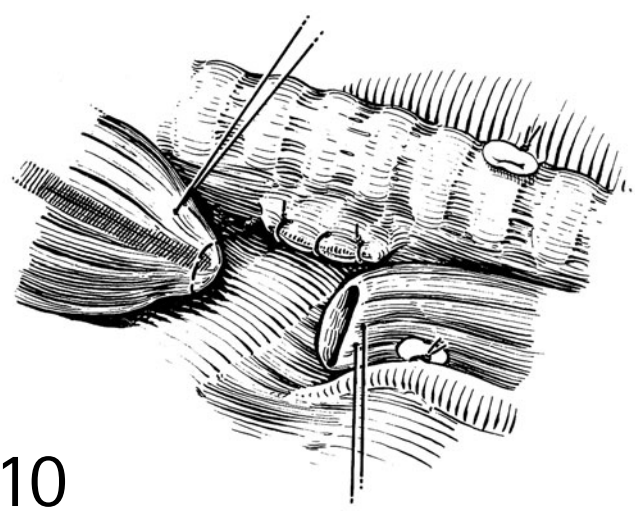
9a



9b

9a,b After carefully defining the junction between the trachea and esophagus, two 5/0 polypropylene (Prolene) sutures are placed in the trachea at the extremities of the fistula and the fistula is divided a few millimeters distal to its entry into the trachea. The trachea is closed with interrupted sutures. The air-tightness of the closure should be tested by instilling a few milliliters of saline into the mediastinum and watching for bubbles on ventilation. An alternative means of closing the fistula is to transfix it close to the trachea with a 5/0 suture.

A small tube is passed through the open end of the distal esophagus into the stomach to ensure that an adequate lumen exists and to enable air distending the stomach to be aspirated. Dissection of the lower esophagus needs care to preserve the vagal attachments and prevent damage to the adjacent thoracic duct and left pleura. A 5/0 stay suture allows traction to be exerted on the lower esophagus without handling with forceps. Dissection should be the minimum required to achieve an anastomosis.



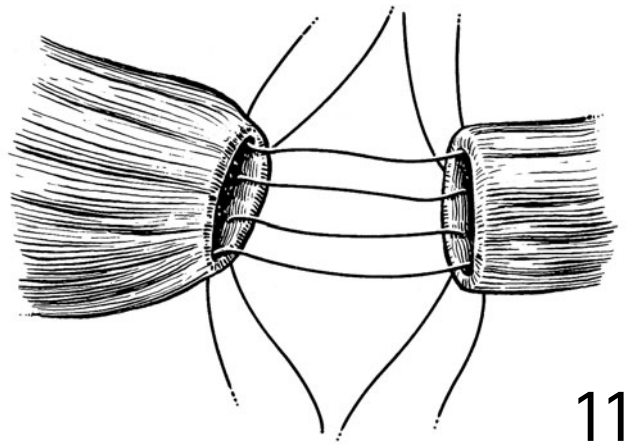
10

IDENTIFICATION OF UPPER POUCH

10 If the upper pouch is not immediately visible, pressure on the Replogle tube by the anesthetist will usually advance it into the mediastinum. Dissection of the upper pouch should be sufficient to allow an opening to be made in the distal end for an anastomosis to be performed. As with the lower esophagus, branches of the vagus supplying the upper esophagus should not be disturbed. Dissection in the plane between the esophagus and trachea should be carried out with extreme care to avoid inadvertently opening the trachea. A stay suture can be placed in the muscular wall of the esophagus to facilitate its exposure and minimize the need for forceps traction. When opening the upper esophagus, care should be taken to ensure that the opening is at the lowermost point; this is most reliably recognized by pushing the Replogle tube down and incising the esophagus over the tip of the tube. The size of the opening in the upper esophagus should correspond to the diameter of the lower esophagus.

ANASTOMOSIS

11 This is achieved using interrupted 5/0 or 6/0 sutures positioned along the posterior aspect of the anastomosis, particular care being taken to ensure that both mucosa and muscle are included in each suture. It is seldom necessary to insert more than four or five sutures. Unless the two ends of the esophagus are very close together, all the sutures are placed in position before they are tied in sequence.



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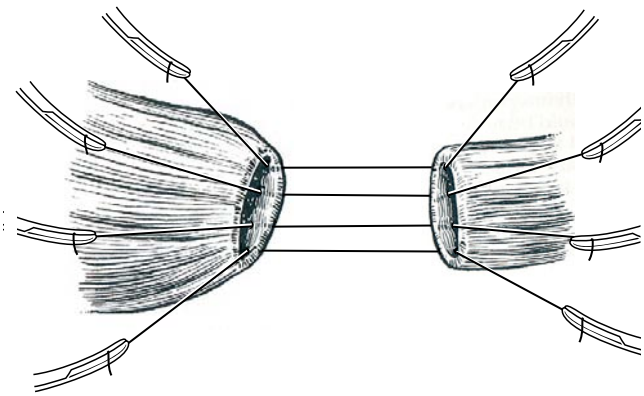
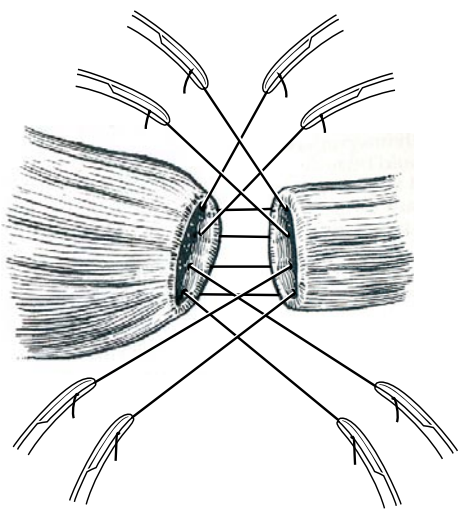


12a

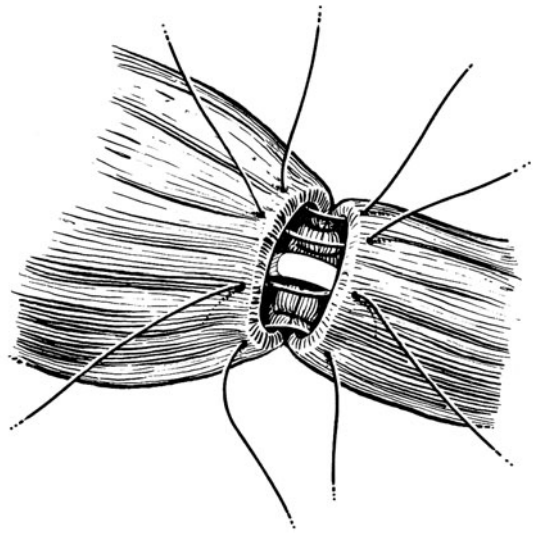


12b

ANASTOMOSIS UNDER TENSION

12a,b The posterior layer of sutures is inserted and the ends of each suture are held separately in artery forceps. The sutures with the attached forceps are crossed over and gradual tension applied, bringing the proximal and distal halves of the posterior esophagus toward each other. While maintaining tension on the sutures, each one is tied in turn, thereby distributing the tension on the sutureline over a wider area.

13 Following completion of the posterior layer of the anastomosis, a fine-bore feeding tube should replace the Replogle tube and it is then advanced across the anastomosis into the stomach. The anterior layer of the anastomosis is then completed over the tube. The use of a nerve hook avoids the need to use forceps on the open ends of the esophagus when placing the sutures. Once the anastomosis is complete, the intraesophageal tube may be withdrawn; if it is to be left in situ for feeding, its mobility should be checked to ensure that a suture has not inadvertently passed through or around it.



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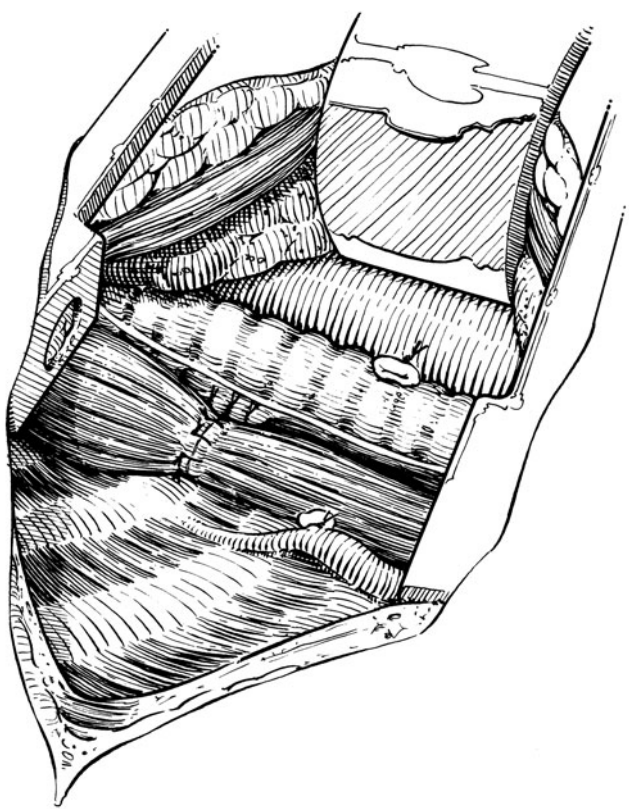

14

WOUND CLOSURE

14 The lung is expanded following the placement of two pericostal 3/0 sutures. The muscles and subcutaneous tissues are closed with a 4/0 suture and the skin with a subcuticular suture.

A chest drain is only used in specific circumstances, e.g., trauma to the underlying pleura and lung, or occasionally in a very tight anastomosis. The tip of the drain should be sutured to the lateral chest wall away from the anastomotic site with 4/0 chromic catgut.

Thoracoscopic repair

POSITION

In the case of a left descending aortic arch, the esophagus is approached from the right, which means that the child is positioned in a left anterolateral decubitus position. In the case of a right descending aorta, the child is positioned in a right anterolateral decubitus position. By using such an anterolateral position, the lung will fall out of the operative area by gravity. A short operating table, tilted in reverse Trendelenburg, is used. The surgeon stands at the ventral side of the patient with the camera person to the left and the scrub nurse at the bottom end of the operating table. The video stack is positioned in front of the patient's face. A second screen is placed opposite to the surgeon at the level of the patient's head.

PORT POSITION AND CREATION OF A WORKING SPACE

The first cannula is inserted 1 cm below and slightly anterior to the inferior angle of the scapula. The technique of insertion is the same as for the introduction of a chest drain. A small incision is made at the upper border of the underlying rib and a small hole is dissected with a Mosquito clamp. Next a cannula with blunt trocar is inserted. After verification of the correct intrathoracic position of the tip of the cannula with the telescope, the hub of the cannula is fixed to the chest wall with a suture. This will prevent the cannula from being pulled out. A sleeve around the cannula prevents the cannula from being pushed inwards.

Carbon dioxide is insufflated at a pressure of 5 mmHg and a flow of 0.1 L/min. This is a critical part of the operation. Desaturation may occur, which requires temporary release of the CO_2 pneumothorax and adjustment of the ventilator settings. It is important to be patient. After 5–10 minutes the child will adjust to the new equilibrium and desaturation will not be a problem. At this stage the lung will often stay collapsed, even without CO_2 insufflation. Once a stable situation is obtained, two more cannulae for the working instruments are inserted slightly more cranial, one behind and one in front of the scapula.

THE OPERATION

A magnified view of the upper chest is obtained. Anteriorly lies the superior vena cava with overlying phrenic nerve. Immediately behind the caval vein is the trachea with the overlying vagal nerve. Posteriorly, there is the vertebral column and rib heads, inferiorly the azygos vein, and superiorly the apex of the chest. If a right descending aortic arch is present, it may be wise to terminate the thoracoscopy on the right and to reposition the child for a left-sided thoracoscopic approach.

The *site of the distal fistula* is usually easy to identify as it bulges during inspiration. When the distal fistula enters the trachea behind the azygos vein, the vein is coagulated with the monopolar hook and divided. The distal esophagus is easily identified by its fleshy color and overlying vagal nerve.

The distal esophagus is dissected as close to the trachea as possible, avoiding any nerve branches. Some vagal branches run close to the fistula. These should be dissected off the fistula and preserved. Once the fistula has been dissected free close to the trachea, a 10 cm long 5/0 polyglycolic acid suture with straightened needle (so that it fits in the cannula) is introduced. The needle is then put through the upper wall of the fistula close to the trachea. The needle is then cut off and removed. The ends of the suture are placed around the fistula and tied. By using a transfixing suture, slippage of the ligature is prevented. The distal esophagus is cut a few millimeters distally to the ligature.

Next, the *upper pouch* is identified. To facilitate this, the anesthetist is asked to put pressure on the Replogle tube. The mediastinal pleura is now opened longitudinally behind the upper pouch, just in front of the vertebral column. There is no need to separate the upper pouch from the trachea in most cases. By limiting the dissection, damage to the nerve supply to the esophagus is kept to a minimum. As there are no nerves posteriorly, posterior dissection of the upper pouch can be performed without damage.

To open the proximal pouch in its most distal part, the anesthetist pushes on the Replogle tube while diathermy is applied right at its tip. Once a small hole has been created, it is extended using scissors. The hole to be made should be large enough to avoid creating a stenosis.

The anastomosis is made with interrupted 5/0 polyglycolic acid sutures. One starts in the middle of the left side of the anastomosis. Once the left-sided part of the anastomosis is completed, the Replogle tube is exchanged for a Fr 6 or 8 nasogastric tube. Finally, the anterior part of the anastomosis is completed. The ends of the sutures can be used to apply retraction, which facilitates the insertion of the next suture. A satisfactory method of tying under tension is to make a true flat knot and then to 'tumble' the knot. If it appears that there is too much traction, the knot should be left as it is and a second suture should be applied. Usually about eight sutures are inserted. There is no need for a chest drain in most cases.

POSTOPERATIVE CARE

The infant is nursed in the neonatal intensive care unit. Intravenous fluids are administered and antibiotic prophylaxis is continued. Feeds via the transanastomotic nasogastric tube may be commenced on the second or third day after operation. Oral feeding is gradually introduced. Regular chest physiotherapy, with nasopharyngeal suction, as required, is carried out to avoid respiratory infection.

If the esophageal anastomosis has been performed under extreme tension, it is recommended that the infant is electively paralyzed and mechanically ventilated for an arbitrary period of 5 days.

COMPLICATIONS

- Early:
  - anastomotic leakage
  - anastomotic stricture
  - recurrence of fistula.
- Late:
  - gastroesophageal reflux
  - tracheomalacia
  - dysmotility.

Anastomotic leaks occur in 15–20 percent of cases, but major disruption is rare. Minor leaks are generally detected on 'routine' contrast studies and can be managed conservatively. Major leaks present in the first 48 hours postoperatively with a tension pneumothorax. Emergency treatment is by insertion of an intercostal drain. Consideration should be given to re-exploration of the anastomosis with the intention of repairing the leak.

Anastomotic strictures develop in 30–40 percent of cases, most responding to one or two dilatations. Risk factors include anastomotic tension, anastomotic leakage, and gastroesophageal reflux.

Recurrent TEFs develop in 5–14 percent of cases and present with choking or cyanotic attacks with feeding or with recurrent episodes of pneumonia.

FURTHER READING

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14

Cervical esophagostomy

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PRINCIPLES AND JUSTIFICATION

Although this procedure is relatively seldom performed, it should still form part of the repertoire of the pediatric surgeon.

Indications

The principal indication is long-gap esophageal atresia, where the atretic segment extends beyond six or eight thoracic vertebral bodies or when attempts at delayed primary anastomosis have failed. Other indications include disruption of a previous primary repair of esophageal atresia, extensive stricture of the esophagus – caustic ingestion or peptic esophagitis – with chronic aspiration, and foreign-body perforation of the esophagus.

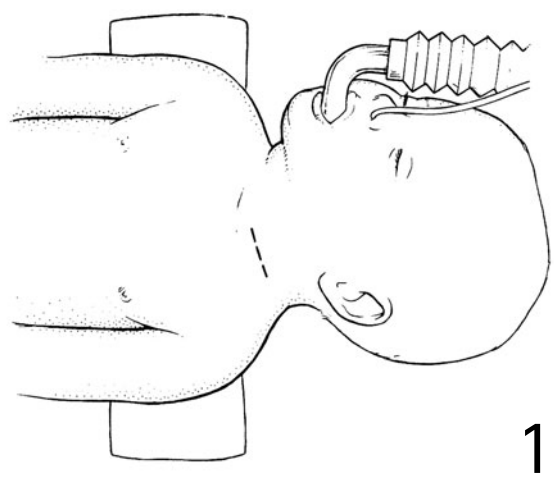
In general, performing a cervical esophagostomy commits the surgeon to carrying out an eventual esophageal replacement.

OPERATION

Position of patient and incision

1 The patient is positioned supine on the operating table under general endotracheal anesthesia. A large-bore nasogastric tube is placed as far distally as possible in the esophagus. A rolled towel is placed under the shoulders, and the neck is moderately extended.

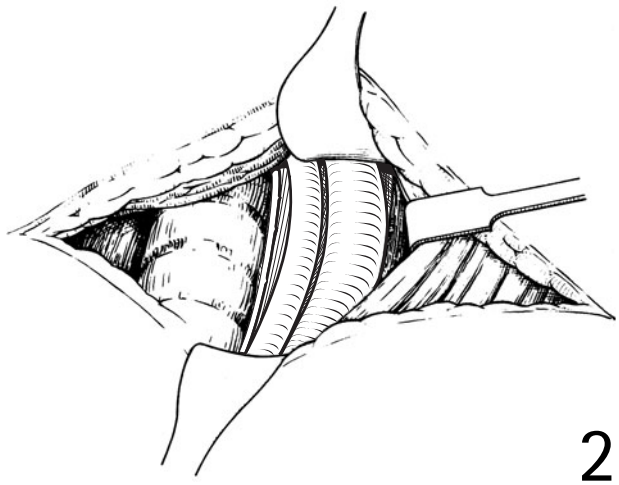
The esophagostomy may be sited on either side of the neck, but the left side is preferred. A transverse incision is made on the left side of the neck 1 cm above and parallel to the medial third of the clavicle.



1

Using diathermy, the incision is deepened through the subcutaneous fat and platysma muscle, ligating and dividing the anterior jugular vein.

2 The clavicular head of the sternocleidomastoid muscle is divided in the line of the incision using electrocoagulation to provide hemostasis. The sternothyroid muscle is similarly divided or retracted posteriorly to reveal the carotid sheath and its contents. The common carotid artery lies medial to the internal jugular vein.



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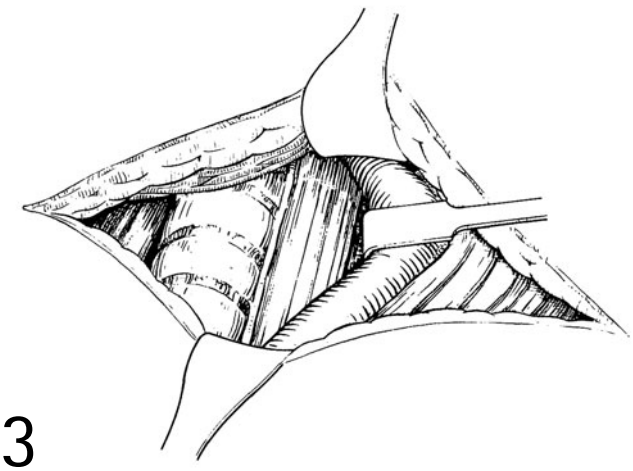


3

Preservation of recurrent laryngeal nerve

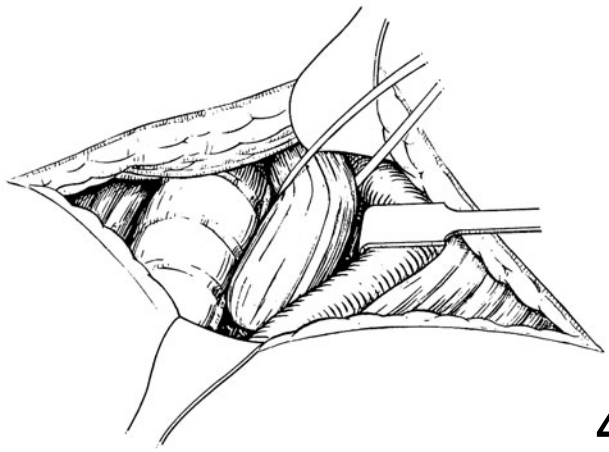
3 The carotid sheath containing the carotid artery, vagal nerve, and internal jugular vein is retracted laterally. This exposes the cartilaginous rings of the trachea anteromedially and the closely applied esophagus lying on its posterior surface. It is important to identify and preserve the left recurrent laryngeal nerve, which runs in the groove between the trachea and esophagus.

The esophagus is clearly identified by palpating the nasogastric tube within its lumen. It may also be helpful to leave a flexible gastroscope in the esophagus. By gradually withdrawing the gastroscope, the esophagus can be accurately identified by viewing the light at the end of the gastroscope as it passes through the neck.

Mobilization of the esophagus

4 The esophagus is carefully separated from the posterior surface of the trachea by blunt dissection, and, remaining close to the esophageal wall, the dissection is continued posteriorly and to the opposite side until the esophagus has been encircled. By keeping the dissection close to the esophageal wall, the risk of traumatizing the right recurrent laryngeal nerve will be minimized.

It is important to mobilize the full thickness of the esophagus. It is easy to enter the submucosal plan and to mobilize a mucosal cuff only. A soft rubber sling is placed around the esophagus, and the dissection continued distally, remaining on the muscle wall.



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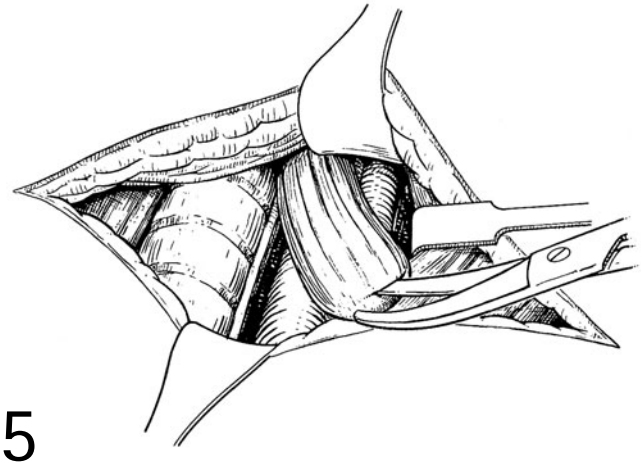
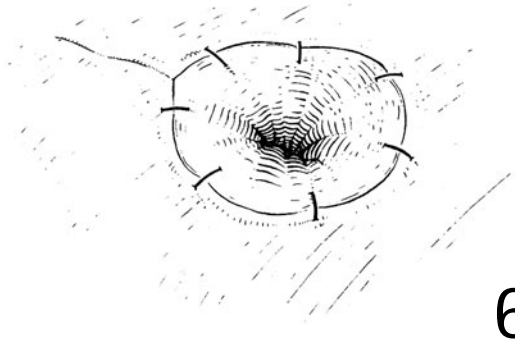


5

5 In isolated esophageal atresia, the proximal esophageal segment is usually short and the blind end soon comes into view. In other cases, e.g., strictures, it is necessary to divide the esophagus at a convenient place, leaving sufficient proximal esophagus to reach the skin incision without tension while permitting accurate closure of the distal esophagus.

Fashioning the esophagostomy

6 The end of the esophagus is brought out of the lateral end of the incision and sutured to the skin edges with interrupted, full-thickness, fine, absorbable sutures. There should be no tension on the suture line, and a nasogastric tube should be able to exit the stoma unimpeded.



6

Wound closure

The remainder of the wound is closed with interrupted sutures to the platysma muscle and subcuticular sutures to skin.

POSTOPERATIVE CARE

The esophagostomy should be covered initially with paraffin gauze and ultimately left open to drain freely onto the surface.

An adhesive plastic bag may be applied to the skin around the esophagostomy site to collect saliva. In infants with esophageal atresia it is vitally important to practice sham feeding pending the esophageal replacement. Failure to do this will result in tremendous difficulty in establishing oral feeds when the esophageal replacement is performed.

COMPLICATIONS

Stricture of the cervical esophagostomy occurs as a consequence of impaired vascularity. Kinking of the esophagus occurs when mobilization has been inadequate and will result in inefficient drainage of saliva.

15

H-type tracheoesophageal fistula

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HISTORY

Described by Lamb in 1873, the first operative success was reported in 1939 by Imperatori.

PRINCIPLES AND JUSTIFICATION

An isolated or H-type tracheoesophageal fistula most commonly presents during the first few days of life, when the neonate chokes on attempting to feed and/or has unexplained cyanotic episodes. The associated gaseous distension of the gastrointestinal tract may be sufficiently severe to mimic that of intestinal obstruction. Older infants and children are likely to present with recurrent chest infections, particularly involving the right upper lobe.

H-type tracheoesophageal fistula constitutes about 3 percent of tracheoesophageal anomalies.

Investigations

The most reliable method of establishing the diagnosis of an H-type fistula is by a prone tube video esophagogram. A small-caliber nasogastric tube is passed into the distal esophagus and contrast medium is gradually injected while the tube is slowly withdrawn. The presence of an H-fistula may be missed in over 50 percent of routine contrast swallows.

Bronchoscopy with esophagoscopy will confirm the presence of a fistula. If performed immediately before ligation of the fistula, it should be possible to pass a fine tube, e.g., a ureteric catheter, through the fistula to aid in its subsequent identification at surgical exploration.

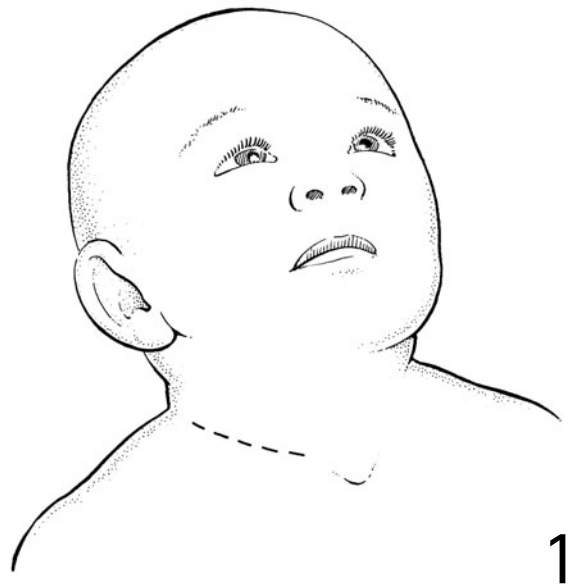
Having accurately identified the position of the fistula, a decision can be made on the most suitable approach. Some fistulas, including a recurrent fistula associated with a previous repair for esophageal atresia, will be best approached through the thorax, but the majority of isolated tracheoesophageal fistulas can be divided through a cervical approach with moderate neck extension.

The thoracic approach to a tracheoesophageal fistula is similar to that previously described for esophageal atresia and tracheoesophageal fistula (see pp. 109–20).

OPERATION

Position of patient

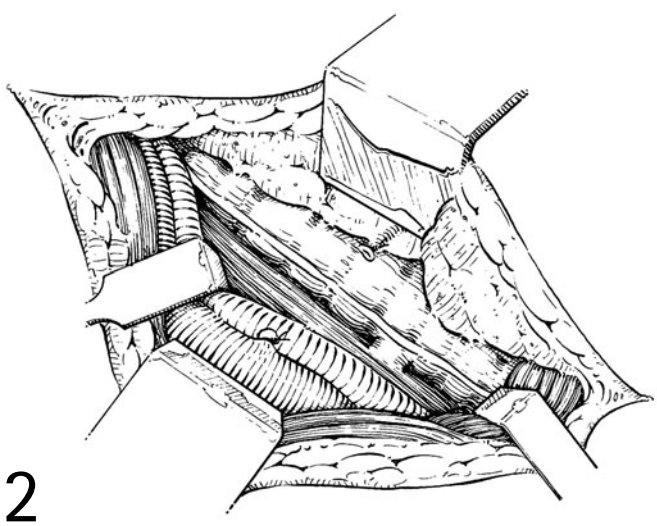
1 The child is placed supine on the operating table with the head turned to the left. Before extending the neck, the site of the incision is drawn with a marker pen in a suitable skin crease 1 cm above and parallel to the clavicle. Failure to mark the site of the incision before extending the neck may result in a cosmetically unsatisfactory incision.



1

A sandbag of appropriate size placed under the shoulders ensures adequate neck extension. An approach through the right side is usually preferred. A nasogastric tube of adequate size should be passed after induction of anesthesia.

In ideal circumstances, a ureteric catheter will have been passed through the fistula under bronchoscopic control. This is invaluable in identifying the fistula site during exploration.



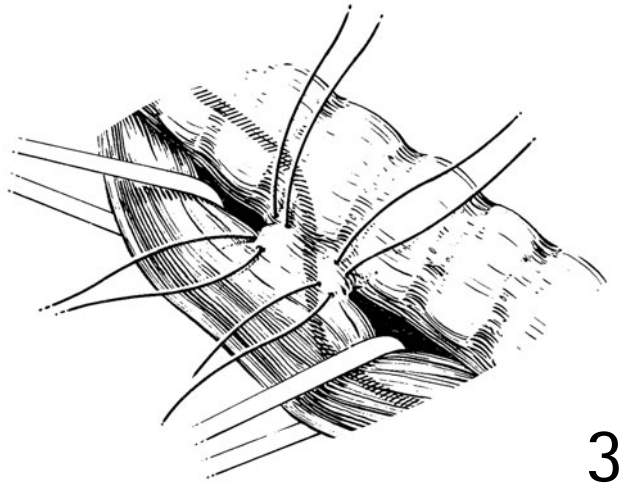
2

Operative approach

2 Having incised the skin and subcutaneous tissues, the sternocleidomastoid muscle is retracted posteriorly, dividing the sternal head of this muscle if necessary to allow adequate exposure. The plane medial to the carotid sheath is identified, and dissection allows the sheath to be retracted posteriorly. The thyroid gland, trachea, and esophagus lie medially. Palpation of the endotracheal and nasogastric tubes facilitates identification of these structures. The inferior thyroid artery and middle thyroid vein are identified crossing the space between the retracted carotid sheath and the thyroid, and division of these vessels may be necessary. The plane between the trachea and esophagus is gently dissected, care being taken to identify and preserve the right recurrent laryngeal nerve.

Dissection of fistula

3 Identification of the fistula requires careful dissection, and it is usually rather higher than anticipated because of the extension of the neck. Slings positioned around the esophagus above and below the fistula will facilitate dissection, but extreme care is required to preserve the left recurrent laryngeal nerve, which lies on the opposite side of the neck and is difficult to visualize.



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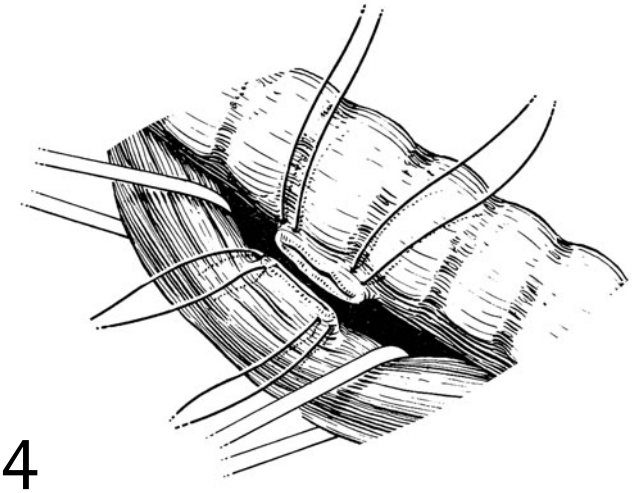

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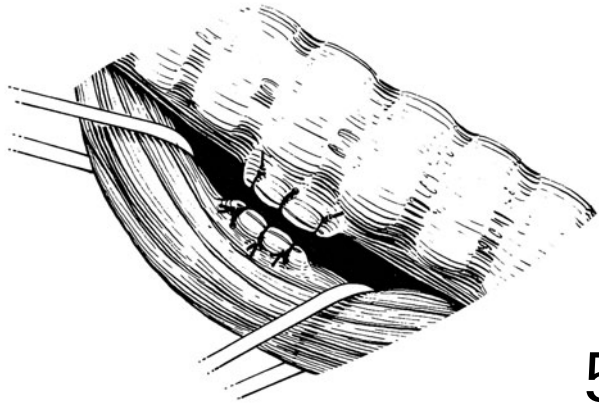
4 Having isolated the fistula, stay sutures are placed on the esophageal side to mark its position because, following division of the fistula, rotation and retraction of the esophageal end may make it difficult to identify. On the tracheal side, a 5/0 polypropylene suture is placed at both limits of the fistula.

5 Following division of the fistula, the tracheal defect is closed with interrupted sutures. The esophageal end of the fistula is closed with one or two interrupted fine polyglycolic acid sutures.



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The retracted tissues will now assume a more normal position. Some surgeons advocate interposing tissue, e.g., fascia or muscle, between the two opposing suture lines in order to reduce the likelihood of recurrence of the fistula. The wound is closed in layers with absorbable sutures and with a subcuticular suture for the skin.

POSTOPERATIVE CARE

Extensive dissection of the trachea and esophagus is often required during this operation. Invariably this produces some tracheal edema, which may be minimal immediately after operation but progresses in severity for up to 24 hours. It is reasonable, particularly in premature infants or in those with pre-existing lung disease, to leave an endotracheal tube in position.

Following extubation, the movement of the vocal cords should be assessed. A proportion of these neonates will require intubation for some considerable time and may have a tendency to stridor, particularly when crying or coughing, for weeks or months afterwards. Feeds can be given through a nasogastric tube.

Recurrence of an H-type fistula is rare.

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16

Recurrent tracheoesophageal fistula

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PRINCIPLES AND JUSTIFICATION

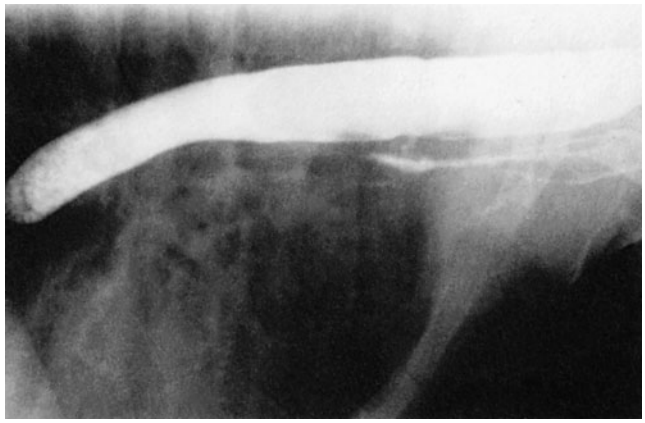
The incidence of recurrent tracheoesophageal fistula ranges from 5 percent to 10 percent. Infants who develop respiratory symptoms following repair of an esophageal atresia, e.g., gagging, coughing, apneic or cyanotic spells during feeding, or suffer from recurrent chest infections should be suspected of having a recurrent fistula.

PREOPERATIVE

Diagnosis

Radiographs of the chest and abdomen may show an air esophagogram or an excessive amount of gas in the gastrointestinal tract. Routine contrast swallows will only detect about 50 percent of recurrent fistulas. Esophagoscopy carries a low diagnostic rate.

1 Cine tube esophagography performed in the prone position is the most reliable method of establishing the diagnosis.

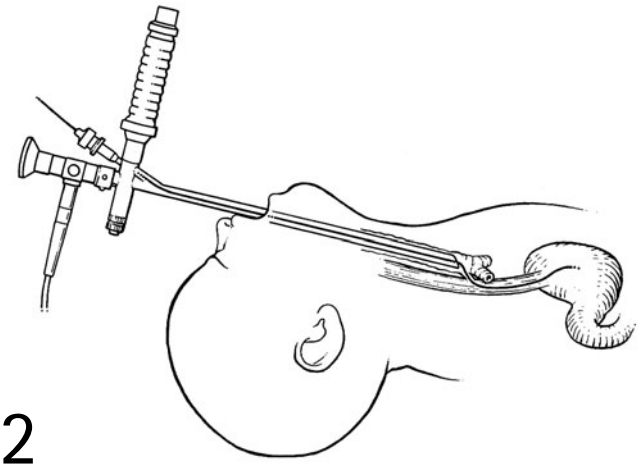


1

Bronchoscopy with cannulation of the fistula is also a reliable diagnostic method and is invaluable in locating the fistula during the operative procedure.

Anesthesia

Anesthesia for repair of recurrent fistula is fraught with danger. Lung function may be compromised by chronic aspiration, and vascular access may have been compromised by previous surgery or the use of parenteral nutrition. Intraoperative ventilation may be difficult because of gaseous escape through a large fistula. Meticulous intraoperative monitoring is mandatory, including continuous oxygen saturation measurements. Hand ventilation with high gas flow rates may be necessary because of changes in pulmonary compliance. Because of pleural adhesions following previous surgery, blood loss may be excessive and should be replaced – volume for volume. Postoperative ventilation and intensive care are often necessary, particularly in young infants because of respiratory stridor.



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OPERATION

Bronchoscopy and cannulation of fistula

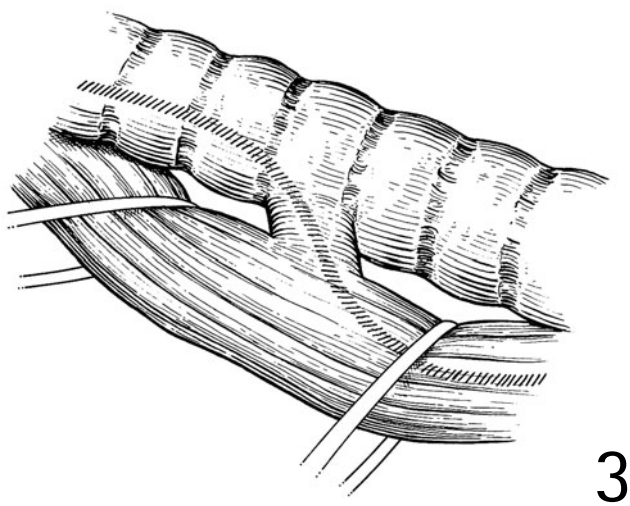
2 A rigid Storz bronchoscope of suitable size for the child's age (2.5–3.0 Fr for infants) is passed and the fistulous opening in the posterior wall of the trachea at or just above the carina is identified. A fine (4–6 Fr) ureteric catheter is passed through the suction channel of the bronchoscope, through the fistula into the esophagus, advanced well down into the esophagus and left in situ.

Incision

A posterolateral thoracotomy via the previous incision is the preferred approach. The fourth or fifth intercostal space is opened in the length of the incision and a small rib spreader is used to widen the thoracotomy. Access to the mediastinum is via a transpleural route as the extrapleural approach is not an option, having been used in the repair of the esophageal atresia.

Mediastinal dissection and mobilization of esophagus

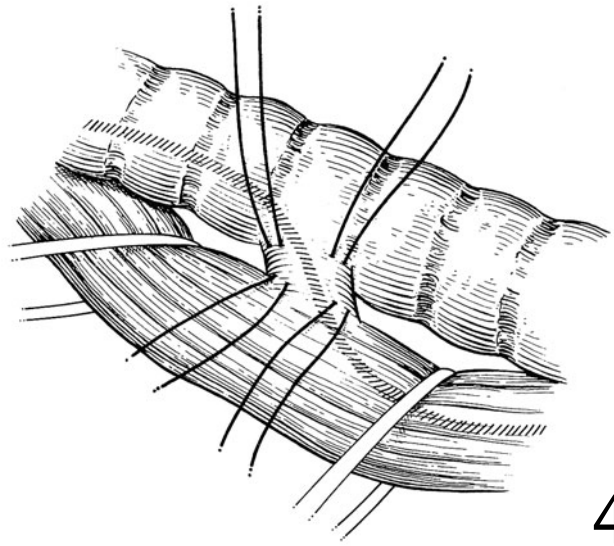
3 The mediastinal pleura is opened longitudinally over the esophagus, exposing its lateral wall proximal and distal to the fistulous site. The esophagus proximal and distal to the fistula (identified by palpating the ureteric catheter within its lumen) is carefully mobilized from the surrounding tissue and rubber slings are passed around it above and below the fistula.



3

Dissection of fistula

4 Having identified and isolated the fistula, stay sutures of 5/0 polypropylene (Prolene) are inserted in the upper and lower walls of the fistula on both the tracheal and esophageal ends. These sutures are invaluable in facilitating accurate closure of the defects following division of the fistula.



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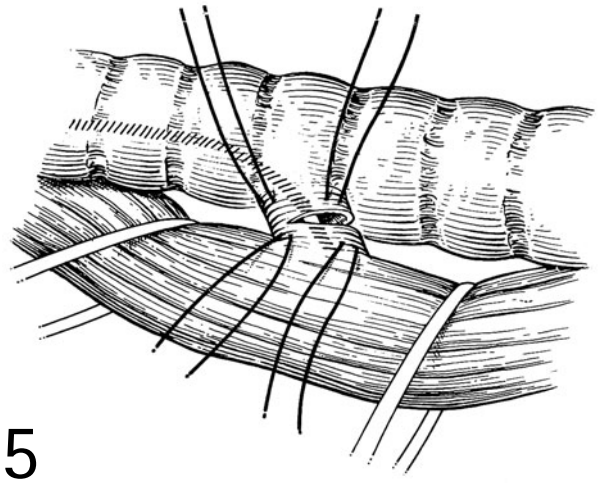

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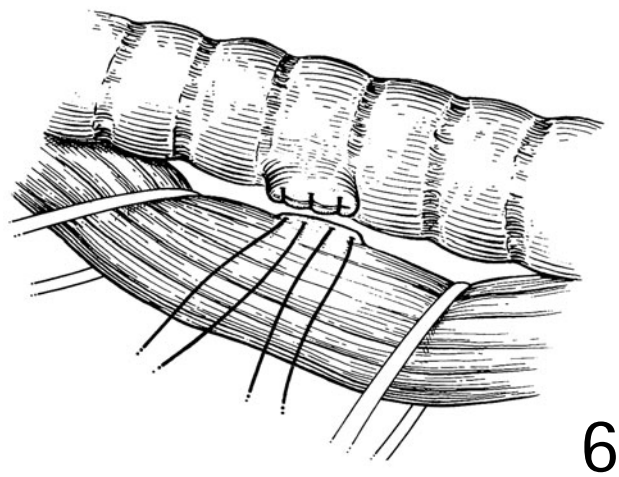
Division of fistula

5 The fistula is now opened. The ureteric catheter should be positively visualized traversing the fistula before being withdrawn through the mouth to allow complete division of the fistula.

Closure of fistulous openings

6 The tracheal and esophageal orifices of the fistula are securely closed with interrupted 5/0 polypropylene sutures.



6

Separation of the suture lines

To prevent the two contiguous suture lines becoming adherent and to prevent refistulization, mediastinal pleura, intercostal muscle, or a flap of pericardium may be interposed between the esophagus and trachea. In the author's experience, once the fistula has been divided and the defects in the trachea and esophagus repaired, the two suture lines tend to rotate away from each other so that interposing tissue between is unnecessary. An intercostal drain is inserted with the tip some distance away from the area of repair.

Wound closure

The thoracotomy wound is closed with pericostal 3/0 polyglycolic acid sutures and the muscles of the chest wall are approximated with continuous 4/0 polyglycolic acid sutures. The skin is closed with a subcuticular suture.

Endoscopic treatment

A variety of endoscopic methods to promote closure of recurrent tracheoesophageal fistulas have been described. The agents used include tissue glues (Histoacryl), fibrin sealant (Tisseel) and lasers. Reported series are small, with variable success rates and short-term follow-up.

POSTOPERATIVE CARE

Endotracheal intubation and elective mechanical ventilation are usually required for 48–72 hours after operation due to tracheal edema. A contrast swallow may be performed on the seventh day after operation and the chest drain removed following confirmation that there is no leak from the esophageal repair.

COMPLICATIONS

These include pneumothorax and esophageal leak with consequent empyema. There is a 10–20 percent risk of fistula recurrence.

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17

Aortopexy

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HISTORICAL BACKGROUND

In 1948, Gross and Neuhauser were the first to recognize and successfully treat innominate artery compression of the trachea by suturing the innominate artery to the sternum. Some 20 years later, Mustard and colleagues reported on 285 patients with innominate artery compression of whom 39 underwent surgery. The operation was performed through a right thoracotomy and the ascending aorta and origin of the innominate artery were sutured to the sternum.

The association of tracheomalacia with tracheoesophageal anomalies and its subsequent management by aortopexy was first reported by Benjamin et al. in 1976. More recently, DeCou et al. (2000) have described thoracoscopic aortopexy.

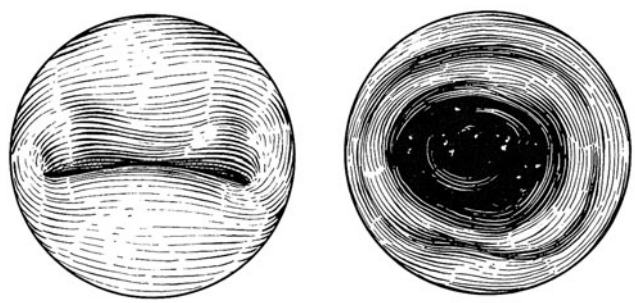
PRINCIPLES AND JUSTIFICATION

Tracheomalacia is suspected when an infant develops expiratory stridor on exertion – feeding and crying. Increasing difficulty with expiration leads to difficulty with feeding, cyanotic spells, and, in extreme cases, apnea and collapse (death attacks). In infants with tracheoesophageal anomalies, symptoms often become pronounced with increasing body mass over the age of about 3 months. Expiratory stridor at rest or biphasic stridor is an ominous sign of severe airway compromise.

The symptoms are similar to features of severe gastroesophageal reflux and recurrent tracheoesophageal fistula, and the three conditions may coexist.

Diagnosis

1a,b The diagnosis is confirmed endoscopically when expiratory collapse of the trachea is seen during quiet respiration. Complete obliteration of the lumen is common under these circumstances. The tracheal collapse in infants previously treated for esophageal atresia and/or tracheoesophageal fistula is usually proximal to the site of entry of the original fistula.



1a 1b

Severe symptoms are unusual when more than 20 percent of the lumen remains open.

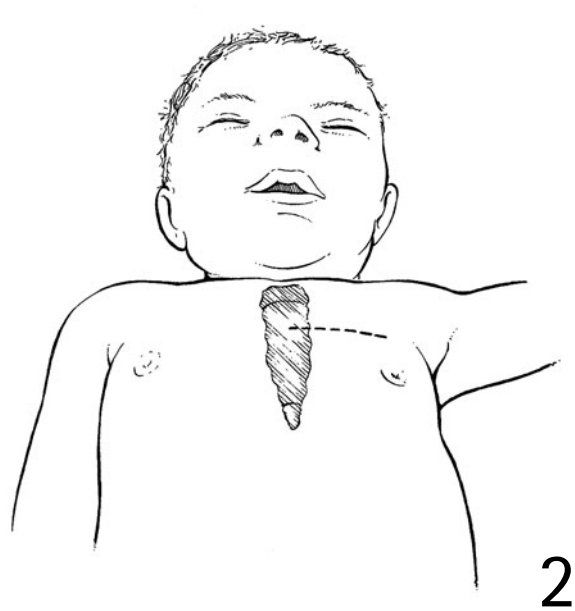
OPERATION

Position of patient

The patient is positioned supine with a small roll beneath the shoulders and the left arm abducted.

Incision

2 The chest is entered through a left anterior thoracotomy through the bed of the third rib. The pectoral muscles are incised medially and split towards the lateral end of the incision.



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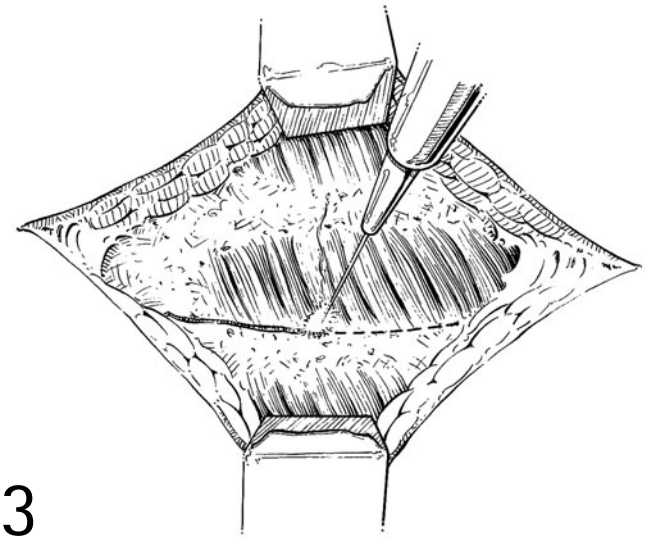

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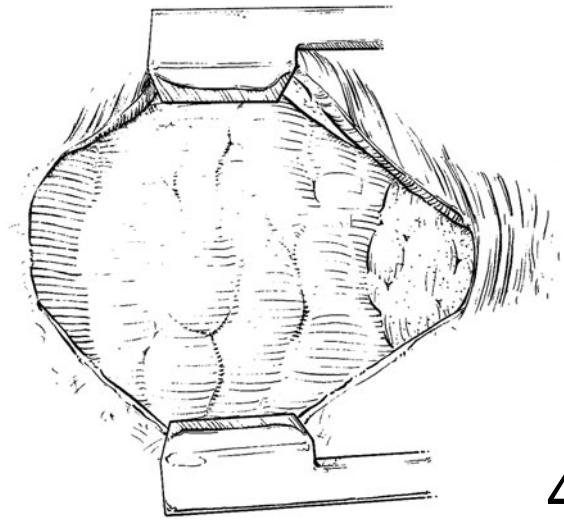
3 The perichondrium and periosteum are incised with diathermy and separated from cartilage and bone respectively. The chest is entered through the bed of the rib and the internal mammary vessels are divided at the medial end of the incision.

4 The thoracotomy is widened by means of an infant-sized chest retractor. The left lung is retracted and held out of the operation field by a moist gauze swab.



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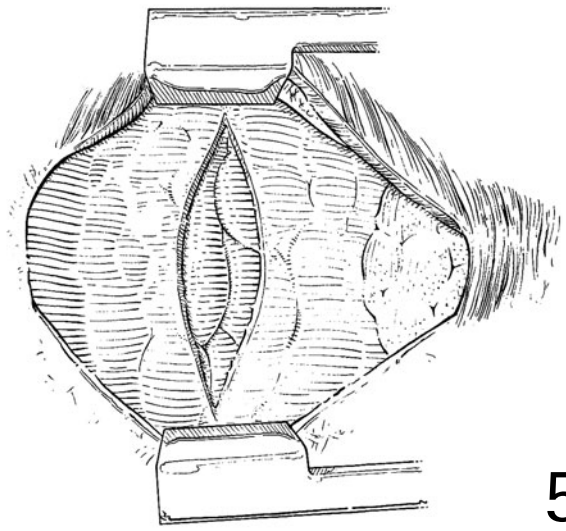
5 The pleura is incised longitudinally over the left lobe of the thymus gland.



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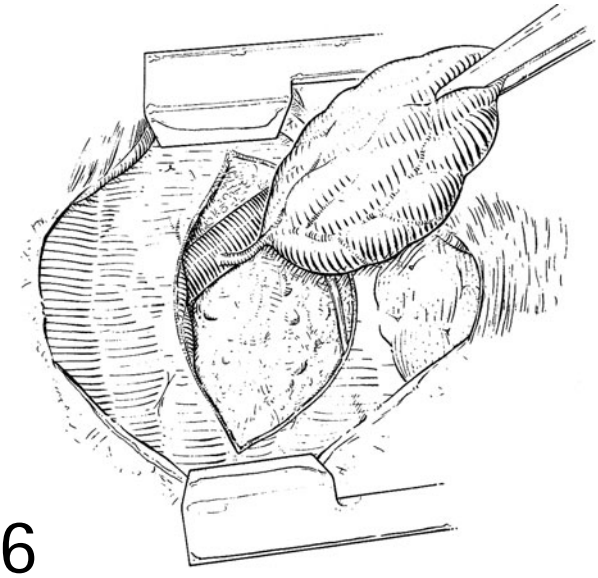

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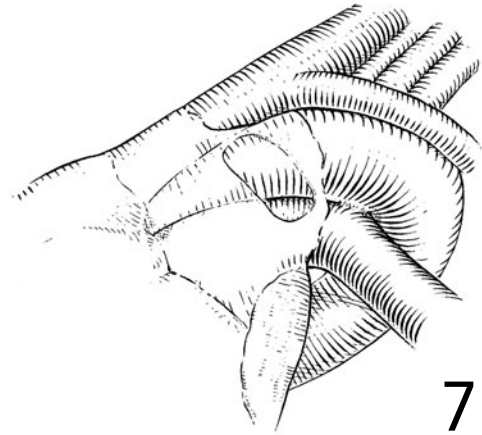
6 The left lobe of the thymus gland is excised with care to protect the phrenic nerve. The thymic vein, which enters the innominate vein, is identified, coagulated, and divided. The great vessels are then in view.

7 The roots of the great vessels are within the pericardium, which is incised transversely just below its reflection on the aorta. Meticulous attention to hemostasis is required.



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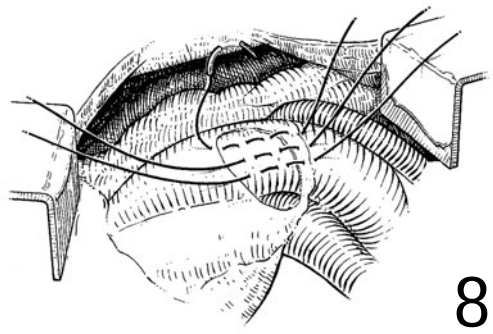
8 Three 4/0 polypropylene sutures are sufficient to perform the aortopexy. The pericardial reflection and aortic adventitia are picked up with two or three bites of each suture. Care is taken to remain superficial to the tunica media of the aorta. The sutures are then inserted into the posterior aspect of the sternum and are left untied until all three are in place.



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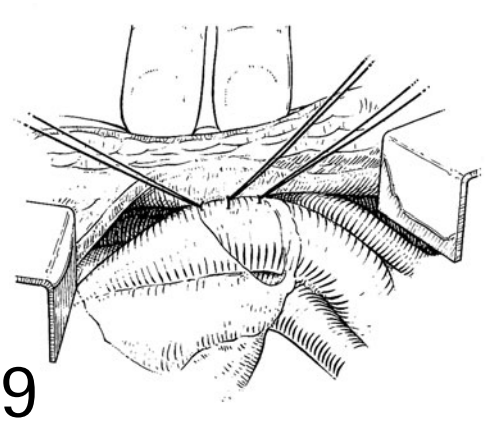

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9 The assistant then depresses the sternum firmly and holds this position until all three sutures have been tied. The sternum is then released and the aorta moves forward with it. The anterior tracheal wall also moves anteriorly to fill the potential space between the aorta and trachea. There are no connections between the aorta and trachea.

Wound closure

The wound is closed in the normal manner. If the operative field is dry, a chest drain is unnecessary.

POSTOPERATIVE CARE

Oral feeding may be commenced the same day and early discharge is possible if symptoms are alleviated.

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18

Esophageal replacement with colon

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HISTORY

Despite a continued emphasis on saving the child's native esophagus and on esophageal replacement operations utilizing the transposed stomach, esophageal replacement with colon remains an important technique for replacing the esophagus in children. It is ideally suited for long-gap esophageal atresia and may be the only choice in patients with congenital microgastria or in whom the stomach is also damaged.

The first description of the anatomic details of esophageal replacement with colon is credited to Vuillet and Kelling in 1911. Von Hacker was the first to perform the procedure 3 years later. For many years, the colon was the primary substitute for a damaged or atretic esophagus. Colon interposition has given way to newer techniques transposing the stomach as the favored procedure for esophageal replacement in the adult population. Many of the pitfalls using the transposed colon in adults stem from problems with vascular supply to the graft. These problems are less commonly seen in the pediatric population, thus esophageal replacement with colon remains an acceptable choice with a high likelihood of long-term success in the pediatric population.

A newer technique, the colon patch esophagoplasty, was described by Hecker and Hollman. Othersen has championed this technique for the correction of persistent, isolated, short strictures of the esophagus. The 'colon patch' avoids many of the functional disadvantages of the segmental colon interposition, which may have a tendency toward redundancy and stasis.

PRINCIPLES AND JUSTIFICATION

The colon is particularly suited for replacement of the esophagus when a long-segment interposition is required or when the stomach is not available as a replacement conduit. In the pediatric population, colon interposition for esophageal replacement is most commonly used in cases of esophageal atresia with inadequate length for primary repair. Colon can also be used to repair esophageal strictures that are too long for simple resection and re-anastomosis. Colon interposition has been utilized for extensive caustic strictures of the esophagus resulting from lye ingestion. In older patients, extensive Barrett's changes of the esophageal mucosa can be an indication for esophageal replacement with colon.

Most authors think that the colonic segment interposed in the esophagus functions by gravity drainage. The normal esophagus contracts with forceful peristaltic contractions in response to initiation of a swallow, while the normal colon contracts in a more complex pattern resulting from distension of the colon both hormonal and neurogenic. The colon interposed into the esophageal bed may preserve its inherent pattern of contractions, but may also need a large volume of distension before emptying. Studies of interposed colon segments in the esophagus show slow transit of solids and an absence of contractions, suggesting that there is no functional advantage of an isoperistaltic right colon interposition over an antiperistaltic left colon interposition. On the other hand, animal studies of isolated segments of colon show more rapid emptying in isoperistaltic versus antiperistaltic loops.

The interposed colon can be placed in the posterior mediastinum or substernally. In some cases, scarring in the native esophageal bed precludes positioning the graft in the native position. If technically possible, however, placement in the esophageal bed may lead to less dysphagia and a more satisfactory long-term outcome.

In children, a right thoracotomy for exposure with a right cervical incision for anastomosis is preferred. Therefore, in patients who require a staged operation with cervical esophagostomy prior to esophageal replacement, the cervical esophagostomy is placed in the right side of the neck. In older children with minimal scarring of the esophageal bed, transhiatal dissection without thoracotomy may be possible.

Variations of technique

Vascular supply to the colon dictates the segment of colon to be transposed into the chest. Right, transverse, or left colon can be mobilized and brought up to the neck in most children without compromise of the blood supply to the graft. An isoperistaltic segment beginning in the right colon can be based on either the right or left colic vessels. The middle colic vessels are divided. A colonic segment based on the left colic vessels affords the greatest mobilization and is well vascularized in children. In adults, a significant incidence of graft failure due to ischemia has led to microvascular techniques that 'supercharge' the interposed segment with an additional vascular anastomosis in the neck. These techniques have not been reported in children and are probably not necessary.

Patients should be informed that stricture, leak, functional problems with emptying of the colonic segment, dysphagia, and respiratory problems can result from this procedure.

PREOPERATIVE ASSESSMENT AND PREPARATION

Assessment

Many authors stress the importance of preserving the native esophagus, if possible. Determining a need for esophageal replacement may be straightforward in cases of long caustic stricture or recurrent stricture after failed esophageal atresia repair, but in cases of newborns with long-gap esophageal atresia, a decision to replace the esophagus may evolve over time. In esophageal atresia with a long gap between the proximal and distal ends, the proximal esophagus can be initially left in place with sump drainage while the child is fed by gastrostomy. In many cases, the gap will shorten and primary anastomosis is possible. Different techniques for esophageal lengthening have been described, with varied reported success.

If esophageal growth is initially inadequate, a right cervical esophagostomy is preferred so that the child can learn to chew and swallow with reduced risk of aspiration. The child can be maintained on gastrostomy feedings until 6–8 months of age. Some surgeons report success with primary esophageal replacement at an earlier age, thereby avoiding the esophagostomy. The goal of establishing a normal pattern of chewing and swallowing at the earliest possible age cannot be overemphasized.

In cases of esophageal stricture, radiographic contrast studies to determine the length of the stricture involved may help the surgeon choose between esophageal replacement and patching with colon. Angiographic studies to investigate the colonic blood supply in children preoperatively are not routinely necessary.

Bowel preparation

Prior to esophageal replacement, the child is admitted and clear liquids given for 24 hours preoperatively. In children beyond infancy, polyethylene glycol electrolyte solution is given at a rate of 25–40 mL/kg per hour until stools are clear.

Antibiotic bowel preparation is accomplished with neomycin 50 mg/kg and erythromycin base 60 mg/kg in three doses, the first two doses given 1 hour apart and a third at midnight before surgery, then nothing enterally.

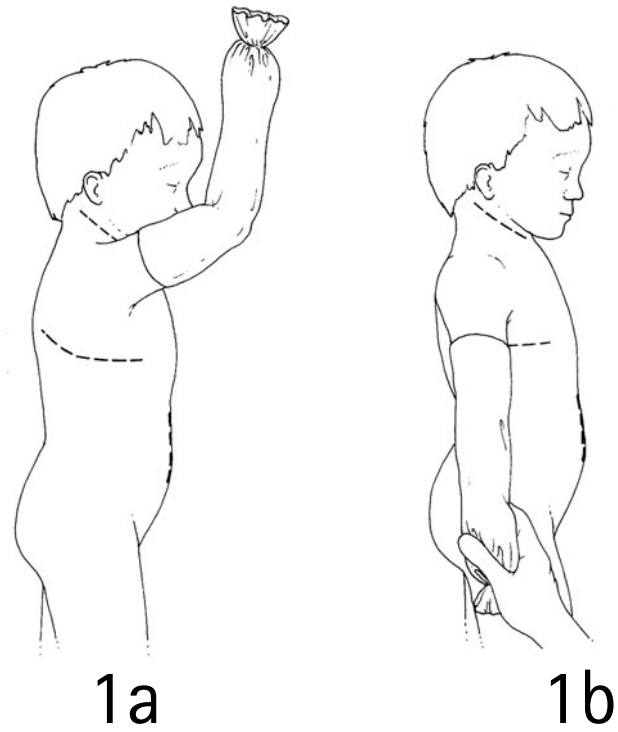
Anesthesia

Colon interposition is accomplished under general anesthesia. An epidural catheter can facilitate the intraoperative anesthetic and postoperative pain control.

OPERATION

Esophageal replacement with colon

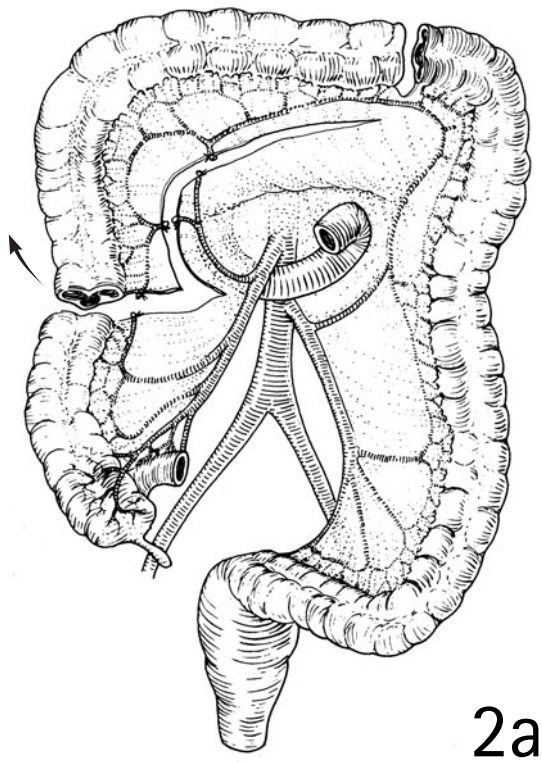
1a,b The patient is positioned in a left lateral decubitus position with the right arm prepped into the operative field. The arm can be raised for the thoracic portion of the procedure. The table can be rolled laterally to afford greater exposure for the abdominal portion of the procedure.



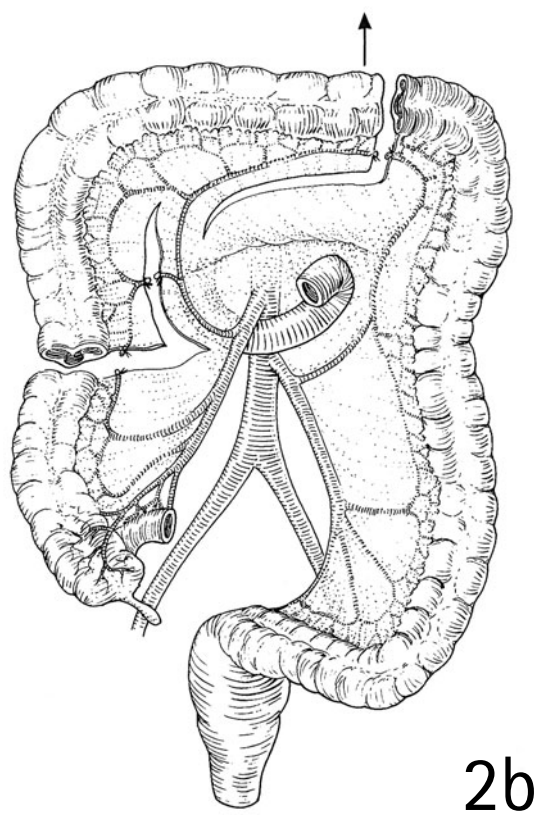
1a 1b

First, a posterolateral thoracotomy incision is made in the sixth intercostal space. The latissimus dorsi is retracted laterally. If there has been no prior thoracotomy, an extrapleural approach is made. A subperiosteal resection of the sixth rib with incision into the extrapleural space through the posterior periosteum may facilitate this approach. A transpleural approach is made in the case of previous thoracotomy. The lung is retracted anteromedially and the proximal esophagus identified with the aid of manipulation of a nasoesophageal tube by the anesthesiologist. The proximal esophagus is mobilized to the thoracic inlet. The distal esophageal segment is mobilized to the esophageal hiatus.

A midline incision is made and the colon examined. The esophageal hiatus is mobilized from the abdomen as well. Finally, an oblique or transverse right neck incision is made. The neck vessels are retracted and the proximal esophagus is mobilized within the thoracic inlet. An umbilical tape is passed from the colonic mesentery through the diaphragmatic hiatus to the level of the proximal esophageal segment in the neck. This tape helps determine the length of colon necessary for replacement.



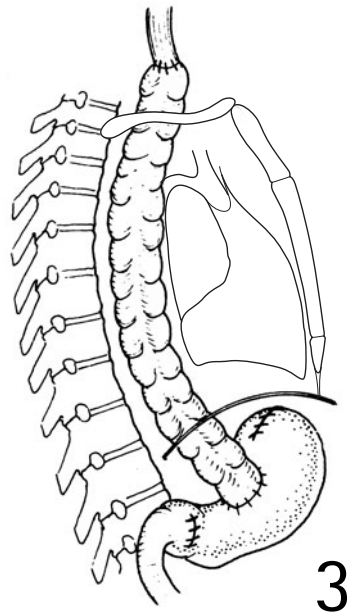
2a



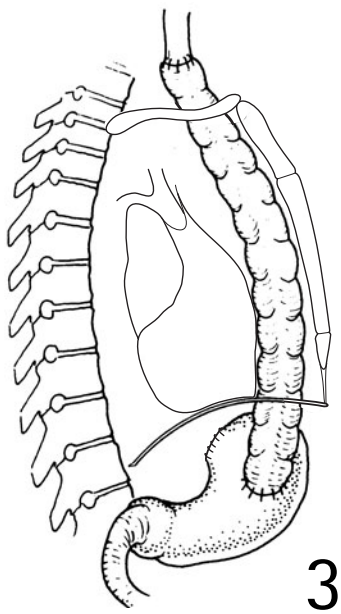
2b

2a,b The selection of the vascular pedicle is crucial. The umbilical tape can be placed along the colon to select the segment for resection and to identify the vessels to be divided in order to mobilize the colon. An isoperistaltic colonic segment of right and transverse colon based on the left colonic vessels is favored. A right colonic segment may also be constructed based on the right colic vessels. Alternatively, an antiperistaltic left colonic segment can be mobilized. For right colon interposition, small vascular clamps are placed on the middle and right colic vessels and on the marginal artery at the proposed level of resection. The perfusion of the proposed graft by the remaining blood supply is assessed after 10 minutes. If the blood supply is adequate, the clamped vessels are divided centrally, preserving collaterals in the marginal artery system of the colon segment. The right colon is then divided distal to the cecum at the appropriate length using a linear gastrointestinal stapling device. A generous length of colon is chosen, so as to avoid tension on the cervical anastomosis. Mobilization of the splenic flexure is undertaken only as much as is necessary to deliver the colon to the neck.

3a,b Once the colon graft is mobilized, the distal esophageal stump is resected, if possible, at the stomach with a linear stapling device to avoid reflux in the stump. In some cases, a normal gastroesophageal junction may be preserved, anastomosing distal esophagus to the colonic interposition. The colonic segment is passed behind the stomach through the esophageal hiatus and placed in the esophageal bed. Alternatively, if scarring in the posterior mediastinal bed is severe, the colon can be placed in a retrosternal or substernal position. Blood supply to the colon is usually excellent in children; both isoperistaltic and antiperistaltic grafts can be successful.

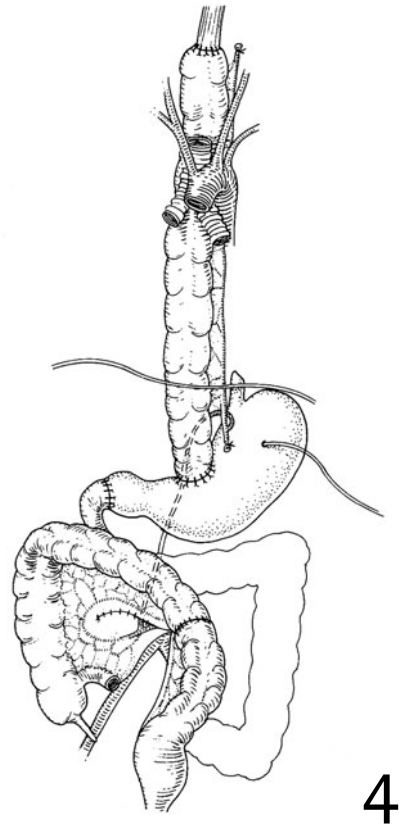


3a



3b

4 The proximal anastomosis between esophagus and colon is created in the neck with a single layer of absorbable sutures. While the anastomosis can be created in the neck or the chest, a cervical anastomosis avoids the potential disaster of an intrathoracic leak, which carries high morbidity and mortality. In most cases, the colonic graft can easily reach the neck. The distal cologastric anastomosis is created on the anterior stomach wall. The anastomosis can be partially wrapped with stomach to limit reflux.



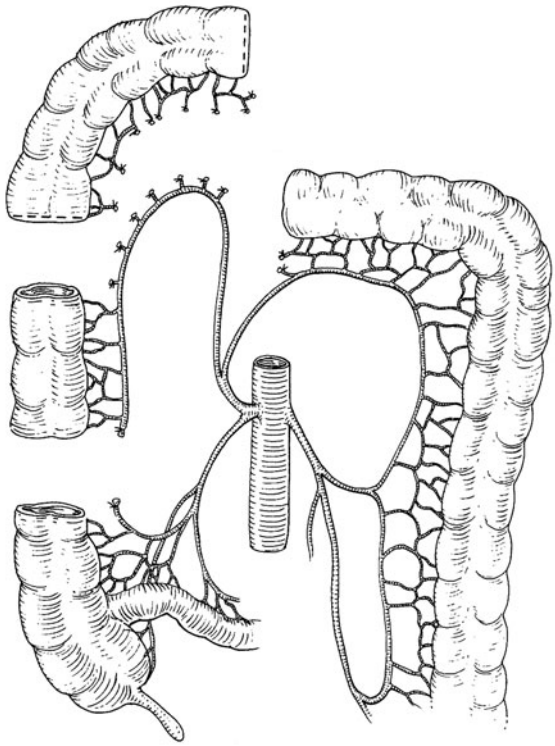
4

If the vagus nerve is divided, a pyloroplasty must be created. Serious consideration should be given to constructing a pyloroplasty in all cases. A temporary gastrostomy is left for gastric decompression and transition to oral feeds.

Finally, the colocolostomy is created anterior to the vascular pedicle to re-establish colonic continuity. The thoracic dissection is drained with intrapleural and extrapleural catheters.

Esophageal patching with colon (colonic patch esophagoplasty)

5a–d Patching of an area of persistent esophageal stricture can be accomplished as an alternative to esophageal replacement. A thoracic approach, either extrapleural or intrapleural, is made, exposing the area of stricture. Via an abdominal incision, the colon is measured carefully and vascular pedicle created as if performing an esophageal replacement procedure. A segment based on the right colic artery is preferred. The esophageal stricture is then opened and the length of stricture to be patched is determined. A segment of colon equal to the length of the esophageal defect is measured. The segment of colon distal to the patch segment but adjacent to the mobilized vascular pedicle is removed. The marginal artery is carefully preserved by dividing vessels close to the wall of the segment of bowel to be discarded.



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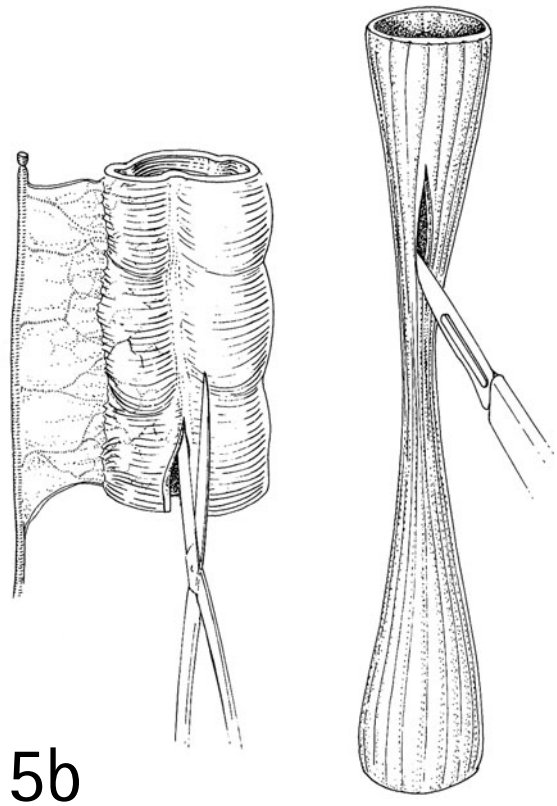

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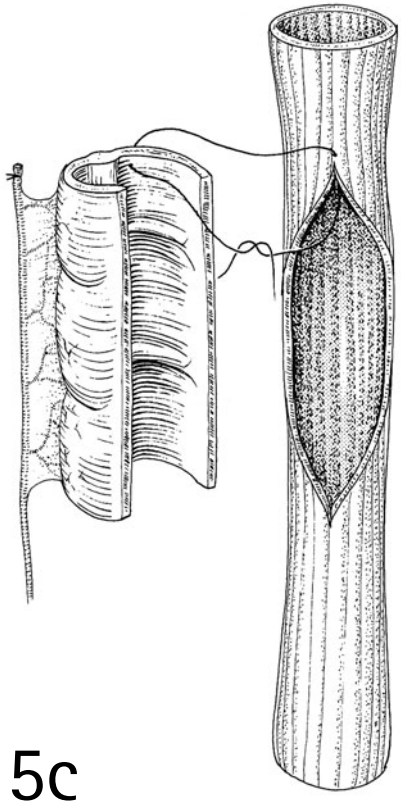

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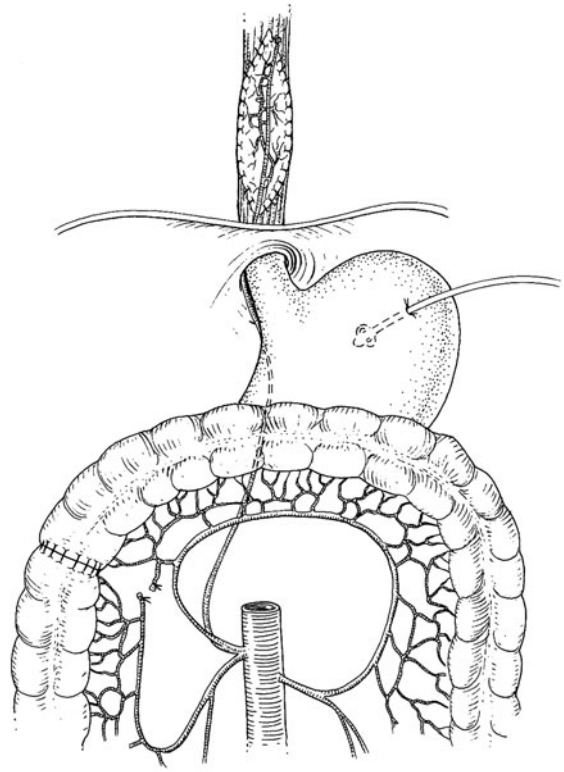

5d

The colon patch segment is opened along its antimesenteric border and the patch is created from a template of the esophageal defect. The patch is then passed behind the stomach through the esophageal hiatus. If the patch is too large, a pseudodiverticulum may develop. A single layer of interrupted or continuous absorbable suture is used to secure the patch in place. As with the cervical anastomosis in esophageal replacement, the integrity of the colon patch suture line can be tested for leaks by injection of an esophageal catheter with saline. The colocolic anastomosis, thoracic drainage, and gastrostomy are performed as in esophageal replacement. A pyloroplasty is not usually necessary.

POSTOPERATIVE CARE

A contrast swallow is performed on the seventh postoperative day. If no leaks are detected, oral feeding may be initiated and thoracic drainage catheters subsequently removed. As the child recovers, clinical evidence of dysphagia or obstruction should prompt a contrast swallow or endoscopy to detect anastomotic stricture. Identified strictures are usually amenable to dilatation with radial balloon expansion or tapered, weighted dilators under fluoroscopic guidance.

OUTCOME

Esophageal replacement with colon is a proven, satisfactory, durable replacement for the esophagus in children. Long-term outcomes after esophageal replacement with colon are good. Most patients report shortness of breath at times, while some suffer from slow transit of food in the colonic segment. Most patients who have undergone esophageal replacement with colon lead an otherwise normal life. Their quality of life is deemed acceptable, but not as good as that following primary anastomosis of the esophagus.

In the pediatric population, ischemic anastomotic strictures are not a common problem, as reported in many adult series. While many surgeons have turned to the stomach as the primary replacement conduit for the esophagus, esophageal replacement with colon maintains several advantages over the stomach. It has an excellent, mobile vascular supply via the marginal artery, provides adequate length to reach up into the chest, is the appropriate caliber for anastomosis, and avoids the problems of acid production in the conduit.

The main disadvantage of esophageal replacement with colon is that the colonic segment does not establish peristaltic contractions coordinated with swallowing as in the native esophagus. Contractions can be measured in the interposed colon in response to distension, but peristalsis coordinated with swallowing is not generally observed. Emptying of the colonic segment is primarily by gravity, and solid transit is slow. Consequently, the colon interposition can become redundant over time, leading to obstruction and stasis.

Unlike the colon interposition, the colon patch esophagoplasty retains the continuity of the esophagus. Injury to the vagus nerve is less common than with esophageal replacement. Problems related to redundancy and difficulty with emptying are not seen. Endoscopic evaluations of patients with colon patch have also demonstrated re-epithelialization and scar regression of the esophagus. This technique is particularly suited for isolated defects or strictures of the esophagus.

Disadvantages of the colon patch include a tendency for it to develop a pseudodiverticulum, but functional impairment has not been seen. The colon patch also includes a long intrathoracic suture line, thereby increasing the potential for perioperative leakage.

Although there is no ideal replacement for the esophagus, both esophageal replacement with colon and colonic patch esophagoplasty are proven, durable methods of establishing esophageal continuity in children.

Acknowledgment

The authors greatly appreciate the advice and mentorship of H. Biemann Othersen in our management of surgical problems of the esophagus and in the creation of this manuscript.

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19

Gastric replacement of the esophagus

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One of the alternatives for replacing the esophagus is gastric transposition involving the whole stomach. This method has the advantage of involving only one anastomosis, which is well vascularized and is associated with a low incidence of leakage.

HISTORY

Kummell in 1922 described the technique of gastric transposition via the mediastinal route in two patients, both of whom died. In 1945 Sweet recorded 12 esophageal resections with esophagogastric anastomosis above the aortic arch. Atwell (1980) reported on six children who underwent gastric transposition, with good long-term results in four. Gastric transposition is currently the procedure of choice for esophageal replacement in adults with esophageal carcinoma.

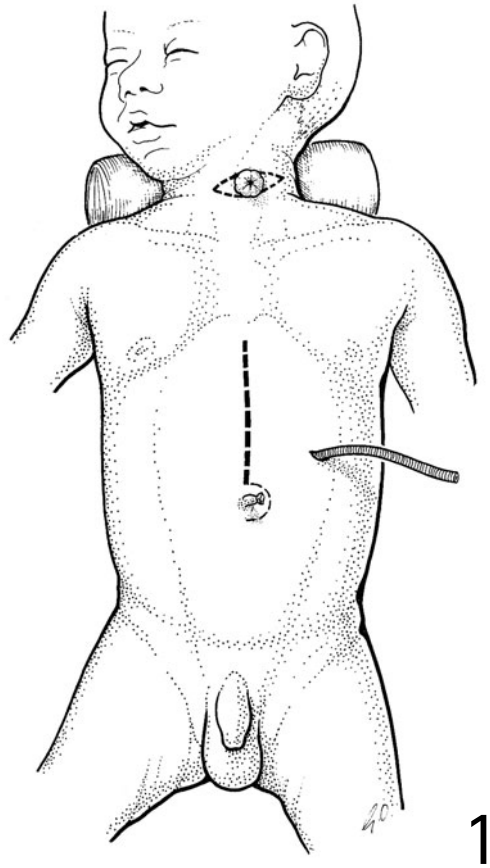
OPERATION

The procedure may be performed either by a thoracoabdominal approach or transhiatally via the posterior mediastinum without having to resort to a thoracotomy. This latter method will be described in detail.

The importance of sham feeds in infants with long-gap esophageal atresia who have undergone a cervical esophagostomy in simplifying the initiation of oral nutrition following the interposition should not be underestimated.

Mediastinal gastric transposition

The initial feeding gastrostomy should ideally have been sited on the anterior surface of the body of the stomach, well away from the greater curvature, in order to preserve the vascular arcades of the gastroepiploic vessels.



1

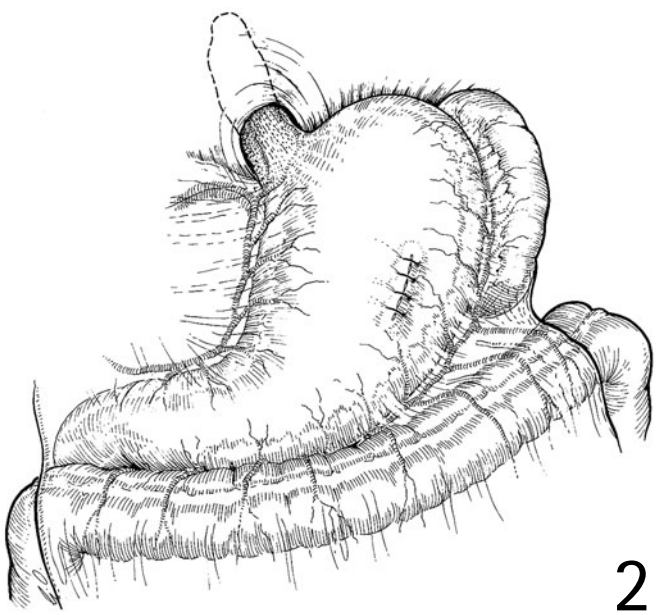
INCISION

1 The preferred approach is via a midline upper abdominal incision extending from the xiphisternum to the umbilicus. Alternatively, a left upper abdominal transverse muscle-cutting incision may be used.

The gastrostomy is carefully mobilized from the anterior abdominal wall and the defect in the stomach closed in two layers with interrupted 4/0 polyglycolic acid sutures.

MOBILIZING THE STOMACH

Adhesions between the stomach and the left lobe of the liver are released, taking care not to damage any of the major blood vessels.



2

2 The greater curvature of the stomach is mobilized by ligating and dividing the vessels in the gastrocolic omentum and the short gastric vessels. These vessels should be ligated well away from the stomach wall in order to preserve the vascular arcades of the right gastroepiploic vessels. Meticulous care must be exercised to avoid damaging the spleen.

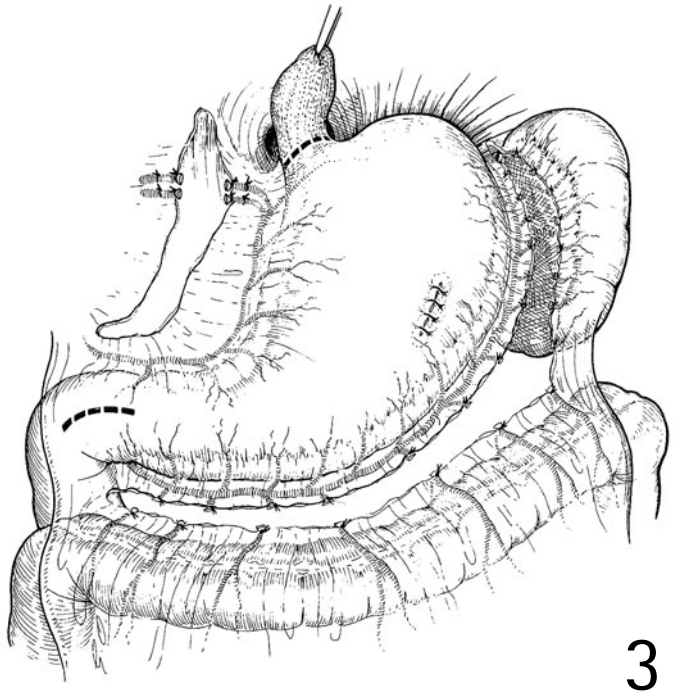
The lesser curvature of the stomach is freed by dividing the lesser omentum from the pylorus to the diaphragmatic hiatus. The right gastric artery is carefully identified and preserved, while the left gastric vessels are ligated and divided close to the stomach. The lower esophagus is exposed by dividing the phrenoesophageal membrane, and the margins of the esophageal hiatus in the diaphragm are defined.

RESECTION OF THE DISTAL ESOPHAGUS

3 The inevitably short, blind-ending lower esophageal stump is dissected out of the posterior mediastinum by a combination of blunt and sharp dissection through the diaphragmatic hiatus. The anterior and posterior vagal nerves are divided during this part of the procedure. The body and fundus of the stomach are now free from all attachments and can be delivered into the wound.

The esophagus is transected at the gastroesophageal junction and the defect closed in two layers with 4/0 polyglycolic acid sutures.

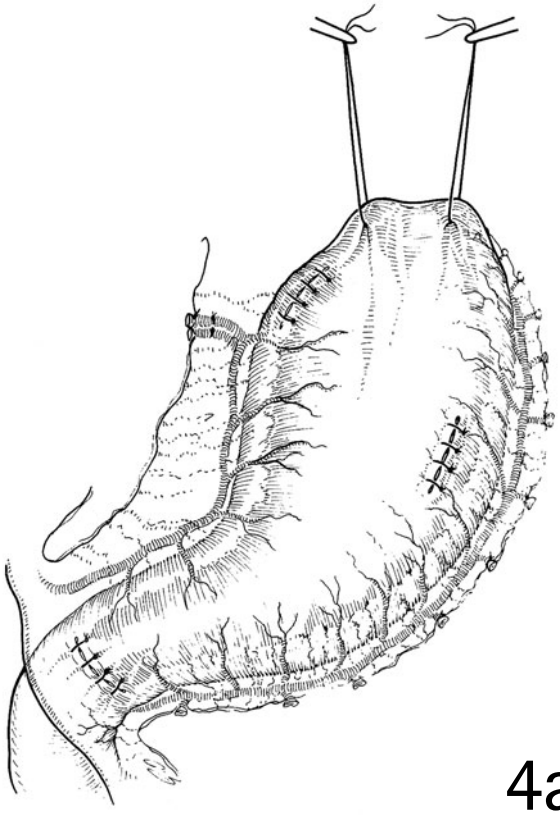
The second part of the duodenum may be Kocherized to obtain maximum mobility of the pylorus.



3

PREPARING FOR GASTROESOPHAGEAL ANASTOMOSIS

4a,b The highest part of the fundus of the stomach is identified and stay sutures of different material are inserted to the left and the right of the area selected for the anastomosis. These sutures help to avoid torsion of the stomach occurring as it is pulled up through the posterior mediastinum into the neck.



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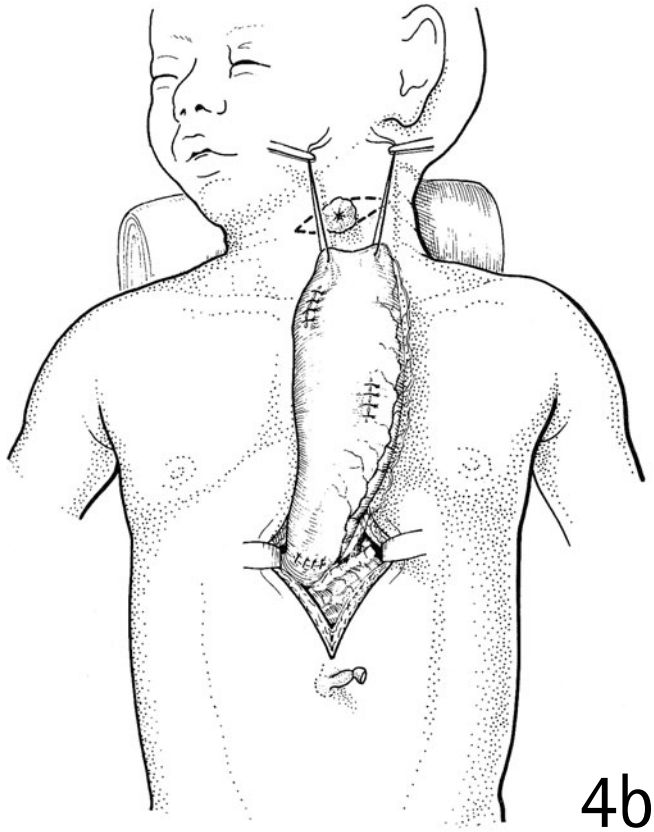


4b

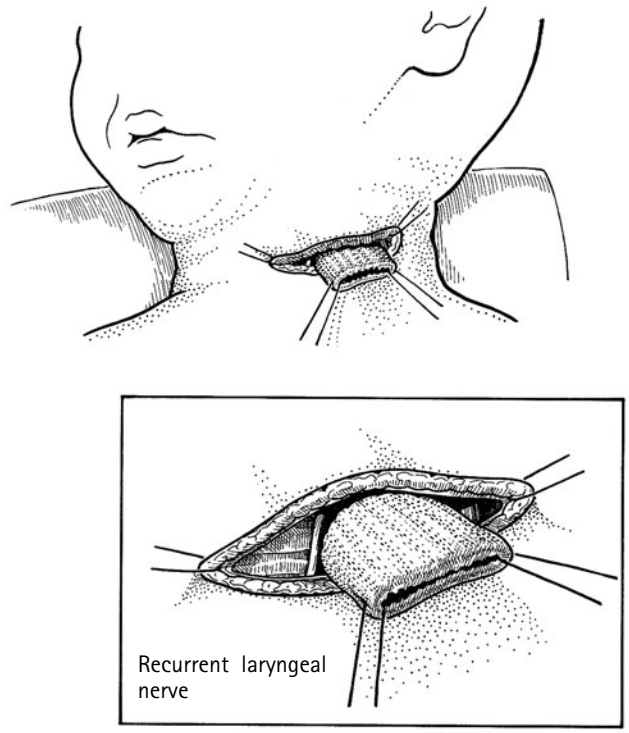
PYLOROPLASTY

A short Heinecke–Mikulicz pyloroplasty is performed, the transverse incision being closed horizontally with interrupted fine polyglycolic acid sutures. An alternative is to perform a short pyloroplasty.

The stomach is now ready for its transposition into the neck.

MOBILIZATION OF THE CERVICAL ESOPHAGUS

5 Attention is now turned to the neck, where the previously constructed cervical esophagostomy (preferably performed on the left side) is mobilized via a 3–4 cm transverse incision, taking care not to damage the muscular coat of the esophagus. The recurrent laryngeal nerve coursing upwards on the posterolateral surface of the trachea is identified and preserved. It is important to mobilize at least 1–1.5 cm full-thickness esophagus to allow a satisfactory anastomosis to take place.



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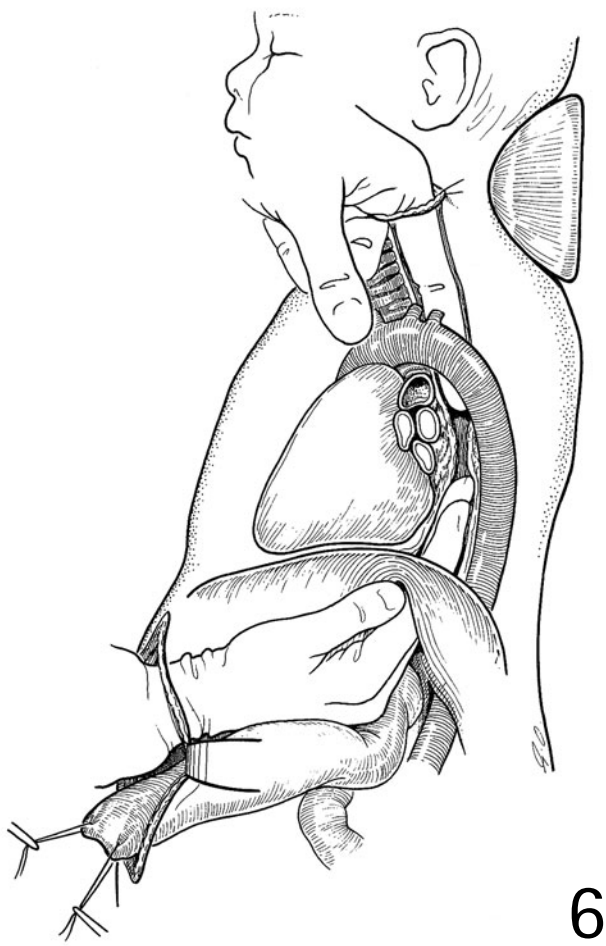


6

PREPARING THE POSTERIOR MEDIASTINAL TUNNEL

6 A plane of dissection between the membranous posterior surface of the trachea and the prevertebral fascia is established, and a tunnel is created into the superior mediastinum by blunt dissection immediately in the midline.

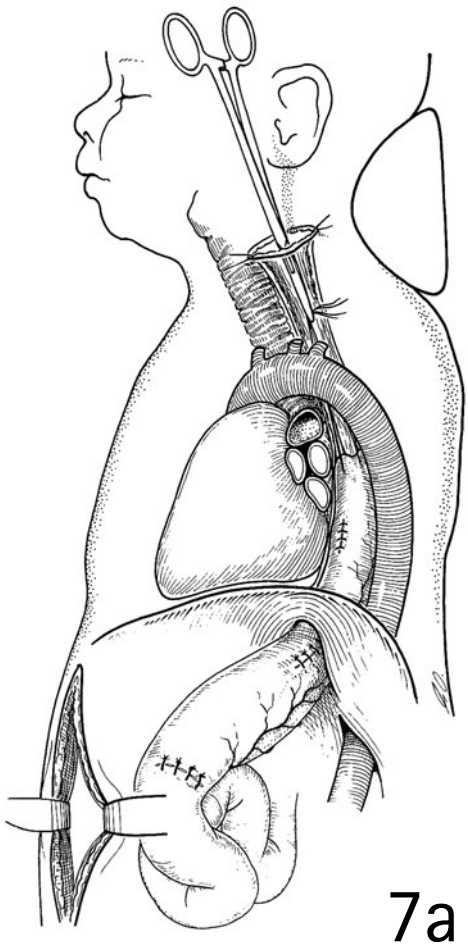
A similar tunnel is fashioned from below in the line of the normal esophageal route, by means of blunt dissection through the esophageal hiatus in the posterior mediastinal space posterior to the heart and anterior to the prevertebral fascia.

When continuity of the superior and inferior posterior mediastinal tunnels has been established, the space to be occupied by the stomach is developed into a tunnel of two to three fingers' breadth.

There will be occasions when fashioning of the posterior mediastinal tunnel by blind dissection is impossible or hazardous due to inflammation, fibrosis from previous surgery, or adhesions following previous perforation or caustic ingestion. Under these circumstances, it is necessary to perform a lateral thoracotomy and for the dissection to be carried out under direction vision.

TRANSPOSING THE STOMACH

7a,b A wide-caliber nasogastric tube is passed through the posterior mediastinal tunnel from the cervical incision to appear via the esophageal hiatus into the abdominal wound. The two stay sutures on the fundus of the stomach are tied to the tube, which is then gently withdrawn, pulling the stomach up through the esophageal hiatus and the posterior mediastinal tunnel into the cervical incision. Orientation of the fundus is checked by realigning the stay sutures in their correct position.



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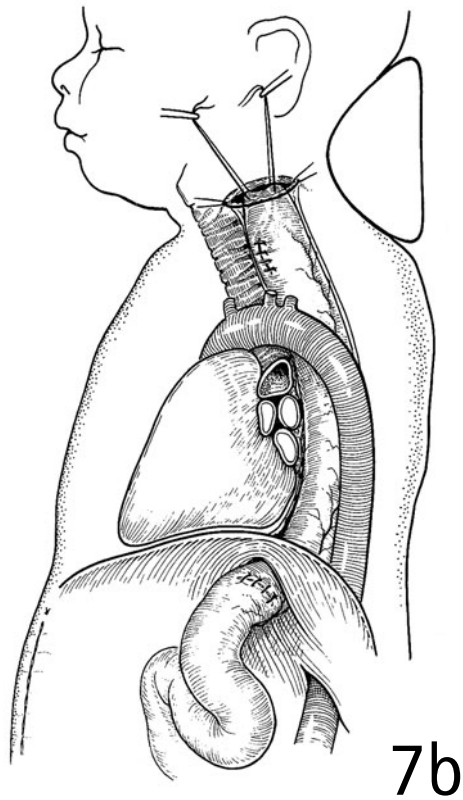
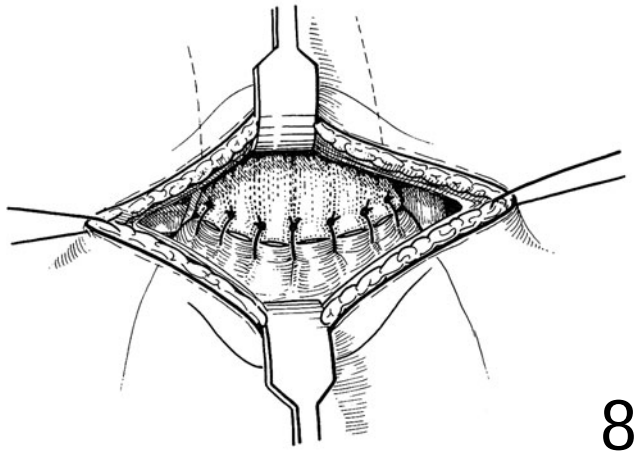


7b

GASTROESOPHAGEAL ANASTOMOSIS

8 The transected end of the cervical esophagus is now anastomosed to the highest part of the stomach using a full-thickness single layer of interrupted 4/0 polyglycolic acid sutures.



8

A 10–12 Fr transanastomotic nasogastric tube is inserted into the intrathoracic stomach before completing the anterior layer of the anastomosis. This is left on free drainage and aspirated at regular intervals to prevent acute gastric dilatation in the early postoperative period.

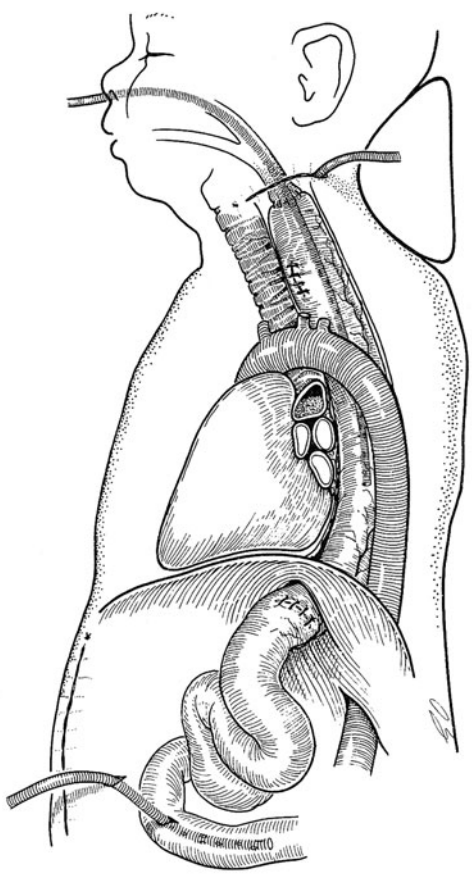
WOUND CLOSURE

A soft rubber drain may be placed at the site of the anastomosis in the neck and the wound is closed in layers.

The margins of the diaphragmatic hiatus are sutured to the antrum of the stomach with a few interrupted sutures – 4/0 polyglycolic acid or braided polyamide (Nurolon) – so that the pylorus lies just below the diaphragm.

A fine-bore feeding jejunostomy has been found to be of considerable value in providing enteral nutrition in the first few weeks following gastric transposition, before full oral nutrition is established.

The abdominal incision is closed en masse or in layers.



9

Final anatomy

9 The gastroesophageal anastomosis is shown in the cervical region with the nasogastric tube passing into the intrathoracic stomach. The pyloroplasty is below the diaphragm and a feeding jejunostomy tube is inserted for postoperative feeding. This is particularly important for infants with esophageal atresia who have previously not acquired the skill of oral feeding. The jejunostomy is a source of potential complications and it is probably wise to omit it in older children who have previously taken full oral nutrition, e.g., caustic strictures.

POSTOPERATIVE CARE

Careful monitoring of vital functions is essential in the early postoperative period. There has been a fairly extensive dissection in the tissues posterior to the trachea, and edema may produce respiratory embarrassment. Elective nasotracheal intubation with assisted ventilation for a few days will simplify the postoperative course and reduce the incidence of respiratory problems.

Jejunal feeds are instituted on the second or third day after operation. The safest method of delivery of these feeds is by a slow continuous infusion rather than as a bolus, which can provoke a 'dumping' effect. A contrast swallow is performed 5–7 days after surgery, and if no leak is identified at the anastomosis, oral feeding may be commenced. The cervical drain is removed when the integrity of the anastomosis has been demonstrated.

COMPLICATIONS

In the period 1981–2005, 192 gastric transpositions were performed at Great Ormond Street Hospital, London. Fifty-two percent were via the posterior mediastinal route without thoracotomy. The mortality rate was 5.2 percent. Anastomotic leakage occurred at the esophagogastric connection in 12 percent, and strictures developed in 19 percent. All leaks except one closed spontaneously and all strictures except three responded to endoscopic dilatation. Strictures were more common (38 percent) after caustic ingestion.

Delayed gastric emptying and dumping syndrome also occurred in some patients, but usually resolved spontaneously within a few months. Feeding problems and recurrent vomiting are commonly encountered in the early postoperative period, but are generally transient.

OUTCOME

Good to excellent results are achieved in 90 percent of patients. Growth and development do not appear to be affected, and respiratory function is not impaired.

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20

Congenital diaphragmatic hernia

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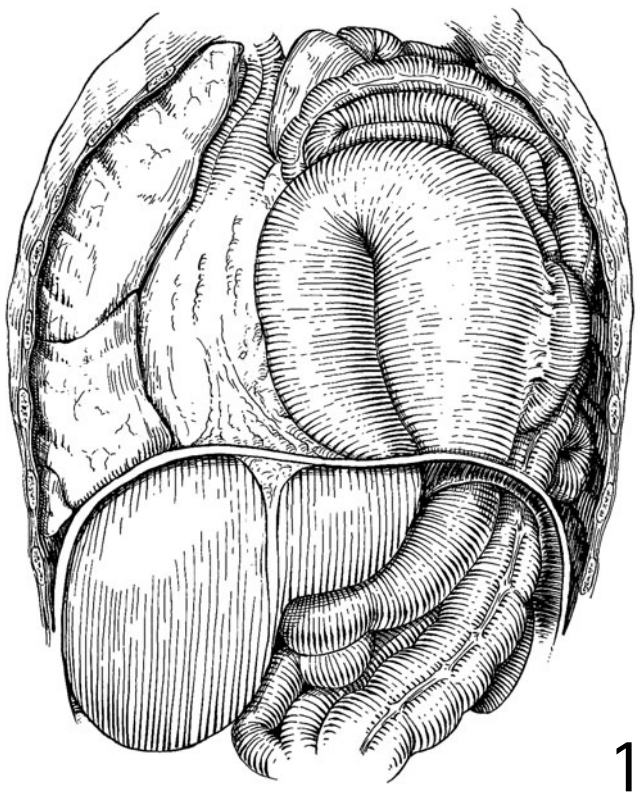
HISTORY, ETIOLOGY, AND PATHOPHYSIOLOGY

Although the earliest descriptions of congenital diaphragmatic hernia were by Paré, the concept of an embryologic etiology was first discussed by Bochdalek in 1848, who erroneously thought that the diaphragm ruptured after formation.

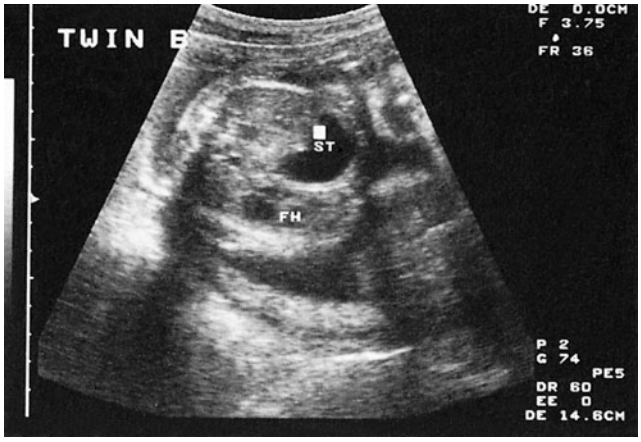
The current view is that the defect is an inherent abnormality of the lungs that results in a secondary abnormality of the diaphragm, or a failure of the diaphragm to separate the pleuroperitoneal canal into the thorax and abdomen before the midgut returns from the umbilicus. Alternatively, the midgut may return too soon. As a consequence, translocation of abdominal viscera into the chest occurs in the first trimester when the lungs are at a very vulnerable lung bud/glandular stage.

The resultant abnormality leads to disordered lung growth. Both lungs are affected, the ipsilateral more so than the contralateral lung. The consequent structural abnormality features compromised bronchiolar and pulmonary arterial divisions, muscular hypertrophy of the intra-acinar arterioles, and a decreased surface available for gas exchange. Affected infants are born with a complex interface of pulmonary hypoplasia and pulmonary hypertension. Pulmonary hypoplasia can be severe enough to preclude life outside the womb, whereas successful management of pulmonary hypertension can lead to a fruitful life.



DIAGNOSIS

2a–c The typical infant with congenital diaphragmatic hernia presents immediately after birth with the stigmata of respiratory distress: tachypnea, grunting, and cyanosis. The infant has usually reached full term, and has a scaphoid abdomen and barrel chest. With increasing frequency, the diagnosis of this condition is made with prenatal ultrasound. Ultrasound examination shows a fetus with mediastinal shift, bowel and/or liver in the chest, no intra-abdominal stomach, or the heart and stomach in the same plane. At birth, the diagnosis is confirmed by a chest radiograph, which will show multiple gas-filled loops of bowel and contralateral shifting of the mediastinum. There is an absence of bowel gas in the abdomen. (ST, stomach; FH, fetal heart; DPH, diaphragm; LUR, liver.)



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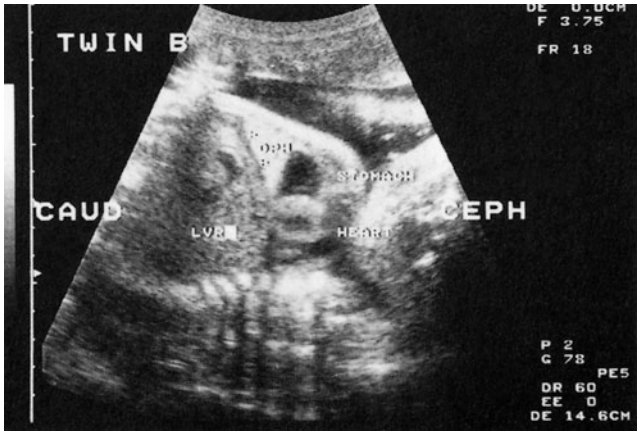

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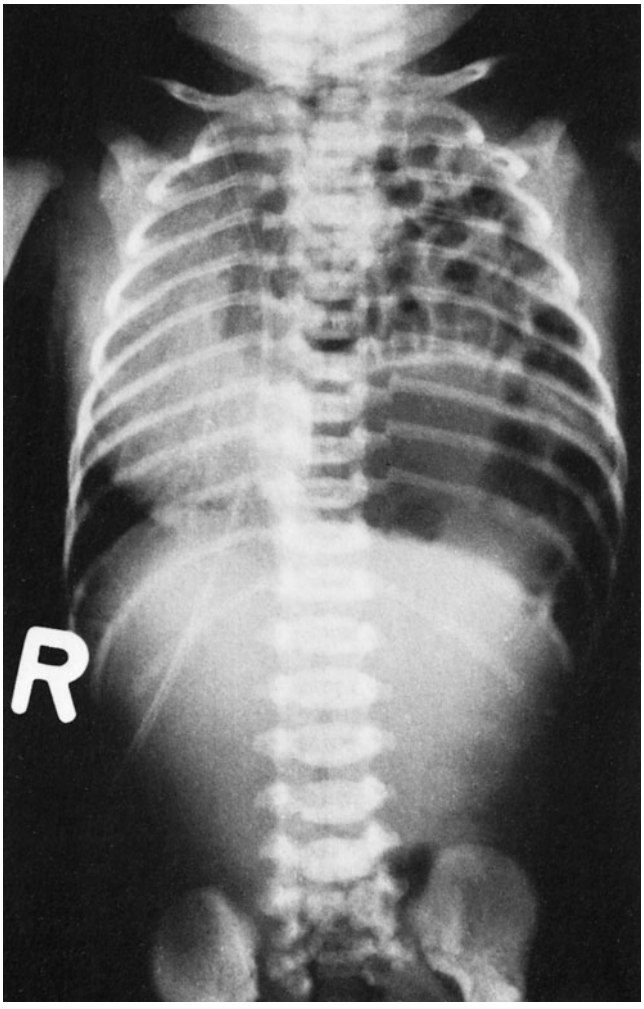


2c

PREOPERATIVE

Resuscitation

When the condition is diagnosed before birth, the mother is transferred to a level III neonatal center capable of appropriate neonatal, surgical, and extracorporeal membrane oxygenation (ECMO) care. If diagnosed after birth, the infant is stabilized and then transferred to the same sort of center before operation.

Traditionally, diagnosis of congenital diaphragmatic hernia made emergency operation mandatory. Recent experience, however, has clearly demonstrated that most infants with this condition can be stabilized over several days and undergo elective surgery. Infants who cannot be stabilized may have pulmonary hypoplasia to a degree incompatible with life, but may be considered for resuscitation with ECMO. The resuscitation strategy is based on prompt intubation, nasogastric decompression, and arterial and venous vascular access. Respiratory care should preclude muscle paralysis and allow spontaneous respiration to minimize barotrauma. Ventilator settings range from low rates and modest peak airway pressure to higher rates with lower peak airway pressures, and to oscillating ventilation. Pharmacologic support may consist of pulmonary vasodilators such as dobutamine, tolazoline, or nitric oxide. An umbilical artery catheter is placed for blood pressure monitoring and blood sampling, and preductal and postductal (Sa_{O_2}) cutaneous monitors are attached. Placement of a preductal arterial catheter is frequently critical. Because most of these infants have significant right-to-left shunting at the ductus arteriosus level, blood gas monitoring from the umbilical artery is often misleading and can lead to premature surgery or inordinate use of ECMO. Assessment of P_{O_2} and P_{CO_2} in the preductal location is a more accurate assessment of the lungs' ability for meaningful gas exchange. Extracorporeal membrane oxygenation is useful in the preoperative infant who has already demonstrated evidence of adequate lung function but who then deteriorates because of pulmonary hypertension. Such an infant is unlikely to tolerate surgery well.

Anesthesia

The airway is controlled with an appropriately sized orotracheal or nasotracheal tube (a nasotracheal tube is preferred because of the potentially lower incidence of airway complications). Volatile anesthesia is administered as needed and is complemented by muscle paralysis and narcotics. Mechanical ventilation is controlled throughout surgery with a pressure-cycled infant ventilator rather than the conventional anesthesia machine. Continuous Sa_{O_2} monitoring, both preductal and postductal, is critical.

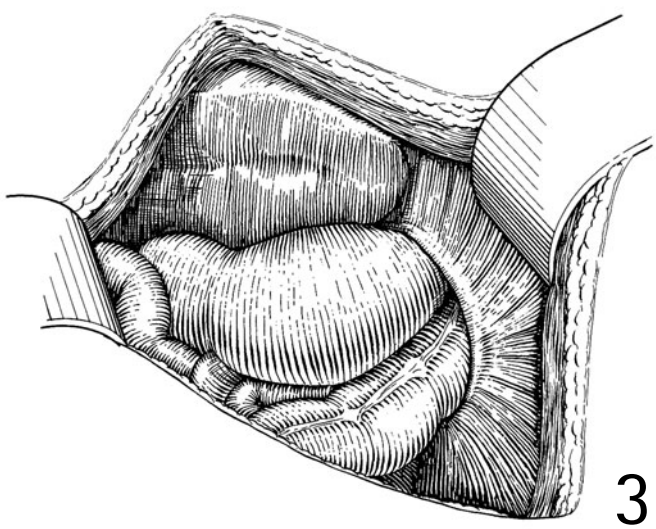
OPERATION

Position of patient

The infant is positioned supine on a heating mattress with a small elevating pad beneath the thoracolumbar spine. The extremities and head are wrapped to minimize heat loss. Both the upper abdomen and chest are prepared as the operating fields. The entire operation is performed as much as possible with electrocautery because of the potential need for ECMO and heparin.

Incision

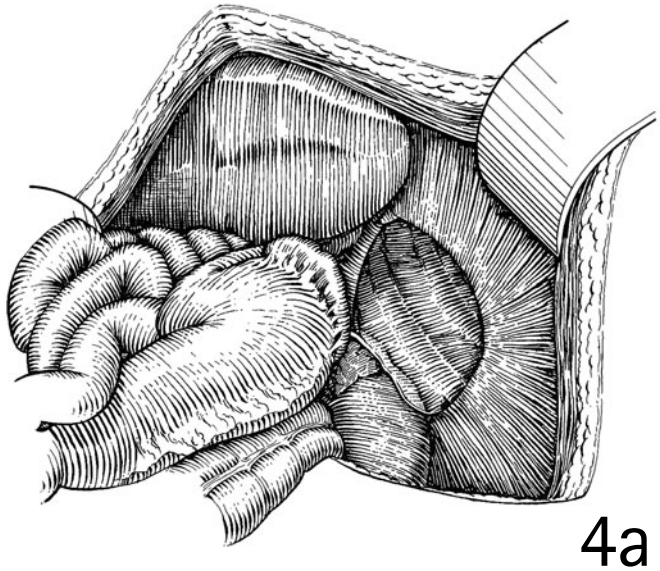
A subcostal incision is made on the side of the hernia.



3

3 The cephalad portion of the incision and the anterior rim of the diaphragm are elevated to expose the defect.

Suction of the stomach is performed via the nasogastric tube, and the viscera are carefully reduced. The unfixed spleen and its tenuous attachments to the colon and pancreas are especially vulnerable. Reduction of the liver may be equally challenging.



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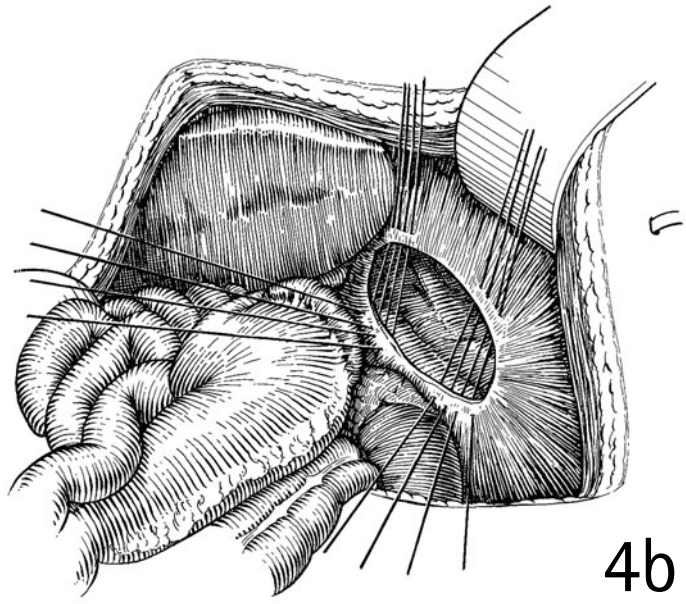

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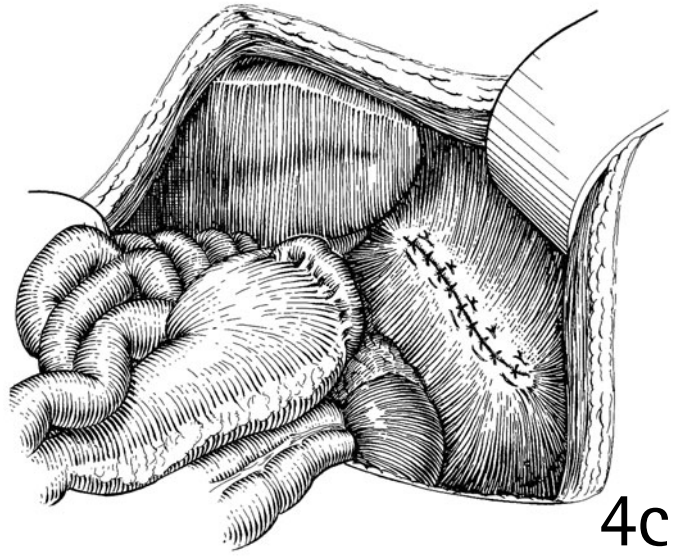


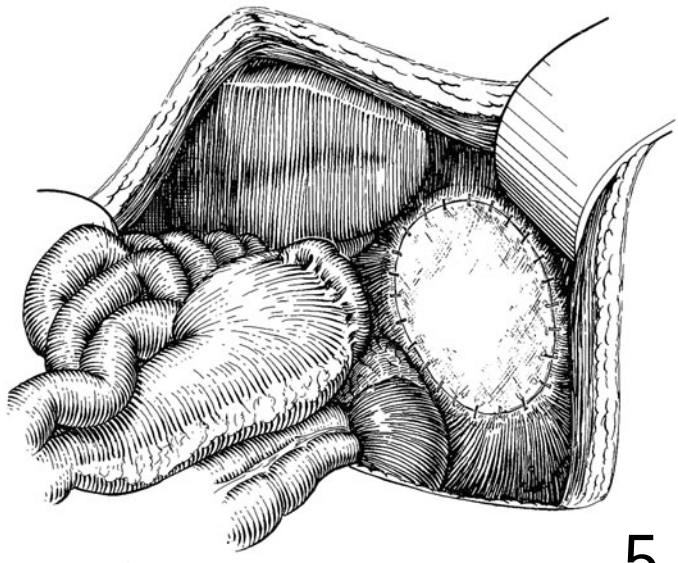
4c

4a–c Once the hernia has been reduced, the viscera are allowed to lie on the abdominal wall. If a true hernia sac is found, it must be excised to ensure proper healing of the defect. The posterior rim can be found by tracing the anterior rim around medially. Its mesothelial covering is sharply incised and carefully mobilized. If primary closure is possible, it is worth pursuing the posterior rim. If a prosthetic patch is required, dissection should be kept to a minimum. Primary repair is accomplished with interrupted simple sutures of a non-absorbable material.

5 A prosthetic patch can be constructed using 1 mm thick Gore-Tex, which is tailored to size and secured with interrupted sutures. Laterally the patch can be anchored to the ribs. A paucity of medial tissue requires the surgeon to be creative.

Once the diaphragm has been constructed, an ipsilateral tube thoracostomy is not needed unless there is bleeding or pneumothorax; it is not required for prophylaxis.

Correction of the unrotated midgut by division of Ladd's bands with or without inversion appendicectomy should be discouraged if ECMO is contemplated or in use unless mechanical obstruction is already present. Postoperative volvulus is rare, but significant bleeding after heparinization may be a major problem.



5

Wound closure

Closure of the abdominal wall can be challenging because of loss of abdominal domain. Vigorous stretching of the abdominal wall should be avoided. If the abdomen cannot be closed safely in layers without compromising venous return, a silo in the manner of omphalocele management is preferred, with subsequent reduction over several days.

POSTOPERATIVE CARE

The therapeutic strategy used in the preoperative period is reinstituted in the postoperative period. Muscle paralysis is discontinued to allow spontaneous respiration. Adequate analgesia with narcotics is mandatory. Sufficient intravenous fluids are given to maintain adequate circulating blood volume and hemoglobin for oxygen delivery. As the infant recovers, it is weaned from a mechanical respirator to nasal prong continuous positive airway pressure. A prolonged recovery will need to be complemented by parenteral nutrition.

EXTRACORPOREAL MEMBRANE OXYGENATION

Although this therapy is discussed in detail in Chapter 21 (see pp. 159–65), there are some special considerations for ECMO in the infant with congenital diaphragmatic hernia. Because ECMO may be appropriate both before and after operation, these infants should be transferred to an ECMO center as soon as possible after diagnosis and the infant (or fetus) is stable enough for transport. It must be remembered that ECMO requires heparin and that any dissected or stretched surface may bleed.

Occasionally an infant with congenital diaphragmatic hernia will need ECMO as part of the preoperative resuscitation. In this case, surgery will be performed in the neonatal intensive care unit while ECMO is in progress. The operative technique is similar, but meticulous hemostasis is critical.

FUTURE DIRECTIONS

Although laproscopic and thoracoscopic repair of congenital diaphragmatic hernia has been done and reported, at this point in time it is certainly not the standard of care in the neonate. On the other hand, it is reasonable to use a minimally invasive approach in the older infant or child.

FURTHER READING

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21

Extracorporeal membrane oxygenation: neonatal vascular cannulation

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HISTORY

The term extracorporeal membrane oxygenation (ECMO) has been used to describe a method of extracorporeal life support (ECLS) using extrathoracic cannulation for cardiopulmonary support. Extracorporeal life support is a supportive rather than a therapeutic intervention. It provides adequate perfusion and gas exchange (venoarterial bypass) or gas exchange alone (venovenous bypass), and so avoids deleterious effects from high oxygen concentration and positive pressure ventilation while allowing resolution of reversible heart and lung pathology.

PRINCIPLES AND JUSTIFICATION

Vascular access for ECLS in the neonate is particularly challenging due to the small vessel size. The route of access depends on the method used. Venoarterial (VA) bypass is indicated if both cardiac and pulmonary support are required, and in neonates where access for venovenous (VV) support cannot be obtained.

Venovenous bypass is the method of choice for pulmonary support and can adequately support most infants, including those with depressed cardiac function from high pressure ventilation used to manage their severe respiratory failure prior to ECMO. It is more physiologic than VA and provides well-oxygenated blood to the pulmonary circulation, which acts as a potent vasodilator to reduce right-to-left shunting. It also obviates the need for arterial cannulation and thereby lowers the risk of arterial embolization and from carotid ligation or repair. Various access sites, including the umbilical, femoral, and carotid/jugular vessels, have been used.

For VA access, the preferred site is the right atrium via the right internal jugular vein for venous drainage, and the aortic arch via the right common carotid artery for arterial infusion. The internal jugular vein and carotid artery are relatively large in the neonate and may be distally ligated without major sequelae.

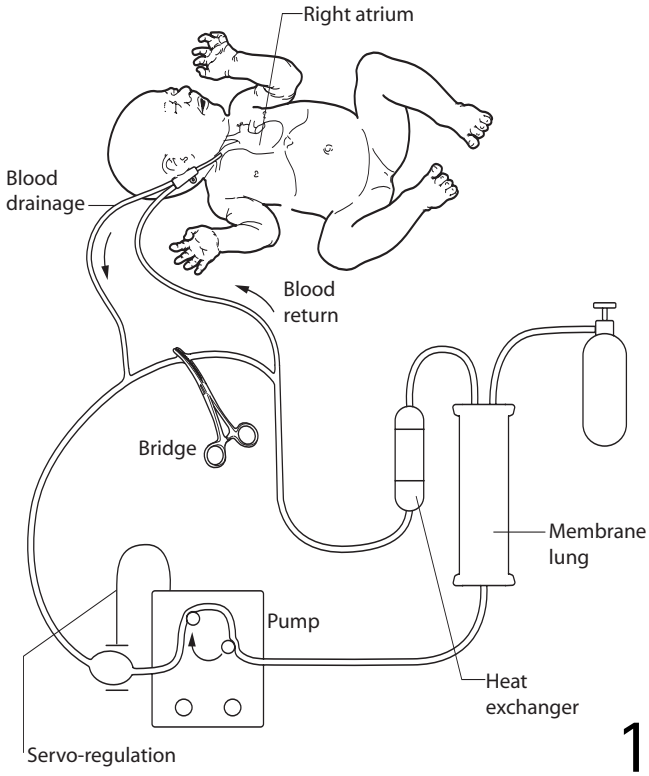
For VV access, a double-lumen cannula is placed into the right atrium via the right internal jugular vein. This technique is limited by the size of the vein, because the smallest cannula currently available is 12 Fr.

Selection of technique

Venarterial bypass requires arterial ligation to prevent distal embolization from flow past the cannula. Venovenous bypass can be performed either using this technique or without vessel ligation via a percutaneous or semi-open technique. Percutaneous access utilizes the Seldinger technique to place the cannula. Because the size of the vessel in relation to the cannula is unknown, vessel disruption is a risk. For this reason, our preferred method is the semi-open technique. This technique requires a small incision to visualize the size of the vein as an aid to selecting the correct cannula size (usually 12 Fr or 15 Fr in a newborn). Cannula insertion can also be visualized through this incision. With this technique, vessel ligation is not utilized. This has several advantages: cephalad flow into the cannula increases the amount of deoxygenated blood available to enter the bypass circuit, the vessel may remain patent after decannulation; and kinking of the cannula at the vessel is reduced.

Extracorporeal membrane oxygenation circuit

1 Blood is drained from the right atrium via the venous cannula into a small bladder by siphon/gravity. As long as venous drainage is adequate to fill the bladder, blood enters the raceway (tubing within the roller pump head) where it is actively pumped into the membrane lung. Here, blood travels in a countercurrent fashion to the sweep gas, separated by a thin silicone membrane. Oxygen enters the blood and carbon dioxide enters the sweep gas by simple diffusion along a concentration gradient. Blood then enters the heat exchanger, where it is warmed to body temperature before entering the arterial cannula. This blood may either enter the arterial (VA bypass) or venous (VV bypass) system.



1

PREOPERATIVE

Vascular cannulation and decannulation are performed in the neonatal intensive care unit under adequate sedation and neuromuscular blockade. Neuromuscular blockade is especially important in preventing the potentially lethal complication of an air embolus during introduction of the venous cannula. The instruments and sterile procedures used are identical to those used in the operating room. Heparin sodium (100 units/kg) is drawn up for subsequent administration.

Anesthesia

Local anesthesia is administered by infiltration of 1 percent lidocaine (lignocaine).

OPERATION

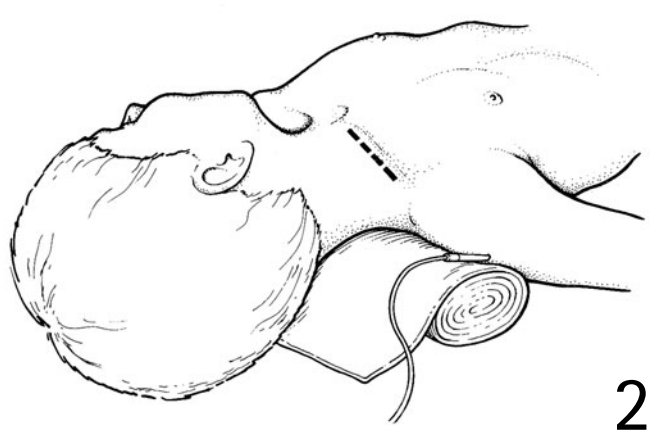
Position of patient

The patient is placed supine with the head turned to the left. A roll is placed transversely beneath the shoulders. Special attention is paid to assuring that the endotracheal tube is positioned to prevent kinking under the drapes during the procedure. This can be accomplished by using a piece of suction tubing split lengthwise and placed over the tube at the connector to prevent kinking. The chest, neck, and right side of the face are aseptically prepared and draped.

Venoarterial/venovenous cannulation: open technique

INCISION

2 A transverse cervical incision approximately 2–3 cm in length is made one finger's breadth above the clavicle over the lower aspect of the right sternocleidomastoid muscle.



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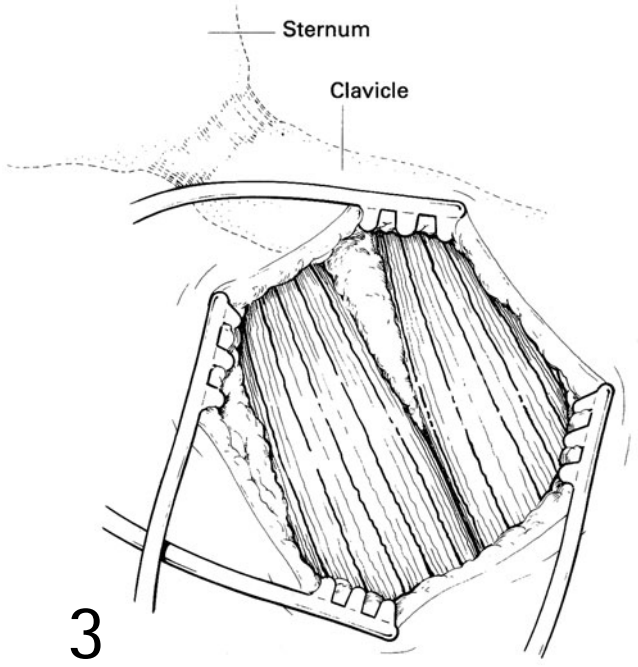

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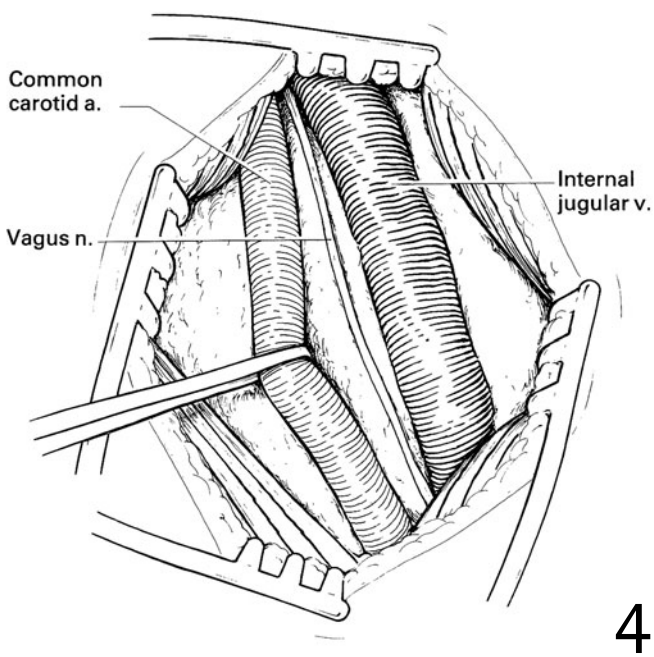
EXPOSURE OF THE CAROTID SHEATH

3 The platysma muscle and subcutaneous tissues are divided with electrocautery and the sternocleidomastoid muscle exposed. Dissection is continued bluntly between the sternal and clavicular heads of the muscle. The omohyoid muscle will be seen superiorly. It may be necessary to divide the omohyoid muscle tendon to expose the carotid sheath. Two alternating self-retaining retractors are placed.

DISSECTION OF THE VESSELS

4 The carotid sheath is opened and the internal jugular vein, common carotid artery, and vagus nerve are identified and isolated. Dissection is progressed proximally and distally along the vessels, dissecting the vein first. Special care should be taken while dissecting the vein to avoid induction of spasm, which makes subsequent introduction of a large venous cannula difficult. Manipulation of the vein should therefore be minimized. There is often a branch on the medial aspect of the internal jugular vein, which must be ligated. Ligatures of 2/0 silk are placed proximally and distally around the internal jugular vein. The common carotid artery lies medial and posterior and has no branches, which makes its dissection proximally and distally safe. Ligatures of 2/0 silk are also placed around the carotid artery. The vagus nerve should be identified.

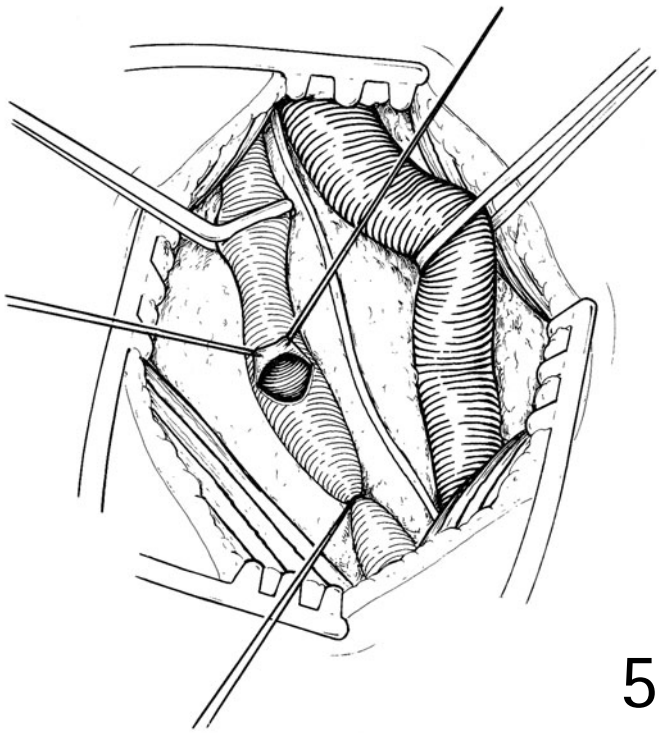
Once vessel dissection is completed, heparin (100 units/kg) is administered intravenously and 3 minutes allowed for circulation. During this waiting period, papaverine is instilled into the wound to enhance vein dilatation.



4

Arteriotomy/venotomy

5 For VA bypass, the arterial cannula is chosen (usually 10 Fr) and marked with a 2/0 silk ligature, left uncut, at a point that will allow the tip of the cannula to lie at the ostium of the brachiocephalic artery (about 2.5 cm). The venous cannula (usually 12–14 Fr) is similarly marked at a point equal to the distance from the venotomy to the right atrium (roughly 6 cm). An obturator is placed into the venous cannula to prevent blood from flowing out through the side holes during introduction into the vessel. The common carotid artery is ligated distally. Proximal control is obtained with the use of an angled ductus clamp. A transverse arteriotomy is made near the distal ligature. Full-thickness stay sutures of 6/0 polypropylene are placed on the proximal edge of the artery to prevent subintimal dissection during cannula insertion. Following arterial cannulation, a venotomy is performed in a similar fashion. Gentle retraction of the caudal ligature around the vein precludes the need for a ductus clamp during venotomy and venous cannulation. Stay sutures are also not routinely necessary for venous cannulation.



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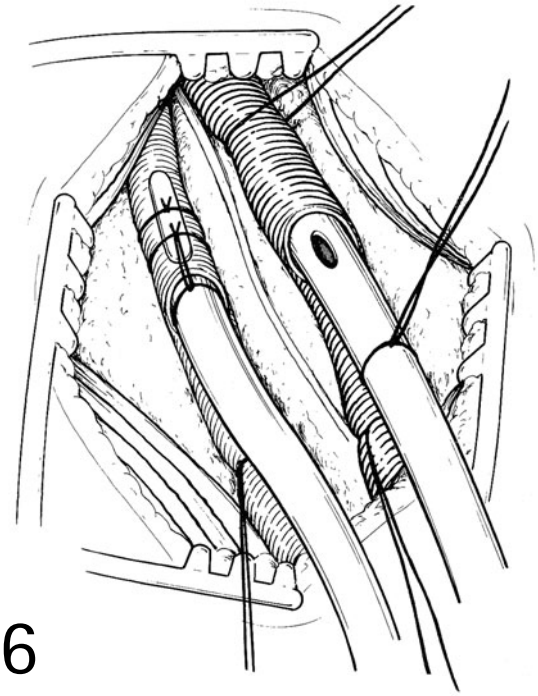


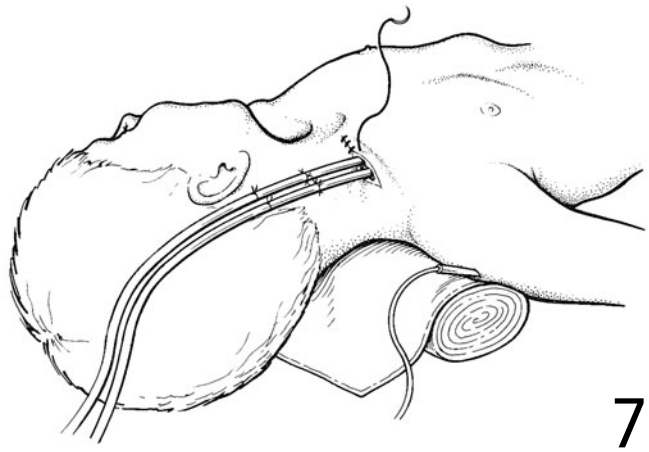
6

CANNULA PLACEMENT

6 The cannulae are carefully placed into the artery and vein and secured using two circumferential 2/0 silk ligatures, with a small piece of silicone rubber vessel loop inside the ligatures to protect the vessels from injury during decannulation when the ligatures are sharply divided. The ends of the marking ligatures are tied to the most distal circumferential ligature for extra security. Immediately after each cannula is secured, it is carefully debubbled via back-bleeding and filling with heparinized saline. For VV bypass, the double-lumen cannula is placed into the venotomy and advanced 5.5 cm. It is crucial to maintain the arterial reinfusion (red) port anteriorly while securing for proper orientation to minimize the recirculation of reinfused blood.

WOUND CLOSURE

7 The wound is irrigated with saline, and hemostasis obtained. The skin is closed with continuous monofilament suture. The wound is dressed with gauze. The cannula is sutured to the skin with several 2/0 silk sutures. Special attention should be directed to affixing the cannulae securely to the bed.



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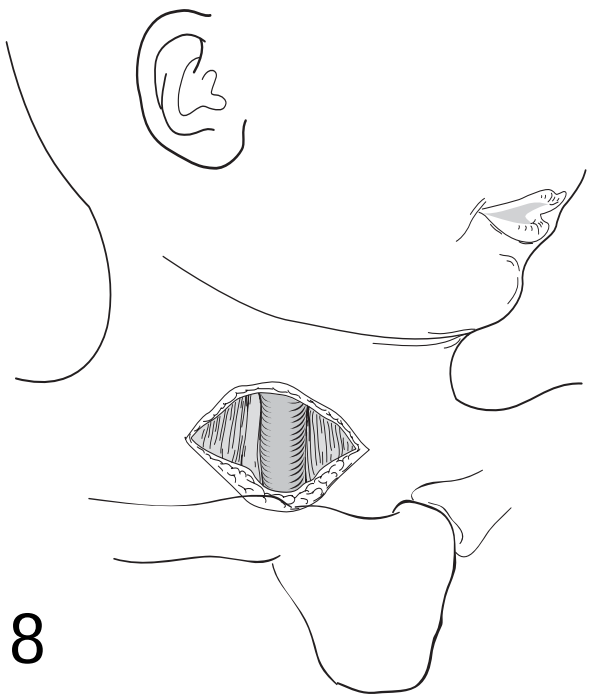


8

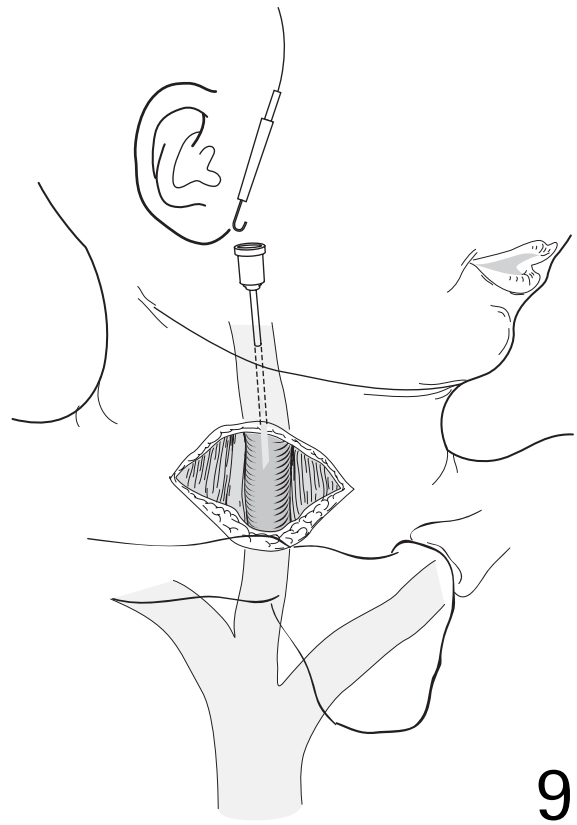
Venovenous cannulation: semi-open technique

INCISION AND VEIN EXPOSURE

8 A transverse cervical incision approximately 1.5–2 cm in length is made 2 cm above the right clavicle between the heads of the sternocleidomastoid muscle. The platysma is divided with electrocautery and the anterior surface of the internal jugular vein is exposed with minimal dissection. The vessel is observed and either a 12 Fr or 15 Fr Origen venovenous ECMO cannula (Origen Biomedical Inc., Austin, Texas, USA) is selected.

GUIDEWIRE PLACEMENT

9 The cannula skin exit position is selected so that the cannula will lie behind the right ear when the head is returned to the midline. The needle/catheter is placed through the skin 2 cm superior to the incision and into the internal jugular vein to enter either under the skin flap or just inside the incision. The needle is removed and a 0.035 inch diameter guidewire is advanced and the catheter is withdrawn. A Teflon guiding obturator is placed over the guidewire into the vessel and right atrium. The skin exit is slightly enlarged with a scalpel.



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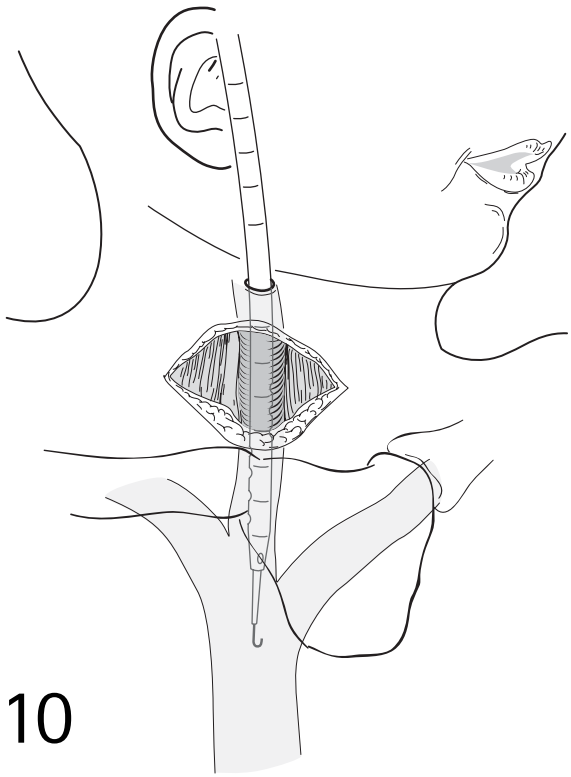
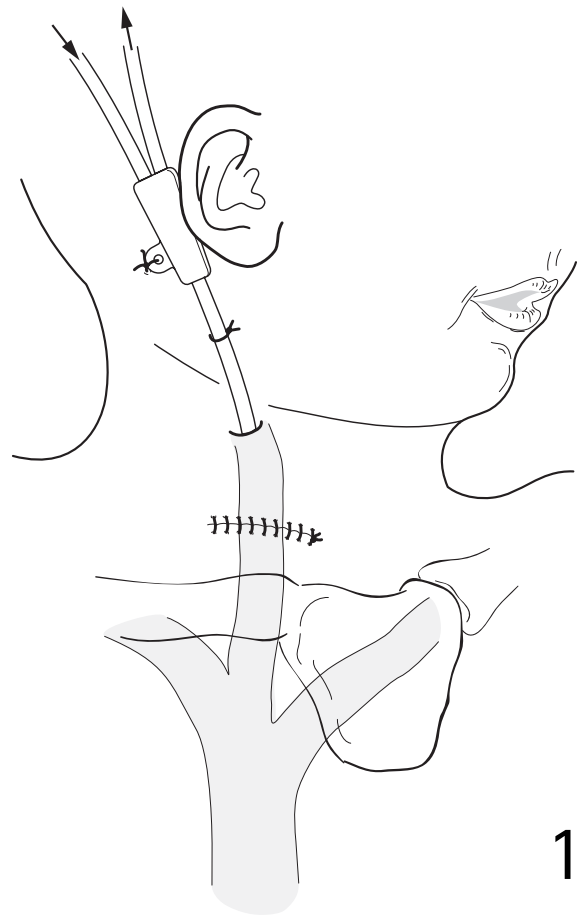


10

CANNULA PLACEMENT

10 Heparin (100 units/kg) is administered and 3 minutes allowed for circulation. The selected cannula is advanced over the Teflon obturator into the vein under direct vision to confirm entrance into the vein. The arterial (red) port of the cannula must be directed anteriorly to allow the arterial blood to cross the tricuspid valve and minimize the recirculation of circuit blood. The tip of the cannula is placed at 6–9 cm from the skin.



11

WOUND CLOSURE AND CANNULA FIXATION

11 The relatively low venous pressure allows adequate hemostasis around the venotomy site without any ligature. This prevents kinking of the thin-walled cannula, which often occurs at the area of a ligature if used around the vessel. Repositioning of the cannula only requires removing the skin sutures, repositioning the cannula, and replacing the skin sutures. The cannula is fixed to the skin with several 2/0 silk sutures. The incision is closed with a monofilament suture.

DECANNULATION

After respiratory failure has resolved, to allow ventilation without extracorporeal support, cannulation can be performed by removing the skin sutures, pulling the cannula, and holding pressure on the catheter exit site for 5 minutes or until bleeding stops. Care must be taken to remove the entire cannula rapidly to prevent air from entering the side holes while the end of the cannula remains in the vessel.

POSTOPERATIVE CARE

The cannulae are connected to the extracorporeal bypass circuit, assuring that no air bubbles are present, and bypass is initiated. Dopamine infusion into the ECLS circuit reinfusion connector is often necessary for inotropic support with the initiation of VV bypass if the patient required this type of support prior to ECLS initiation. A chest radiograph should be obtained following the procedure to verify optimal cannula placement distal to the aortic arch (arterial cannula) and inferior aspect of the right atrium (venous cannulae). Bleeding from the wound is controlled by lowering activated clotting times, platelet transfusions, the administration of fresh frozen plasma, and local instillation of fibrin glue. Bleeding not controlled by these maneuvers should be investigated and controlled by operative exploration after the open technique. Placement of a purse-string suture around the catheter exit site controls bleeding after the semi-open technique.

OUTCOME

More than 15 000 neonates have been treated since 1972, with an overall survival rate of 84 percent. The four most common diagnoses requiring ECLS in neonatal patients are meconium aspiration syndrome, congenital diaphragmatic hernia, pneumonia/sepsis, and persistent pulmonary hypertension. Survival rates of 93 percent, 58 percent, 76 percent, and 83 percent, respectively, have been achieved.

FURTHER READING

Peek GJ, Firmin RK, Moore HM, Sosnowski AW. Cannulation of neonates for venovenous extracorporeal life support. *The Annals of Thoracic Surgery* 1996; **61**: 1851–2.

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22

Eventration of the diaphragm

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HISTORY

Eventration of the diaphragm refers to the radiographic finding of an abnormally elevated hemidiaphragm. The physiologic consequences of the loss of diaphragm function include reduced lung volume, decreased tidal volume, increased work of breathing, and respiratory insufficiency that may preclude ventilator weaning. The mobility of the mediastinum of an infant may result in significant respiratory embarrassment when the normal hemidiaphragm descends during inspiration, the mediastinum shifts to the normal side, and the eventration side paradoxically elevates.

The term diaphragm eventration includes several distinct abnormalities. Acquired eventration (paralytic eventration) is easily understood on the basis of injury to the phrenic nerve, most commonly occurring at the time of intrathoracic surgery. Congenital eventration of the diaphragm is less well understood and probably includes a number of different entities. Phrenic nerve injury from birth trauma is similar to operative injury in that the diaphragm is developmentally normal, with a normal distribution of muscle and a normal central tendinous area. Other congenital eventrations (nonparalytic eventration) of the diaphragm are associated with anatomic abnormalities of the diaphragm. The diaphragm muscle is thinned and may be entirely absent from a portion that is normally muscular. In its most extreme forms, eventration is indistinguishable from a congenital diaphragmatic hernia with a hernia sac.

Eventration of the diaphragm was initially recognized in infants only as a post-mortem finding. The first successful surgical correction, using a form of diaphragm plication, was reported in 1947. Surgical treatment is based on removing the laxity of the abnormal diaphragm leaf to prevent paradoxical motion. This abnormal laxity may be removed by excision and closure of the resultant defect, suture of a portion of the diaphragm to the chest wall, or pleating of the lax muscle to create a taut diaphragm.

PRINCIPLES AND JUSTIFICATION

Diaphragm eventration may be discovered incidentally as an elevated hemidiaphragm on a chest radiograph obtained for other reasons, or it may be the cause of severe symptoms such as respiratory failure or pneumonia. The most common presentation of diaphragm eventration is postoperative respiratory failure as a result of phrenic nerve injury following an intrathoracic operation.

No rigid criteria exist for recommending repair of eventrations. Small eventrations with minimal compromise of lung volume may be monitored by serial radiographs. A pneumonia responding to antibiotic therapy or mild respiratory distress that responds to supportive care, such as chest physiotherapy and supplemental oxygen administration in association with eventration, need not be treated surgically if there is complete symptomatic resolution. Recurrent symptoms or respiratory distress that requires mechanical ventilation are indications for surgical correction. When the possibility of reversible phrenic nerve injury exists, such as that due to birth trauma or operative injury, a period of observation is indicated. If there is no improvement in diaphragm function after a reasonable period of observation (2–4 weeks), diaphragm plication is performed. If function has not returned in that period of time, it is

likely that prolonged mechanical ventilation will be required and surgical correction will be beneficial, allowing the discontinuation of mechanical ventilation. Some function may eventually return to the previously paralyzed diaphragm, but this may require many months. Operative plication does not preclude some recovery of diaphragm function.

Several techniques have been used for correction of eventration, including transabdominal and transthoracic approaches. The excess length of diaphragm has been dealt with by excision or various methods of 'gathering' the excess tissue with sutures. All of these procedures have in common the creation of a taut diaphragm that is mechanically resistant to elevation when negative intrathoracic pressure is created during spontaneous breathing. Specific abnormalities lend themselves to repair by the different methods. Video-endoscopic techniques may be used to perform diaphragm plication as well. Both thoracoscopic and laparoscopic techniques have been used.

When concurrent abdominal pathology that requires operative correction is present, the transabdominal approach is used. A single abdominal incision allows correction of both problems. Likewise the rare bilateral eventration may be approached through a single incision. The transthoracic approach is used for isolated unilateral eventration. It allows better visualization of the course of the phrenic nerve. For right-sided eventrations, the transthoracic approach avoids the need to mobilize the liver for visualization of the diaphragm.

No portion of the diaphragm needs to be excised in cases of acquired eventration. The muscular diaphragm may ultimately regain some function, and excision only increases the risk of additional injury to the intradiaphragmatic portion of the phrenic nerve. In congenital eventrations with muscular aplasia or atrophy, the thinned portion of the diaphragm may be excised when the thoracic approach is used and the course of the phrenic nerve is visualized. This allows the edges that will be brought together to be visualized precisely. Full-thickness sutures may be placed without fear of injury to intra-abdominal organs. Pledgets and non-absorbable sutures provide the most secure closure. Since diaphragmatic defects come in all shapes and sizes, the precise orientation of the plication procedure and the decision to excise some or all of the eventration must be made on an individual basis.

PREOPERATIVE

Assessment and preparation

Unilateral eventration is suspected when the right hemidiaphragm is greater than two rib levels higher than the left, or the left hemidiaphragm is more than one rib level higher than the right. A rare bilateral eventration is suspected when respiratory failure is present in association with radiographic demonstration of bilateral diaphragm elevation. Chest radiographs may be misleading in the patient on positive pressure mechanical ventilation. A non-functioning diaphragm may not be elevated. The most convincing diagnostic tests are those that allow dynamic visualization of diaphragm function during spontaneous respiration. These include fluoroscopy and ultrasonographic imaging. Ultrasonography has the advantage that it can be performed easily at the patient's bedside in the intensive care unit. Absent or paradoxic elevation of the hemidiaphragm during spontaneous inspiratory effort is diagnostic.

Preoperative evaluation includes physical examination to determine the presence of other anomalies or stigmata of chromosomal abnormalities. An echocardiogram should be performed in the presence of congenital eventration to rule out associated structural cardiac abnormalities. Diaphragm eventration has been associated with intestinal rotational abnormalities and gastric volvulus. It is noted in rare genetic conditions. Upper gastrointestinal contrast studies or computed tomography (CT) scanning may be indicated if associated malformations are suspected.

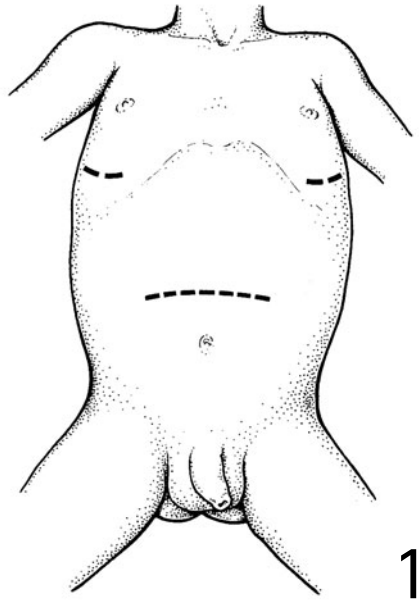
Anesthesia

General endotracheal anesthesia is used. Unilateral intubation improves exposure, but, if difficult to perform or not well tolerated, it is not mandatory because the lung is easily retracted. Unilateral ventilation is more important in the thoracoscopic approach. Bronchoscopic guidance of endotracheal tube placement has expanded the capabilities of pediatric anesthesiologists to provide unilateral ventilation in infants and small children and may be useful. Intraoperative orogastric intubation with regular gastric aspiration is important since a dilated stomach is at risk of injury when diaphragmatic sutures are passed. An epidural catheter for intraoperative anesthesia and postoperative analgesia may be helpful.

OPERATIONS

Incision

1 A transverse upper abdominal incision is used for bilateral eventration or in cases of unilateral eventration with malrotation. A lateral muscle-sparing seventh intercostal space thoracotomy is used for isolated left or right eventration.



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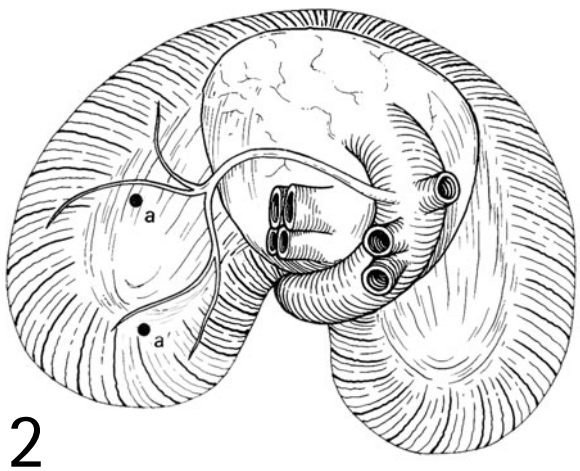
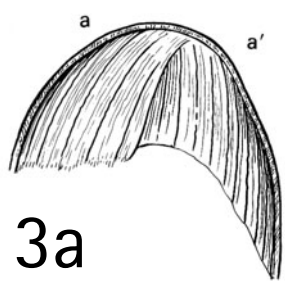


2

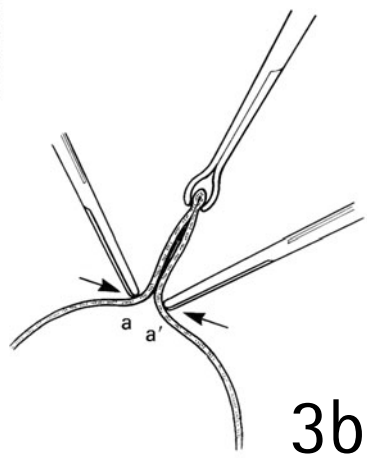
Transthoracic repair of left-sided acquired eventration

2 The main phrenic nerve on each side divides into an anterior and posterior division. Subsequent divisions usually include a sternal branch immediately off the anterior division and a bifurcation of the posterior division. The branches run in a medial to lateral orientation, allowing sutures to be placed to minimize the risk of injury to the muscular branches of the nerve. Viewed from above in this figure, the points *a* and *a'* represent the portions of the diaphragm that will be brought together by the plication.

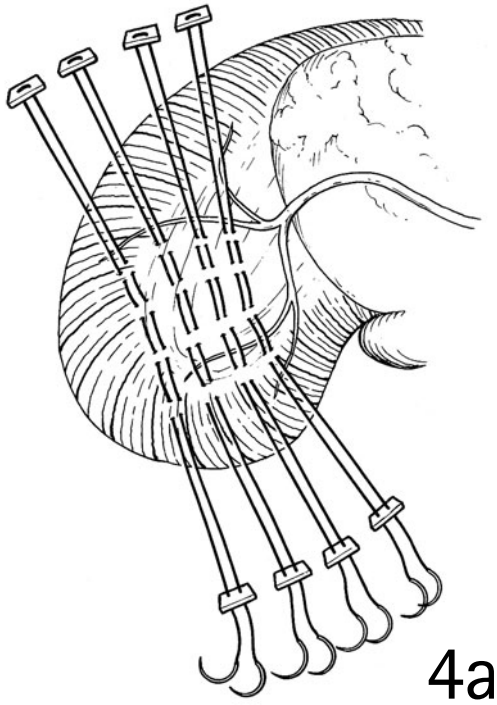


3a

3a,b The diaphragm is grasped and manipulated to determine the amount that must be included in the plication to create a taut closure. This is conveniently performed by grasping the center of the diaphragm with a non-crushing clamp (Babcock). The extent of the plication, determined by manipulations with the two forceps, is marked with a surgical marker.

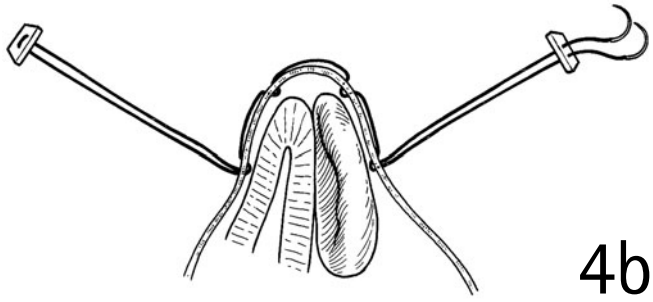


3b



4a

4a,b Several non-absorbable sutures are placed to bring the marked portions of the diaphragm together. The sutures are passed through the intervening diaphragm muscle three or four times. This maneuver is referred to as gathering, reefing, or pleating. Pledgeted mattress sutures as shown. Care must be taken that the sutures are passed adequately through muscle but not deeply enough to penetrate adjacent abdominal viscera.



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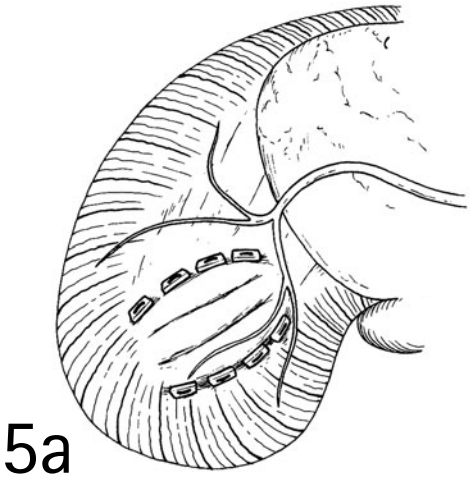
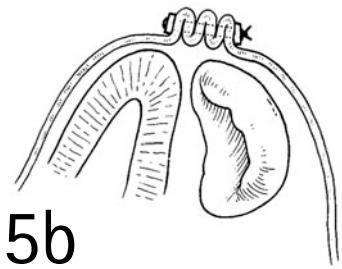


5a

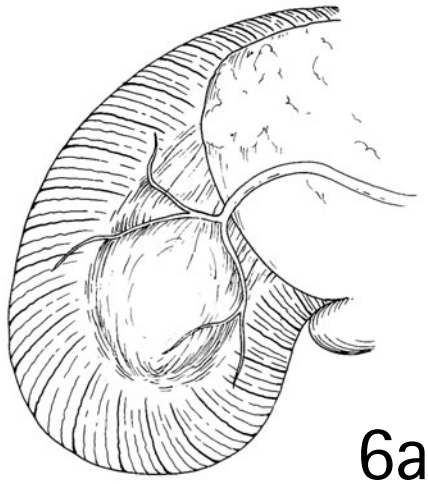


5b

5a,b The final result of the plication creates a taut diaphragm.

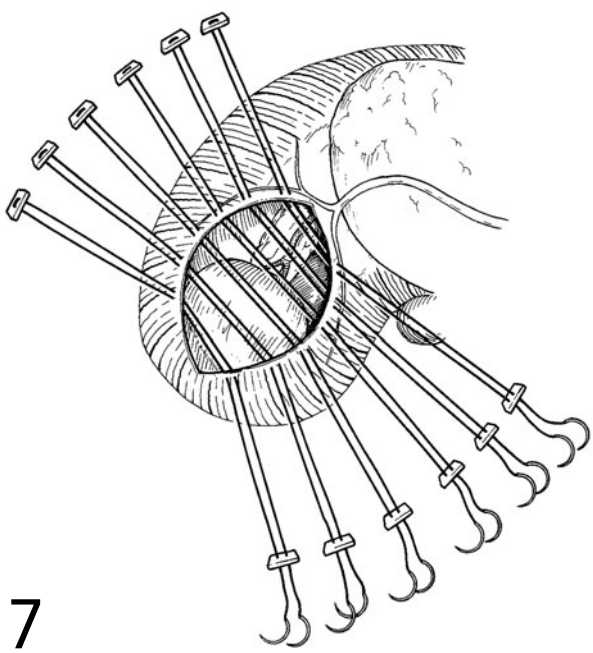
Transthoracic repair of left-sided congenital eventration

6a,b In this condition, the central portion of the diaphragm is 'thinned out'. Although the diaphragm may be 'gathered' in a fashion similar to the repair of an acquired eventration, excising the thin central portion of the diaphragm allows the edges to be visualized clearly so that sutures may be placed in normal muscularized tissue and the abdominal viscera avoided.



6a

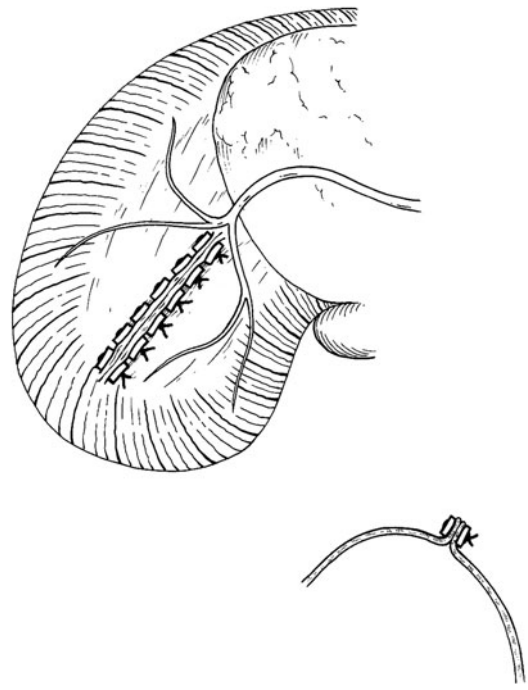
6b



7

7 Non-absorbable, pledgeted mattress sutures are placed and oriented to close the defect in a transverse fashion.

8 The final result brings muscularized diaphragm together. If excessive tension is required to bring the tissues together, a prosthetic patch may be used to close the defect. This is rarely necessary.



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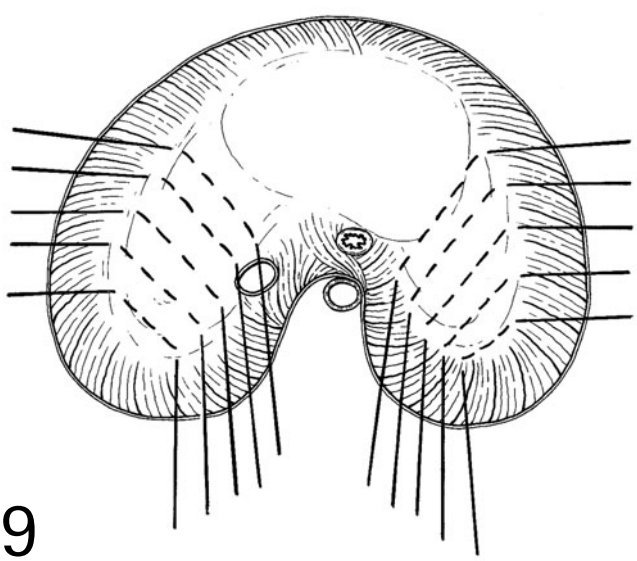
Transabdominal repair of bilateral eventration

Rotational abnormalities of the intestine are addressed if present.



9

9 The undersurface of the diaphragm is exposed on the left by mobilizing the liver as necessary. The stomach and spleen are retracted and mobilized to give complete exposure. The right lobe of the liver is mobilized if needed. Congenital eventration with thinning of the diaphragm will require little mobilization.

Plicating sutures are arranged to avoid the phrenic nerve based on its expected location. The plication is oriented anteromedial to posterolateral, identical to the transthoracic approach.

POSTOPERATIVE CARE

Response to operation may be immediate, allowing prompt weaning and extubation. In patients who have required prolonged mechanical ventilation prior to operation, slower ventilator weaning is performed. Intrapleural drainage is used briefly after surgery and the drainage tube is usually removed within 2–3 days.

OUTCOME

Although death may result from chronic respiratory failure and pneumonia, outcome is largely dependent on the presence of associated conditions such as pulmonary hypoplasia or congenital heart disease. Long-term survival is variably reported as 69 percent to 100 percent. Children with bilateral eventration fare less well.

Surgical correction is durable and recurrence requiring repeat plication rare. Inadequate plication may result if the diaphragm is not made taut at the initial operation, and the plication process will need to be repeated.

Transdiaphragmatic injury may occur with either open or throacoscopic techniques. Intra-abdominal injuries such as gastric perforation or colon perforation require immediate operative attention. Herniation into the site of the repair can occur.

Many patients examined years after diaphragm plication have evidence of appropriate although diminished movement of the involved side.

There are no reports comparing video-endoscopic and open techniques of diaphragm plication. Small technical differences, such as the use of pledgets, and the ability to assess the completeness of the plication ('tautness') must be considered. In a condition that is relatively rare, surgeons should be cautious to make sure that the video-endoscopic procedure they perform is as safe and effective as the open approach.

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23

Lung surgery

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PRINCIPLES AND JUSTIFICATION

Removal of lung tissue is done less often in children than it is in adults. Acquired lesions, such as carcinoma or chronic infections, are much less common in childhood. Most operations are performed for congenital problems, and less commonly for infectious etiologies. Table 23.1 lists the indications for pulmonary resection in children.

In general, the principles of lung surgery are similar in adults and children. Children usually have a greater reserve and physiologically withstand resections better than adults. In the first 7 years of life, there is ongoing alveolar development, which may lessen the physiologic impact of lung resection. Depending on the indications for pulmonary resection, the approach and operative principles utilized will have to be adjusted. Minimally invasive techniques are being used more frequently in children for thoracic procedures, and there is growing experience in thoracoscopic pulmonary resections. However, in cases of severely altered anatomy such as inflammatory conditions, an open procedure is usually indicated.

Complications of lung surgery in children are generally fewer than in adults. Major bronchial stump air leaks are rare in healthy pediatric patients. Scoliosis and/or the development of chest wall deformities are potential long-term complications unique to children who have undergone thoracotomy. Children with inflammatory conditions such as lung abscess, pneumonia, and bronchiectasis are at greater risk of postoperative complications.

PREOPERATIVE ASSESSMENT AND PREPARATION

If the patient meets the indications for having lung surgery, pulmonary function should be optimized and infections should be controlled as much as possible with preoperative antibiotics. Lobectomy and non-anatomic resections less than a lobectomy are well tolerated in healthy children. In patients with cystic fibrosis or other generalized pulmonary diseases in which pulmonary reserve is reduced, pulmonary resection, especially lobectomy, is less well tolerated. Preoperative work-up should include measurement of forced expiratory volume in the first second (FEV1) and forced vital

Table 23.1 *Pediatric lung conditions requiring surgery*

Congenital	Acquired
Sequestration	Acquired lobar emphysema
Cystic adenomatoid malformation	Bronchiectasis
Lobar over-expansion (emphysema)	Apical (or other) blebs
Lung cysts (single, multiple)	Metastatic lesions
Hamartomas	Fungal infections (lobar)
Congenital pulmonary insufficiency (needing biopsy for diagnosis)	Diffuse pulmonary disease

capacity (FVC) to assess the effects of resection. All patients should have a minimum baseline measurement of hemoglobin, and a type and screen with blood rapidly available if needed, especially in complex resections. This laboratory work may be done after the induction of anesthesia prior to the initiation of the operative procedure to avoid unnecessary pain for the younger child or infant.

See Chapter 12 for additional discussion.

Anesthesia considerations

Many lung operations in children can be successfully performed using a standard endotracheal tube without single lung ventilation. Single lung ventilation can be accomplished in infants and young children by either selective intubation of one bronchus or the use of bronchial blockers. In the older child, the use of double-lumen tubes (when at least a 7 Fr endotracheal tube can be utilized) will allow for single lung ventilation. For many thoracoscopic procedures, gentle insufflation at pressures of 5–10 mmHg will be enough to collapse the lung sufficiently, and single lung ventilation may not be needed. In certain cases or at certain intervals during the case, it is preferable to have the lung inflated during surgery for visualizing blebs and some surface metastases and for identifying air leaks.

Postoperative pain relief after a thoracotomy is another important consideration, and the use of thoracic epidurals should be considered for difficult cases. Epidurals facilitate anesthetic delivery intraoperatively and decrease requirements for intravenous analgesics in the postoperative period. If an epidural is not done, the surgeon can inject local anesthetic at multiple intercostal levels to perform nerve blocks or, alternatively, place a pleural catheter for delivery of local anesthetic. Intercostal nerve blocks can be completed very accurately with thoracoscopy.

Monitoring of children undergoing major lung surgery intraoperatively may involve the placement of an arterial line and a central venous line, but these are dependent on the patient's preoperative condition, and on surgeon and anesthesiologist preference, and are not considered mandatory.

OPERATIONS

These are discussed in two broad categories – the general principles of pulmonary resection and specific approaches to various lobectomies, and the indications and operations needed for specific conditions.

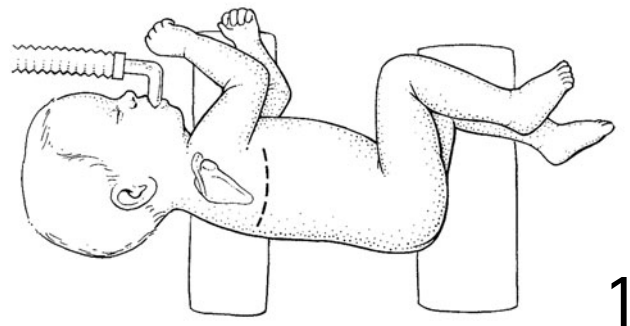
Principles of pulmonary resection

ACCESS TO THE LUNG

Operations on the lung can be performed in two major ways: open thoracotomy and thoracoscopically. Thoracotomy can be performed via one of three approaches. The first is an anterolateral approach mostly used for open lung biopsies, or wedge resections. The second approach is via posterolateral thoracotomy, which is the most common method used for open lung resections. A third approach through a prone position is employed to reduce spilling of infected secretions into the contralateral lung during resection. However, with modern anesthesia techniques allowing single lung ventilation as well as more effective antimicrobial therapy, this approach is used less commonly. Median sternotomy is used by some surgeons when bilateral wedge resections are required, such as in osteosarcoma metastases.

The anterolateral thoracotomy is performed by placing the patient supine with a roll just below the side to be explored to elevate it 30–45° from the table. The ipsilateral arm is allowed to fall back on the table. After appropriate and wide preparation of the field, the incision is performed below the level of the nipple in the fourth, fifth, or sixth interspace, taking care not to injure the underdeveloped breast bud complex. The incision may be extended along the ribs toward the axilla as required. The pectoralis and intercostal muscles are then divided to enter the pleural cavity. After performing the desired resection or biopsy, a chest tube is placed a couple of interspaces below the incision and directed to the apex. The ribs are approximated loosely with an absorbable pericostal stitch (polygalactin). The muscle fascia is approximated with a running absorbable suture, followed by subcutaneous closure with either a running or interrupted suture and subcuticular closure of the skin (both with absorbable material).

1 Posterolateral thoracotomy is performed with the patient in a lateral decubitus position. An axillary roll is used and appropriate padding of the legs is done to avoid pressure damage and injury to the brachial plexus. The upper arm is allowed to lie on the same side with support to prevent excessive stretching of the arm as well as the brachial plexus. A wide preparation is done from the vertebral column posteriorly to the sternum anteriorly. The nipple and areola are marked, as is the tip of the scapula, to help guide the incision. A gently curved thoracotomy incision is performed in the interspace chosen. In most cases, a muscle-sparing approach can be employed (see Chapter 12 for details), in which the serratus anterior is retracted forward and the latissimus dorsi is reflected posteriorly. These may be partially or completely divided as needed to gain wider access during the operation. Care is taken not to divide the paraspinal muscles, but to free them up longitudinally. This move, as well as avoiding division of the trapezius and rhomboids, may help in reducing the development of scoliosis. The ribs are held apart with a self-retaining metal retractor (Finochietto). After the resection is performed, a chest tube is placed a couple of interspaces below the incision and near the apex and secured with a non-absorbable suture. Pericostal sutures are placed in an interrupted fashion and appropriately secured to avoid excessive approximation of the ribs. Fascia and skin are closed as described previously.



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Thoracoscopic approaches are now being employed increasingly. Lung biopsies, wedge resections, and lobectomies are being performed using this approach. Improving optics with high-definition cameras and brighter light sources have made visualization excellent, and the development of devices such as the Ligasure® electrocoagulator for dissection and control of vessels up to 7 mm in diameter has allowed these resections to be performed safely. The clear benefits in cosmesis, less pain, decreased length of stay, and potentially much less scoliosis have been the driving forces, but have yet to be proven definitively. For thoracoscopic resections of any kind, the patient is placed in a decubitus position as described previously. The table may be rotated to the right or left as needed for exposure. Some surgeons prefer to have the patient in the position described for the anterolateral thoracotomy. It is important to use gravity as a retractor during thoracoscopy and the surgeon should vary the position accordingly. Single lung ventilation may be used if needed to facilitate exposure, although insufflation usually obviates the need for this. The chest is prepared as for a thoracotomy and a 5 mm incision made in the mid-axillary line in the fourth to sixth interspace. A Veress needle is placed into the chest carefully just above a rib to avoid the neurovascular bundle. Insufflation with CO_2 to a pressure of 3–7 mmHg, at least initially, facilitates exposure and creates a functional working space. A 5 mm port is placed and the video-thoracoscope is introduced. The remaining port sites are then chosen based on the specific anatomy and which lobe is to be operated on. Usually, two additional ports are placed – one anteriorly and one posteriorly – to facilitate triangulation and dissection of the fissure and lobe. A fourth access site may be used for retraction if necessary. If an endoscopic stapling device is to be used for a biopsy, a 12 mm port will need to be placed. One of the port sites may need to be enlarged slightly for removing the specimen. Upon completion of the procedure, the chest tube is placed under thoracoscopic vision through one of the dependent port sites, directed towards the apex and secured. The fascia may be closed with an absorbable stitch and the skin approximated by suture or other means.

Lung biopsy and wedge resection

This procedure is performed for diagnostic or therapeutic purposes. Indications for biopsy include an infectious process, diffuse parenchymal disease of unknown etiology, and discrete parenchymal process for the diagnosis of inflammatory or malignant process. Those patients who have diffuse disease may not tolerate single lung ventilation, and in these cases the procedure will have to be performed with the lung expanded. Thoracoscopic procedures allow visualiza-

tion of the entire lung, and a small wedge can be excised efficiently with a stapling device. An open operation with a small anterolateral thoracotomy is also a reasonable technique for a wedge biopsy with a stapling device. A persistent air leak after non-anatomic resection can be controlled by a second firing of the stapling device after removing the knife blade, or oversewing the suture line. Chest closure is performed in the same way as described previously.

Non-anatomic wedge resections may also be performed for metastatic disease to the lung. In general, an open thoracotomy is preferred for osteosarcoma metastatic disease, to allow palpation of the lung parenchyma and to remove as many lesions as possible. Although not definitively proven, some advocate performing either a staged bilateral thoracotomy or a median sternotomy to evaluate and treat both lungs with osteosarcoma.

In other tumors it may be reasonable to utilize a thoracoscopic approach. Smaller or deep parenchymal lesions may not be accessible via thoracoscopy. Some authors have described using computed tomography (CT) guided marking of the lesions immediately preoperatively to facilitate localization and resection with thoracoscopy.

Apical bleb disease leading to recurrent or persistent pneumothorax can also be treated thoracoscopically. The apex is visualized and wedges of parenchyma with blebs are removed using an endoscopic stapling device. In most cases it is best to use these devices with the vascular load, as they have smaller staples and may cause less bleeding. Again, if there is an air leak from the staple line, a further fire of the device after removing the blade is helpful. Fibrin glue or Tisseal® can be applied over the staple line as an adjuvant to help control air leaks.

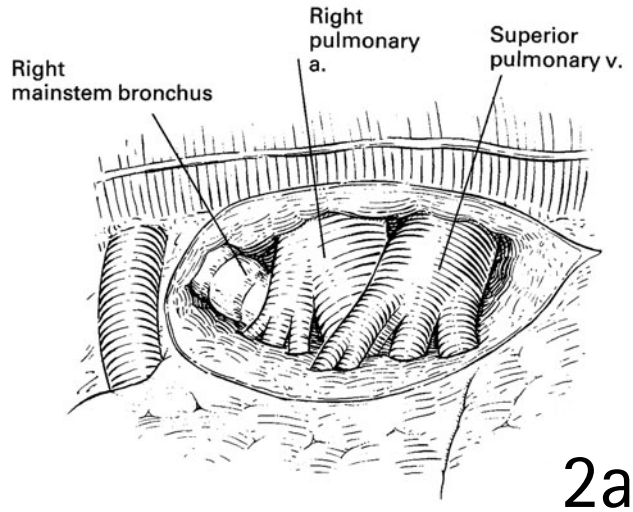
Lobectomy

The principles of lobectomy are similar in children and adults. The most important principles are adequate visualization and exposure of the hilar structures – the blood vessels and bronchus. In most instances it is better to dissect out and control the pulmonary arterial branches first, venous drainage second, and the bronchus last. Occasionally, as in the case of a severe purulent infectious process, it is better to divide the bronchus first.

The anatomic considerations involved in the removal of different lobes are now discussed in detail. The surgeon must have a complete understanding not only of normal anatomy but also of the variations in anatomy that are frequently encountered during lung resections.

RIGHT UPPER LOBECTOMY

2a,b The patient is positioned for either a posterolateral thoracotomy via the fifth interspace or a thoracoscopic approach. The lung is retracted posteriorly and the pleura covering the hilum of the right lung is opened from below the level of the azygous vein and around the hilus superiorly and posteriorly to a level below the right mainstem bronchus. Care is taken to avoid injury to the right phrenic nerve. It should be noted that considerable variation in vascular anatomy occurs, but the usual pattern is shown in the illustration. Bronchial anatomy is more standard. The main pulmonary artery is identified behind the vena cava, and dissection is carried out peripherally to expose the superior and inferior pulmonary arterial trunks. The superior pulmonary arterial trunk, with its branches to the anterior and apical segments, is exposed and then often individually ligated, especially distally. The superior lobe vein is identified and dissected laterally, usually exposing three segmental pulmonary veins. The middle lobe veins entering the superior pulmonary vein must be identified and preserved. It is often easier and safer to ligate the individual venous branches peripherally. The oblique fissure is then opened between the upper and lower lobes. This permits the dissection and isolation of the posterior ascending segmental pulmonary arterial branch, which ordinarily comes off the pulmonary artery after the middle lobe branches. All the vessels are best managed by ligation with non-absorbable suture as well as suture ligation proximally. However, when performing the operation thoracoscopically, the Ligasure® device can be used to coagulate a portion of the vessel and divide it. Most of the electrosurgical devices are approved to control vessels up to 7 mm in diameter. The sequence of dissection is similar to that for the open approach.



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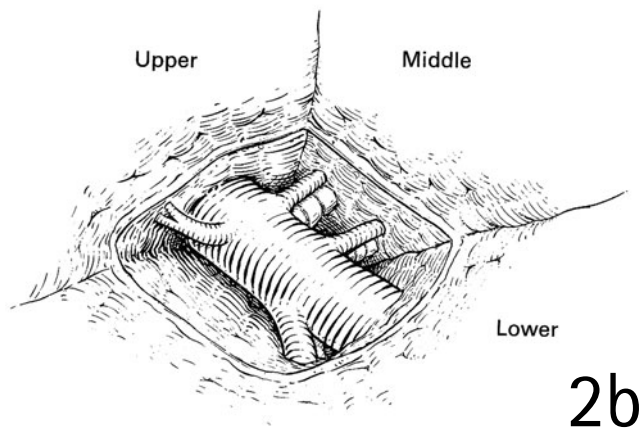


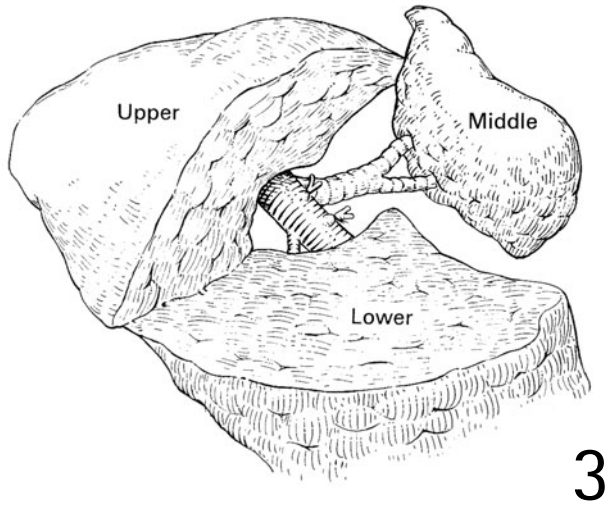
2b

There are usually three veins draining the right upper lobe, which should be ligated just before their junction with the superior pulmonary vein. These are visualized by retraction of the lung posteriorly in order to gain wider exposure of the anterior hilum.

After division of the arterial and venous branches of the right upper lobe, adventitial tissue surrounding the bronchus is cleared away. The bronchus only needs to be cleared to show its origin, as further dissection may compromise the blood supply and delay healing of the bronchial stump. Stay sutures of non-absorbable material are placed on either side of the bronchus and it is divided about 1 cm or 2 cm from the main stem in order to avoid a long stump, which may accumulate secretions. After division of the bronchus, the stump is closed with interrupted sutures of non-absorbable material, or with a stapling device. Thoracoscopically, the same choices exist for handling the bronchus, with most surgeons preferring interrupted sutures, especially in small children. After closure, warm saline is poured in the chest and the stump is tested for a leak by applying pressure of 30–40 cmH_2O with the ventilator. Surrounding pleura may be used to reinforce the stump and possibly promote healing. A chest tube is placed and secured as described previously.

RIGHT MIDDLE LOBECTOMY

3 The arteries to the right middle lobe are best exposed through the oblique fissure between the upper, middle, and lower lobes. After development of the interlobar fissure, one or two middle lobe vessels are usually encountered, which are ligated as described previously. The lung is retracted posteriorly to expose the anterior hilum, which is also fully dissected, and one or two right middle lobe veins are found joining the right superior pulmonary vein. At times, the interlobar fissure is incomplete and dissection is performed to the vasculature by a stapling device or ligasure cautery. Once the middle lobe has been separated from the upper lobe and both arterial and venous branches divided, the lobe is retracted anteriorly and the bronchus is divided. The stump is controlled in the fashion described previously. The potential for injury to the ascending posterior segmental artery of the upper lobe makes this operation somewhat more technically demanding than upper and lower lobectomies.



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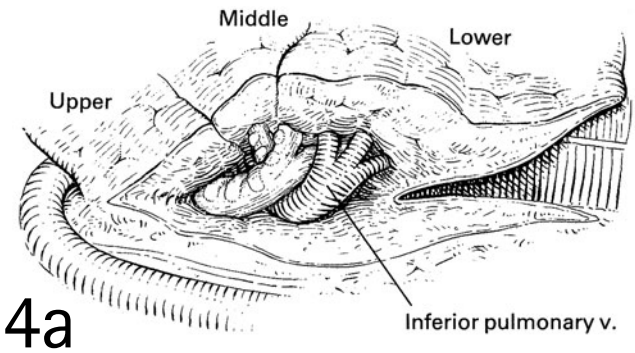

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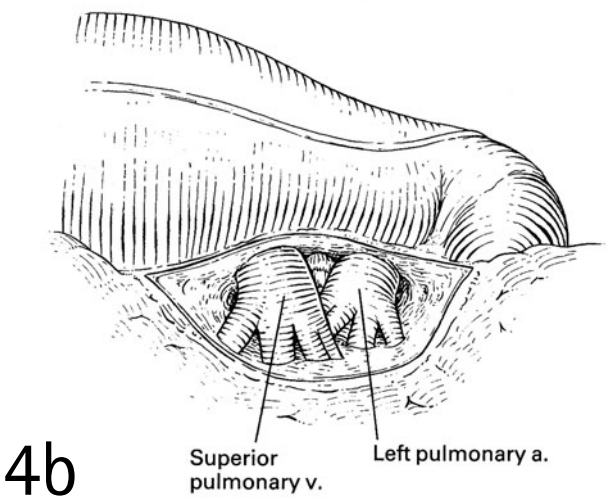


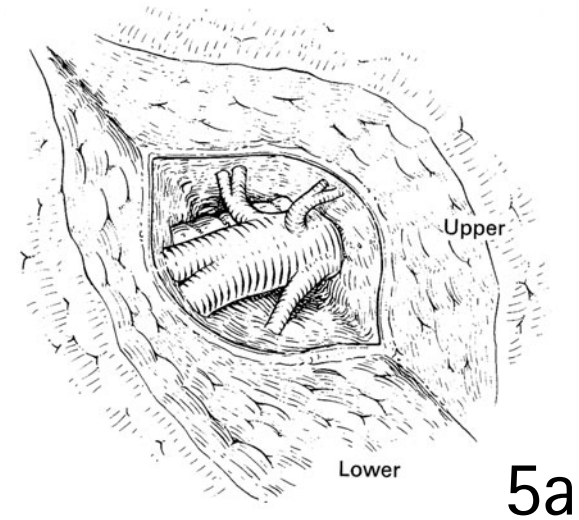
4b

RIGHT LOWER LOBECTOMY

4a,b For performing a right lower lobectomy, a lower intercostal space is used for thoracotomy. For thoracoscopy, the ports may need to be placed slightly lower, although this is not mandatory. The interlobar fissure is exposed by retraction of the upper and middle lobes superiorly and the lower lobe inferiorly. The branches of the interlobar portion of the right pulmonary artery are exposed and carefully identified. Just beyond the middle lobe arteries and opposite them, one or two superior segmental arteries supplying the superior segment of the lower lobe are encountered. These are divided after ligation. There is a remaining vessel to the basilar segments, which should be identified and similarly ligated. The lobe is then retracted anteriorly to expose the posterior hilum. The inferior pulmonary vein is exposed by opening the inferior pulmonary ligament and carrying the pleural dissection upwards to isolate and facilitate ligation of the inferior pulmonary vein. After this, the right lower lobe bronchus can be easily identified by posterior retraction. Care is taken to keep the bronchial stump length short.

LEFT UPPER LOBECTOMY

5a,b After positioning the patient in the appropriate right lateral decubitus position, either a posterolateral thoracotomy is performed or access is obtained by thoracoscopy. As for the right lung, the pleura overlying the hilus of the lung anteriorly is incised and carried superiorly and posteriorly below the level of the left mainstem bronchus. The left pulmonary artery is best identified anteriorly first, and then found as it courses superiorly and posteriorly to the upper lobe bronchus. Four to six branches of the left pulmonary artery to the upper lobe can be noted. Anteriorly, the anterior, apical, and posterior segmental arteries are seen. The apical segmental artery may be encountered superiorly, and anterior segmental and lingular segmental branches are usually seen in the interlobar fissure. After ligation of all these branches, the lung is retracted posteriorly and the left superior pulmonary vein is ligated just before it divides. Occasionally, the left superior and inferior pulmonary veins form a common vein, so before ligating the superior vein on the left side, the inferior vein should be identified separately. With anterior retraction, it is possible to see the bronchus to the upper lobe and lingula and divide these close to the origin as described.



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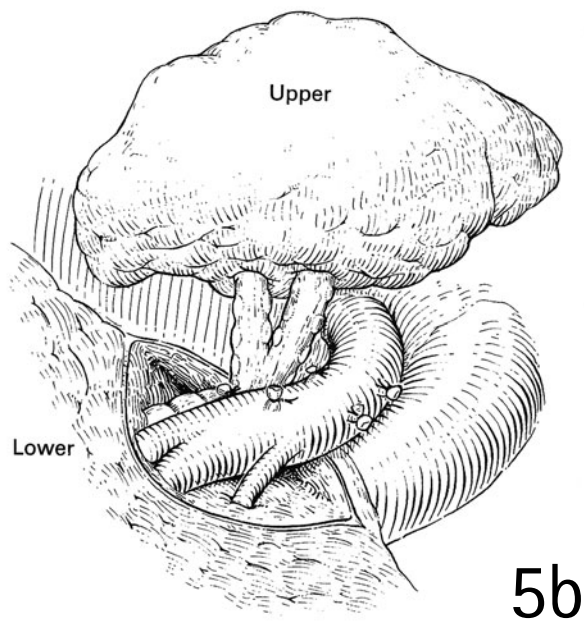

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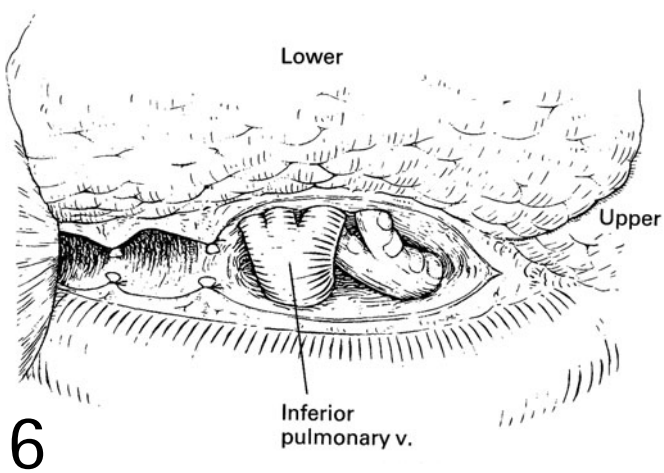


6

LEFT LOWER LOBECTOMY

6 After gaining access to the chest, the interlobar fissure is exposed to identify the arteries. One or two arteries supply the superior segment, but care must be taken as the superior segmental artery may arise proximal to the lingular vessels. Thus the lingular arteries must be identified in the course of this dissection. Following ligation of these vessels, the basilar portion of the left pulmonary artery may be divided just distal to the lingular arteries. After this, the lung is retracted anteriorly to expose the posterior hilum. The inferior pulmonary ligament is divided to expose the inferior pulmonary vein. The vein is then divided as described, taking care that the superior lobe has good venous drainage. After vascular division, the lobe is retracted posteriorly and the bronchus isolated and divided as described previously.

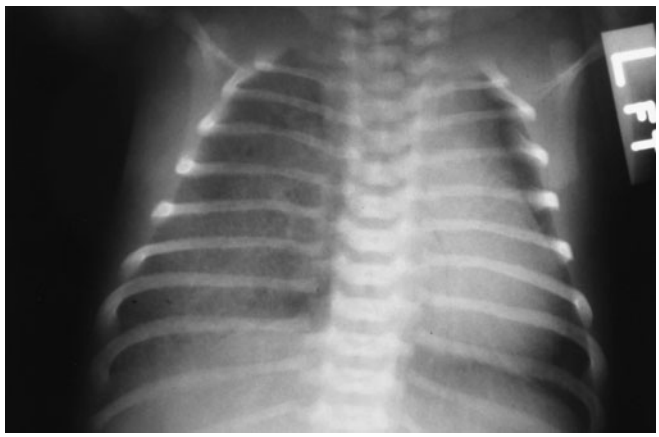
Next we discuss pediatric lung surgery in the context of the specific common conditions congenital lobar over-expansion (emphysema), cystic adenomatoid malformations, and sequestration. As mentioned, the principles of lung surgery, including those for specific lobectomies, will apply to each condition.

Lobar over-expansion (emphysema)

7a,b Congenital lobar over-expansion or acquired problems can be found in neonates and infants. Acquired problems arise from either mucous plugs or structural problems causing an obstruction of the airway. Congenital lobar over-expansion is caused by absence of the bronchial cartilage in 35 percent of cases, leading to ball-valve-type air trapping. Occasionally, extrinsic compression (e.g., congenital heart anomalies) can lead to distal air trapping and over-expansion. Bronchogenic cysts are also seen in association with this problem. One-third of cases are from alveolar hyperplasia in a specific lobe of the lung.

The symptoms arise from compression of other thoracic and mediastinal structures, which may occur acutely or chronically. Some infants do not require surgical intervention and remain stable with the lesion. Of those who need operation, 50 percent will develop symptoms within a few days of birth, while the remaining present a few months later.

Chest X-ray will show a hyperlucent area in the affected chest with a variable degree of compression of the mediastinal structures. The diaphragm is flattened on the affected side. Involvement is usually restricted to the upper lobes (left upper 42 percent, right upper 21 percent, right middle 35 percent), with less than 1 percent involving the lower lobes. Differential diagnosis includes pneumothorax, cystic adenomatoid malformation (CAM), pneumatocele, and atelectasis.



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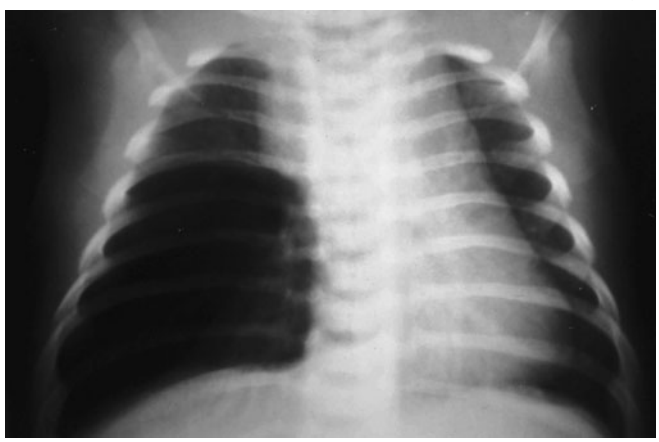


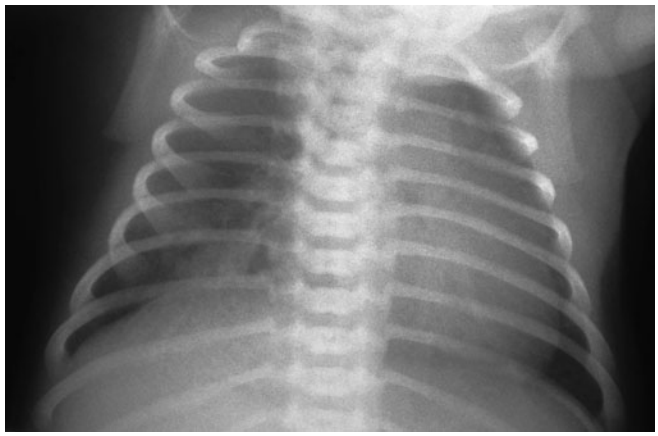
7b

Additional studies are usually unnecessary, but ventilation perfusion scans are occasionally used and show delayed uptake and poor vascular supply to the affected lobe. In some instances, it is necessary to perform an emergency thoracotomy for life-threatening compression. The lobe will usually herniate out as soon as the chest is opened, with immediate clinical improvement. The anesthesia team should not overventilate the patient, but should use frequent small tidal volumes.

Resection is carried out as described. In situations where there is less urgency, a bronchoscopy should be done before thoracotomy to ensure that there are no intrinsic bronchial problems that may be treated, thus avoiding a lobectomy.

Cystic adenomatoid malformations

Cystic adenomatoid malformations are lesions that are frequently now diagnosed prenatally by ultrasound. They form about 25 percent of all congenital lung malformations. Some become large enough that they lead to secondary lung hypoplasia and may impede the vena cava and the heart and lead to polyhydramnios and hydrops fetalis. Some of the affected fetuses will be stillborn. In a few centers in the USA, fetal surgery with thoracotomy and resection has been undertaken in patients who develop hydrops, some of whom have survived. Other methods of fetal intervention have not been successful. The CAM is found in the left lower lobe in 25 percent, left upper lobe in 20 percent, right lower lobe in 19 percent, and right upper lobe in 10 percent.



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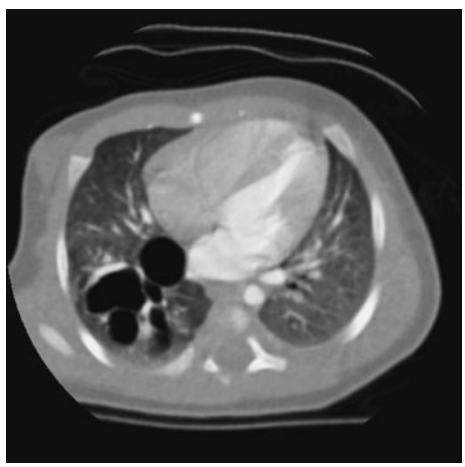


8b

8a,b In those neonates who are live born, some are symptomatic at birth, with distress, and can present with severe pulmonary hypertension requiring extracorporeal membrane oxygenation (ECMO) for stabilization prior to resection. Chest X-ray will often reveal an irregular cystic mass in the affected lung, which may involve the entire chest and have mediastinal shift. In some cases, the chest X-ray is normal, and chest CT is performed to confirm the prenatal diagnosis. A few rare cases have been reported to involute after birth. In patients who are in no distress after birth, resection is usually delayed until the child grows older. The risks of infection and cancer remain the major indications for elective resection. Waiting until the child is close to 1 year of age before proceeding with resection is reasonable, and some have even advocated long-term follow-up without surgical intervention, although there are no data to support this. Bronchoalveolar carcinoma, pleuropulmonary blastoma, and rhabdomyosarcoma have been reported to arise in these congenital lesions, and close surveillance is required.

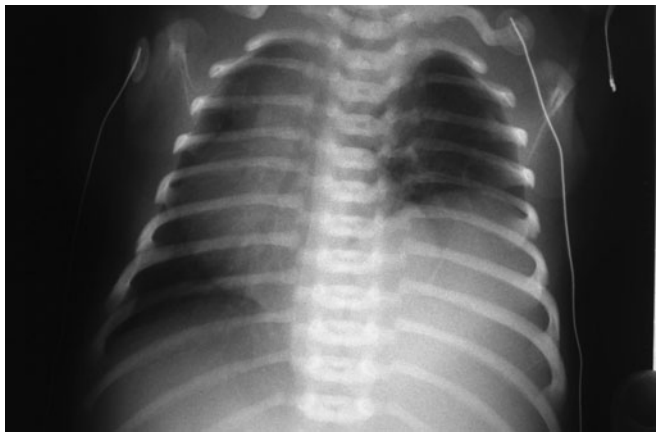
In symptomatic patients, resection should be done as soon as possible. The commonest procedure is a lobectomy, although segmentectomy has been reported as well. It is vital to resect the entire CAM otherwise it may recur or have a prolonged leak. In some cases the CAM may involve multiple lobes, and in very rare cases pneumonectomy may be needed. Mediastinal shifts after pneumonectomy can be symptomatic, and filling of the ipsilateral chest with saline or a tissue expander may be required.

Sequestration

9a,b A bronchopulmonary sequestration is a congenital malformation in which that portion of the lung receives systemic arterial supply and has no bronchial communication. There are two types – intralobar and extralobar sequestrations. Most large series describing sequestrations note a preponderance of extralobar lesions. These may coexist with other malformations such as bronchogenic cysts, CAM, and congenital heart lesions. Extralobar sequestrations are also associated with diaphragmatic hernias and eventrations. They may also arise in the abdomen – usually in the vicinity of the left adrenal gland. Most intralobar sequestrations are found in the lower lobes, with only 15 percent in the upper lobes.

Presentation of intralobar lesions is usually in the form of recurrent pneumonias. Therefore the diagnosis is usually after 2 years of age and a chest X-ray may reveal a consolidation. Recently, there have been a large number of cases detected prenatally similar to the experience with prenatal diagnosis of CAM. Extralobar sequestrations are usually discovered during repair of a congenital diaphragmatic hernia or eventration. They also may be found incidentally as a posterior mediastinal mass, with pneumonias less likely. It is important to remember that there may be a communication of the sequestration with the foregut (esophagus or stomach) due to the embryologic origins. During resection, these communications should be sought and controlled if present.

Resection follows the same principles as described previously, with a few key differences. The vascular supply arises from the abdominal aorta in 85 percent of cases and needs to be carefully sought and ligated at the outset. It is usually found in the inferior pulmonary ligament. There is no bronchial attachment, which makes the resection easier. There is usually a clear demarcation in intralobar sequestrations from the normal lung parenchyma and this forms a good plane of dissection. Lobectomy for intralobar lesions is the procedure of choice.



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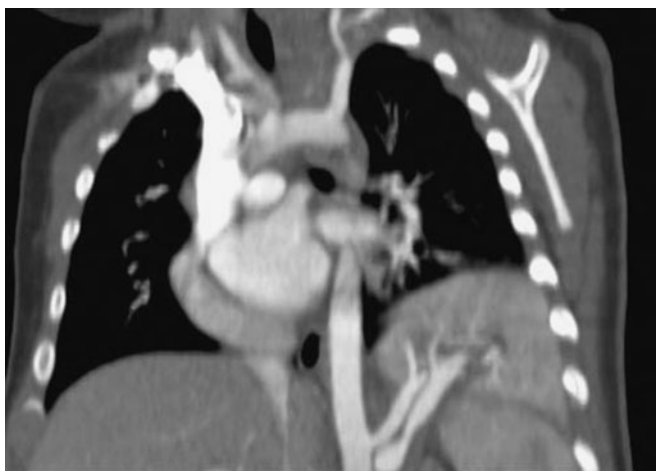

9b

POSTOPERATIVE CARE

Important principles of postoperative care after lung surgery involve ensuring adequate pulmonary toilet. Pain control is critical to optimizing postoperative pulmonary lung function and may be provided by a thoracic epidural. The epidural, if used, is usually maintained for 48–72 hours postoperatively. A Foley catheter for bladder drainage may be needed with epidural pain relief. Pain may not be a prominent feature following thoracoscopic procedures, but often the chest tube appears to be a major component of postoperative pain and should be removed as soon as possible. The chest tube is maintained for 2–4 days, initially on suction then under water seal, and removed once there is no air leak and the drainage is minimal.

Antibiotics are given for 24 hours and then usually discontinued. Some surgeons continue antibiotics until the chest tube is removed, although there are few data to support this practice. Narcotics should be used judiciously, and non-narcotic analgesia incorporated into the algorithms for pain control. Feeding is usually resumed the day after surgery.

Intensive care is required for patients with significant underlying lung disease and reduced pulmonary function. Most patients are able to go home within 3–4 days, although older children may need to stay longer because of pain issues. Patients with thoracoscopic procedures may have shorter hospital stays.

OUTCOMES

Mortality following lung resection including lobectomy or bilobectomy should be minimal with modern surgical techniques and postoperative care. The only group at increased risk is those with significant other comorbidities such as congenital heart disease, or those with additional pulmonary disease.

Mortality and morbidity after lung biopsy are most dependent on the underlying disease process. The risk of prolonged air leak with diffuse parenchymal disease also exists and can add significant morbidity.

Infectious complications are not common following lung resection for congenital lobar over-expansion, uncomplicated CAM, or sequestrations. When there is a pre-existing infection or abscess, the risk is higher. Bronchial stump leaks are more common in adults, and very rare in children.

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Empyema

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INTRODUCTION

The accumulation of purulent material in the pleural space is termed empyema or empyema thoracis. Usually a complication of inflammation or infection within or adjacent to the pleural space, empyema rarely resolves spontaneously because host defenses are limited by the anatomy and physiology of the pleural space. In children, the majority of cases occur as a result of pneumonia; however, an infection in the chest wall or a subphrenic abscess can also lead to empyema.

Empyema develops by progression through three well-recognized stages. Initially, sterile pleural fluid accumulates in the pleural space as an inflammatory response to neighboring infection. In this stage, known as the acute or exudative phase (Stage I), a parapneumonic effusion develops that is characterized by clear, low-viscosity pleural fluid with normal pH and glucose levels, and low lactate dehydrogenase (LDH) levels. The visceral and parietal pleurae in this stage are not fused. The transitional or fibropurulent stage (Stage II) is characterized by turbid pleural fluid with an increasing accumulation of leukocytes and decreasing pH and glucose. The LDH of the fluid begins to increase and a fibrinous peel develops along both pleural surfaces that may limit full expansion of the lung. Finally, the chronic or organizing phase (Stage III) is characterized by the ingrowth of capillaries and fibroblasts into the fibrinous peel, resulting in fusion of the visceral and parietal pleural surfaces. This typically occurs 4–6 weeks following the onset of the infection. At this end stage of disease, the pleural fluid has a pH of less than 7 and a glucose level of less than 40 mg/dL.

PRINCIPLES AND JUSTIFICATION FOR TREATMENT

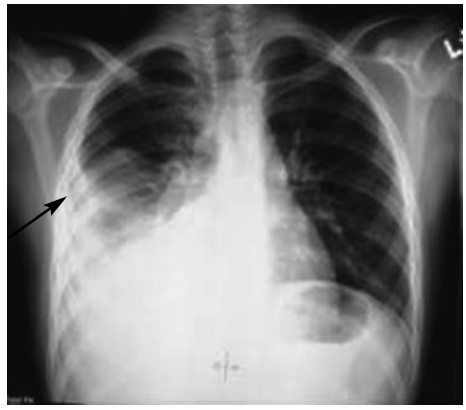
Stage I

Traditionally, parapneumonic effusions associated with Stage I empyema are treated by tube thoracostomy and intravenous antibiotic therapy. This approach is reserved for children with simple effusions that are not loculated. In our experience, the effusion can be adequately assessed by either plain chest radiography with decubitus films or, more effectively, by ultrasonography (US) or chest computed tomography (CT). We do not advocate simple aspiration of the fluid in these cases, as there is a high rate of recurrence.

Stage II

Progression to the second stage of empyema with the development of fibrinous adhesions and loculations is best treated by debridement of the pleural space. Instillation of fibrinolytic agents into the pleural space for management of a complicated (loculated) parapneumonic effusion has been successfully employed in adults and recently advocated in select pediatric patients. However, one must be cautious when extrapolating adult experience into pediatric practice. The attractiveness of fibrinolytic therapy in adults often stems from the significant comorbidities that make operative intervention relatively prohibitive. In the pediatric population, significant comorbidity is rare, and the overwhelming majority of patients with empyema are previously healthy. In a multicenter, randomized, placebo-controlled trial, the addition of urokinase did not reduce hospitalization when compared to placebo. Moreover, the length of hospitalization for children treated with urokinase is still uniformly longer than that of children treated by early decortication and debridement. Thus, the use of fibrinolytic therapy is employed in the rare case in which operative risks outweigh the benefit.

1 To prevent the development of end-stage empyema, we advocate minimally invasive debridement of the pleural space when Stage II empyema is confirmed by US or CT scan. On this CT scan, a large amount of fluid (arrow) with consolidation and collapse of the right lung parenchyma is seen. The chest radiograph 1 month postoperative from thoracoscopic debridement and decortication is almost normal (right).



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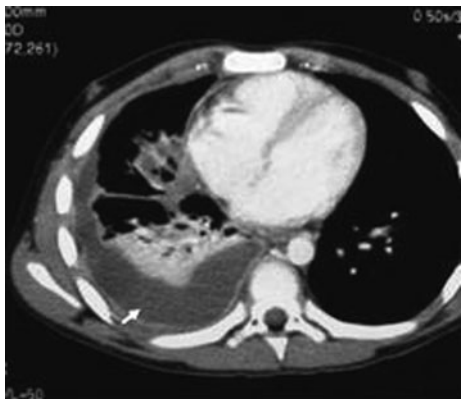

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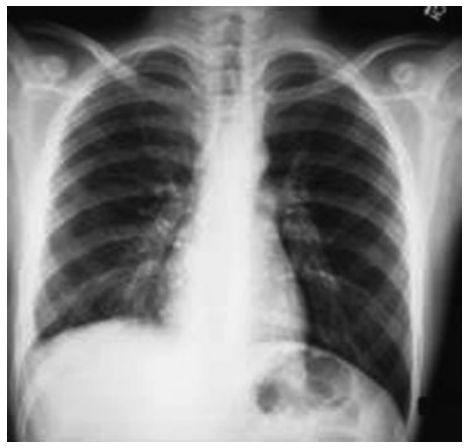


1c

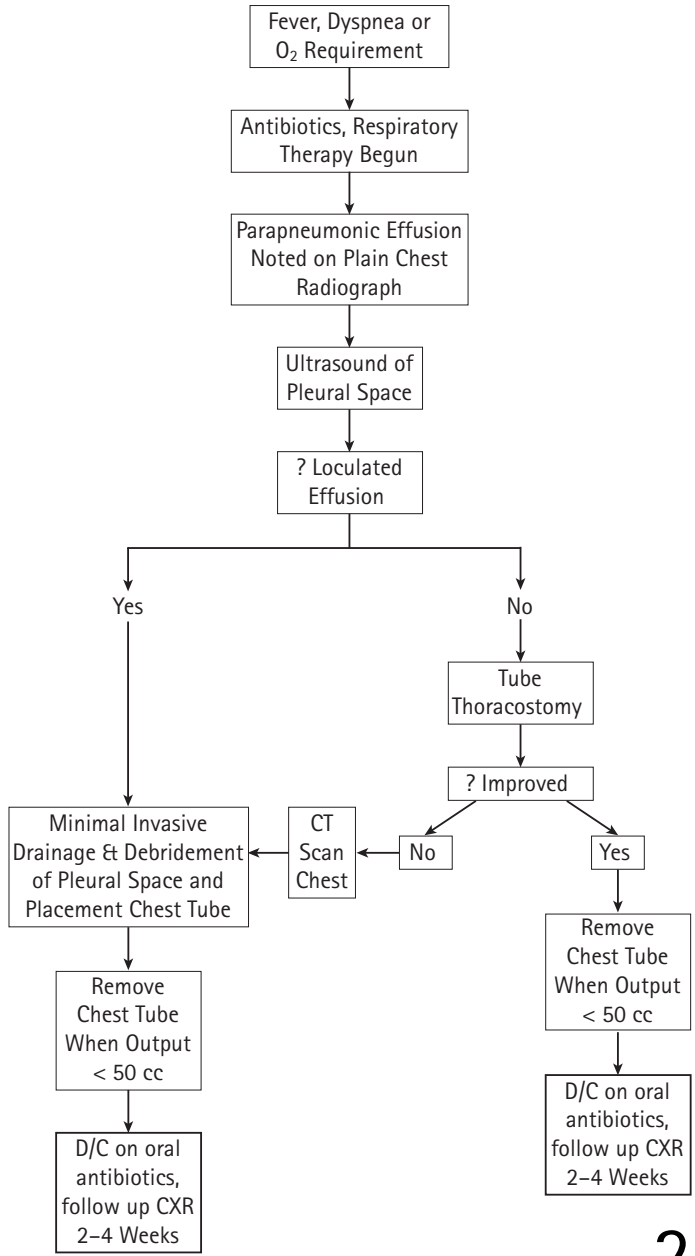
Continued conservative therapy with intravenous antibiotics and tube thoracostomy with or without the addition of fibrinolytics risks the development of Stage III empyema and subsequent lung trapping, resulting in respiratory dysfunction. The majority of children undergo thoracoscopic-assisted debridement and drainage of the pleural space, after which appropriately placed chest tubes are inserted under direct visualization. Infants and very small children can also be effectively managed by pleural debridement through a muscle-sparing mini-thoracotomy. When minimally invasive techniques are not available or feasible, open thoracotomy and pleural debridement and/or decortication are required.

Stage III

Minimally invasive techniques are nearly impossible to accomplish safely when the visceral and parietal pleural surfaces have fused in Stage III empyema. Patients with entrapped lung and persistent pulmonary dysfunction benefit from open decortication to liberate the lung from the thickened visceral and pleural peel and allow for re-expansion of the underlying pulmonary parenchyma.

PREOPERATIVE ASSESSMENT AND PREPARATION

2 An algorithmic approach to patients with empyema is employed. This algorithm depicts our preferred management strategy for patients presenting with suspected empyema. Either an ultrasound or a CT scan is performed to evaluate for loculations within the parapneumonic effusion. If loculations are present, the patient proceeds along the pathway to thoracoscopy as soon as possible. However, if there is no evidence of loculations within the parapneumonic effusion, tube thoracostomy is initially performed, with either resolution of the disease process or thoracoscopic debridement if the empyema persists. Children are started on intravenous antibiotic therapy consisting of a third-generation cephalosporin (ceftriaxone) and a macrolide (clindamycin). For the rare pediatric patient with hospital-acquired pneumonia, Gram-negative coverage should be added.

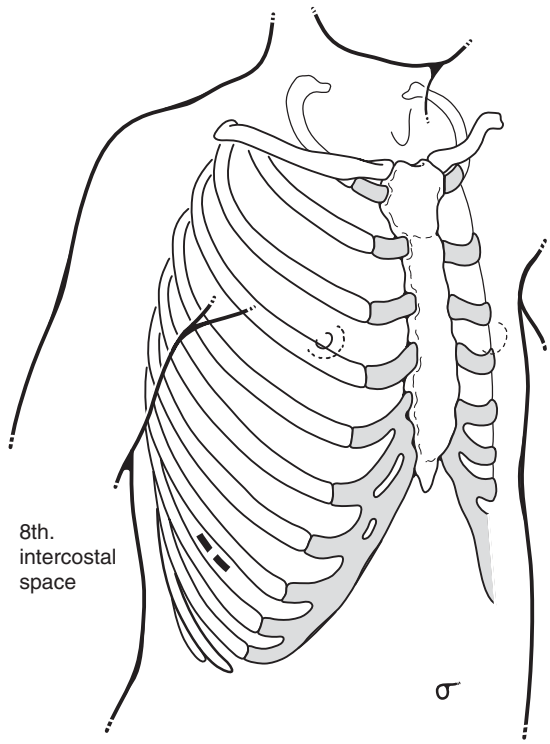


2

THERAPEUTIC OPTIONS AND MANAGEMENT

Tube thoracostomy

3 Patients requiring tube thoracostomy are sedated with midazolam, and intravenous narcotics are used for pain control. Cardiorespiratory monitors are placed. The skin and soft tissues about the eighth intercostal space in the mid-axillary line are infiltrated with either 0.25 percent bupivacaine or 1 percent lidocaine (1 mL/kg maximum). A 1 cm transverse skin incision is made in the skin of the intercostal space below which the tube will pass. Using blunt dissection, a soft tissue tunnel is created over the rib and into the pleural space just cephalad to the rib. Blunt or sharp dissection is then used to open the pleura. A large chest tube is secured to the end of a hemostat, which is used to insert the tube into the pleural space, directing it posterior and cephalad. The tube is sutured to the skin with a non-absorbable suture, and a sterile occlusive dressing gauze is applied. The tube is then connected to suction. A post-procedure chest radiograph is obtained to ensure proper placement of the tube.



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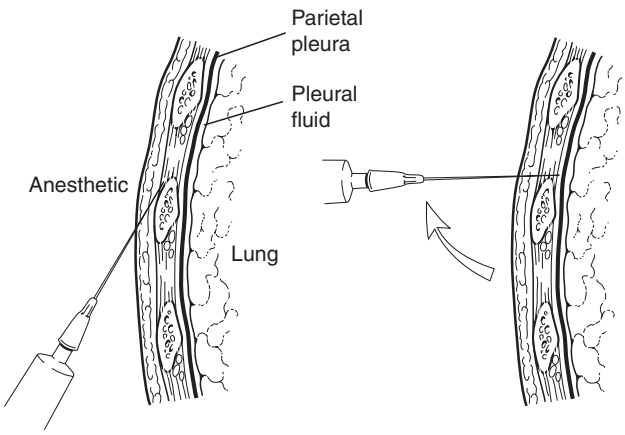

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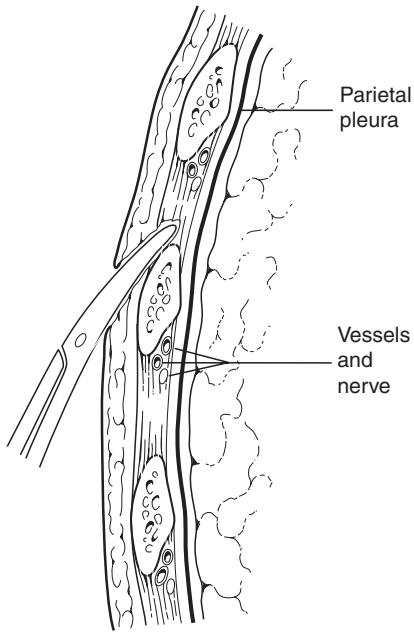

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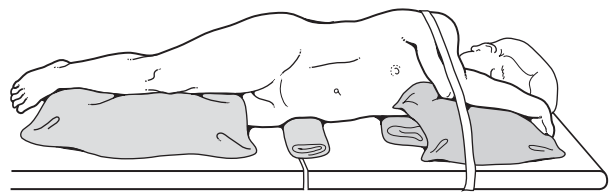
In the rare case in which fibrinolytic therapy is indicated, 20 000 U of urokinase and 20 mL of sterile water are instilled through the chest tube, which is subsequently clamped for 2 hours. During that time, the patient's position is frequently changed to facilitate pleural distribution of the agent. Treatment is repeated three times a day for 1–3 days, if necessary. Efficacy of treatment is determined by increased chest tube drainage and/or improvement in the radiographic appearance.

Anesthesia

Except for tube thoracostomy, the debridement and decortication operation is completed under general anesthesia with tracheal intubation and is facilitated by single lung ventilation, if possible. To this end, we employ either double-lumen endotracheal intubation or tracheal intubation with ipsilateral bronchial blockade.

Position of patient

4 The patient is positioned in the lateral decubitus position with the affected side up, and an axillary roll is placed. We prefer to position the patient's iliac crest just inferior to the kidney rest. When raised, the intercostal spaces are widened for access to the thoracic cavity.



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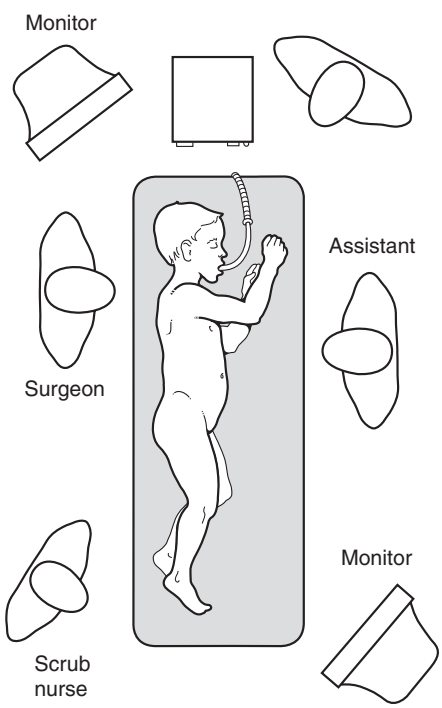

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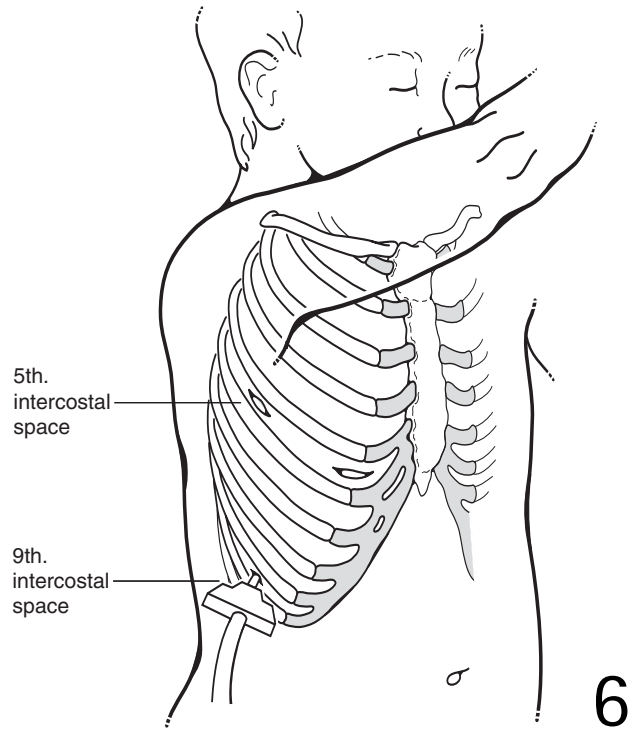
Thorascopically assisted minimally invasive pleural debridement

5 Prior to preparation and drape, if single lung ventilation is possible, the surgeon asks the anesthesiologist to occlude ventilation to the affected lung. This diagram depicts positioning of the personnel for performance of a right thoracoscopic debridement and decortication. After placing the patient in a lateral decubitus position and elevating the kidney rest, the patient is prepped and draped as widely as possible. The surgeon and assistant stand opposite each other with the scrub nurse to the surgeon's right. It is helpful to have a video monitor at the head of the table and one at the foot of the table as the telescope is often rotated between the three small incisions. Part of the dissection is usually in the more cephalad aspect of the thoracic cavity, while the majority of it is in the caudal aspect of the affected thoracic cavity.

6 As almost all empyemas affect the caudal portion of the thoracic cavity, initial access to the affected chest cavity is gained through a 10 mm incision in the anterior axillary line at the level of the fourth or fifth intercostal space. A 10 mm valved cannula with a blunt tip is then introduced through this incision into the thoracic cavity. Valved cannulas are used initially to allow positive pressure insufflation up to 6 torr to help compress the lung and create a working space within the rigid thoracic cage. A 10 mm angled telescope is inserted through the cannula and is used to create a working space by sweeping the adhesions and lung away from the chest wall. (We initially used a 5 mm angled telescope, but now use a 10 mm telescope because of the increased light emitted through this larger endoscope.) Once the underlying lung is adequately freed, a second 10 mm incision and cannula are inserted in the same interspace in the posterior axillary line. The third incision and cannula are positioned in the mid-axillary line in the ninth or tenth intercostal space. This incision will be the site for exteriorization of the chest tube. With this arrangement, a working triangle is formed.



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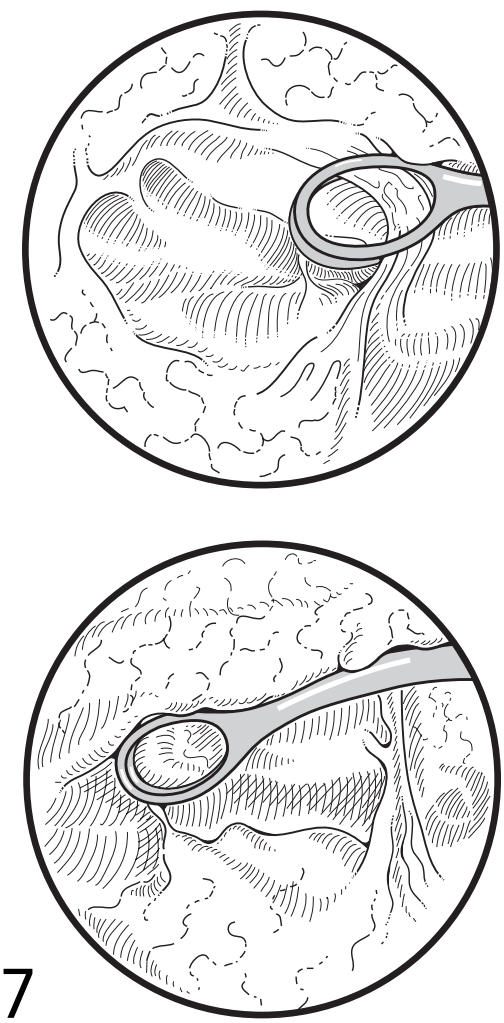

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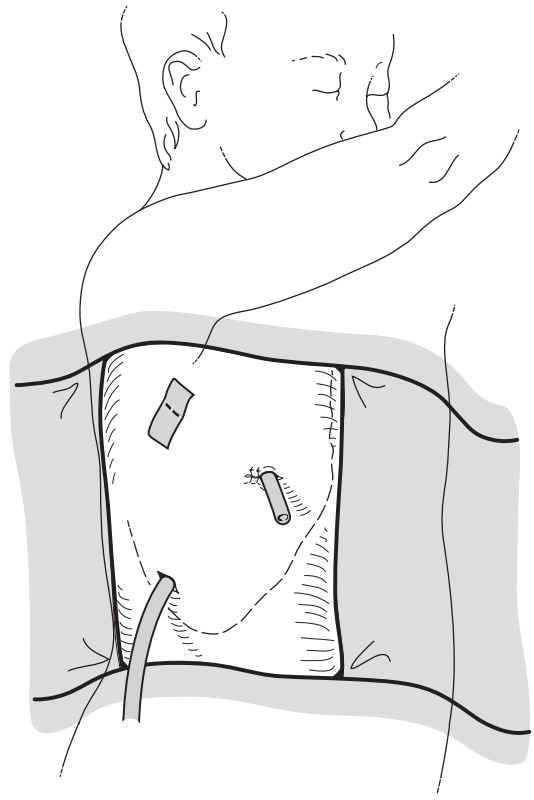
7 Once the lung is collapsed and an adequate working space is achieved, the cannulas can be removed and curved ring forceps introduced directly through the chest wall incisions to debride the pleural space. The use of these forceps enables large portions of the inflammatory debris to be removed without injury to the underlying lung. The telescope is rotated from one port to another while the surgeon and assistant work through the other two sites to debride the pleural space. Using the ring forceps, the parietal and visceral pleural peel is removed under telescopic visualization.

Great care is taken not to enter the lung parenchyma, but small parenchymal tears will probably heal spontaneously. Samples for Gram stain and culture are routinely sent by attaching a Lukens trap to the suction device. Once all pockets of fluid have been drained and the majority of pleural debris removed, a single chest tube is placed through the most caudal incision into the pleural space under direct vision.



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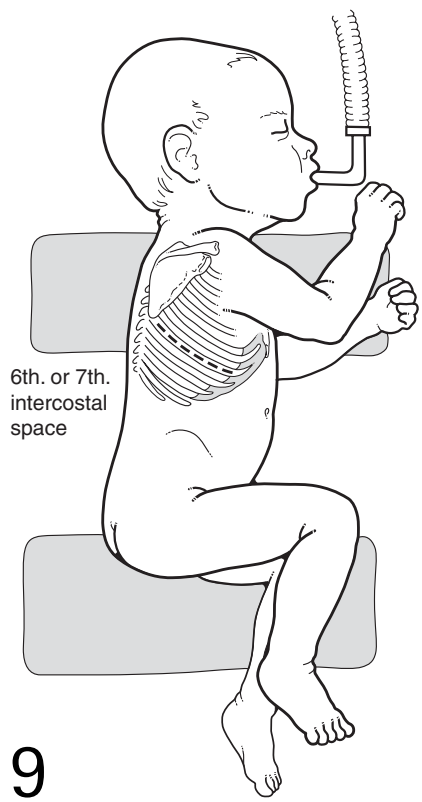
8 To lessen the possibility of a pneumothorax developing when the tube is removed, the tube is tunneled over the rib space cephalad to the incision through which it is going to exit. The incisions are closed with absorbable suture, and sterile dressings are applied. If a chest tube was placed prior to the thoracoscopy, a small portion of a latex drain is inserted in this incision to help prevent a wound infection at this site.



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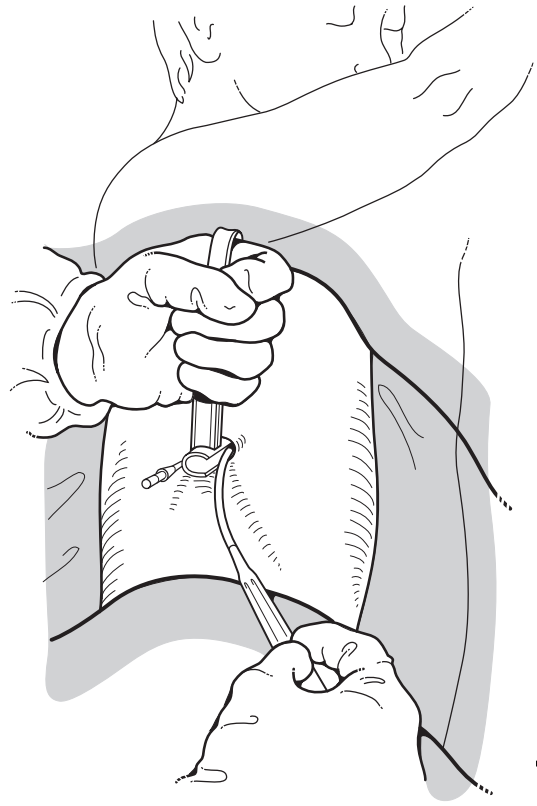
Mini-thoracotomy pleural debridement

9 When videoscopic expertise is not available or the patient is an infant, pleural debridement through a small thoracotomy incision can be easily performed. A small (3–4 cm) incision is created in the mid-axillary line at the level of the fifth or sixth intercostal space.

10 Use of an appropriately sized mediastinoscope with a light source facilitates visualization of the pleural space. Pleural debridement is then accomplished under direct vision. A suction device can be inserted through the mini-thoracotomy, by means of which loculations can be lysed and infected debris removed. Also, ring forceps can be inserted through this incision for debridement, with visualization achieved through the mediastinoscope. An appropriate-sized chest tube is tunneled over the ribs inferior to the incision. The intercostal muscles are re-approximated with absorbable suture and the chest tube secured to the skin with non-absorbable suture.



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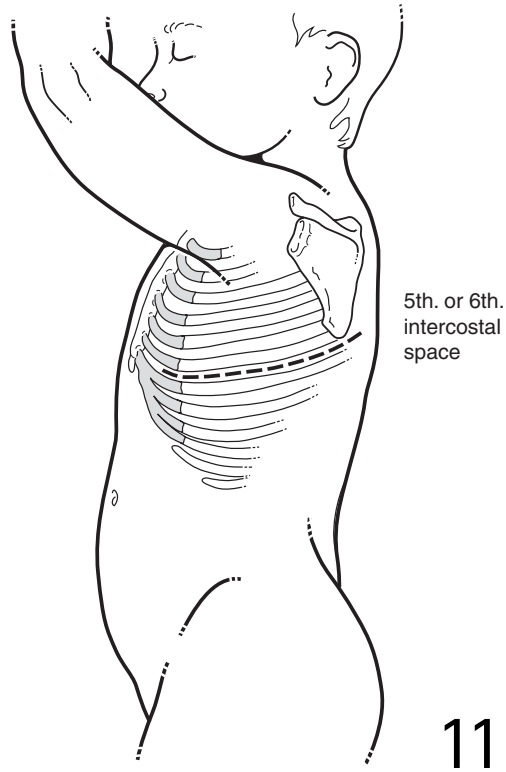
Thoracotomy

Pleural debridement via a posterolateral thoracotomy is reserved for patients in whom thoracoscopy or mini-thoracotomy is unsafe or has proven ineffective. This is rarely necessary. Open decortication is reserved for patients with persistent pleural thickening (usually symptoms > 6 months) and persistent abnormalities in pulmonary function or evidence of entrapped lung on CT scan.

11 Through a posterolateral thoracotomy in the fifth or sixth intercostal space, the pleural space is developed, and the visceral pleural peel is dissected away from the pulmonary parenchyma. This tedious and meticulous dissection is facilitated by incising the peel in its thickest portion, and teasing it away from the lung with a Kitner. This dissection can be quite bloody and proceeds until the peel has been removed in its entirety.



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The lung is then observed for air leak. Large air leaks can be controlled with a suture ligature or, alternatively, with fibrin sealant. Small areas can be covered with either fibrin sealant or observed, as most will resolve spontaneously. A tunneled thoracostomy tube is placed caudal to the incision, and the ribs are re-approximated with large absorbable suture. The lung is observed for full re-expansion prior to final closure of the chest.

Complicated empyema in critically ill patients who may not tolerate thoracoscopy or a lengthy open debridement may be managed by rib excision and tube drainage of the cavity. This is accomplished by excising a small segment of one to three ribs at the most dependent portion of the thoracic cavity followed by insertion of a large thoracostomy tube, which is secured to the skin and connected to suction. Thereafter, the tube is gradually withdrawn after the tube is trimmed, and drainage is controlled with an ostomy appliance. Fortunately, this approach is seldom necessary in children.

POSTOPERATIVE CARE

The chest tube is initially placed on 20 cmH_2O suction. A postoperative chest radiograph is obtained in the recovery room to ensure that the lung has expanded completely and that the chest tube is in the appropriate position. The chest tube is maintained on suction until drainage is less than 50 mL in a 24-hour period. Thereafter, it is placed to water seal, and removed when there is no evidence of air leak, the lung is completely expanded, and there is no significant re-accumulation of pleural fluid on chest radiography. Patients are discharged when they are afebrile and the chest tube has been removed. In most cases, discharge occurs between the fourth and sixth postoperative day. Patients are discharged on oral antibiotics to complete a 14-day course. Rarely, the antibiotics can be targeted to a specific organism. If not, a broad-spectrum agent such as amoxicillin/clavulonic acid is used. A follow-up outpatient evaluation and chest radiograph are recommended 3–4 weeks following discharge.

OUTCOME

Between 2001 and 2003, we treated 25 children with Stage II empyema by thoracoscopic pleural debridement. The mean postoperative hospitalization was 5.5 days, with no child returning with recurrence. This is similar to other reports in the literature showing early thoracoscopic intervention leads to shorter hospitalization, reduced cost, and fewer instances of patients proceeding to Stage III disease.

FURTHER READING

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Mediastinal masses

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PRINCIPLES AND JUSTIFICATION

Mass lesions of the mediastinum have multiple origins and may appear at any age throughout infancy, childhood, and adolescence. The mass may be cystic or solid and of either congenital or neoplastic origin. The symptoms produced by a mediastinal mass are almost as diverse as the underlying pathology of these lesions, but most symptoms are due to the 'mass effect' of the lesion, which may compress the airway, esophagus, or the lung. Occasionally they present with pain resulting from inflammation produced by infection or perforation of a cyst. Invasion of the chest wall by a malignant tumor will also result in pain. Many mediastinal lesions, in fact, are found as a radiographic abnormality on a study obtained for symptoms unrelated to the mass. Respiratory symptoms of expiratory stridor, cough, dyspnea, or tachypnea require urgent investigation. Cystic or solid lesions located at the carina may produce major airway obstruction. These lesions are often 'hidden' in the normal mediastinal shadow and may not be apparent on the anterior–posterior or lateral chest radiographs. Orthopnea and venous engorgement from superior vena caval syndrome are found with extensive involvement of the anterior mediastinum and are harbingers for respiratory obstruction upon induction of a general anesthetic. Less frequently, dysphagia from pressure on the esophagus is the presenting symptom. Neurologic symptoms from spinal cord compression or Horner's syndrome are seen with neurogenic tumors arising in the posterior mediastinum.

INDICATIONS FOR RESECTION

Management of these lesions is determined by the presumed diagnosis. Cystic lesions in the anterior mediastinum are generally resected. Acute enlargement in thymic cysts has been noted following viral respiratory illnesses. Teratomas, because of their possible malignant degeneration, are also resected. Lymphangiomas may secondarily involve the mediastinum, with their predominant component in the cervical–facial area. Isolated mediastinal involvement is seen in less than 5 percent of cases. Pericardial cysts are the most innocent of these lesions and if well demonstrated on scans and radiographs, often are simply followed, because they rarely increase in size and are unlikely to compress any vital structures.

The solid lesions require establishment of a histopathologic diagnosis. The most common solid tumor in the anterior mediastinum is Hodgkin's disease, followed by non-Hodgkin's lymphoma. Primary treatment of these tumors is by chemotherapy or radiotherapy; the surgeon's role is to establish the diagnosis. The primary treatment of the malignant germ-cell tumors is also chemotherapy. Surgical resection is not generally recommended for any of these lesions.

Teratomas or dermoids are the only neoplastic lesions that require resection as they may become secondarily infected or undergo malignant degeneration. Retrosternal thyroid goiters are resected through the neck.

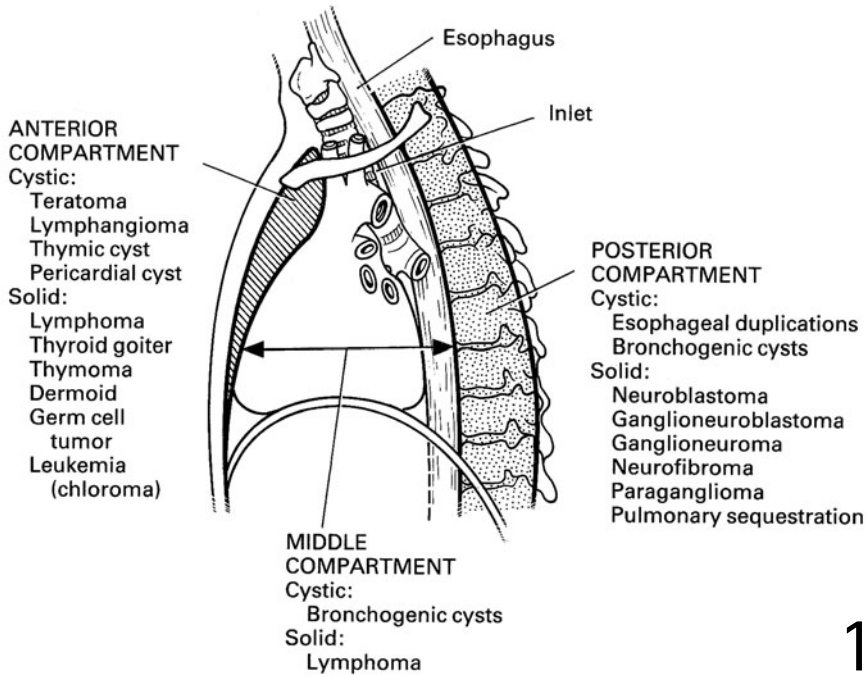
Bronchogenic cysts and esophageal duplications arise in the middle and posterior mediastinum. They develop in the embryo during division of the aerodigestive systems. Bronchogenic cysts are generally lined by respiratory epithelium and esophageal duplications by intestinal mucosa, but ectopic mucosa may be present in both lesions. These lesions should be resected because of their potential for growth with accumulation of secretions. They can also become secondarily infected or develop malignancy. Lesions with gastric mucosa can erode into the bronchus, esophagus, or pleural cavity.

Solid tumors in the posterior mediastinum should be resected. Resection of a thoracic neuroblastoma is a major component of its treatment. The requirement for further treatment with radiation or chemotherapy will depend on the age of the child, the presence of metastatic disease, and the cytogenetic findings of the tumor, particularly amplification of the *N-myc* oncogene or normal ploidy, both of which

suggest an aggressive tumor. Ganglioneuroma, while a benign tumor, may grow locally, may erode the ribs, and may extend into the spinal canal producing neurologic symptoms. While these benign lesions are often found when asymptomatic, resection generally is recommended to establish the diagnosis and prevent local extension. A paraganglioma (extra-adrenal pheochromocytoma) should be removed to control the systemic manifestations of neuropeptide production. The patient should be well prepared for surgery with alpha-blocking and beta-blocking agents and volume repletion. Pulmonary sequestrations are generally resected to obtain a definitive pathologic diagnosis, to prevent an arteriovenous shunt, and to avoid infection in the intralobar sequestrations.

DIAGNOSIS

The preoperative diagnosis of a mediastinal mass can be obtained quickly with only a few studies. Anterior–posterior and lateral chest radiographs will demonstrate the area of the mediastinum in which the mass arises. The location and knowledge of whether the mass is cystic or solid and the age of the patient will often allow an accurate diagnosis to be made.



1 Lesions occurring in each of the three compartments of the mediastinum are shown, grouped by their cystic or solid nature. Further investigation is determined by the location of the lesion.

1

Masses in the anterior compartment

Masses in the anterior mediastinum may produce respiratory symptoms and cause compression of the trachea. Computed tomography (CT scan) is generally best for the evaluation of masses in this area: it defines the cystic or solid nature of the lesion and most accurately demonstrates the extent of tracheal compression. The extent to which the trachea is compressed will determine the safety of anesthesia required for further diagnosis or resection.

Cystic lesions may be differentiated by their structure and location. A teratoma generally has both cystic and solid components, with areas of varying density that are well demonstrated on the CT scan. Most lymphangiomas have multiple cystic areas with very thin walls, which often extend up into the neck. Only a small proportion are limited entirely to the anterior mediastinum. Thymic cysts are often single, thin-walled, and contiguous with the thymus. Pericardial cysts arise in the inferior portion of the chest adjacent to the pericardium.

Solid anterior mediastinal lesions are also easily assessed on a CT scan. A dermoid or entirely solid teratoma has areas of varying fat and water density and often calcification. A substernal thyroid goiter arises from the thyroid gland and extends into the retrosternal space. Thymomas are extremely rare in children. Lymphomas involve multiple nodal sites. The CT scan also defines lymph node enlargement in the pulmonary hilum and pulmonary parenchymal lesions. Germ-cell tumors are uncommon, arising primarily in teenagers or young adults. These tumors are usually diagnosed with serum markers.

Masses in the middle compartment

A bronchogenic cyst at the carina may be 'hidden' in the mediastinal shadow on the chest radiograph despite significant respiratory distress. Fluoroscopy of the infant will demonstrate compression and anterior displacement of the airway, and ingestion of barium into the esophagus will demonstrate displacement of the esophagus posteriorly and confirm the presence of a space-occupying lesion. While CT scan and magnetic resonance imaging (MRI) will demonstrate these lesions more definitively, the sedation required for these studies in infants may be dangerous if respiratory compromise is significant.

Masses in the posterior compartment

The main cystic lesions in this area are bronchogenic cysts and esophageal duplications, which are typically ovoid in shape and may be diagnosed on routine radiographs.

The solid neural tumors have a fusiform shape and are based in the posterior sulcus between the vertebral bodies and the ribs. The age of the patient will give some hint of the diagnosis: neuroblastomas and ganglioneuroblastomas arise more often in infants. Ganglioneuromas occur in older children and are generally asymptomatic, but can extend into the spinal canal and produce neurologic symptoms. Neurofibromas arise primarily in conjunction with neurofibromatosis (von Recklinghausen's disease) and are often associated with scoliosis. Paragangliomas may arise in the posterior mediastinum, although they are rare. They often present with symptoms related to catecholamine secretion, particularly paroxysmal hypertension, diaphoresis, and palpitations. Extralobar pulmonary sequestrations arise in the posterior mediastinum with arterial supply from the aorta. They can generally be distinguished by a triangular shape.

Magnetic resonance imaging is often used to evaluate patients with masses in the posterior mediastinum, because it provides a better definition of possible extension of the tumor into the spinal canal than does a CT scan. It is important to identify this extension prior to surgical resection. In infants and young children in whom neuroblastoma is a major diagnostic concern, evaluation of metastatic disease is also important. This should include a bone marrow biopsy and bone scan, and urine should be collected to measure catecholamines, which are elevated in 95 percent of infants and children with neuroblastoma. The majority of esophageal and bronchogenic cysts are not associated with vertebral anomalies, but some rare cases in which large duplication cysts originate from the stomach, pancreas, or duodenum and extend into the thoracic cavity will demonstrate abnormalities of the vertebrae.

PREOPERATIVE

Solid lesions require further histopathologic diagnosis. The most common solid tumor in the anterior mediastinum is Hodgkin's disease, followed by non-Hodgkin's lymphoma. Other areas of lymph node involvement besides the mediastinum, particularly the neck, should be sought where biopsy could be more easily performed. In those rare instances where only the mediastinum is involved, a germ-cell tumor should be suspected and serum alphafetoprotein and human chorionic gonadotrophin levels (hCG) should be obtained. In cases where there is no extrathoracic tumor, either a needle biopsy with radiographic guidance or a limited anterior thoracotomy may be required to establish a tissue diagnosis. The rare chloroma of leukemia presenting as a mediastinal mass can be diagnosed with the initial complete blood count and bone marrow biopsy.

Preparation for surgery

The child should be prepared for surgery after completion of the diagnostic studies. If a bronchogenic cyst is compressing the airway sufficiently to produce pneumonia or respiratory distress, no undue delay should occur. Appropriate antibiotic coverage and physiotherapy should be instituted for pneumonia. Preliminary bronchoscopy should be avoided in these patients, because a tenuous airway in an infant or child will be further damaged by manipulation. Catecholamine-secreting tumors, primarily paraganglioma, require the institution of alpha-blocking and beta-blocking agents and volume repletion. Direct involvement of the bronchus is very rare, and compression of the airway can be defined most safely radiographically. In the occasional case of thymoma and associated myasthenia gravis, the neuromuscular deficit should be minimized as much as possible prior to surgical intervention.

Anesthesia

Anesthesia is of major concern primarily for solid lesions of the anterior compartment, which often compress the airway. A cross-sectional tracheal area of less than 50 percent of that expected for age or a peak expiratory flow rate of less than 50 percent of predicted suggests that a child is at significant risk for respiratory collapse on induction of anesthesia. Children with either of these two findings must be limited to local anesthesia with sedation; general anesthesia (particularly paralytic agents) must be avoided at all costs. Bronchogenic cysts in the area of the carina may also cause significant airway obstruction in infants, but the endobronchial tube can generally be passed down one of the mainstem bronchi to provide adequate ventilation until the pressure is relieved. This maneuver may not be feasible in children with a solid mass compressing the airway.

Appropriate monitoring of these patients requires transcutaneous oximetry and, in those children requiring extensive resections, central venous as well as arterial pressure monitoring. Uncuffed endotracheal tubes are routinely used in younger children to avoid any injury to the airway from pressure. An 'air leak' should be present around the endotracheal

tube to confirm that pressure on the subglottic mucosa, the narrowest segment of an infant's upper airway, is not excessive.

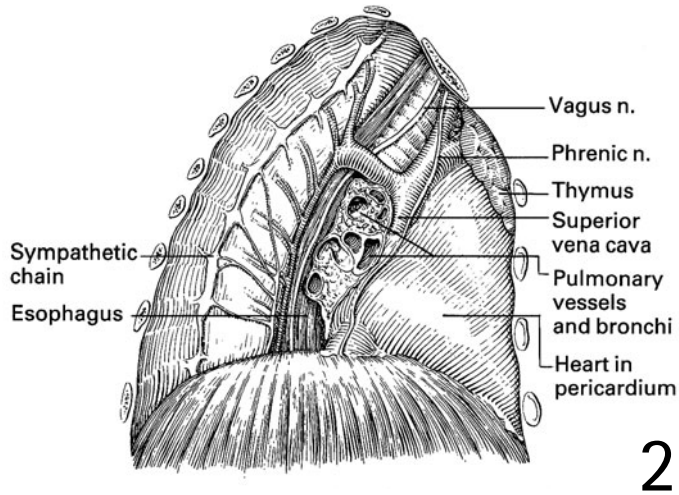
For thoracoscopic procedures, single lung ventilation will often facilitate the resection. Double-lumen endotracheal tubes are available down to 26 French caliber, which may be used on children of over 25 kg. In smaller children or infants, mainstem intubation of the contralateral bronchus or placement of a balloon catheter (bronchial blocker) will allow deflation of the lung to facilitate exposure.

Choice of approach and applied anatomy

The approaches to these masses are based primarily on their location and nature; most may be resected through a posterolateral thoracotomy. If a teratoma or a dermoid is primarily located in the midline, it may be most easily resected through a median sternotomy. Often these lesions are asymmetric and prolapse into one of the hemithoraces, allowing them to be resected from that side. This is also true of a thymic cyst. Extensive lymphangiomas, if they extend into the thoracic cavity, are also best dealt with through a thoracotomy. Sternotomy should be avoided for suspected lymphomas because compression of the airway can occur when the sternum is closed after biopsy of the mass. A posterolateral thoracotomy is the procedure of choice for lesions of the posterior compartment. Extension into the spinal canal from benign tumors requires a preliminary laminectomy with resection of the tumor or a combined laminectomy and thoracotomy. Swelling of the residual segment of tumor in the canal after resection of the thoracic component could produce neurologic sequelae.

Many of these lesions can now be approached with thoracoscopic techniques. The anatomic considerations and steps of the procedures are identical for either method.

2 The major structures of concern on the right side of the mediastinum are shown. Particular care should be taken to preserve the phrenic nerve, avoiding loss of diaphragmatic function. The upper mediastinum and carina are most readily approached from the right side because of the aortic arch and its branches on the left.



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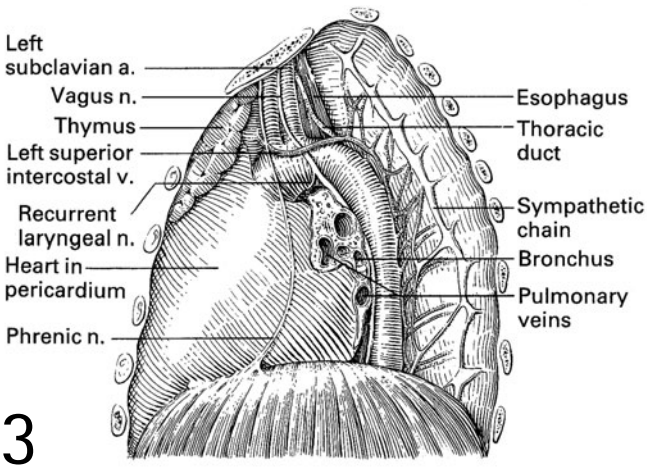

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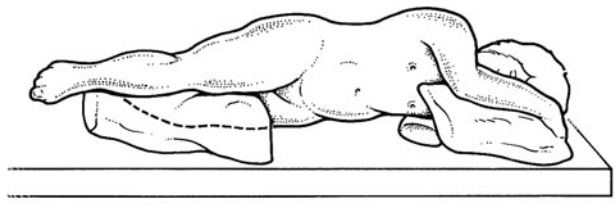
3 On the left side, in addition to the phrenic nerve, attention must be paid in the upper mediastinum to the course of the vagus nerve and the recurrent laryngeal nerve, which loops around the aortic arch before it ascends to the larynx.

OPERATION

Posterolateral thoracotomy and thoracoscopy

POSITION OF THE PATIENT

4 The patient's back should be perpendicular to the ground. The lower leg should be flexed and the upper leg straight, with a pillow placed between them. The axilla should be padded. The uppermost arm should be angled at 90° and brought anterior to the chest. Greater extension should be avoided, as traction injury to the brachial plexus may occur. Adequate padding of all weight-bearing areas on the table is critical, particularly for extended procedures. For a midline sternotomy the child is placed supine. The head must be adequately extended to provide ready access to the sternal notch without dislodging the endotracheal tube.



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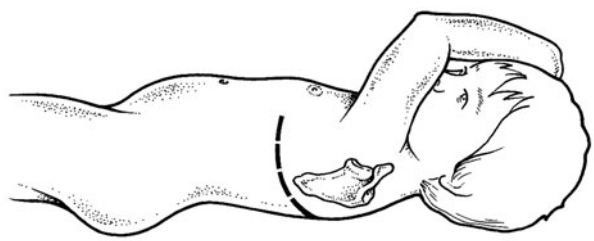


5

INCISION

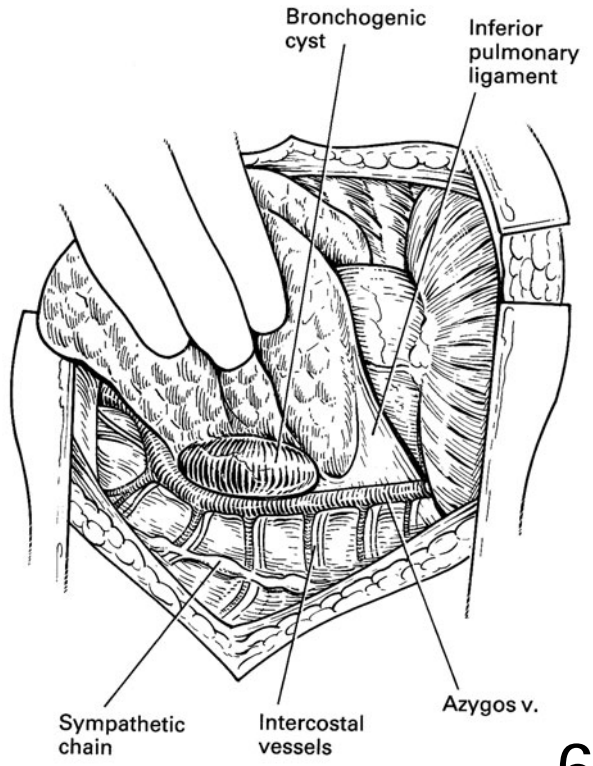
5 The incision curves from below the nipple in the estimated inframammary crease to a point two finger breadths below the tip of the scapula, traveling superiorly to a point midway between the scapula and the spinous processes. This incision does not have to extend very far superiorly in children because of the mobility of the scapula. The latissimus dorsi muscle is divided with electrocautery. The serratus muscles can generally be mobilized adequately anteriorly and are not divided.

The intercostal space is then entered. If the mass is in the superior mediastinum, the chest is best entered in the fourth intercostal space; if the mass is lower, the fifth intercostal space is used. Neurogenic tumors with an inferior location near the diaphragm are approached through the sixth or seventh intercostal space. A thoracoabdominal incision is occasionally required for extensive neurogenic tumors with abdominal and thoracic components. The surgeon should take care not to be 'trapped' through an incision that is too low and does not allow access to the apex of the mass. It is rarely necessary to remove a rib in children for adequate exposure. The pleura is opened and the chest is entered.

For thoracoscopic approaches, at least three and occasionally four port sites are required: one for the camera, one or two sites for the traction instruments, and one for the dissection instrument. These sites are often placed along the potential line for the incision should conversion to an open procedure be required. Often the middle of these sites is placed below the line for the incision for use with the camera and for a postoperative chest tube if one is required. The sites can be shifted anteriorly or posteriorly based on the location of the lesion, but adequate distance must be maintained between the sites to provide working space between the instruments.

Bronchogenic cysts

6 The thoracic cavity is explored to identify the extent of the mass to be resected and its relationship with the vital intrathoracic structures. The lung is retracted anteriorly to expose the mass. During resection of cystic lesions, aspiration is unnecessary unless the airway is compressed or the lesion is too large for safe dissection. Keeping the cyst filled with secretions actually facilitates its dissection. These lesions can be resected by either open or thoracosopic methods.



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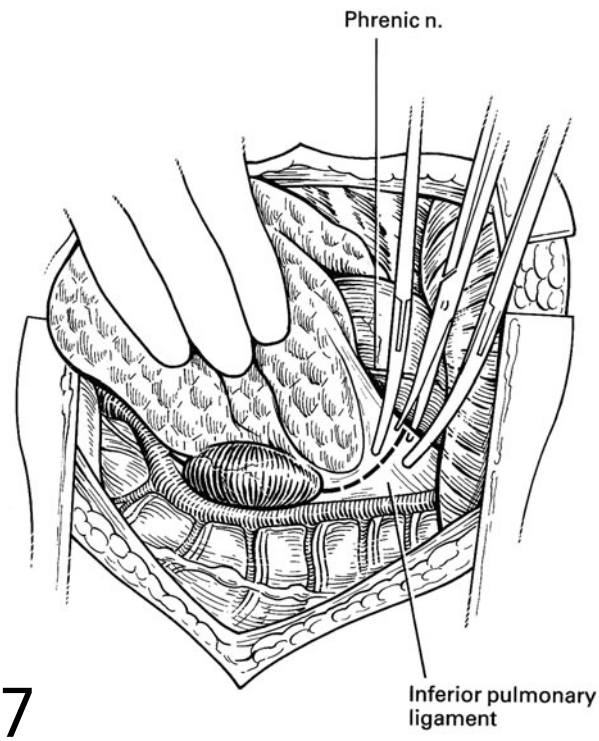
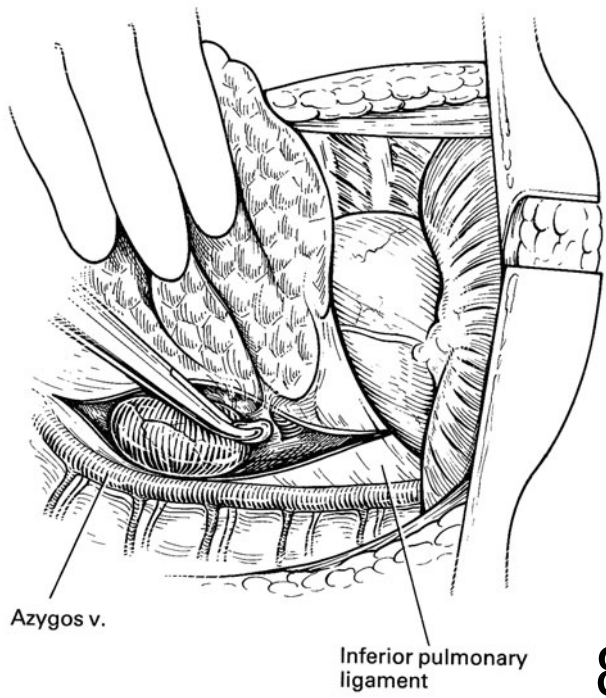


7

7 The pleura around a cystic lesion is first incised.

8 Bronchogenic cysts generally lie adjacent and posterior or lateral to the bronchus or trachea, but direct communication is extremely rare. These lesions can be easily dissected away from surrounding structures and removed intact. Extreme caution must be taken to avoid injury from dissection or cautery of the membranous part of the trachea and bronchus. Delayed recognition of this injury has been reported after thoracoscopic resections.



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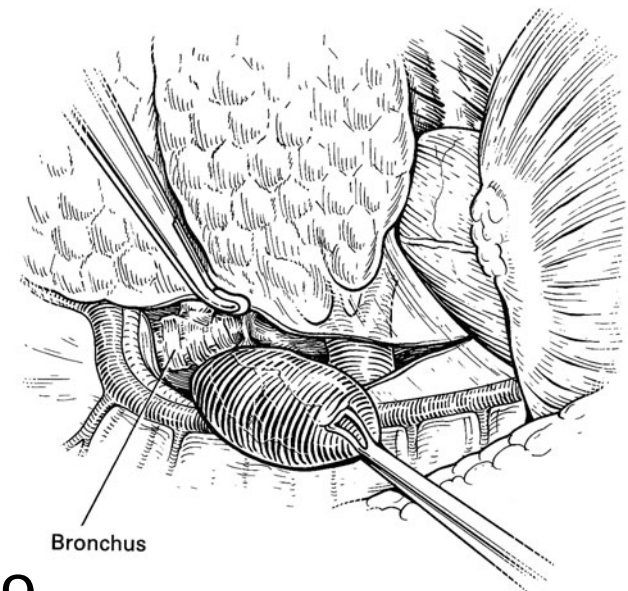


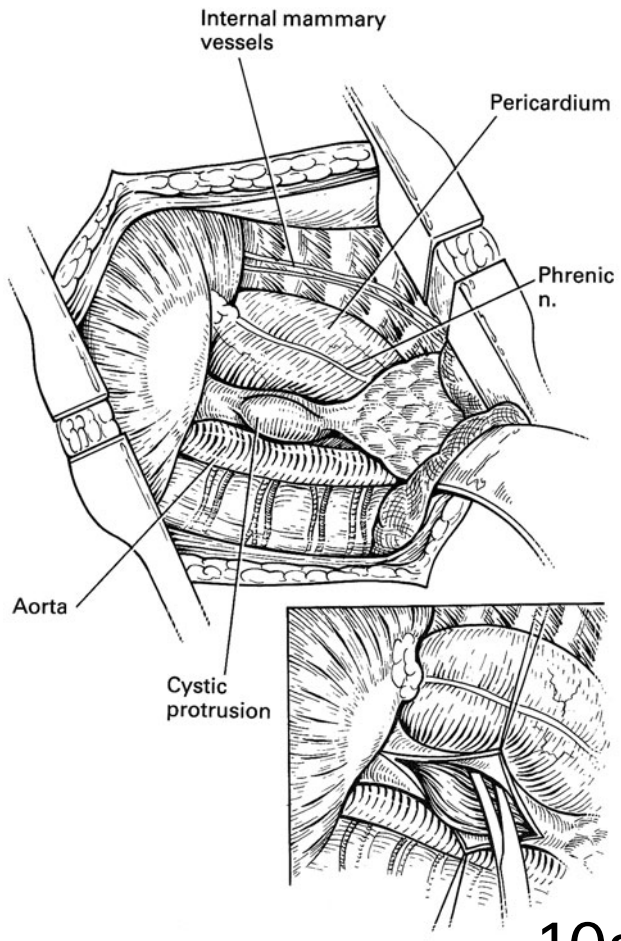
9

9 Cysts associated with the right bronchus, carina, and central left bronchus are best approached through the right chest; only more peripheral lesions of the left bronchus lateral to the aortic arch are resected through the left chest.

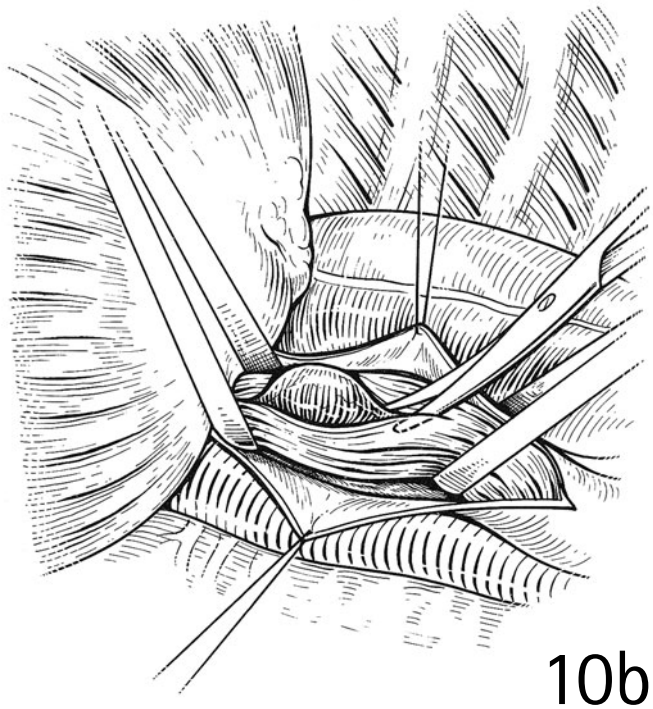
Inflammatory reaction around the cyst suggests ectopic gastric mucosa within the cyst or secondary infection of the cyst. Only rarely, when acid produced by the gastric mucosa has eroded through the cyst wall, will it be densely adherent to either the bronchus or esophagus. Significant hemorrhage or pulmonary reaction can occur in this situation. Cysts with gastric mucosa eroding into the bronchus or the esophagus may present with hemoptysis, hematemesis, or pain.

Esophageal duplications

10a,b Esophageal duplications are often surrounded by esophageal muscle. They are best approached through the side of the thorax into which they protrude. These can also be resected by either open or thoracoscopic methods. The pleura is incised first over the esophagus and then the muscle overlying the cystic lesion is opened longitudinally. It is particularly helpful to keep the cyst intact in these patients.



10a



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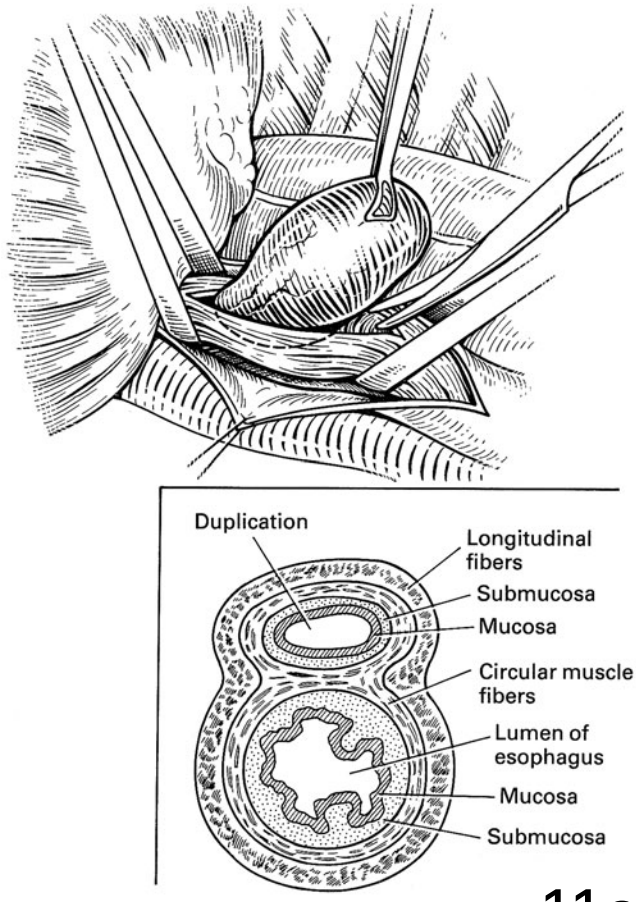
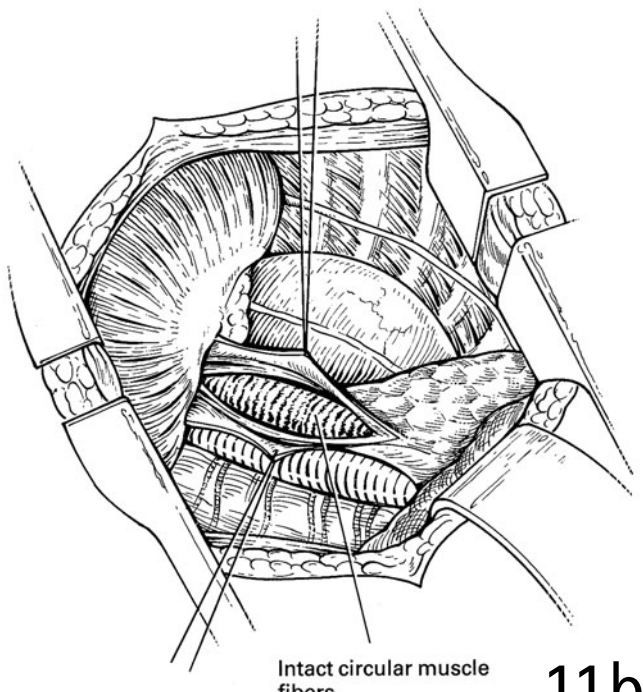


11a

11a,b Submucosal resection is the safest plane of dissection of the cyst from the esophagus to avoid an untoward entrance into the esophageal lumen. A single muscular layer comprises the common wall between the esophagus and the duplication.

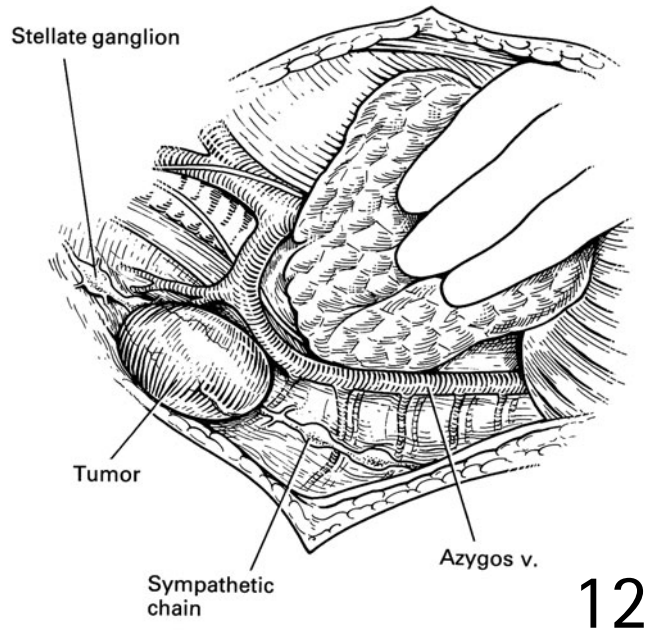


11b

Bronchogenic cysts and esophageal duplications should have their mucosa removed in its entirety. Any remaining mucosa may cause the cyst to recur. Aspiration or sclerosis of these lesions is not recommended because the mucosal surface will regenerate and produce a recurrent mass. The risk of development of malignancy in the mucosa must always be considered.

Solid posterior masses

12 Solid posterior mediastinal lesions are broad based and adhere to the ribs, intercostal muscles, and sulcus of the vertebral bodies. They commonly arise from the sympathetic chain and may involve the stellate ganglion. Resection of this ganglion with an apical tumor will produce Horner's syndrome with apparent ptosis, miosis, and anhidrosis. The family and child should be forewarned of this possibility.



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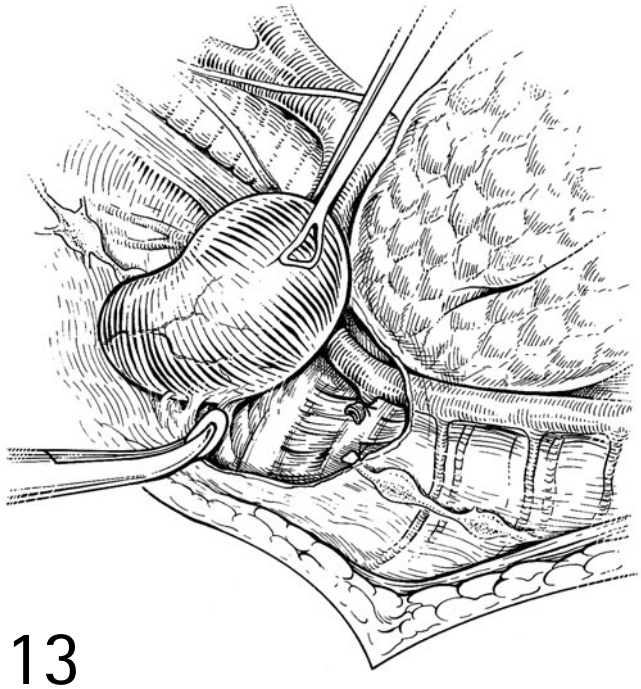
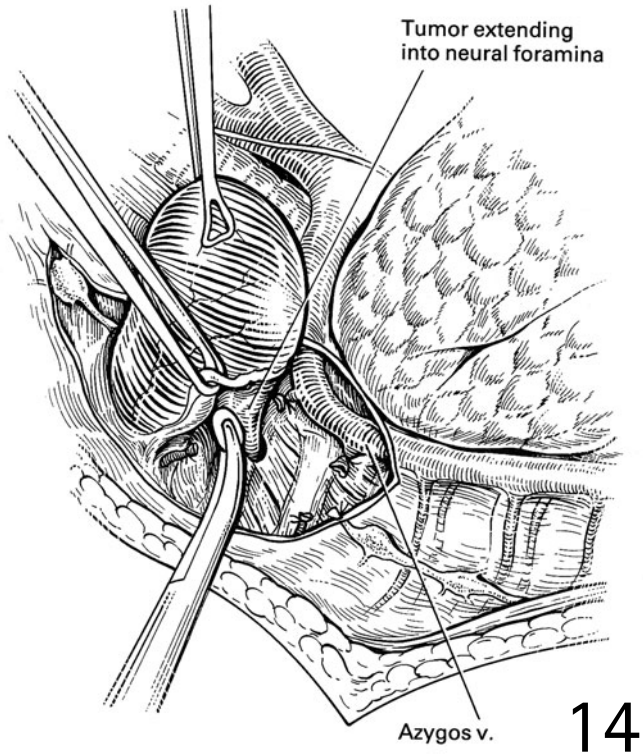


13

13 After incision of the pleura around the periphery, blunt dissection is used to elevate the tumor off the ribs. This will facilitate identification of the plane between the tumor and the intercostal muscles. These lesions may be densely adherent to the chest wall, making thoracoscopic resection challenging.

14 The most difficult part of this dissection occurs at the sulcus, where the tumor may extend into the neural foramina. An artery and vein accompany the nerve from each foramen. The use of bipolar cautery in this area avoids the risk of conduction of the current to the spinal cord. The aorta and esophagus, when involved, can generally be dissected from the anterior aspect of the tumor easily, as direct involvement of the tumor is rare. As the aorta or azygous vein is dissected forward, each of the intercostal arteries should be controlled and ligated or clipped or cauterized for thoracoscopic resections. The tumor rarely extends through the periosteum of the vertebral bodies. A combination of blunt and cautery dissection is utilized to mobilize the tumor off each of the vertebral bodies.



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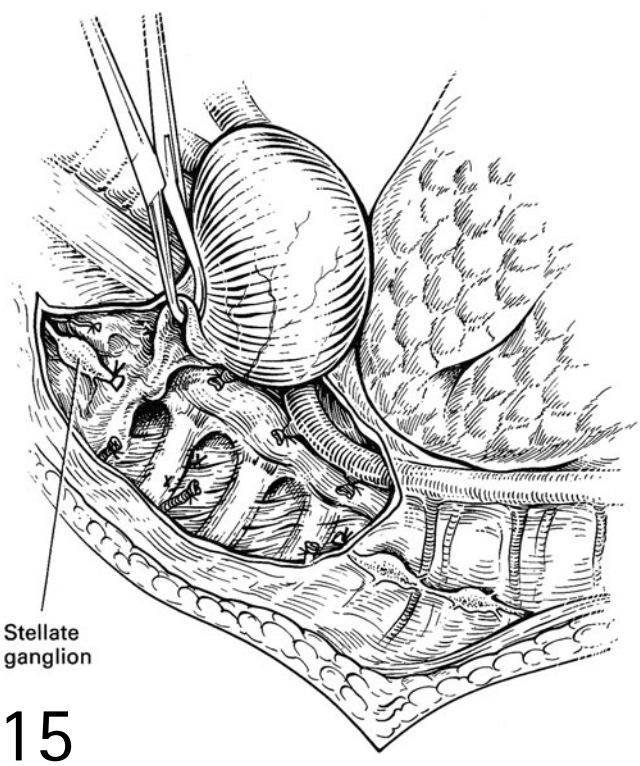

15

15 Dissection proceeds around the mass, working where progress can most readily be achieved. The sympathetic chain is divided superior and inferior to the tumor. The stellate ganglion should be preserved if possible to avoid Horner's syndrome. Ganglioneuromas and neurofibromas do not have an extensive blood supply and are firm and fibrous and easy to keep intact during the dissection. The neuroblastomas are much softer and more vascular and rupture should be avoided. If preliminary chemotherapy is administered, the neuroblastomas become much firmer, fibrotic, and also less vascular. A nasogastric tube in the esophagus will facilitate its identification and dissection from a large tumor.

Once the mass, whether cystic or solid, has been completely resected, the area should be inspected for any ongoing bleeding or evidence of lymphatic leak in the posterior lesions. These should be controlled and an intercostal chest tube inserted.

The bed of a resected neuroblastoma should be marked with radio-opaque tantalum clips to facilitate radiation therapy if it is required, particularly if gross residual tumor is present, which should be rare in the mediastinum, except for tumor extending through the neural foramina.

Wound closure

It is rarely possible to cover the defect with pleura, particularly in the solid tumors, where a significant amount of pleura is resected. A chest tube is placed in the mid-axillary line two intercostal spaces below the incision so it can be brought up through the muscle in a superior trajectory. It can also be brought out through an appropriately positioned port site. It is secured with non-absorbable sutures. The ribs are then approximated with pericostal sutures and each of the musculofascial layers is re-approximated with absorbable sutures. Subcuticular polyglycolic acid absorbable sutures are used to re-approximate the skin edges. For the thoracoscopic port sites, absorbable sutures are used to approximate the musculofascial layer and the dermis.

POSTOPERATIVE CARE

Management of the airway

Most children can be extubated following surgery unless the procedure has been particularly long or required significant volume replacement. In infants and children, a humidified oxygen tent will often be better tolerated than a facemask. In very small infants, a short period of postoperative ventilation will allow equilibration and progressive withdrawal of respiratory support. If a child is to remain intubated, an 'air leak' must exist around the endotracheal tube; this confirms that the uncuffed tube is not producing undue pressure on the tracheal or subglottic mucosa, with a risk of developing a postoperative stricture.

Intercostal drainage

Suction is employed for 24–48 hours and if drainage is insignificant, the tube can then be removed. Air leakage is rare following these resections. The intercostal tube is removed once it is confirmed that there is full expansion of the lung with no air leak. Serous drainage is common but does not usually require extended intercostal drainage. A chest radiograph should be obtained once the drain is removed to document continued full expansion of the lung.

Analgesia and sedation

Young children and adolescents require analgesia for the first few days after surgery. For major resections through a full posterolateral thoracotomy, an epidural catheter may provide optimum pain relief, particularly in adolescents. Infants require a shorter duration of analgesia. Sedation is also necessary in all infants and children during mechanical ventilation.

FURTHER READING

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26

Surgical treatment of chest wall deformities

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PECTUS EXCAVATUM

PRINCIPLES AND JUSTIFICATION

Pectus excavatum is the most common of chest wall deformities in infants, children, and adolescents. Its incidence is estimated at between 1 and 400 live births and 7.9 per 1000 births and it has a male-to-female ratio of 3:1. The exact etiology of pectus excavatum is unknown. It clearly has a genetic predisposition, with patients having a family history of chest wall deformities in 37 percent of cases. What is remarkable is that patients may have a family history of a carinatum or protrusion deformity as well as of a depression deformity. The pectus excavatum depression is created by two components. First is a posterior angulation of the sternum, generally at the level of the insertion of the second or third costal cartilages. Second is the posterior angulation of the costal cartilages to meet the sternum. In older teenagers, there may be posterior angulation of the most medial part of the osseous rib as well as the cartilaginous component. The pectus excavatum depression may be symmetric or asymmetric. In the asymmetric deformities, the more acute and severe depression is primarily on the right side. The rarest configuration of pectus excavatum is a combination of ipsilateral depression and a contralateral carinate protrusion. In approximately 90 percent of cases, the excavatum deformity is noted within the first year of life. This is in marked contrast to a carinatum deformity, where almost half of the patients have the protrusion noted after they enter the pubertal growth spurt.

Indications

Children with pectus excavatum may present with symptoms of shortness of breath during strenuous exercise and rapid development of fatigue. Symptomatic improvement after repair is frequently noted. Multiple studies have demonstrated a 'restrictive' defect with a decrease in the vital capacity, total lung capacity, and maximum breathing capacity. Despite the symptomatic relief often seen in patients and improvement in their exercise tests, the pulmonary function tests are not consistently improved after repair. Several recent studies have suggested that the symptomatic improvement may be produced as much by the relief of anterior compression of the heart, particularly on the right ventricle, as from the pulmonary components. It has also been demonstrated that the chest wall deformity as well as the physiologic components can have a significant adverse psychologic impact on patients. Therefore consideration for surgical repair must include both the physiologic and psychologic aspects of this congenital deformity. Children and their parents must be apprised of the risks and benefits of the surgery prior to proceeding with repair.

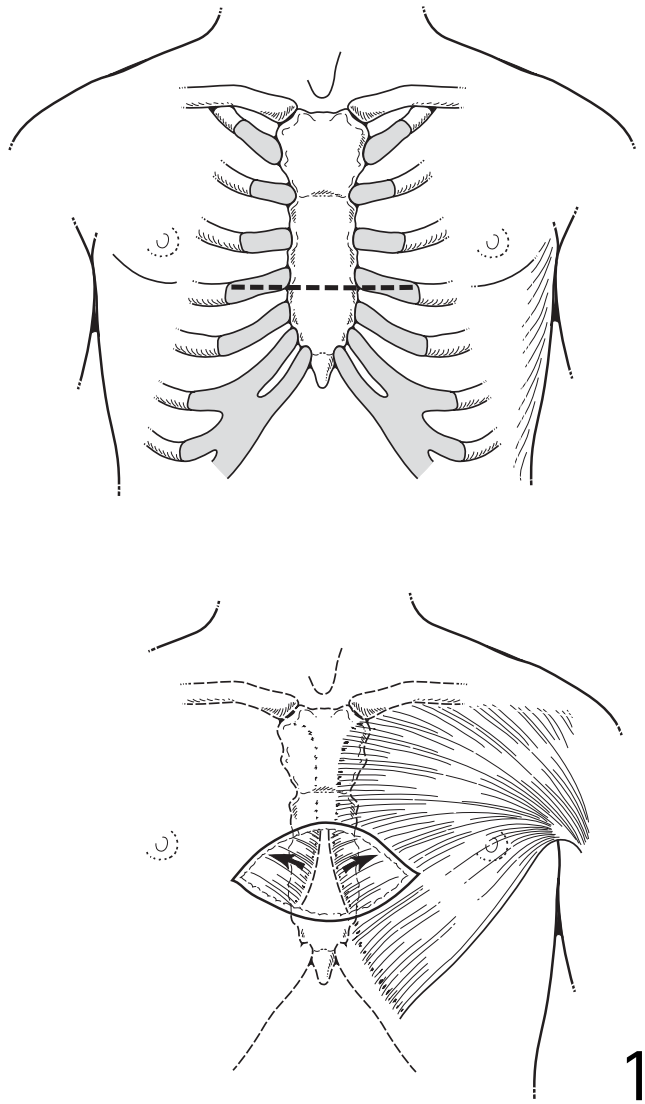
OPERATIONS

Multiple repairs have been developed for the correction of pectus excavatum, but none has been uniformly accepted as the optimal procedure. This would suggest that none of the alternatives provide perfect results. The current standard open repair is frequently attributed to Ravitch. His initial

description included resection of the costal cartilage and the perichondrium with anterior fixation of the sternum with Kirschner wires. Welch and Baronofsky subsequently stressed the vital importance of preservation of the perichondrium to achieve optimal regeneration of the cartilage after repair.

Ravitch procedure

1 In the standard open procedure a transverse incision is made below and well within the nipple lines and, in females, at the inframammary crease. The pectoralis major muscle is elevated from the sternum along with portions of the pectoralis minor and serratus anterior bundles.



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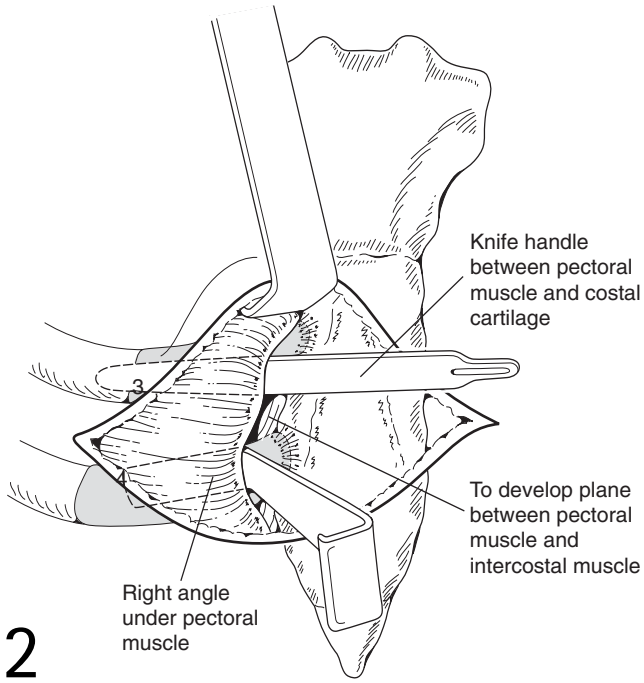

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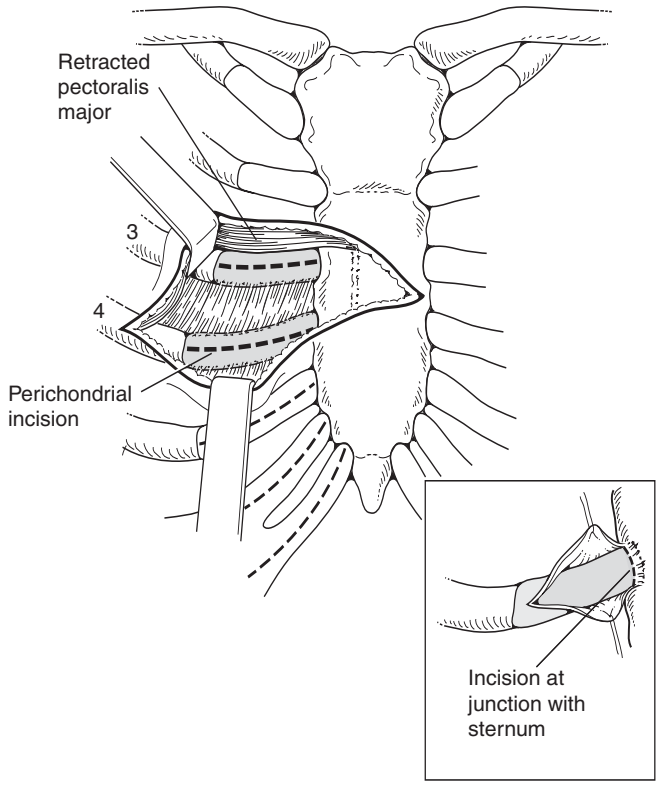
2 The correct plane of dissection of the pectoral muscle flap is defined by passing an empty knife handle directly anterior to a costal cartilage after the medial aspect of the muscle has been elevated with electrocautery. The knife handle is then replaced with a right-angle retractor, which is pulled anteriorly. The process is then repeated anterior to an adjoining costal cartilage. Anterior distraction of the muscles during the dissection facilitates identification of the avascular areolar plane and avoids entry into the intercostal muscle bundles. Elevation of the pectoral muscle flaps is extended bilaterally to the costochondral junctions of the third to fifth ribs and a comparable distance for ribs six and seven or to the lateral extent of the deformity.

3 Subperichondrial resection of the costal cartilages is achieved by incising the perichondrium anteriorly. The perichondrium is then dissected away from the costal cartilages in the bloodless plane between perichondrium and costal cartilage. Cutting back the perichondrium 90° in each direction at its junction with the sternum (inset) facilitates visualization of the back wall of the costal cartilage.



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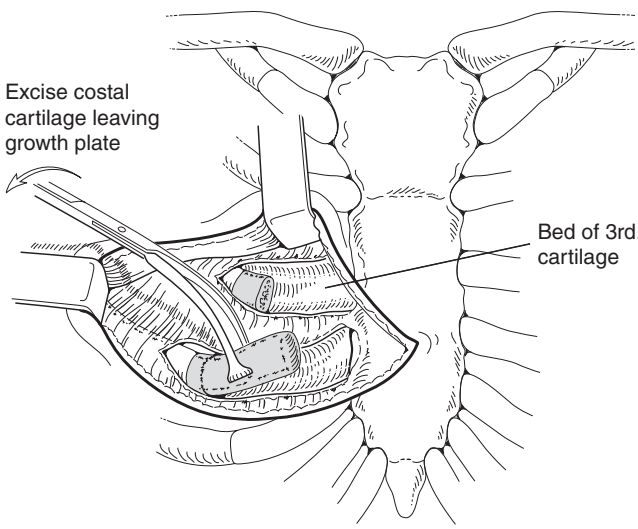

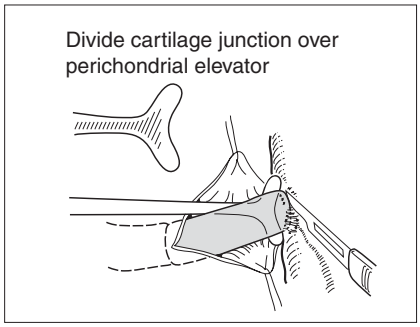

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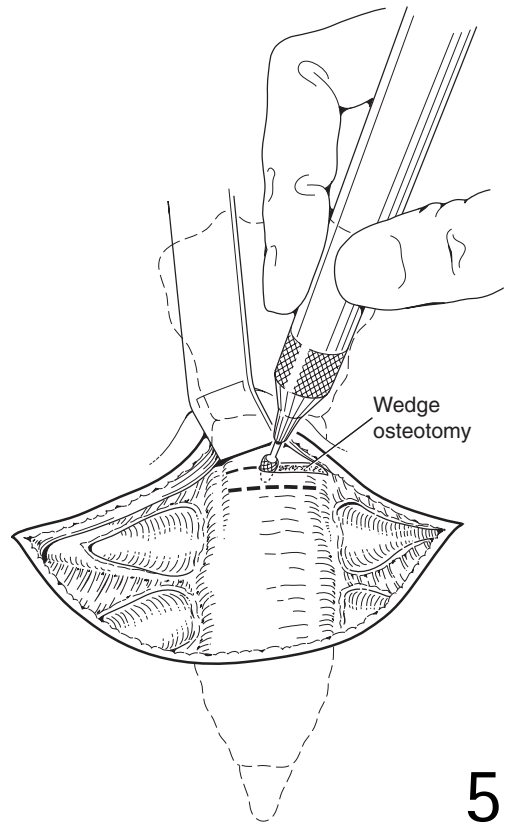
4 The cartilages are sharply divided at their junction with the sternum as a Welch perichondrial elevator is held posteriorly to elevate the cartilage and protect the mediastinum (inset). The divided cartilage can then be held with an Allis clamp and elevated. The costochondral junction is preserved by leaving a segment of costal cartilage on the osseous ribs by incising the cartilage with a scalpel. Costal cartilages three through seven are generally resected, but occasionally the second costal cartilages must be removed if posterior displacement or funneling of the sternum extends to this level. Segments of the sixth and seventh costal cartilages are resected to the point where they flatten to join the costal arch. Familiarity with the cross-sectional shape of the medial ends of the costal cartilages facilitates their removal. The second and third cartilages are broad and flat, the fourth and fifth are circular, and the sixth and seventh are narrow and deep.

5 The sternal osteotomy is created at the level of the posterior angulation of the sternum, generally at the level of the insertion of the third cartilage, but occasionally the second. Two transverse sternal osteotomies are created through the anterior cortex with a Hall air drill 3–5 mm apart, and the wedge of bone is partially mobilized. The base of the sternum and the rectus muscle are elevated with two towel clips, and the posterior plate of the sternum at the osteotomy is fractured. The xiphoid can be divided from the sternum if its anterior angulation produces an unsightly bump below the sternum when it is elevated in its corrected position. The insertion of the rectus muscle into the sternum can generally be preserved by dividing the xiphoid with electrocautery through a lateral approach. Preservation of the attachment of the perichondrial sheaths, and xiphoid to the sternum, avoids an unsightly depression that can occur below it. When a strut is not used, the osteotomy is closed with several heavy silk sutures as the sternum is elevated by the assistant.



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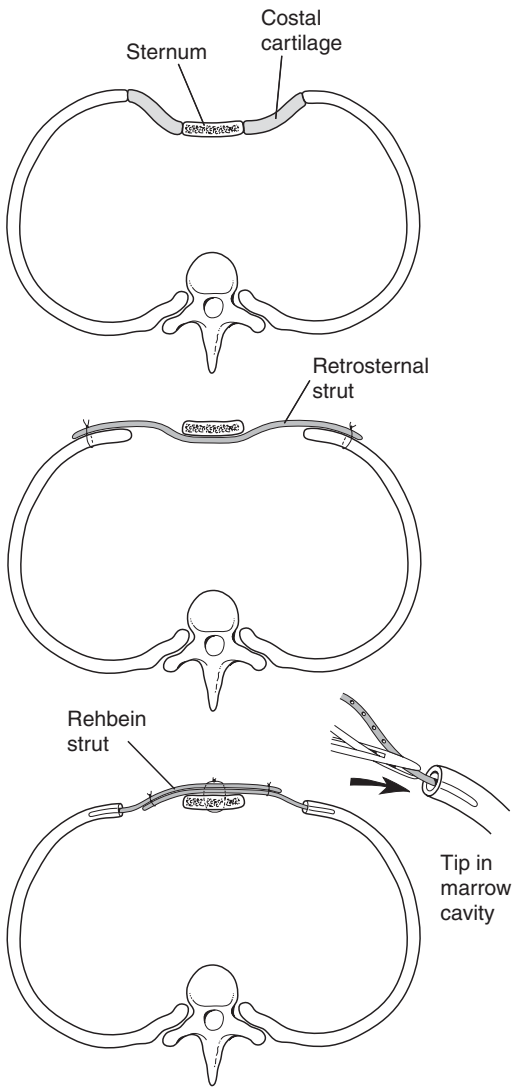

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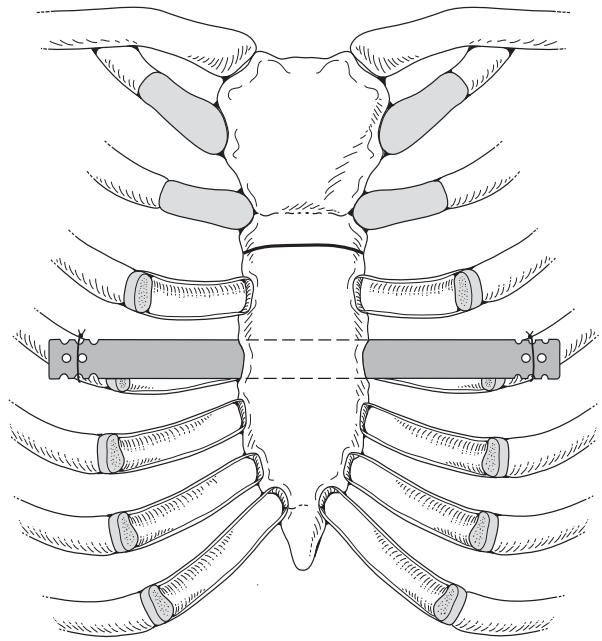
6 This figure demonstrates the use of both retrosternal struts and Rehbein struts. The retrosternal strut is placed behind the sternum and is secured to the rib ends laterally to prevent migration. Rehbein struts are inserted into the marrow cavity (inset) of the third or fourth rib, and the struts are then joined medially with stainless-steel wire to create an arch to secure it in its corrected position.

7 Anterior depiction of the retrosternal strut. The perichondrial sheath to either the third or fourth rib is divided from its junction with the sternum, and the retrosternal space is bluntly dissected to allow passage of the strut behind the sternum. It is secured with two pericostal sutures at each end to prevent migration. The wound is then flooded with warm saline and cefazolin solution to remove clots and inspect for a pleural entry. A single-limb medial Hemovac drain is brought through the inferior skin flap and placed in a parasternal position.



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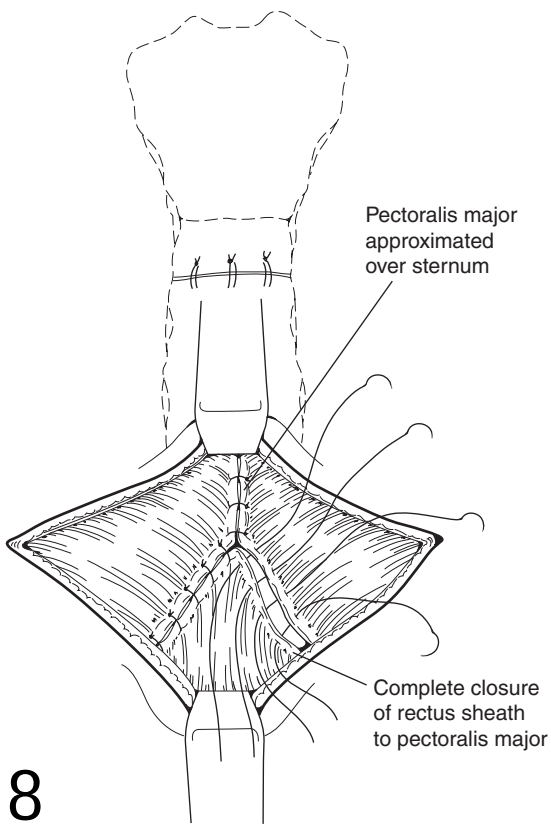

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8 The pectoral muscle flaps are secured to the midline of the sternum, advancing the flaps inferiorly to obtain coverage of the entire sternum. The rectus muscle fascia is then joined to the pectoral muscle flaps, closing the mediastinum.

POSTOPERATIVE CARE

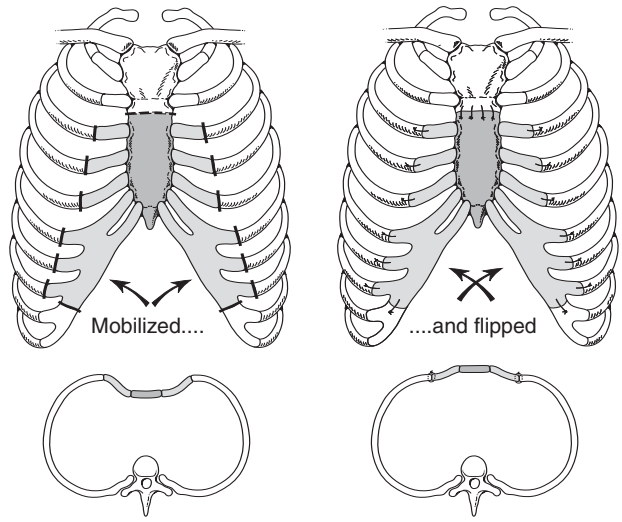
Perioperative antibiotics are utilized by most surgeons. Pain control is managed by either an epidural catheter or intravenous narcotics in the immediate perioperative period. Most surgeons utilize closed drainage early after repair to minimize the risk of hematoma or seroma development under the flaps. Children are encouraged to ambulate early after repair. Contact sports are generally avoided during the first 6 months after repair with either suture or strut elevation of the sternum.

OUTCOME

Generally successful repair of the chest wall deformity is reported. It is critical that children are followed until they reach their full stature, for it is during the period of rapid growth in puberty that recurrences may occur. No randomized studies between sternal fixation with struts and suture fixation have been performed. Both techniques report comparable outcomes. One long-term series from a single institution, however, reports improved outcome in those patients with retrosternal strut fixation.

Sternal turnover

The 'sternal turnover' approach for repair of pectus excavatum was first proposed by Judet and Judet and Jung in the French literature. It has been utilized primarily in Japan, with a large series reported by Wada and colleagues.

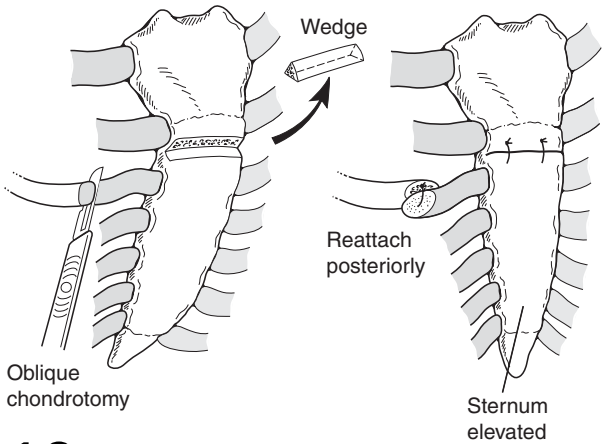


9

This technique involves the use of a free graft of the sternum and costal cartilages, which is divided en bloc and rotated 180° and secured to the costal cartilages and sternum from which it has been divided. This technique can result in major complications, including wound infection, dehiscence, and necrosis of the sternum. Because of these complications, Taguchi proposed a modification that preserved the internal mammary artery in an effort to avoid ischemic osteonecrosis and its sequelae. Other authors have suggested revascularization of the turnover by microvascular reconstruction of the internal mammary arteries. The sternal turnover is generally considered as a radical approach for children with pectus excavatum, given the acceptable alternative methods.

Tripod fixation

Haller developed the technique known as tripod fixation.



10

The subperichondrial resection of the abnormal cartilages is performed followed by a posterior sternal osteotomy. The lowest normal cartilages are then divided obliquely in a posterior/lateral direction. Elevation of the sternum rests the sternal ends of the cartilage on the costal ends, to which they are secured, providing further anterior support of the sternum.

Nuss technique for minimally invasive repair of pectus excavatum (MIRPE)

INDICATIONS

A complete history and physical examination are performed on all patients and include documenting photographs. Patients who have a mild to moderate pectus excavatum are treated with an exercise and posture program in an attempt to halt progression of the deformity. The exercises are designed to strengthen the chest and back muscles and improve exercise tolerance. These patients are followed up at 6–12-month intervals.

Patients who have a severe deformity or who have documented progression are also treated with the exercise and posture program, but in addition they undergo objective studies to see whether their condition is severe enough to warrant surgical correction. These studies include a thoracic computed tomogram (CT scan), pulmonary function tests, and a cardiac evaluation that includes an electrocardiogram (ECG) and an echocardiogram.

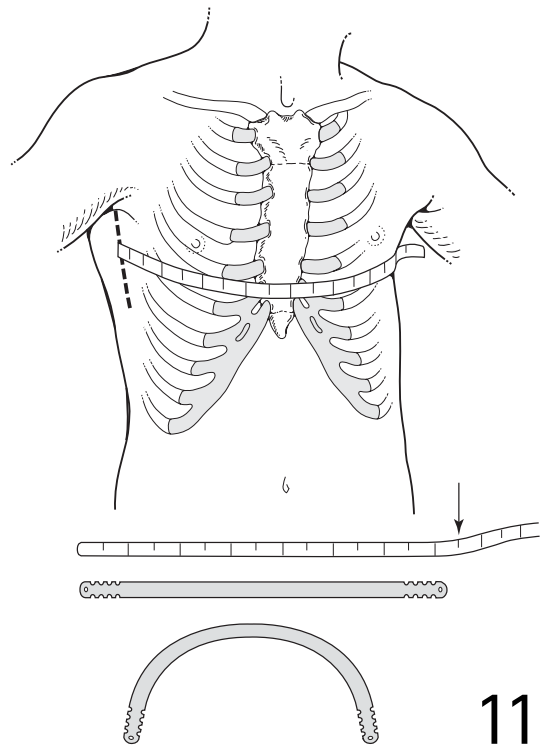
Determination of a severe pectus excavatum and the need for repair include two or more of the following criteria: (1) a Haller CT index greater than 3.25; (2) pulmonary function studies that indicate restrictive and/or obstructive airway disease; (3) a cardiology evaluation demonstrating the compression is causing murmurs, mitral valve prolapse, cardiac displacement, or conduction abnormalities on the echocardiogram or ECG tracings; (4) documentation of progression of the deformity with associated physical symptoms other than isolated concerns of body image. When utilizing these criteria, less than 50 percent of patients are found to have a deformity severe enough to warrant surgery.

Our experience has shown that the optimal age for repair is 7–14 years, as the patients' chests are still soft and malleable, and show quick recovery, a rapid return to normal activities, and have excellent structural results. After puberty, the flexibility of the chest wall is decreased, which often necessitates the insertion of two bars. It also takes the patients longer to recover when they are older. However, we have performed the procedure in patients up to the age of 29 years with equally good long-term results.

TECHNIQUE

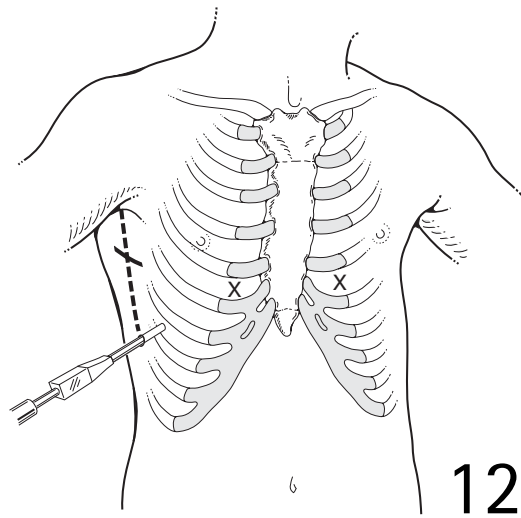
A first-generation cephalosporin is used for antibiotic coverage and continued until discharge. Standard endotracheal anesthesia is used for surgical correction, and epidural analgesia is used as an adjunct during surgery and is continued for pain control for an average of 3 days postoperatively. The arms are abducted at the shoulders and care is taken to pad all pressure points and keep the upper extremities extended without tension on the brachial plexus.

11 The length of the pectus bar is determined by measuring the distance from the right mid-axillary line to the left mid-axillary line and subtracting 2 cm or 1 inch, because the bar takes a shorter course than the tape measure. The measurement is done over the area of the deepest depression that is still part of the sternum. The bar is bent to the desired convex configuration, bearing in mind that the center of the bar should be flat for 2–4 cm to allow greater stability.



11

12 A thoracoscope is inserted into the right chest two intercostal spaces below the planned bar placement to check that the internal anatomy corresponds with the external markings and to look for unexpected pathology. If all is well, the lateral thoracic incisions are made in the region of the mid-axillary line and subcutaneous tunnels are created to the greatest apex of the pectus deformity (X). These Xs represent the entrance and exit sites of the bar from the chest. They are in the intercostal space that is in the same horizontal place as the deepest depression and care should be taken that they are placed medial to the greatest apex of the chest.



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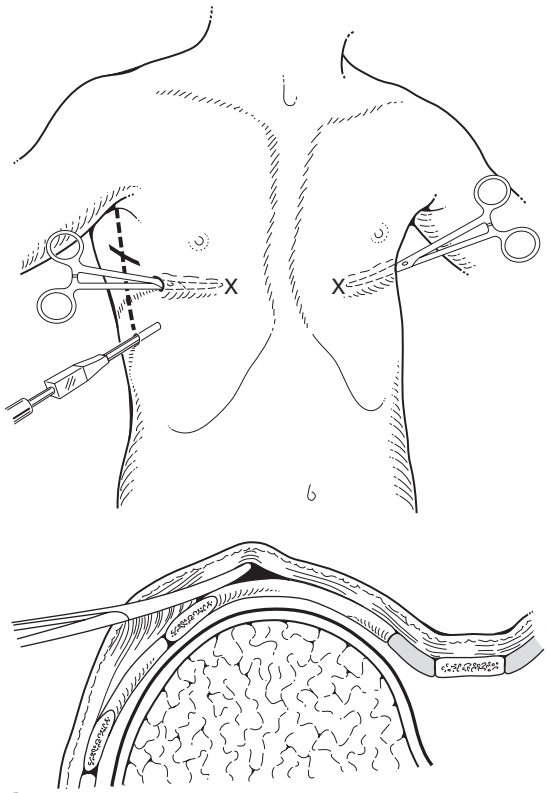
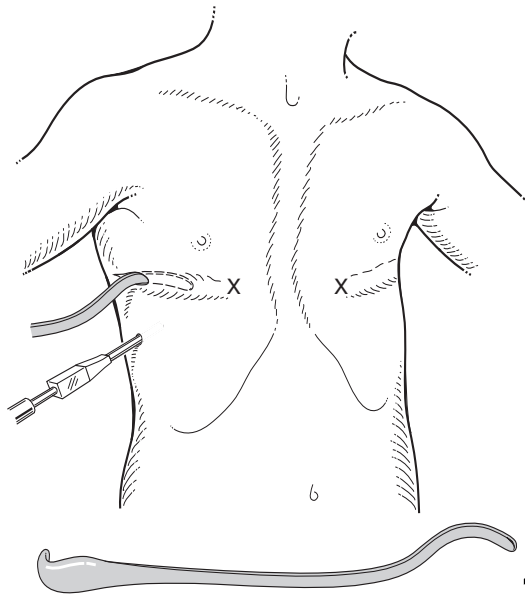


13

13 Skin tunnels are created above the muscle starting from each of the lateral thoracic incisions to the top of the pectus ridge on each side. Subcutaneous tunnels are created so that the entry and exit sites of the bar from inside the chest are medial to the top of the pectus ridge on each side. With the thoracoscope in place, a tonsil clamp is inserted into the subcutaneous tunnel on the right and a blunt thoracostomy is created at the site marked X, taking care not to injure the intercostal vessels, lung, or pericardium.

14 Under continued thoracoscopic visualization, a Lorenz (W. Lorenz Surgical, Jacksonville, FL) introducer is inserted into the chest through the right tunnel and thoracostomy site at the top of the pectus ridge. With great care and thoracoscopic guidance, the pleura and pericardium are dissected off the undersurface of the sternum, creating a substernal tunnel. The introducer is slowly advanced across the mediastinum and brought out through the corresponding intercostal space on the left and advanced out of the incision on the contralateral side. Again, this exit site is medial to the top of the pectus ridge.



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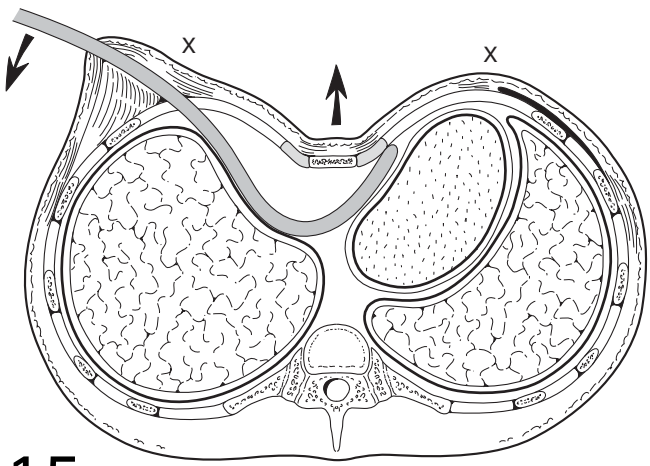
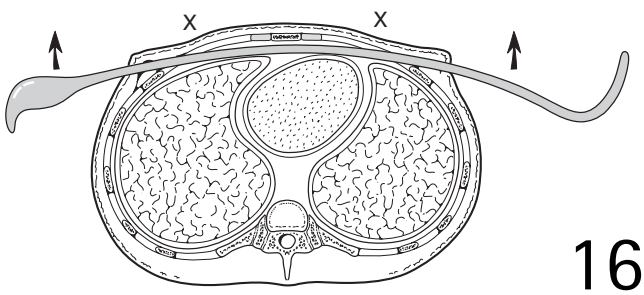


15

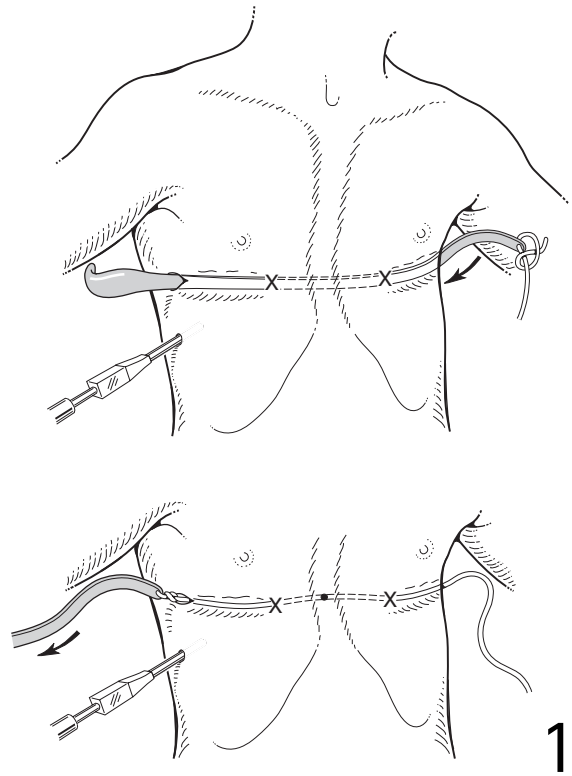
15 A 30° scope facilitates visualization during the substernal dissection, and care is taken to keep the point of the dissector underneath the sternum at all times to push the heart out of the way of the dissection plane. During the dissection, the ECG monitor should be turned to maximum volume to listen for any ectopy or arrhythmias.

16 The introducer is pushed out of the thorax through the previously marked intercostal space 'x' on the left and advanced out through the corresponding tunnel and incision. When the introducer is fully in place, the sternum is elevated by lifting the introducer on each side, thus correcting the pectus excavatum. The sternum is lifted out of its depressed position with the introducer numerous times. This is facilitated by pressing down on the lower chest wall while lifting the introducer.



16

17 Once the sternal depression has been corrected, umbilical tape is attached to the introducer, which is slowly withdrawn from the chest cavity with the umbilical tape attached.



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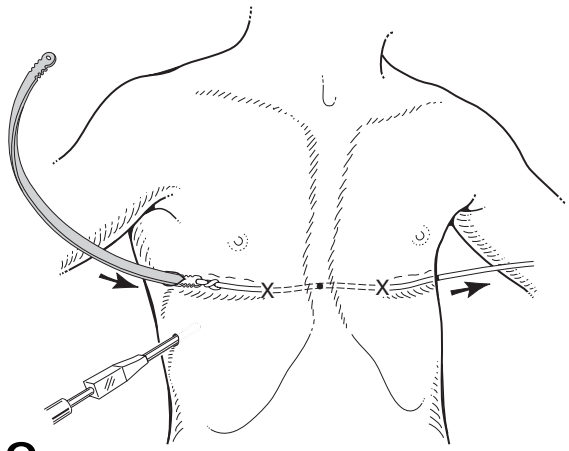
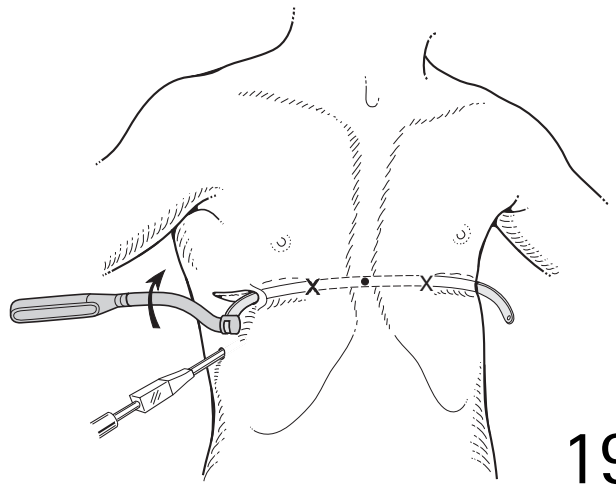


18

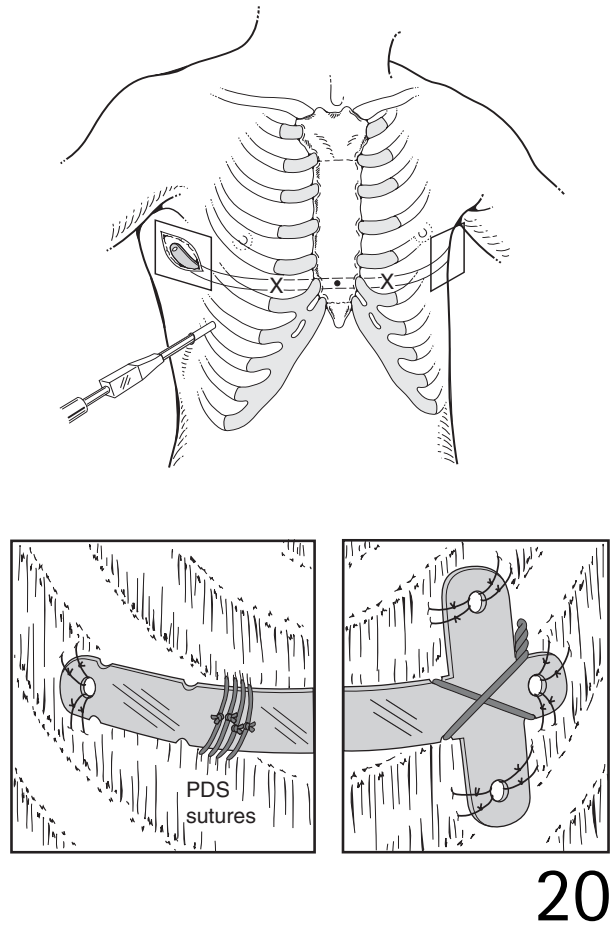
18 The pectus bar that was previously bent into a convex shape is then attached to the umbilical tape and slowly guided through the right subcutaneous tunnel under thoracoscopic visualization and through the substernal tunnel with its convexity facing posteriorly until it emerges on the contralateral side.

19 The pectus bar is positioned inside the chest with its convexity facing posteriorly and an equal amount of bar protruding on each side. Using the specially designed Lorenz bar flipper, the bar is rotated 180° giving instant correction to the pectus deformity. The sides of the bar should be resting comfortably against the musculature and should not be too tight or too loose. If the bar does not fit snugly on each side because of pressure on the middle, it can be re-flipped and molded as necessary while still in place in the chest.



19

20 The bar is stabilized by attaching a stabilizer to its left end and wiring the bar and stabilizer together with No. 3 surgical steel wire. The stabilizer and bar are also secured by placing numerous interrupted absorbable sutures through the holes in the bar and adjacent fascia. An additional stabilizing technique utilizes a laparoscopic 'auto-suture' needle to place multiple '0' PDS or Vicryl sutures around the bar and underlying ribs with thoracoscopic guidance.



20

POSTOPERATIVE CARE

Perioperative antibiotics are continued until discharge on the fourth or fifth day. An epidural catheter is used for 3–4 days and then transitioned to intravenous and oral pain medications. Patients are discharged on a combination of narcotic, anti-inflammatory, and muscle relaxant medications.

For the first 6 weeks, patients are prohibited from playing sports, but are encouraged to do deep breathing exercises and to ambulate. At 6 weeks post-repair they are encouraged to resume normal activities, and at 3 months they may resume competitive sports. Heavy contact sports such as American football, boxing, and ice hockey are prohibited until the strut has been removed, but other aerobic sports are encouraged.

OUTCOME

In 1997 we published our initial 10-year experience with 42 patients utilizing the minimally invasive technique for pectus repair. As of 1 January 2004, we have performed 618 primary procedures and 51 redo procedures – 28 Ravitch recurrences and 23 Nuss recurrences, 3 of which were our own patients. Bars have been removed in 343 patients, with good to excellent long-term results in 92 percent of patients.

There have been no deaths at our institution, with minimal morbidity. The most common early complication is a pneumothorax, which usually resolves spontaneously and rarely requires a chest tube. The infection rate is less than 1 percent. Bar displacement has been decreased to 0.8 percent with the advent of wiring the stabilizers to the bar on the left side and using PDS sutures around the bar and underlying rib on the right side.

Silastic implants

A technique for superficial correction of the depression utilizes implantation of silastic molds into the subcutaneous space to fill the deformity. Although this approach may improve the contour of the chest, it fails to increase the intrathoracic volume, with its potential benefit for both pulmonary and cardiac function.

PECTUS CARINATUM

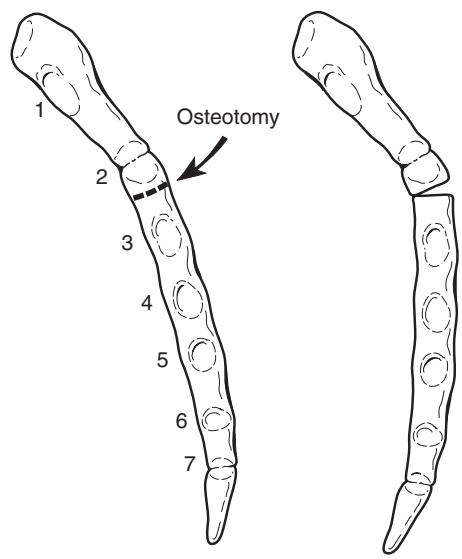
The carinatum deformity is much less common than the depression deformity, with approximately a 1:5 ratio of occurrence. It is more common in boys than in girls (4:1), as

is pectus excavatum. As noted previously, the deformity often appears after the eleventh birthday. In those children in whom a protrusion is noted at birth or in childhood, it often worsens during the period of rapid pubertal growth. The etiology of this deformity is no better understood than is that of pectus excavatum. A family history is identified in 26 percent of patients, suggesting a genetic predisposition. Scoliosis occurs in conjunction with pectus carinatum in 15 percent of children, suggesting a diffuse abnormality in connective tissue development. Consideration for repair of this deformity is based entirely on the severity of the anterior protrusion. Cardiopulmonary abnormalities are rarely identified. Correction of the deformity by bracing has been attempted, but while its efficacy has been demonstrated, compliance with the compression regimen is low.

OPERATION

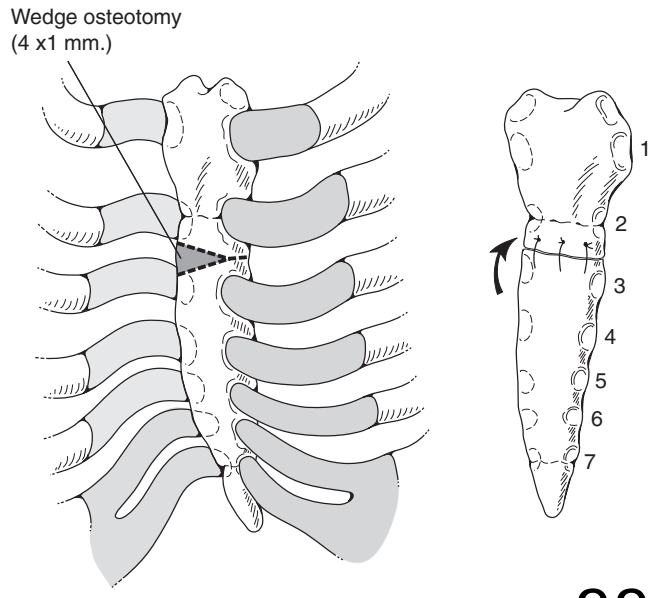
Repair of pectus carinatum has had a colorful past. Early attempts at repair included such maneuvers as resection of the anterior table of the sternum and removal of the distal half of the sternum, with re-attachment of the rectus muscle higher up on the body of the sternum. Current techniques stress the need for preservation of the perichondrial sheath with sternal osteotomies tailored to the specific deformity of the sternum. Pectus carinatum presents in three primary configurations. The most common is protrusion of the body of the sternum (gladiolous) and costal cartilages and is termed the chondrogladiolar protrusion. This protrusion can be asymmetric, producing a keel-like protrusion along one side of the sternum. As mentioned previously, some children can have a 'mixed' deformity with ipsilateral carinatum and contralateral depression components. The sternum is often rotated to one side in these cases. The rarest configuration is the chondromanubrial protrusion, also referred to as a 'pouter pigeon breast'. In this deformity, the manubrium and the first and second costal cartilages protrude and there is relative depression of the body of the sternum.

The exposure of the sternum and costal cartilages is identical to that of the open repair for pectus excavatum. The protruding costal cartilages are resected with attention to maximum preservation of the perichondrial sheaths.



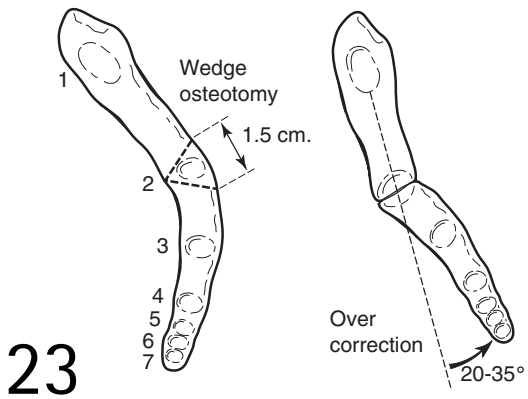
21 A single or double transverse osteotomy with fracture of the posterior plate after resection of the costal cartilages allows posterior displacement of the sternum to an orthotopic position.

22 The mixed pectus deformity is corrected by full and symmetric resection of the involved costal cartilages, followed by transverse offset (0–10° wedge-shaped sternal osteotomy). Closure of this defect achieves both anterior displacement and rotation of the sternum.



Mixed deformity (anterior view) 9.2%

22



23

23 In the chondromanubrial deformity, both the protrusion of the manubrium and the depression of the body of the sternum must be corrected. A broad, wedge-shaped sternal osteotomy is placed through the anterior cortex of the obliterated sternomanubrial junction. Closure of the osteotomy after fracture of the posterior cortex achieves posterior displacement of the superior portion of the sternum, which is attached only at its junction to the first rib. The lower portion of the sternum is overcorrected 20–35° and is secured in position by strut or suture fixation correcting both components of the abnormality.

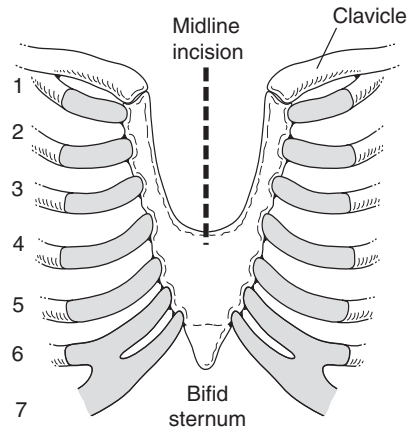
OUTCOME

Generally excellent results are achieved with repair of pectus carinatum. Repair at a young age may result in development of a protrusion of cartilages that have not been resected or, in cases of a unilateral abnormality, contralateral protrusion may occur. For this reason, correction of pectus carinatum is generally deferred until children have completed the majority of their pubertal growth.

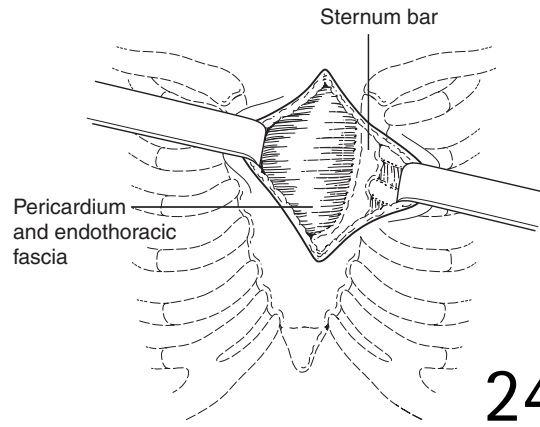
STERNAL CLEFT

The sternal cleft deformity is the least severe of the sternal abnormalities, which also include thoracic and thoracoabdominal ectopia cordis. A sternal cleft results from incomplete fusion of the sternal bars in the fetus. The heart is in an orthotopic position. While intrinsic cardiac anomalies are common in thoracoabdominal and thoracic ectopia cordis, they are rare in infants with a sternal cleft. Repair of the sternal cleft relieves the paradoxic motion often seen in this anomaly. The sternal cleft may be complete, but in most cases the inferior base of the sternum is fused. Repair is best achieved in the newborn period or within the first few months of life when the chest wall is most flexible and primary repair is tolerated by the infant.

OPERATION

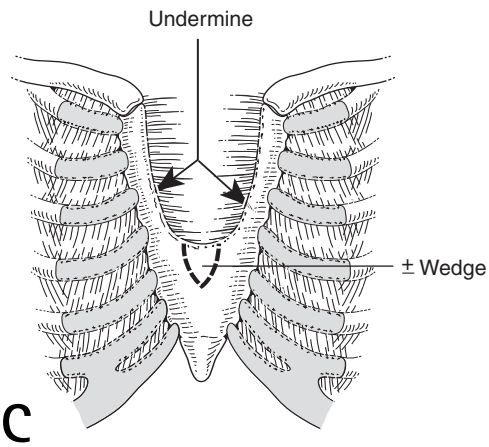


24a

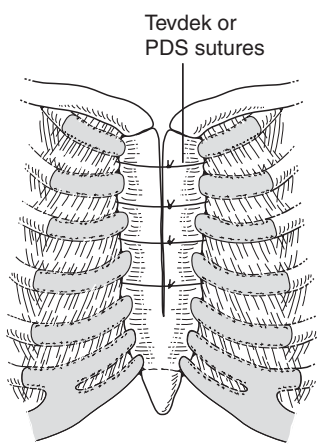


24b

24a,b (A) Repair of bifid sternum is best performed through a longitudinal incision extending the length of the defect. (B) The sternal bars are encountered directly beneath the subcutaneous tissues. The pectoral muscles insert lateral to the bars.



24c



24d

24c,d (C) The endothoracic fascia is mobilized off the sternal bars posteriorly with blunt dissection to allow safe placement of the sutures. In many cases, excision of a wedge from the most inferior portion of the defect will facilitate approximation of the two sternal halves during suture closure. (D) Closure of the defect is achieved with 2-0 Tevdek or PDS sutures.

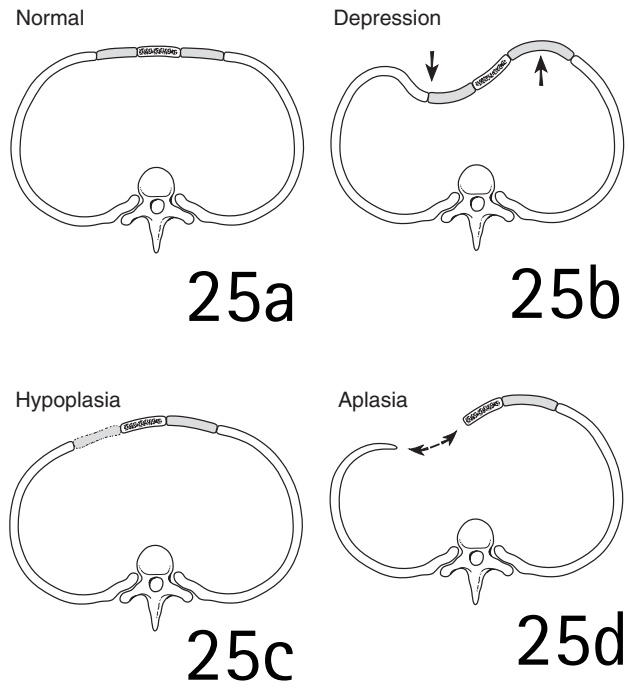
More complex repairs are required in older children, including bilateral oblique incisions through the costal cartilages to increase their length and allow midline approximation of the sternal halves or division of the cartilages to allow them to be swung medially to cover the defect.

OUTCOME

Repair at an early age is most satisfactory. Closure with cartilage or rib grafts often leads to instability of the chest with progressive growth.

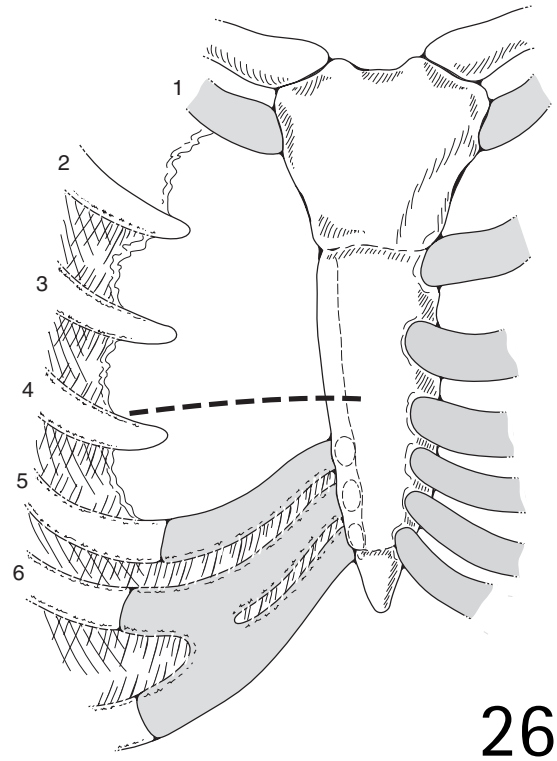
POLAND'S SYNDROME

Poland's syndrome is defined as a constellation of anomalies including absence of the pectoralis minor muscle, absence of the costal portion of the pectoralis major muscle, hypoplasia of the breast and nipple or complete absence of the breast and nipple, and brachysyndactyly of the digits. A variable deformity of the chest wall occurs, ranging from hypoplasia to a severe excavatum deformity to aplasia of several ribs.



25a–d These figures depict the spectrum of thoracic abnormality seen in Poland's syndrome. (A) Usually, an entirely normal thorax is present, and only pectoral muscles are absent. (B) Depression of the involved side of the chest wall, with rotation and often depression of the sternum. A carinatum protrusion of the contralateral side is frequently present. (C) Hypoplasia of ribs on the involved side but without significant depression may be seen. It does not usually require surgical correction. (D) Aplasia of one or more ribs is usually associated with depression of adjacent ribs on the involved side and rotation of the sternum.

This deformity has its gravest implication in females, in whom significant abnormality of the breast is seen. In females, repair of the underlying chest wall depression facilitates correction of the absence or hypoplasia of the breast with a prosthetic implant. Often simultaneous rotation of the latissimus dorsi muscle is performed to create a more natural texture to the breast. In males, repair of the aplastic segment is undertaken in adolescence to avoid impairment of growth resulting from surgery at an early age.

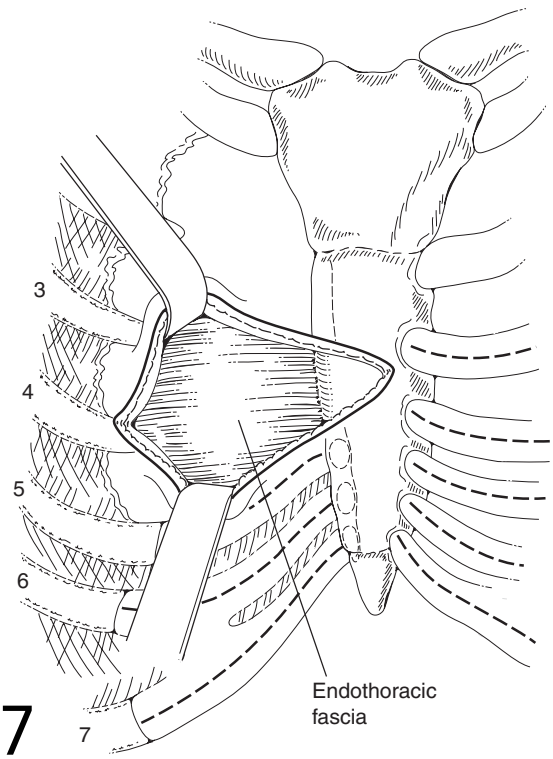


26

26 Schematic depiction of a severe deformity with rotation of the sternum and aplasia of three ribs. Ironically, the contralateral side of the chest may have a carinatum protrusion, which accentuates the depression on the ipsilateral side. The rotation of the latissimus dorsi muscle that is occasionally utilized in males has the potential drawback of decreasing the strength of the shoulder.

OPERATION

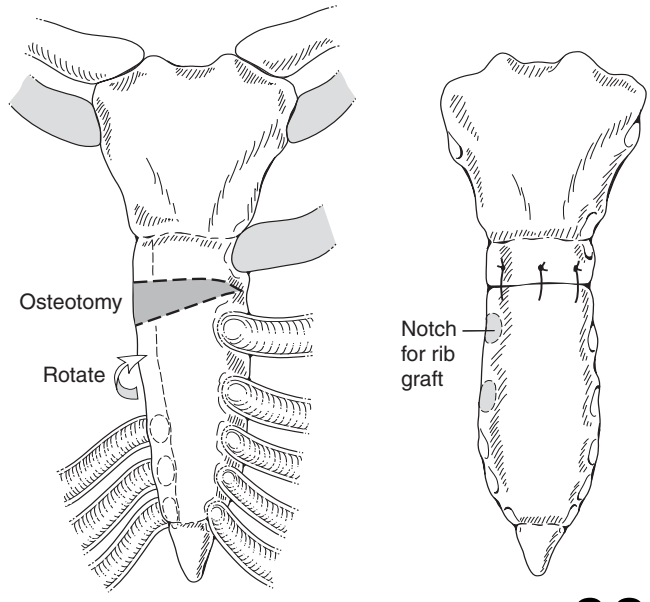
The incision for repair is similar to that in pectus excavatum and carinatum. A transverse incision is placed below the nipple lines and, in females, in the inframammary crease.



27

27 In patients with aplasia of the ribs, the endothoracic fascia is encountered directly below the attenuated subcutaneous tissue and pectoral fascia. The pectoral muscle flap is elevated on the contralateral side and the pectoral fascia, if present, on the involved side. Subperichondrial resection of the costal cartilages is then carried out, as shown by the bold dashed lines. Rarely, this must be carried to the level of the second costal cartilages.

28 A transverse, offset, wedge-shaped sternal osteotomy is created below the second costal cartilage. Closure of this defect with heavy silk sutures or elevation with a retrosternal strut corrects both the posterior displacement and the rotation of the sternum.



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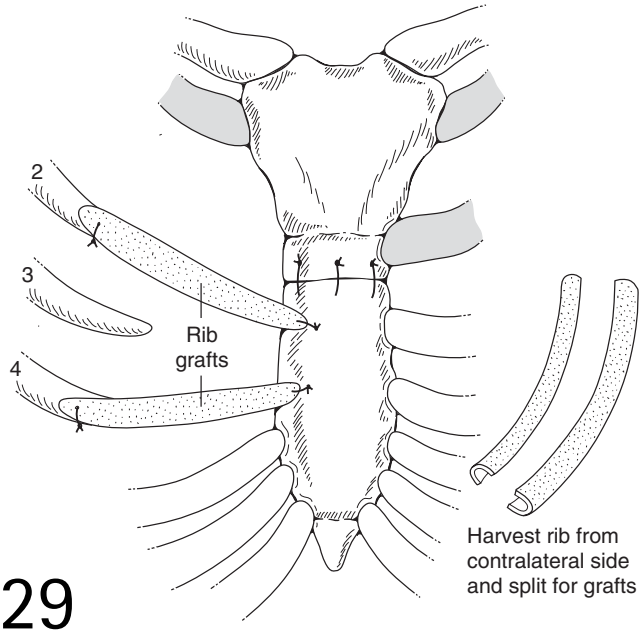

29

29 In patients with rib aplasia, split rib grafts are harvested from the contralateral fifth or sixth rib and then secured medially with wire sutures into previously created sternal notches and with wire to the native ribs laterally. Ribs are split as shown along their short axis to maintain maximum mechanical strength.

OUTCOME

A broad spectrum of chest wall abnormalities is seen in Poland's syndrome, and surgical intervention must be appropriately tailored to the severity of the deformity. Hypoplasia of the chest without a localized posterior depression does not require repair. A significant posterior depression, particularly in females, warrants repair in adolescence.

Acknowledgments

Figures 1–5 and 8 reproduced with permission from Shamberger RC, Welch KJ. Surgical repair of pectus excavatum. *J Pediatr Surg* 1988; **23**:615.

Figure 4 adapted from an original figure.

Figures 6 and 7 from Shamberger RC. Chest wall deformities. In: Shields TW (ed.), *General Thoracic Surgery*,

5th edition. With permission from Lippincott Williams & Wilkins.

Figures 9 and 10 from Shamberger RC. Depression deformities: pectus excavatum. In: *Pediatric Surgery and Urology: Long-term Outcomes*. With permission from WB Saunders Co. Ltd.

Figures 21 and 22 reproduced with permission from Shamberger RC, Welch KJ. Surgical correction of pectus carinatum. *J Pediatr Surg* 1987; **22**:228; 48.

Figure 23 reproduced with permission from Shamberger RC, Welch KJ. Surgical correction of chondromanubrial deformity. *J Pediatr Surg* 1988; **23**:319.

Figure 24 reproduced with permission from Shamberger RC, Welch KJ. Sternal defects. *Pediatr Surg Int* 1990; **5**:156.

Figures 25 and 29 reproduced with permission from Shamberger RC, Welch KJ, Upton J III. Surgical treatment of thoracic deformity in Poland's syndrome. *J Pediatr Surg* 1989; **24**:760.

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27

Patent ductus arteriosus

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PRINCIPLES AND JUSTIFICATION

The ductus arteriosus of many preterm infants remains patent at birth. Patency approaches 90 percent under 28 weeks of gestation, 75 percent at 28–31 weeks, 45 percent at 31–33 weeks, and 21 percent at 34–36 weeks. When correlated with weight, there is 83 percent patency under 1000 g; this falls to 47 percent in those weighing 1000–1500 g at birth and 27 percent in neonates weighing more than 1500 g. While spontaneous closure is directly related to arterial oxygen tension, there are many additional factors that affect functional and anatomic closure, and re-opening, in both the premature and full-term infant.

Indications

Approximately 70 percent of infants delivered before 28 weeks of gestation will need the ductus closed, medically or surgically. A patent ductus arteriosus results in increased blood flow to the lungs, exposing them to systemic blood pressures. The left-to-right shunt decreases organ perfusion, which correlates with such clinical problems as necrotizing enterocolitis and lower glomerular filtration rates.

Clinical findings are of a left-to-right shunt include collapsing pulses (widened pulse pressure), cardiomegaly, and a continuous murmur. Practically, this produces a continuing dependence on mechanical ventilation and an increasingly or persistently high oxygen requirement. Traditional therapy includes restricting intravenous fluids, administering digitalis, diuretics, and dopamine, and adding positive end-expiratory pressure. All of these may influence the degree of shunting but do not directly promote closure.

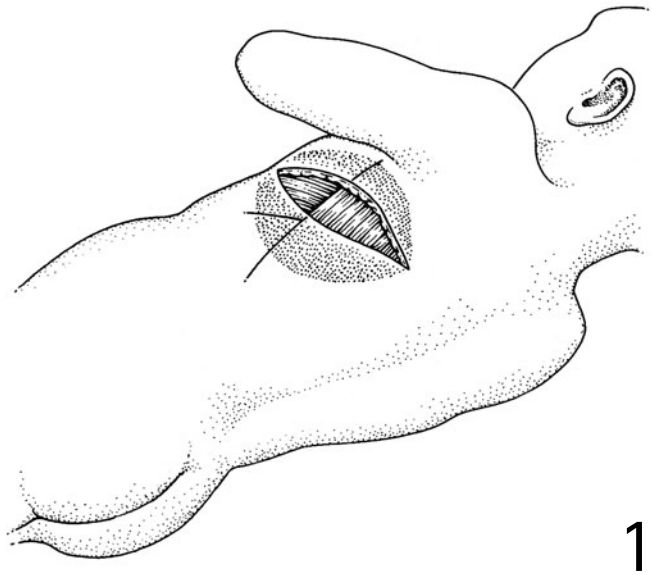
Diagnosis is confirmed by two-dimensional echocardiography, which is invaluable in assessing both patency and pressures. The hemodynamic significance of a patent ductus arteriosus, rather than its patency, determines the course of treatment. The decision about when to close a patent ductus arteriosus by medical or surgical means remains a clinical one.

NON-SURGICAL CLOSURE

The treatment of choice for non-surgical closure of a patent ductus arteriosus in most neonatal intensive care units is indomethacin. Two or three courses are usually required to promote closure, and further administration beyond this rarely improves the success rate. Indomethacin tends to be less effective as the infant grows older. Contraindications to giving indomethacin include sepsis, necrotizing enterocolitis, azotemia, and prolonged coagulation. When these conditions exist, surgical intervention is necessary.

OPERATION

1 The infant is placed in the lateral decubitus position with the right side down. The surgeon stands on the right side, facing the infant's abdomen. A headlight provides valuable illumination of the small operative field. A lateral incision is made and the subcutaneous layer is mobilized from the underlying muscles. The flimsy attachment of the anterior margin of the latissimus dorsi, which overlies the inferior edge of the serratus anterior, is incised. The latissimus is elevated and retracted posteriorly, and the serratus is freed and retracted anteriorly. It is unnecessary to divide any muscle to achieve exposure.



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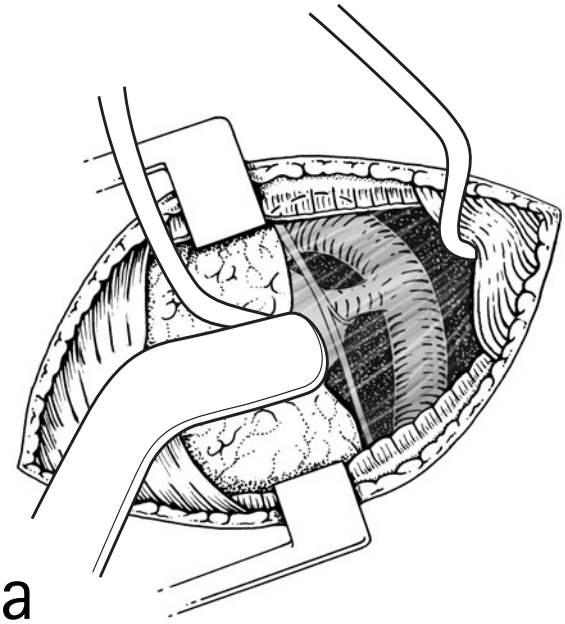

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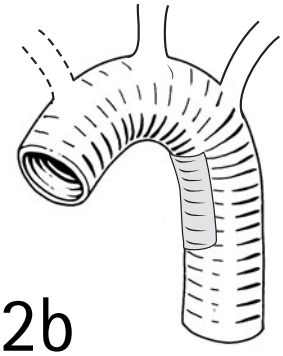

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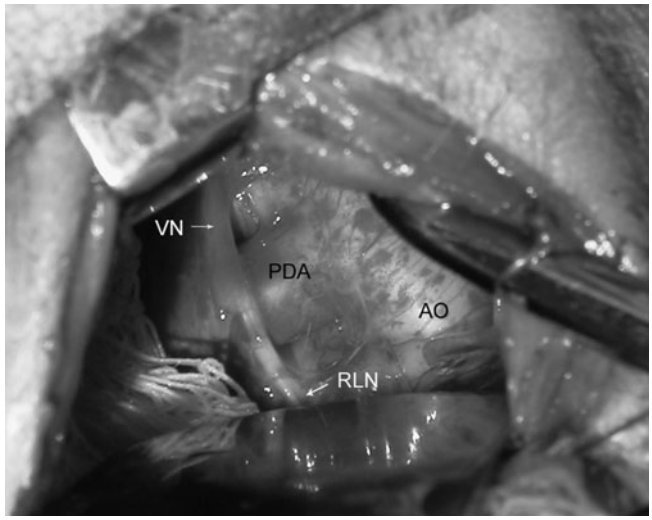
2 The third or fourth intercostal space is entered and a small self-retaining retractor (Finochetto) is placed. A second retractor (Gelpi) retracts the muscles in an anterior–posterior direction. The ductus arteriosus may be as large as, or larger than, the aorta. Its exact position is variable; as it arises from the ventral side of the aorta, it may be located just inferior to or more distal from the aortic arch (inset). When mediastinal edema exists and in the tiniest babies, the usual landmarks can be obscure. Identification of the vagus nerve and its recurrent branch always leads to the ductus arteriosus (photo). (VN, vagus nerve; PDA, patent ductus arteriosus; AO, aorta; RLN, recurrent laryngeal nerve.)

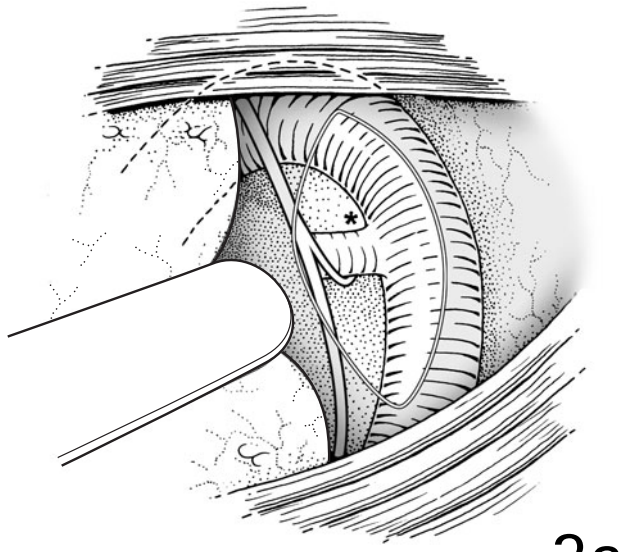


2c

3 The mediastinal pleura overlying the aorta is opened; a large superior intercostal vein often overlies the ductus, and if present must be divided. A malleable retractor over a moist sponge holds the lung anteriorly and inferiorly for exposure, but in unstable patients it is often necessary to allow intermittent lung expansion during surgery. The vagus nerve and recurrent branch are visible and are reflected anteriorly with the pleura.

Adequate exposure of the ductus is achieved by incising and spreading the tissue just above and below the ductus. This delicate areolar tissue can be grasped with forceps, but care should be taken to avoid direct handling of the ductus, which is extremely friable. The use of tenotomy scissors enhances the accuracy of this dissection. The superior angle between the aorta and the ductus arteriosus (*) is particularly vulnerable, and dissection here must be done with great care.

It is not necessary completely to dissect and encircle the ductus unless it is too large for a metallic clip and requires ligation. The dissection is complete once most of the circumference can be visualized; the posterior shaded area (inset) remains undissected.



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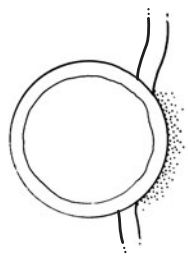

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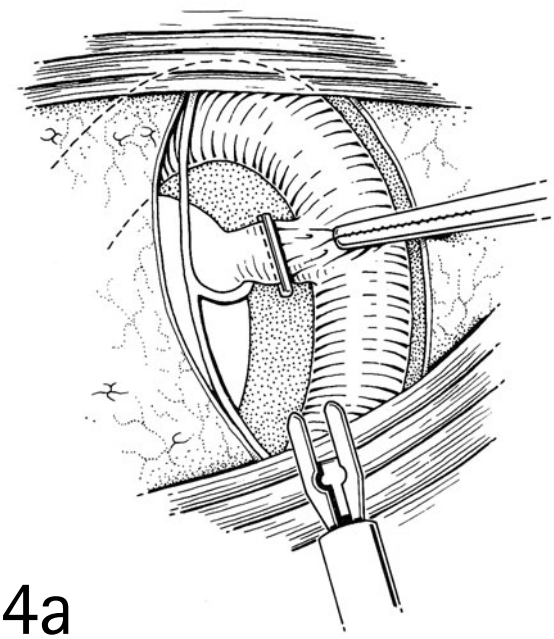

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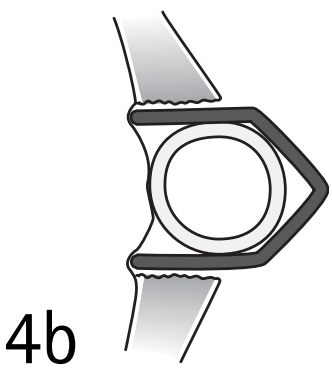


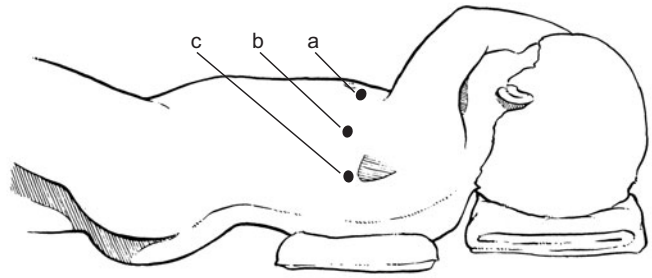
4b

4 The adventitia of the aorta can be grasped and pulled away from the surgeon when necessary to provide enough length to place the clip. The tip of the metallic clip must extend beyond the margin of the ductal wall to ensure complete occlusion. The appropriate-sized metallic clip is placed closer to the aortic end of the ductus arteriosus (but not flush against the aorta). When ligation is necessary, a heavy ligature is safest, as it decreases the likelihood of cutting through the wall. It should be passed around the ductus arteriosus using a right-angled clamp.

Several absorbable pericostal sutures are placed to appose the ribs and close the intercostal space gently. Since a muscle-sparing incision is used, no muscular sutures are needed. The lung is gently re-expanded with the ventilator and, although rarely needed, a small rubber catheter or thoracostomy may be placed into the pleural cavity and gentle suction applied. The catheter may be removed once the pericostal sutures have been tied, unless there is an obvious air leak. The intercostal muscle is not sutured, and a very fine skin closure is performed.

Thoracoscopic ductal ligation

5 Thoracoscopic patent ductus arteriosus ligation is an alternative to open thoracotomy. It offers the theoretical advantages of better chest wall compliance, decreased risk of scoliosis, and cosmetic improvement. In this approach, three small skin sites in the fourth intercostal space are used. A 3 mm port can be placed in the mid-axillary line for the scope. An additional site anterior to this is used for lung retraction with a cotton-tip applicator and a posterior 5 mm site is used to introduce a dissector and then the clip applier. Lung compliance and reduced visualization due to bleeding are the main reasons for conversion to an open approach.



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POSTOPERATIVE CARE

In many centers, the operation is performed in the neonatal intensive care unit, as transportation of these tiny, unstable infants can be challenging. The operation should be completed in less than 30 minutes. After surgery, lung re-expansion is confirmed by chest radiography. The infant should recover quickly from the short-acting intravenous agents. The hemodynamic response of interrupting a large left-to-right shunt is immediate, but the gratifying increase in blood pressure may require prompt treatment to avoid central nervous system hemorrhage.

COMPLICATIONS

Postoperative complications are uncommon, but chylothorax, transient vocal cord paralysis, and incomplete occlusion have been reported, in addition to the usual sequelae of any thoracic surgical operation. If the ductus is torn, or there is inadvertent ligation of the left pulmonary artery or aorta, the baby is unlikely to survive.

OUTCOME

Improvement in cardiopulmonary function starts immediately, depending on the volume of flow through the left-to-right shunt and the preoperative status of the heart and lungs. Many studies have tried to compare the results of indomethacin versus surgical ligation but fail to be convincing because the patient groups are different, and many babies undergo surgery only because medical therapy has failed. Because the method of ductal closure appears to have little influence on overall survival, accurate comparison of these modalities is difficult.

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28

Thoracoscopic sympathectomy

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BACKGROUND

Hyperhidrosis is a condition of unknown etiology whereby excess sweating occurs from the eccrine glands in the palms, axillae, and soles of the feet. Excess axillary sweating does not occur in prepubertal children.

Onset may be as early as infancy, and a family history of the condition is not unusual.

Severe palmar sweating may render schoolwork almost impossible. As a general rule, the condition worsens during adolescence.

The only permanent cure is by interruption of the sympathetic supply to the eccrine glands by sympathectomy. At the present time, plantar hyperhidrosis is not treated, and treatment is confined to the management of palmar and axillary sweating.

Topical medication is ineffective in those who are severely affected. Injection of botulinum toxin provides temporary relief in axillary hyperhidrosis.

Endoscopic thoracic sympathectomy was first performed in 1942 and has since become the standard treatment for the condition.

OPERATION

Position of patient

The patient lies supine on the operating table with the thorax elevated about 20°. Both arms are abducted to 90°.

Single-tube endotracheal anesthesia is employed. The aim is to deal with both sides at one operation.

We have employed a two-port technique, using one port for the camera and the second for the diathermy probe.

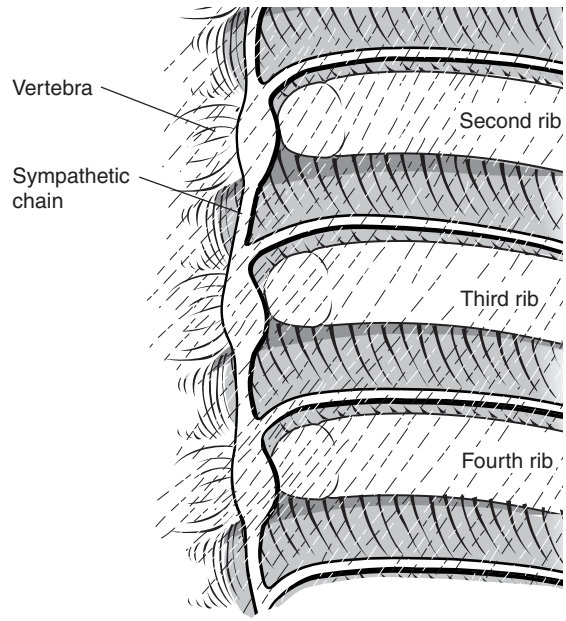
Incision

Two incisions are made, both in the mid-axillary line. The initial incision is 1 cm in length in about the sixth or seventh interspace. Local anesthesia is employed initially, followed by blunt dissection with an artery forceps until the pleura is entered. Subsequently, a Veress needle is utilized to achieve a pneumothorax. Pressure greater than 10 mmHg is unnecessary.

A blunt Hasson trocar and cannula is inserted and subsequently a 5 mm telescope (0° or 30°).

Procedure

1 Once lung collapse had been achieved, a stab incision is made about three intercostal spaces above the initial incision. A 3 mm or 5 mm diathermy probe is then introduced under vision. The sympathetic trunk is identified abutting the heads of the ribs. The first rib is not visible as it is obscured by Sibson's fascia. The most proximal rib seen is the second rib.



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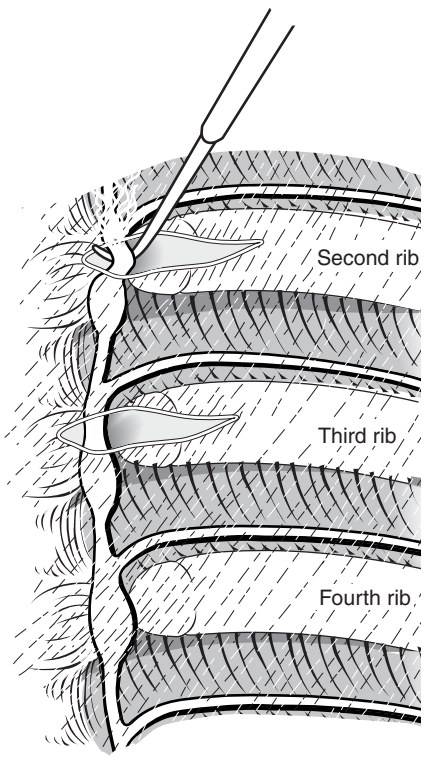

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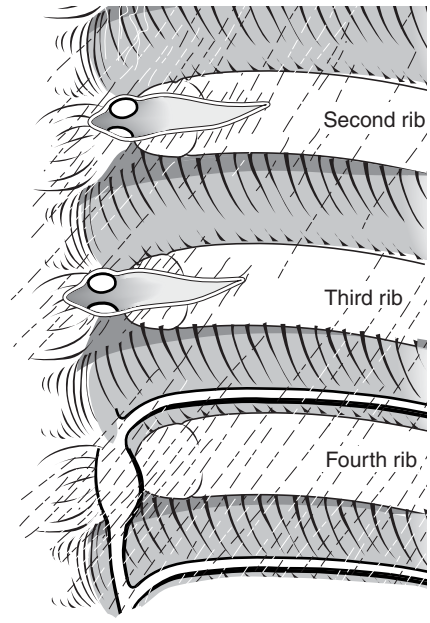
2 The sympathetic trunk is divided over the second and third ribs, incising all tissue down to bone. For those with axillary hyperhidrosis, the trunk is divided over the fourth rib as well. The length of the incision is about 2 cm in each case.

3 Once the sympathectomy is complete, the lung is inflated under vision and the procedure is repeated on the contralateral side. Chest drains are not used routinely. Skin closure is achieved with tissue glue.



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SIDE EFFECTS

Horner's syndrome has been reported and occurs in perhaps 1–2 percent of cases.

Compensatory trunk sweating is very common in adult practice (> 80 percent), but seems much less problematic in prepubertal children. Occasionally this symptom is disabling in adults.

No adverse long-term consequences have been reported in children who have undergone this procedure.

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SECTION IV

Abdominal

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29

Hernias in children

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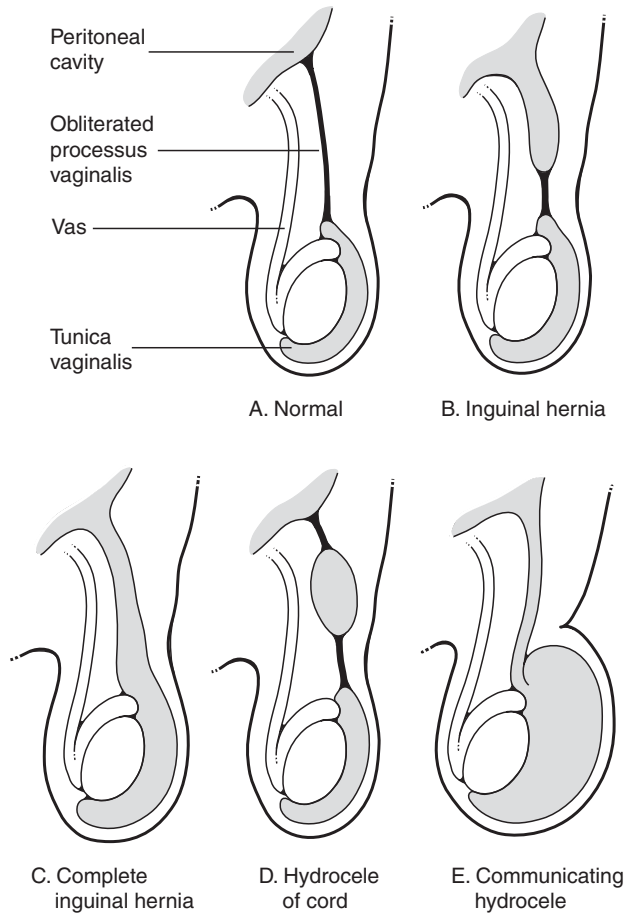
INGUINAL HERNIA AND HYDROCELE

HISTORY

The first reference to hernia repair in children is credited to Celsus, who, in AD25, recommended removal of the hernia sac and testes through a scrotal incision. Pare recommended treatment of childhood hernia; however, the first accurate description was made by Pott in 1756. Czerny performed high ligation of the hernia sac through the external ring in 1877. Ferguson recommended that the spermatic cord should remain undisturbed during inguinal hernia repair in 1899. In 1912, Turner documented that high ligation of the sac was the only procedure necessary in most children. Herzfield was the first advocate of outpatient surgical repair of inguinal hernia in children in 1938. Early repair in infancy was recommended by Ladd and Gross in 1941. The concept of bilateral inguinal exploration was promoted by Duckett, Rothenberg, and Barnett, among others. Advances in neonatal intensive care have resulted in improved survival of premature infants, who have a high incidence of hernia and an increased risk of complications. These cases have stimulated great interest in considerations regarding the timing of operation and choice of anesthesia. With the advent of laparoscopy, many have challenged the necessity of routine bilateral inguinal exploration.

PRINCIPLES AND JUSTIFICATION

The occurrence of congenital inguinal hernia is related to descent of the testis, which follows the gubernaculum testis as it descends from an intra-abdominal retroperitoneal position to the scrotum. Those factors affecting descent (androgenic hormonal influences for the abdominal descent phase, and local hormonal release from the genitofemoral nerve for the scrotal descent phase) are beyond the scope of this chapter. However, as the testis passes through the internal ring it drags with it a diverticulum of peritoneum on its anteromedial surface referred to as the processus vaginalis. In girls, the persistent processus vaginalis extends into the labia majora in the canal of Nuck. The layers of the processus vaginalis normally fuse in more than 90 percent of full-term infants, obliterating the entrance to the inguinal canal from the peritoneal cavity. Failure of obliteration may result in a variety of inguinal–scrotal anomalies, including complete persistence resulting in a scrotal hernia, distal processus obliteration and proximal hernial patency, complete patency with a narrow opening at the internal ring referred to as a communicating hydrocele, hydrocele of the canal of Nuck in girls or inguinal canal in boys, and a hydrocele of the tunica vaginalis.



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Clinical presentation

The majority of inguinal hernias in infants and children are indirect hernias. Boys are more commonly affected than girls, in a ratio of 9:1; 60 percent present on the right side due to later testicular descent and obliteration of the processus vaginalis on the right, 25 percent occur on the left side, and 15 percent are bilateral. The diagnosis is often made on the basis of a bulge that is apparent in the groin with crying or straining. Scrotal enlargement and frequent change in scrotal size resulting from transfer of fluid (or bowel) between the peritoneal cavity and the sac may be noted. Physical examination will often confirm these observations; however, diagnosis may depend on visualization of these events by the referring pediatrician or parent.

Inguinal hernia is a high-risk hernia, particularly in early infancy, as it is frequently complicated by incarceration, occasionally leading to intestinal obstruction and strangulation. In young infants with undescended testes and associated hernia, the testis is sometimes at risk of torsion or atrophy caused by compression of the vascular supply by a hernia sac filled with bowel compressing the testicular vessels at the level of the internal inguinal ring. The incidence of incarceration is highest in the youngest patients, particularly premature infants and infants under the age of 1 year, in whom an incarceration rate of 31 percent has been reported. The incarceration rate in children up to 18 years of age is 12–15 percent.

Indications

Because of the high rate of complications associated with inguinal hernia, there is no place for conservative management except in instances of an isolated hydrocele of the tunica vaginalis. The natural history of this particular abnormality is spontaneous involution at 6–12 months of age. As long as the hydrocele does not change in size, it can be observed. All other inguinal scrotal anomalies require surgical intervention. In addition to instances of incarceration seen in boys, girls can present with a mass in the labia majora due to a sliding hernia of the ovary and fallopian tube. This may be associated with a risk of torsion of the ovary in the hernia sac.

PREOPERATIVE ASSESSMENT AND PREPARATION

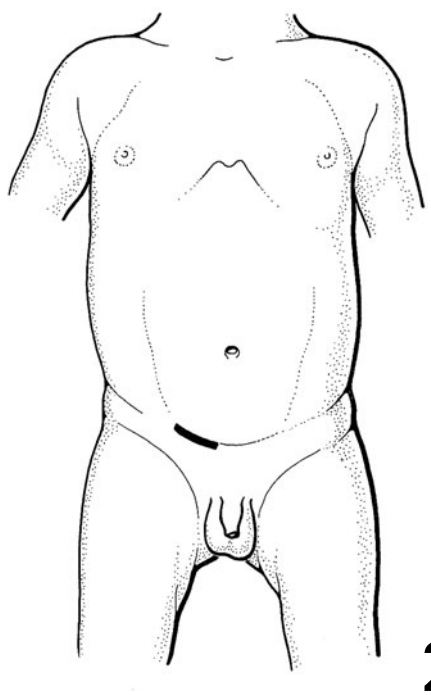
The operation is usually performed shortly after the diagnosis is made. Attempts to reduce an incarcerated hernia using sedation and manual reduction are successful in more than 80 percent of cases. An elective operation is then typically carried out within 24 hours of the reduction as the recurrence rate for incarceration is noted to be as high as 15 percent if repair is delayed beyond 5 days. In the case of symptomatic hernias in small premature infants already hospitalized in the neonatal intensive care unit because of other illnesses, elective repair is carried out just before discharge and/or at a weight greater than 2.0 kg. For infants diagnosed after discharge from the hospital who require ventilatory support or experience episodes of apnea and/or bradycardia in the neonatal period, elective repair is usually delayed until 50 weeks of corrected conceptual age. Although most infants and children can be managed in an ambulatory setting, infants with bronchopulmonary dysplasia, anemia, or who required ventilator support at the time of birth should be observed after operative repair in an extended observation (23 hours) center and monitored for episodes of apnea and/or bradycardia.

ANESTHESIA

The operation is usually performed under general anesthesia, although some surgeons prefer spinal anesthesia in very premature infants. The lower abdomen, inguinal scrotal area, perineum, and thighs are prepared with a sterilizing solution of choice and then draped appropriately for herniotomy.

OPERATION: OPEN APPROACH

2 A transverse incision is made in the lowest inguinal skin crease above the external inguinal ring of the affected side. One must be aware of the superficial epigastric vein to avoid bleeding and subsequent ecchymoses in the superficial wound in the postoperative period. Scarpa's fascia is incised and the external oblique fascia identified. The external oblique fascia is exposed and traced laterally to the inguinal ligament. The inguinal ligament is traced inferiorly to expose the external inguinal ring. This minimizes the risk of opening the inguinal canal too medially.



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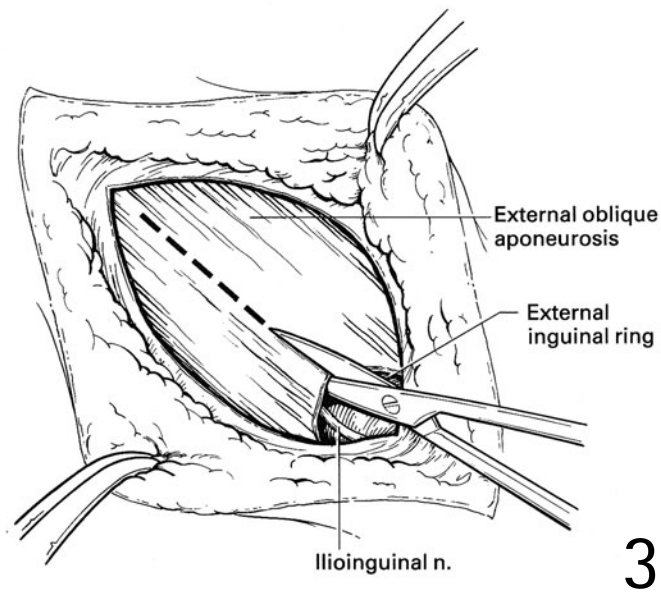
3 Once the superficial inguinal ring has been identified, the external oblique fascia is opened superiorly in the long axis of its fibers, perpendicular to the ring, for a distance of 1–2 cm (near the deep inguinal ring). Care is taken during this maneuver to minimize accidental injury to the ilioinguinal nerve.



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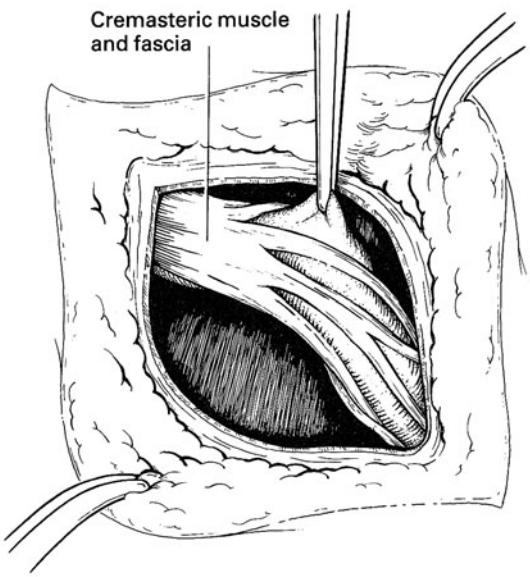

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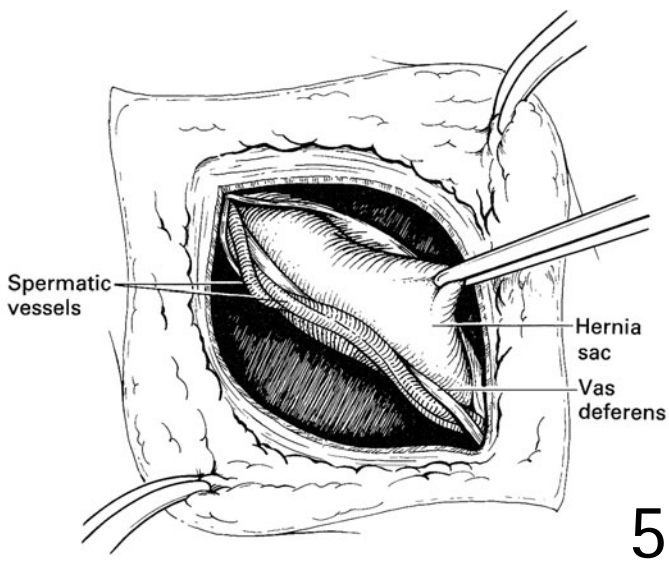
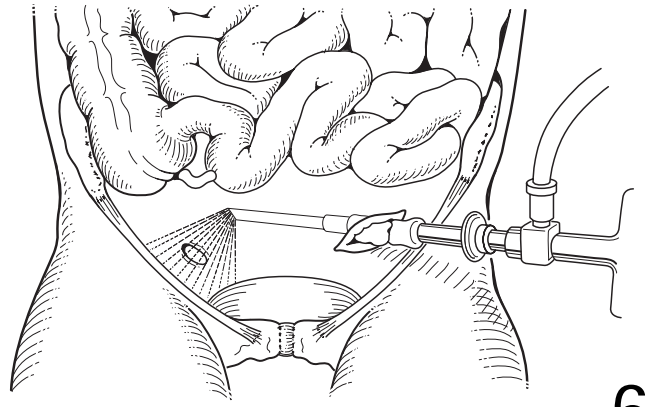
4 The undersurface of the superior leaflet of the external oblique fascia is gently dissected free from the internal oblique and abdomen's transverse muscle. The inferior leaflet is mobilized down to the inguinal ligament. During this mobilization, the iliohypogastric and ilioinguinal nerves are located. The ilioinguinal nerve can be seen on the outer vestment of the spermatic fascia. The cremasteric muscle is teased open by blunt dissection on the anteromedial surface of the cord and spread to expose the glistening peritoneum of the indirect hernia sac.

5 The sac is elevated anteromedially and the spermatic vessels are identified and carefully dissected free from the diverticular structure of the inguinal hernia sac. Once the spermatic vessels are mobilized away from the sac, the vas deferens is visualized. The vas deferens is often intricately adherent to the sac and should never be grasped directly with forceps or a clamp during the dissection, as this can result in an injury. The hernia sac often extends to the testicular area. Once the vital structures (vessels and vas deferens) are identified and cleared laterally, the hernia sac can be divided between clamps and the upper end dissected superiorly to the level of the internal (deep) inguinal ring. The proper extent of the superior dissection is identified by the presence of retroperitoneal fat at the neck of the sac.



5

6 At this point the sac can be opened on the most distal end and a 3 mm or 5 mm trocar advanced into the abdomen and secured with a tie to minimize insufflation leak. The abdomen is insufflated with CO_2 to 8–14 mmHg pressure and a 70° laparoscope is advanced through the trocar into the abdomen to view the opposite internal ring.



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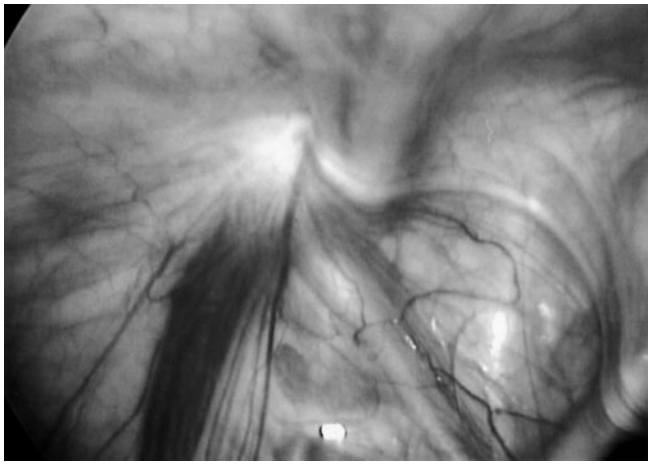
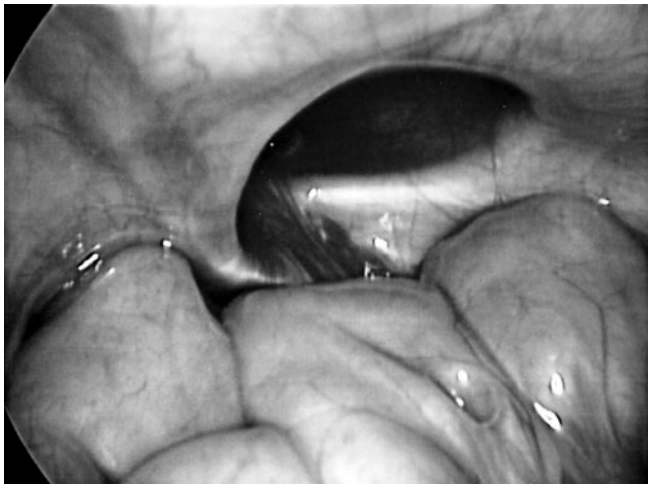


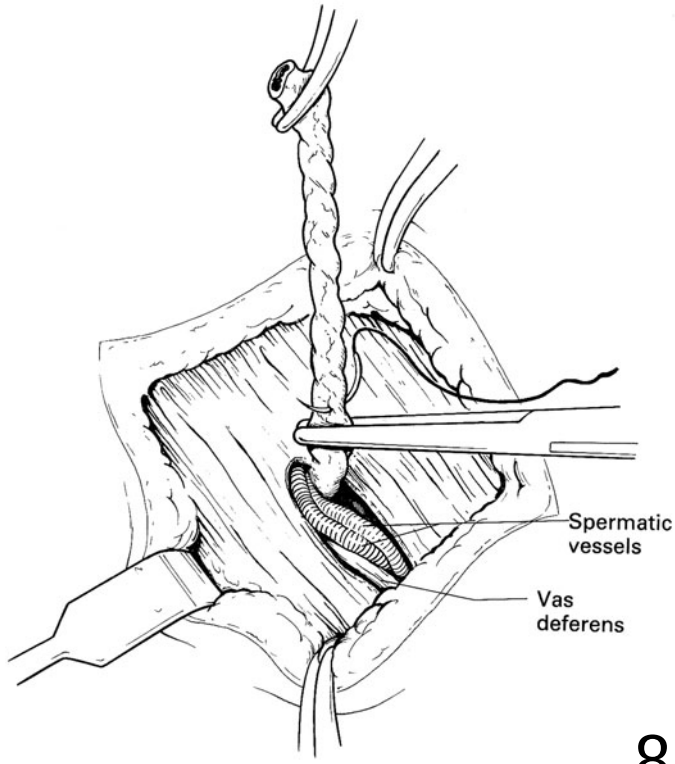
7a



7b

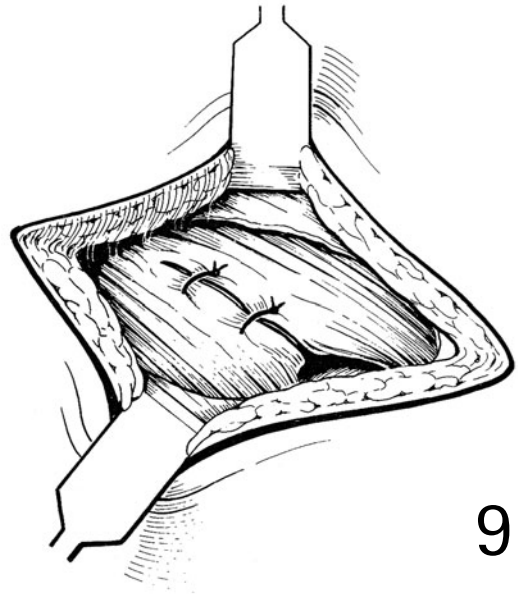
7a,b Viewing the left internal ring via the right inguinal hernia sac, one can note in Figure 7A a normal internal ring with the vas deferens medially and the spermatic cord vessels located laterally. There is no inguinal hernia evident. In Figure 7B, while viewing the right internal ring via the left inguinal hernia sac, a large indirect inguinal hernia is noted with weakening of the inguinal floor. When a contralateral hernia is noted, a similar repair can be performed on the opposite groin during the same anesthetic with limited morbidity. This technique avoids unnecessary routine contralateral exploration for all patients without a recognizable bulge on the opposite side and limits repair to those babies who have a demonstrable hernia on laparoscopy. The authors have employed this technique in most children less than 24 months of age.

8 Following laparoscopy, the trocar is removed and any contents in the sac should be reduced into the peritoneal cavity. The base of the sac may be twisted to ensure that all of the contents are fully reduced. If contents seem to remain within the sac, one should suspect a sliding component within the posterior wall of the sac. Sliding hernias often present as a more chronic hernia with the bulge often noted for a prolonged period of time prior to referral. In these instances a viscus is usually noted as a component of the sac. The sac may contain bowel – partly intraperitoneal and partly extraperitoneal. The bladder can occasionally be found medially, and a slider of the cecum might be noted on the right lateral side. The pelvic colon may be part of the sac on the left lateral side. In girls, the fallopian tube and uterus can be part of the sliding component. Unwise dissection may result in an accidental injury to the intestine or the bladder. Similarly, failure to recognize the sliding component may result in injury if the surgeon attempts to perform high suture ligation of the sac and includes the viscus in the suture. The best treatment in boys usually involves the separation of the spermatic vessels and vas deferens from the sac, opening the sac, and delicately dissecting the intra-abdominal structures off it. Following reduction of the sliding component into the abdomen, a high ligation of the sac at the internal ring can then be safely carried out. Forceps (or a slotted spoon) may then be placed at the base of the sac to protect the cord structures. The neck of the sac is transfixed twice with a non-absorbable 4/0 (or 3/0 in older children) suture ligature. A free tie is avoided because of the risk of postoperative abdominal distension and the possibility of dislodgment of the free tie from the peritoneum. If the dissection has been sufficient, retraction of the sutured hernia sac stump into the preperitoneal space will occur. The distal end of the hernia sac is then opened on its anterior surface and excess tissue excised. If a separate hydrocele is present, this should be excised at the same time. If the internal ring is excessively large, it can be made smaller inferior to the cord vessels with an interrupted 4/0 non-absorbable suture placed across the transversalis fascia. The floor of the inguinal canal is usually normal and requires no specific repair. During the procedure the surgeon should avoid any injury to the transversalis fascia. High ligation of an infant hernia is usually all that is required. In rare cases where there is an associated direct inguinal hernia, or the internal ring is excessively large, this can be repaired by inserting a few non-absorbable sutures between the conjoined tendon and Poupart's ligament. If the direct component comprises a majority of the floor of the canal, a Cooper's ligament repair may rarely be necessary. The testis should be returned to a normal intrascrotal location at the end of the procedure. Administration of a local anesthetic (e.g., bipivacaine 0.25 percent, use 0.25 mL/kg per hernia repair side for a maximum of 3 mg/kg dose) along the ilioinguinal and iliohypogastric nerves will reduce postoperative pain.



8

9 Wound closure is accomplished with an interrupted or running absorbable 4/0 suture, approximating the external oblique fascia leaflets to the external ring.



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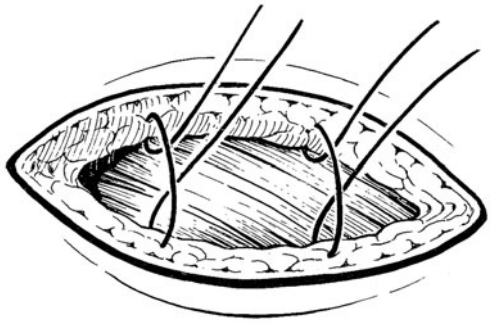


10

10 Scarpa's fascia is closed with one or two interrupted 4/0 absorbable sutures. The skin edges are opposed with either interrupted or running subcuticular 4/0 or 5/0 absorbable sutures. The skin edges are approximated with a Dermabond dressing in infants if they are not toilet-trained. Alternatively, Mastisol and sterile skin closure strips (e.g., Steristrips) and a semipermeable adhesive film dressing (e.g., Opsite) are applied in older children.

OPERATION: LAPAROSCOPIC APPROACH

The abdomen and groin area are prepped with a sterile solution and draped with linens. A 3 mm or 5 mm trocar is placed through an umbilical incision and the abdomen insufflated with CO_2 to approximately 8–14 mmHg pressure. A 3 mm or 5 mm laparoscope is advanced into the abdomen to view both groins for the presence of hernias. Evaluation for indirect, direct, and femoral hernias is performed. If no hernia is noted, the procedure is completed with closure of the umbilical defect. If a hernia is present, two 2 mm or 3 mm needle drivers are inserted through the lateral abdominal wall with or without trocar use to complete the repair. The neck of the sac is closed with a 4/0 monofilament suture in an interrupted or purse-string fashion. The instruments are removed along with the trocar, and the umbilical defect is the only wound that typically warrants fascial closure. The skin is approximated with Steristrips on all wounds. The duration of the procedure has been reported to be approximately 16 minutes for a single-sided repair and 22 minutes for a bilateral repair. The reported recurrence rate is 3.4 percent with up to a 7-year follow-up.

POSTOPERATIVE CARE

With the exception of infants who require extended observation, most patients are discharged from the day surgery room within 2 hours after operative repair. Oral intake may be resumed when the child awakens. Tylenol with codeine is used for analgesia for approximately 48 hours following the procedure. Baths can be resumed on the third postoperative day. There are no activity restrictions for infants, but older children should refrain from bicycle riding or other vigorous physical activity until their pain has subsided. Treatment of routine inguinal hernias usually includes a postoperative

clinic visit. Prospective assessment of the necessity for the traditional approach has shown that there is no difference in overall satisfaction with the care received when given a follow-up clinic visit or no follow-up and a detailed instruction sheet. Accurate postoperative instructions and open access to follow-up when required are as effective as the traditional postoperative clinic visit, especially for families that would have to travel great distances.

OUTCOME

Injury to the spermatic vessels or vas deferens is unusual but may occur in approximately 3 per 1000 cases. If the vas deferens is divided, it should be repaired with interrupted 7/0 or 8/0 monofilament sutures. The use of magnifying lenses or an operating microscope will make the repair more precise.

Intraoperative bleeding is also an unusual complication unless the floor of the canal is weakened and requires repair. Needle-hole injury to the epigastric vessels or the femoral vein can usually be controlled by withdrawal of the suture and direct pressure.

Postoperative complications include wound infection, scrotal hematoma, postoperative hydrocele, and recurrent inguinal hernia. The wound infection rate at most major pediatric centers is quite low (1–2 percent). An increased incidence of infection might be expected in patients presenting with an incarcerated hernia.

Recurrent inguinal hernia is a relatively uncommon complication in children, with recurrence rates of less than 1 percent having been reported by experienced pediatric surgeons. Of these, 80 percent are noted within the first postoperative year. The major causes of recurrent inguinal hernia in children include: (1) a missed hernial sac or unrecognized peritoneal tear; (2) a broken suture ligature at the neck of the sac; (3) failure to repair (snug) a large internal inguinal ring; (4) injury to the floor of the inguinal canal, resulting in a direct inguinal hernia; (5) severe infection; (6) increased intra-abdominal pressure (patients with ventricular–peritoneal shunts and continuous ambulatory peritoneal dialysis); (7) cystic fibrosis and chronic cough; and (8) connective tissue disorders (i.e., Ehlers–Danlos syndrome). Most surgeons would approach a first-time recurrent inguinal hernia repair through the previous inguinal site. However, a preperitoneal repair (Cheatle–Henry type) or traditional laparoscopic repair should be considered in selected cases with multiple recurrences.

Postoperative hydrocele may occur rarely after high ligation of the proximal hernial sac and incomplete excision of the distal portion. To avoid this complication, the anterior and lateral aspects of the sac are partially resected. The postoperative hydrocele often resolves spontaneously. Rarely, long-term persistence of the hydrocele may require a hydrocele aspiration and possible formal hydrocelectomy.

Testicular atrophy has been observed after repair of incarcerated hernias and in instances of large, acute, tense hydroceles in young infants, but is very rare after a typical hernia repair.

FEMORAL HERNIAS

PRINCIPLES AND JUSTIFICATION

A femoral hernia is a protrusion of preperitoneal fat or viscus through a defect in the femoral canal. It is the least common hernia occurring in the inguinal region in infants and children. However, the diagnosis must be considered when examining a swelling in the inguinal region, especially if the bulge or mass presents inferior to the inguinal ligament. Femoral hernia may occasionally be confused with an enlarged, swollen, infected lymph node near the saphenofemoral junction (lymph node of Cloquet) just inferior to the inguinal ligament. Femoral hernias are often misdiagnosed and treated as inguinal hernias. Careful examination for a lower extremity focus of infection should be performed. Femoral hernias are most common in girls between 5 and 10 years of age. Occasionally, a femoral hernia may be noted shortly after an ipsilateral inguinal hernia repair. This may represent either a missed femoral hernia or operative damage involving the femoral canal. Some individuals advocate the use of laparoscopy in the child with presumed recurrent inguinal hernias because of the concern for a missed diagnosis. The anatomic boundaries of the femoral canal can be divided into anterior, posterior, lateral, and medial. The anterior border involves the iliopubic tract and/or the inguinal ligament. The posterior boundary includes the pectineal ligament (Cooper's) and iliac fascia. The lateral boundary involves a connective tissue septum and the femoral vein. Medially, the canal is bordered by the aponeurotic insertion of the transversus abdominis muscle and tranversalis fascia.

Indications

The presence of a femoral hernia is an indication for operation. Conservative management is contraindicated because of the risk of incarceration and strangulation. The fixed margins of the femoral ring result in early compression of swollen tissues and increase the risk of visceral compromise when incarceration occurs.

Choice of procedure

There are four possible methods to repair a femoral hernia: (1) the lower infra-inguinal ligament procedure of Langenbeck, (2) transinguinal Cooper's ligament repair (McVay procedure), (3) abdominal extraperitoneal repair (Cheatle–Henry), and (4) laparoscopic repair. Although femoral hernia repair has also been performed using endo-

scopic techniques through the laparoscope in adults and there are limited reports in children, the authors have not used this technique. It may have a role in instances of recurrent hernias.

An inguinal or extraperitoneal approach should be used in instances of strangulated obstruction; however, since this is rarely seen in infants and young children, the low infra-inguinal repair is preferred. If a concomitant inguinal hernia is present, an inguinal incision and McVay repair are recommended.

PREOPERATIVE ASSESSMENT AND PREPARATION

In elective cases, the infant or child is kept without oral intake for 4–6 hours before the anticipated procedure.

ANESTHESIA

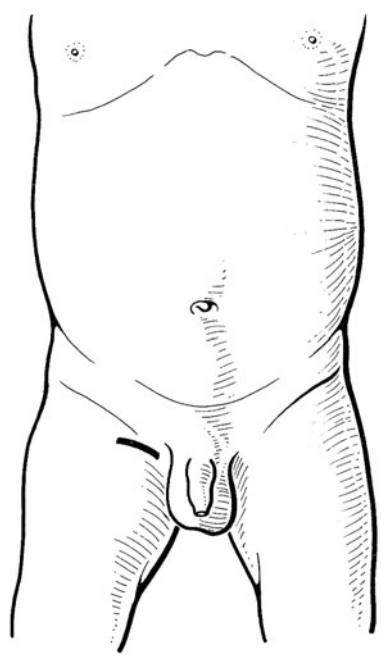
In elective cases, an outpatient operation may safely be carried out. Mild sedation is administered preoperatively (midazolam 0.5–1.0 mg/kg orally 15–20 minutes prior to going to the operating room) and general endotracheal anesthesia is employed. Following appropriate skin cleansing of the lower abdomen, inguinoscrotal (or labial) area, thigh, and perineum, sterile drapes are applied.

OPERATION

The infra-inguinal (Langenbeck) repair is described, as the other approaches are dealt with in the section on inguinal hernia.

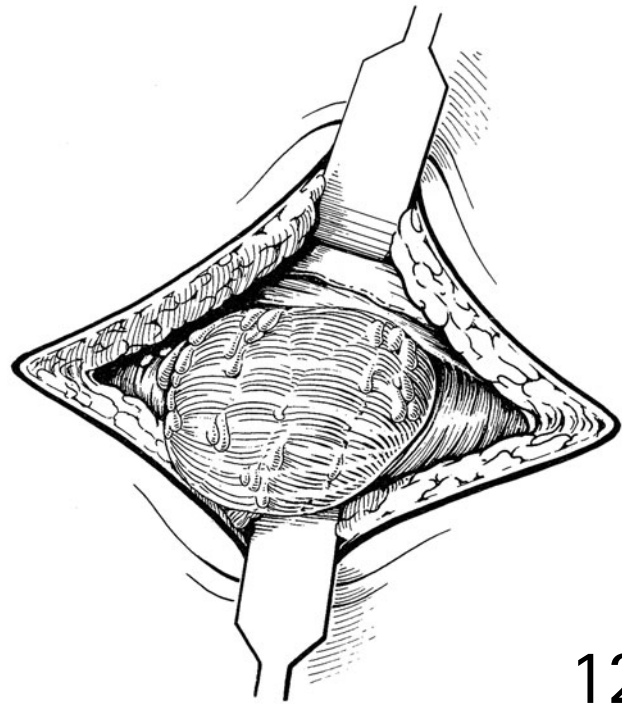
Infra-inguinal (Langenbeck) repair

11 A transverse incision is made in a skin crease over the mass from a point just inferior to the pubic tubercle medially, extending laterally just past the palpable pulsation of the femoral artery.



11

12 Hemostasis is effected with an electrocoagulator and the wound is deepened to expose the hernia sac bulge. The sac is covered by cribriform fascia and groin fat and may overlie the femoral vein and extend upwards over the inguinal ligament.



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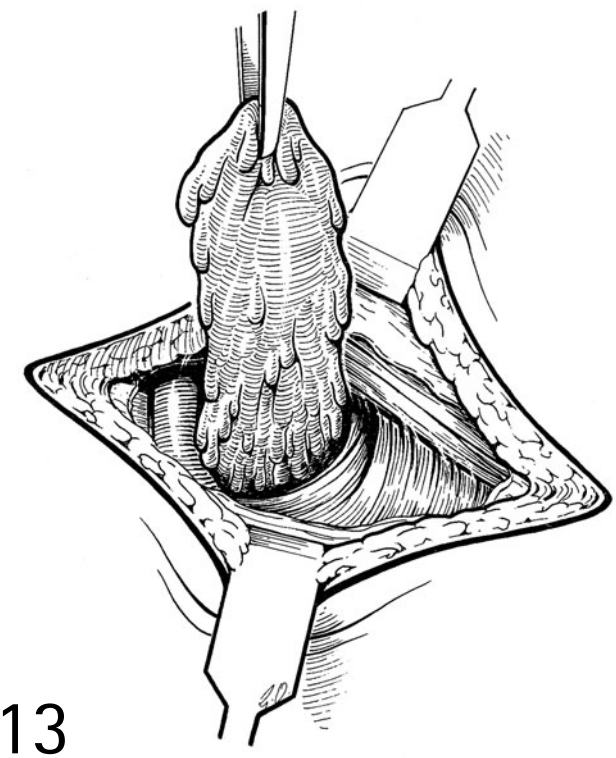
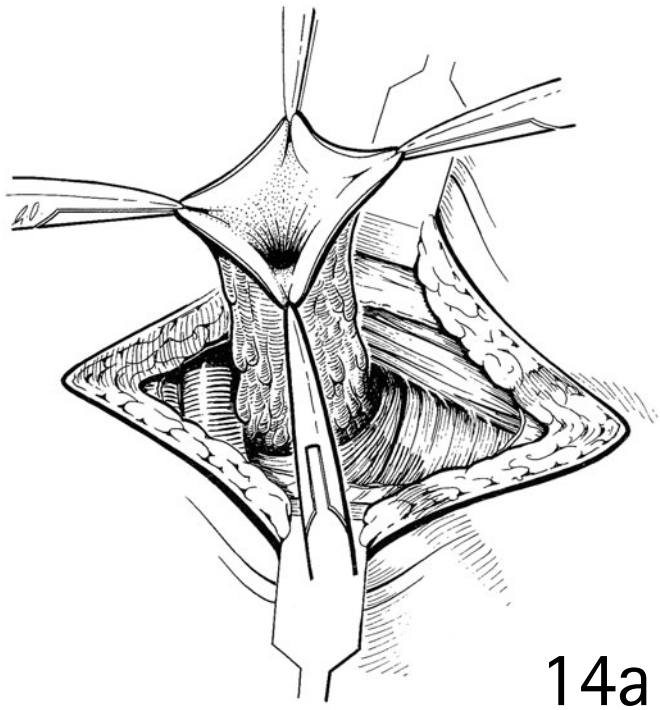


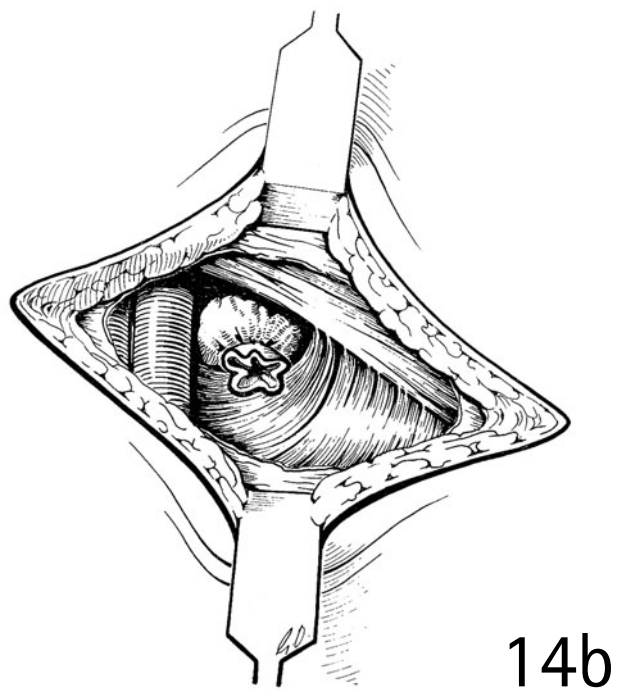
13

13 The cribriform fascia and fat layers are incised, exposing the femoral hernia sac. Note that the femoral sac protrudes into the femoral canal medial to the femoral vein, inferior to the iliopubic tract, above the Cooper's ligament, and lateral to the reflected fibers of the iliopubic tract. The sac should be carefully palpated for visceral contents, which should be gently reduced through the defect. Occasionally, when incarceration is present and reduction is difficult, the sac is released by carefully incising the insertion of the iliopubic tract into the Cooper's ligament at the medial margin of the femoral ring.



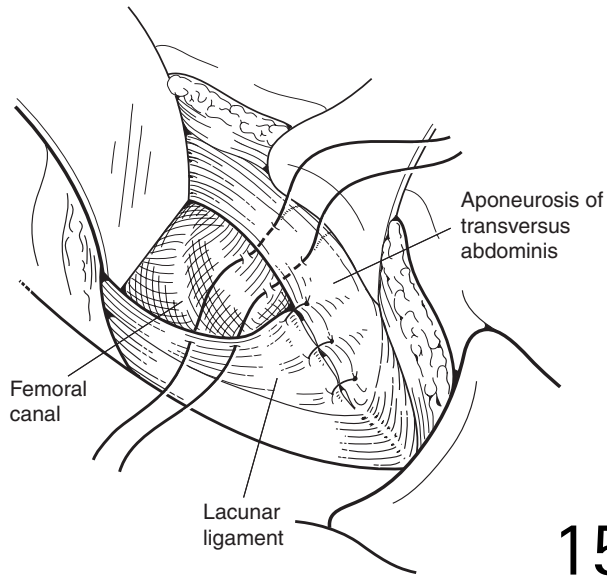
14a

14a,b Often, the peritoneal sac is small; however, it may have a considerable amount of retroperitoneal fatty tissue at its base. The sac can be opened (to ensure reduction of contents) if there is any question about the reduction having been complete. Hernial repair is initiated with high ligation of the sac similar to that for an inguinal hernia. This is accomplished with 3/0 non-absorbable suture ligatures. The sac may be bulky, however, and an alternative method of sac closure is inversion and reduction of the intact sac, placing one or two 3/0 non-absorbable purse-string sutures at a level just above the femoral defect.



14b

15 Repair of the femoral defect is facilitated by placing a mall retractor to raise the inguinal ligament superiorly in order to expose the pectineus fascia and Cooper's ligament. If this is not performed, the femoral vessels may be traumatized by suture placement. The hernia repair is completed by suturing these two structures together, thereby obliterating the femoral canal medial to the femoral vein. All sutures are placed between Cooper's ligament and the inguinal ligament under direct vision before tying. The authors use interrupted 3/0 non-absorbable suture material in older children and 2/0 suture in adolescents. Special attention is given to avoid either injury to the femoral vein by the needle or its compression when the sutures are tied. Gentle lateral retraction of the vein during suture insertion is useful. In the posterior approach, the external iliac and femoral veins are easily seen and can be maintained out of the line of suturing. In addition, one can maximize the degree of closure of the femoral canal without compressing the vein.



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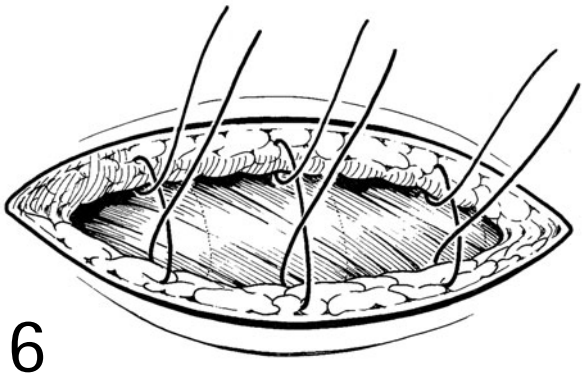

16

16 Prior to closing, it is important to evaluate the rest of the groin to ensure there is no evidence of another hernia, as a missed hernia may present as a recurrent femoral hernia. Wound closure is accomplished with a few interrupted, inverting 4/0 absorbable sutures. The wound edges are approximated with 5/0 subcuticular absorbable suture material and the skin is sealed with Steristrips and a semipermeable adhesive film dressing (e.g., Opsite) in children or with Dermabond in infants who are not toilet-trained.

POSTOPERATIVE CARE

Oral intake may be resumed when the infant is alert. Acetaminophen (paracetamol) with codeine may be used for pain control for 24–48 hours. Most young children return to normal activity within a few days. The family is instructed to keep the wound dry for the first few days to minimize wound complications. In older children or teenagers, avoidance of competitive athletics and bicycle riding is advised until the pain has subsided.

OUTCOME

In most cases, recovery from elective femoral hernia repair is uncomplicated. A superficial wound infection may develop in 1 percent of cases and should be recognized promptly and the wound opened or the administration of oral antibiotics should be initiated early to avoid possible extension of a closed infection to the deeper tissues. A wound hematoma is rarely observed and may be caused by an unrecognized tear in a saphenous/femoral venous branch.

Pain and ipsilateral leg swelling may be the result of compression of the iliac or femoral vein, which can be obviated or minimized with meticulous attention while placing the hernia repair sutures as previously described.

Since a femoral hernia is relatively rare in children, data concerning recurrence are not available. A recurrence rate of 1–2 percent in children might be expected. In the event of a recurrence, one must rule out an associated hernia. Some have adopted the preperitoneal approach for recurrent repair due to the higher rate of secondary recurrences. Because recurrences may be the result of a localized collagen defect, and direct re-approximation of the weakened tissue may not be appropriate, the use of a prosthetic buttress may be indicated. The posterior approach will avoid re-operation

through a previous surgical site, allow evaluation and exclusion of all groin-related hernia defects, facilitate the repair by easy visualization of Cooper's ligament and the iliopubic tract, and if a prosthetic buttress is chosen, allow the even distribution of intra-abdominal pressure to the prosthetic patch.

UMBILICAL HERNIA

PRINCIPLES AND JUSTIFICATION

Umbilical hernias are commonly encountered in infants and young children. The hernia sac protrudes through a defect in the umbilical ring due to a failure of complete obliteration at the site where the fetal umbilical vessels (umbilical vein and the two umbilical arteries) are joined to the placenta during gestation.

Approximately 20 percent of full-term neonates may have an incomplete closure of the umbilical ring at birth. However, 75–80 percent of premature infants weighing between 1.0 kg and 1.5 kg may show evidence of an umbilical hernia at birth. Umbilical hernia is more common in girls than in boys. Black children have a higher incidence than white children.

The umbilical bulge becomes more apparent during episodes of crying or straining, or even during defecation, and may result in considerable protrusion of the sac and, at times, visceral contents through the ring. The hernial protrusion is composed of peritoneum adherent to the undersurface of the umbilical skin. The hernia often causes considerable parental anxiety and frequent requests for operative repair in early infancy.

Although rupture or incarceration of an umbilical hernia occurs, this is an exceptionally rare event in the authors' experience. The hernia is rarely a cause of pain or other symptoms. Almost 80 percent of umbilical hernias will decrease in size and close spontaneously by 4–5 years of age. Careful counseling will usually allay unnecessary parental anxiety and fear.

Indications

As the majority of these very low-risk hernias close spontaneously, it is safe to wait until the child is 4 years of age (particularly if the umbilical ring is less than 1.5 cm in diameter) before attempting repair. In contrast, defects of more than 1.5–2.0 cm diameter rarely close spontaneously. Since there is a significant risk of complications, including incarceration and strangulation in adults with umbilical hernia, those hernia defects that do not close by 4–5 years of age should be electively repaired. The umbilical defect can also be repaired in children under the age of 4 years with a ring of more than 1.5–2.0 cm in whom a general anesthetic is anticipated for another condition.

PREOPERATIVE ASSESSMENT AND PREPARATION

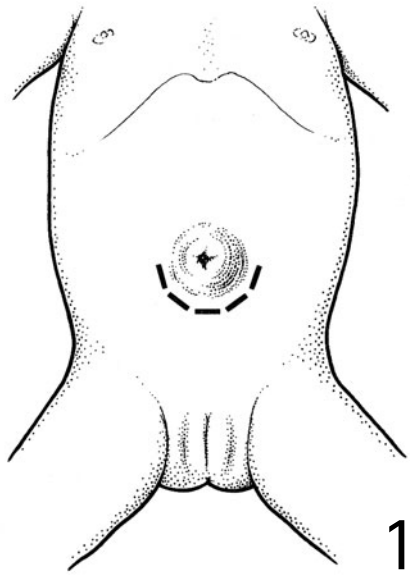
The child is kept without oral intake for 6 hours before the anticipated time of the procedure. The operation can safely be carried out on an outpatient basis. Careful preparation of the skin is essential, as the umbilicus is often a repository of surface debris, lint, etc., and is not always kept immaculate. Preoperative cleansing with cotton applicator sticks may be useful.

ANESTHESIA

After the administration of a mild preoperative sedative (oral midazolam), the procedure is carried out under general anesthesia via endotracheal intubation or laryngeal mask airway.

OPERATION

17 After appropriate skin preparation and application of sterile linen drapes, a curved ('smile') incision is made in a natural skin crease immediately below the umbilicus. A supraumbilical incision is also acceptable, especially if a supraumbilical defect is encountered. Placement of four-quadrant traction by the assistant on the abdominal wall and slight upward traction of the defect allows selection of the site of the incision. The curved incision should typically not extend beyond 180°.



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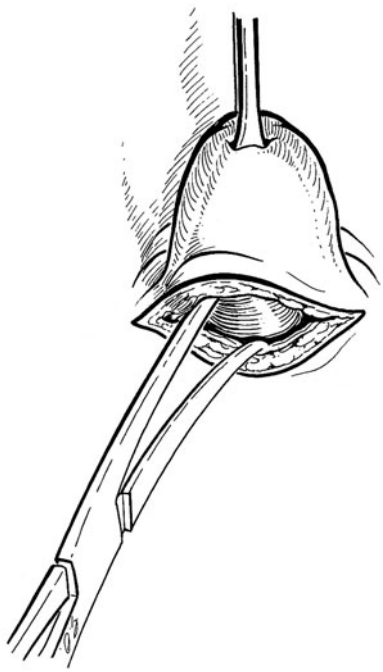

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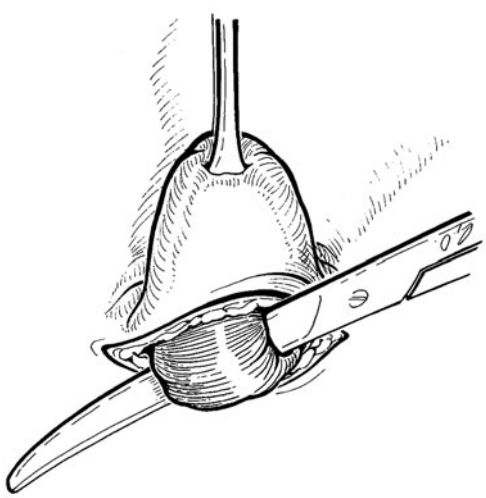
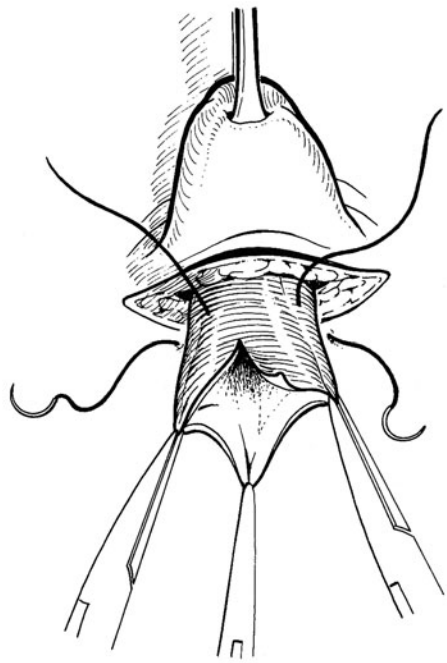


18b

18a,b The subcutaneous tissue is incised and bleeding points controlled with a fine tip electrocoagulator. With upward traction on the inner margin of the upper lip of the incision, dissection is carried out down along the sac to the level of the anterior abdominal wall fascia. By blunt dissection with a mosquito clamp, a plane is developed on either side of the sac, extending superiorly to gain control of the entire circumference of the sac. Any contents in the sac should be reduced into the peritoneal cavity. If the sac is large, the surgeon or assistant places an index finger in the skin defect to evert the sac where it is attached to the skin. The sac is dissected free from its skin attachments, preserving the umbilical skin for an umbilicoplasty. Separation of the sac may require its transection near the skin to preserve the umbilicus for cosmetic purposes.

19a,b The entire sac is elevated by mosquito clamps to maintain control of the edge of the defect and to have direct visualization during placement of sutures to avoid visceral injury. The sac is opened and any contents reduced. The rim of the defect is identified and the sac incised to allow placement of sutures starting at the corner farthest from the surgeon. Continuous or interrupted 3/0 (infants and young children) or 2/0 (older children and teenagers) non-absorbable sutures are placed. The sutures are elevated to maintain upward traction on the abdominal wall. The sac is partially excised at the level of the abdominal wall as more sutures are placed. A traction suture may also be placed at the corner of the transverse wound closest to the operating surgeon to offer exposure as the remaining sac is excised and sutures placed. If a lot of redundant tissue is present or the initial tissue layer seems sparse, a layer of fascia can be imbricated over the initial line of repair with interrupted or continuous absorbable suture.



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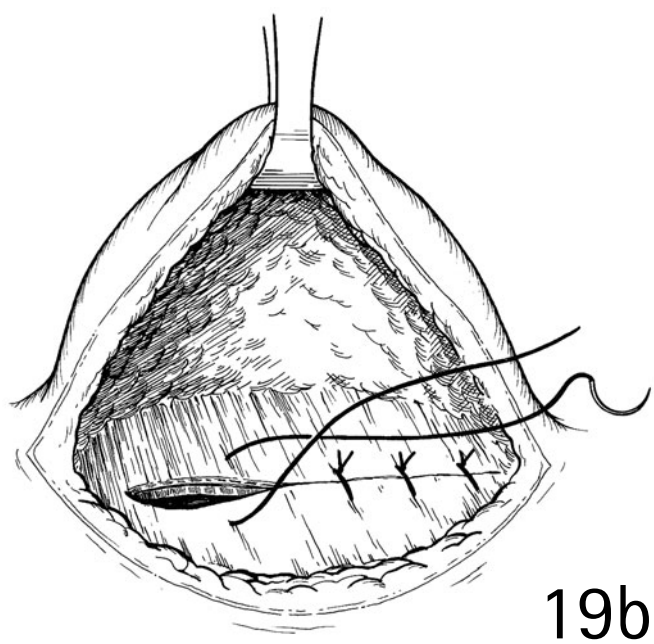
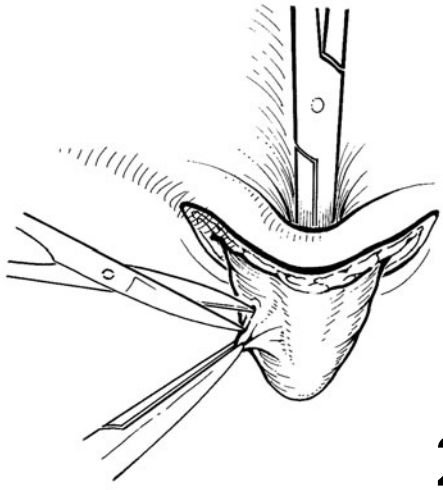
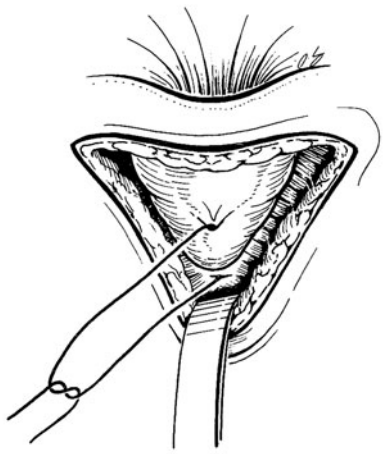


19b

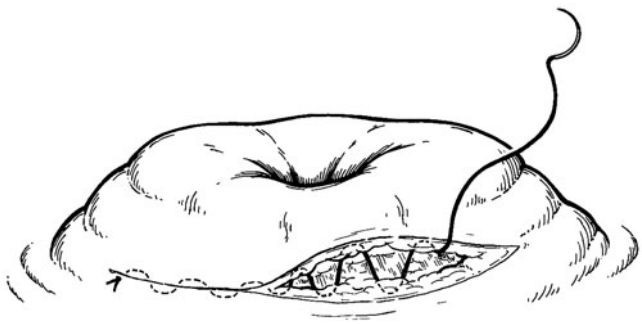
20a,b An umbilicoplasty is performed for cosmetic purposes by inverting the undersurface of the redundant umbilical skin to the anterior abdominal wall fascia with one or two interrupted 4/0 absorbable sutures. Any remnant of the peritoneum on the umbilical sac that is adherent to the skin may be safely left behind, excised, or cauterized. In children with very large protuberant hernias with redundant skin following the removal of the sac and fascial closure, the umbilicoplasty can sometimes be frustrating to the surgeon and patient. The volume and variety of literary material on the repair of umbilical defects attests to the fact that there exists no single ideal approach. The management of the excess umbilical skin can be completed using numerous methods such as purse-stringing, complicated V-Y advancement procedures, four equilateral triangular skin flaps, or the Mercedes-Benz umbilicoplasty.



20a



20b



21

21 The wound is closed by a few interrupted, inverted 4/0 absorbable sutures in the subcutaneous fascia. The skin edges are opposed with a running subcuticular suture. A Dermabond dressing may be applied or one may use Steristrips and an Opsite dressing in older patients.

POSTOPERATIVE CARE

Oral fluids can be offered when the infant or child is alert. Acetaminophen with codeine may be used for pain control for 24–48 hours. Postoperative activity restrictions are similar to those for an inguinal hernia repair.

OUTCOME

Complications are unusual and are limited to wound infection (1 percent) or an occasional wound hematoma. Bowel injury is the rarest complication. Recurrence is rare as well, and those that do recur are typically in children with associated comorbidities such as long-term continuous ambulatory peritoneal dialysis, ventricular peritoneal shunts, or a connective tissue disorder.

SUPRAUMBILICAL AND EPIGASTRIC HERNIAS

PRINCIPLES AND JUSTIFICATION

A number of defects may be observed along the linea alba between the xiphoid process and the umbilicus. Failure of fixation of the medial borders of the rectus abdominis muscles at the linea alba results in a large, bulging defect – a diastasis recti – which is of virtually no consequence and resolves spontaneously as the linea alba becomes a firm structure.

Epigastric hernias usually occur in the midepigastrium. The actual defect may be small; however, it is often symptomatic. Preperitoneal fat can incarcerate and cause pain. In some cases a tender mass of incarcerated fatty tissue can be palpated in the defect. Since epigastric hernias do not close spontaneously and are often symptomatic, they should be repaired on an elective basis. As the hernial defect may be small, it is wise to mark the skin over the exact site before surgery with the child awake and in a standing position prior to the procedure.

PREOPERATIVE ASSESSMENT AND PREPARATION

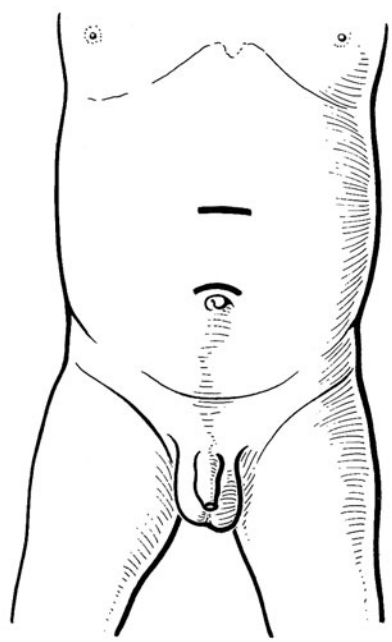
Repair can be performed on an outpatient basis.

ANESTHESIA

General anesthesia is typically required. The abdomen is prepared with a cleansing solution and sterile drapes applied.

OPERATION

22 A transverse or vertical incision is made directly over the pre-marked area identifying the site of the hernia defect, or, in the case of a supraumbilical defect, a supraumbilical frown incision may be made to maximize cosmesis.



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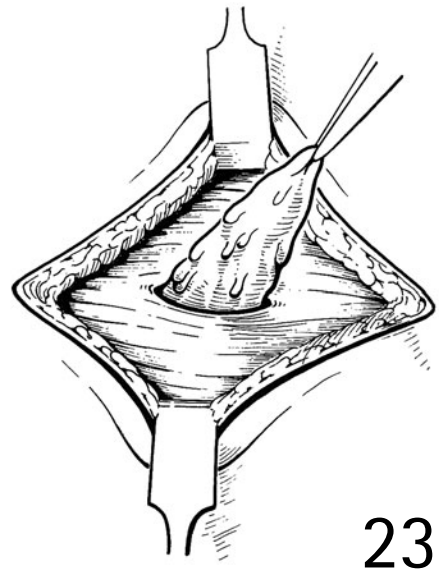
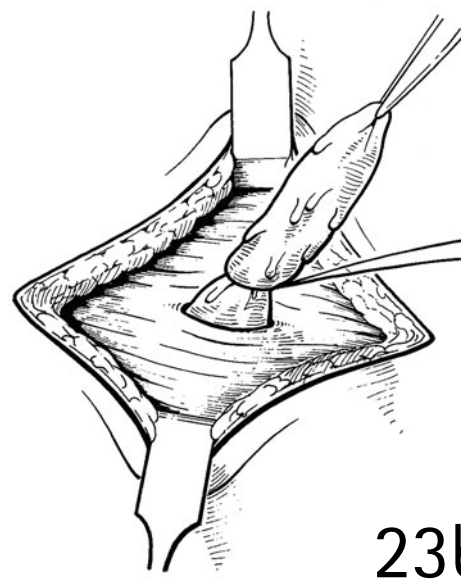


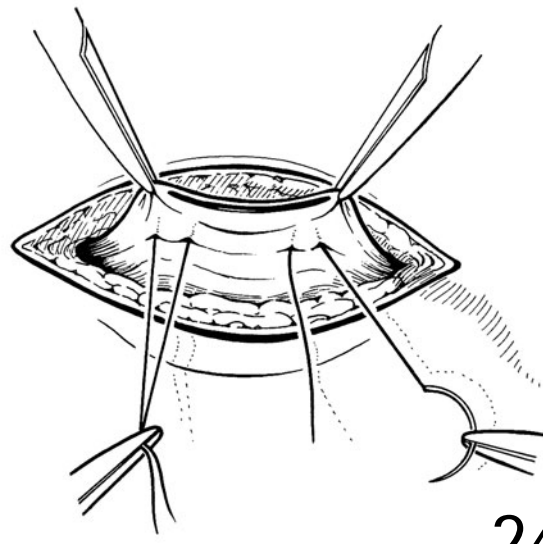
23a

23a,b Hemostasis is controlled with an electrocoagulator. There is often no peritoneal sac identified in instances of epigastric hernia. A fatty mass protruding through the defect in the linea alba is observed and may be suture ligated and excised or inverted and reduced into the preperitoneal space.



23b

24a,b The small defect is repaired with interrupted 3/0 absorbable sutures. Wound closure is accomplished with a running subcuticular 5/0 absorbable suture, and the skin edges are approximated with either Steristrips or Dermabond dressing.



24a

24b

POSTOPERATIVE CARE

Postoperative care is similar to that described for umbilical hernias.

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30

Omphalocele/exomphalos

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HISTORY

Ambrose Paré, the famous French surgeon, first described an infant with omphalocele in 1634. Although small omphaloceles were subsequently successfully repaired, there were few reported survivors of larger abdominal wall defects until the 1940s, when Gross described a two-stage closure of an omphalocele using skin flaps followed by ventral hernia repair. Further advancement in the treatment of massive omphalocele defects occurred when Schuster devised an extracelomic 'pouch' to house eviscerated bowel temporarily. This was later modified by Allen and Wrenn, who devised the additional innovation of staged reduction of abdominal contents to allow gradual enlargement of the abdominal cavity. The development of total parenteral nutrition in the 1960s allowed vigorous nutritional support of infants with abdominal wall defects, in whom a period of 1–3 weeks of intestinal dysfunction is expected after the operation. The basic principles of occasional non-operative therapy, primary closure when possible, and staged reduction with a temporary Silastic 'pouch', remain the mainstays of contemporary therapy for these congenital defects.

EMBRYOLOGY

Although controversy continues regarding the similarities, relationships, and embryological events surrounding omphalocele and gastroschisis, it is probably most reasonable to consider these as separate entities. Omphalocele is basically a persistence of the body stalk in the midline, where somatopleure normally develops. Failure of return of the normally herniated midgut at the twelfth fetal week either causes or aggravates the condition. Unless ruptured, omphaloceles are covered with a sac consisting of inner peritoneum and outer amnion. Infants with omphalocele frequently have associated anomalies (cardiac, renal), chromosome abnormalities (trisomy 13-15, 16-18), or recognizable syndrome associations (Beckwith–Wiedemann, Cantrell's pentalogy, caudal regression).

PRINCIPLES AND JUSTIFICATION

Because of the possibility of serious, life-threatening, or even lethal associated anomalies, infants with omphalocele are occasionally treated non-operatively. Painting the sac with agents designed to induce eschar formation, followed by gradual epithelialization from the base of the defect upwards, will eventually produce a covered ventral hernia. Early use of alcohol, iodine, and mercury-containing compounds produced toxicity due to systemic absorption of these compounds, and they have therefore been largely replaced by silver nitrate solutions or silver sulfadiazine cream.

The operative treatment of choice for small to medium-sized omphaloceles is excision of the sac, with primary closure of fascia and skin. If fascia closure increases intra-abdominal pressure sufficiently to cause respiratory embarrassment, skin closure alone, with later repair of the ventral hernia, is advisable. For giant omphaloceles, frequently containing liver as well as bowel, attaching a Silastic 'pouch' to the fascia allows gradual (10–14 days) reduction of the contents into the abdominal cavity, with eventual skin flap closure. Occasionally a prosthetic patch is needed to close the fascial defect in this setting, but skin must be mobilized sufficiently to cover the prosthesis.

PREOPERATIVE ASSESSMENT AND PREPARATION

Newborn infants with omphalocele must be placed immediately in a warm, aseptic environment to prevent evaporative

fluid loss, hypothermia, and infection. Warm, soaked sterile gauze can be placed on the defect, covered by transparent plastic wrap. Alternatively, a transparent plastic drawstring 'bowel bag' may be used, which can be kept sterile in the delivery room. The lower two-thirds (to the axillae) of the neonate can be placed within the bag. Safe transport of the infant to a center that is experienced in the management of these complex infants can then be accomplished.

Infants with large omphaloceles, especially when the liver is in the sac, should be positioned on their side to prevent twisting of the inferior vena cava from the sac 'tipping' to one side. Alternatively, rolls can be used to support the sac if the infant is placed supine.

Intravenous access must be established soon after birth to replace evaporative fluid loss and administer broad-spectrum antibiotics. Placement of the intravenous line above the diaphragm is preferable because of the possibility of inferior cava compression and partial obstruction as the eviscerated bowel and/or liver are reduced. An oral gastric or nasogastric tube should be placed to prevent gastric distension. Preoperative endotracheal intubation is reserved for premature infants or those with significant respiratory distress. The latter is occasionally encountered because pulmonary hypoplasia can be associated with omphalocele. All infants with omphalocele should undergo complete cardiac and renal evaluation before they are subjected to operative repair.

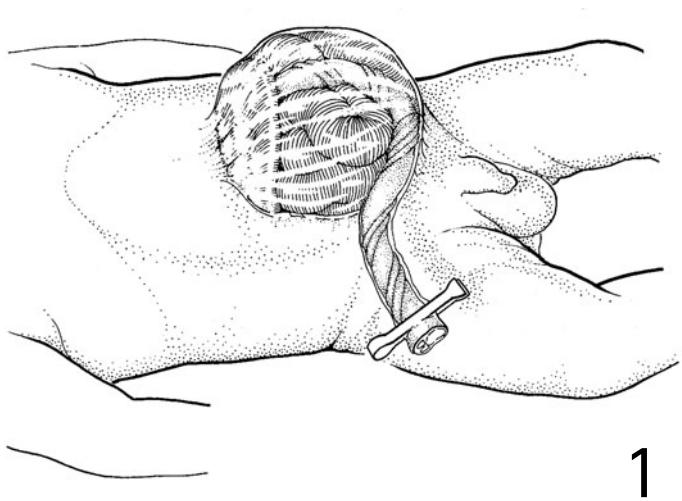
Anesthesia

General, endotracheal anesthesia, with complete muscle paralysis, is recommended for all infants with omphalocele. As stated above, infants with omphalocele who have serious or life-threatening associated anomalies, especially cardiac, should probably be treated non-operatively with application of escharotic agents to the sac.

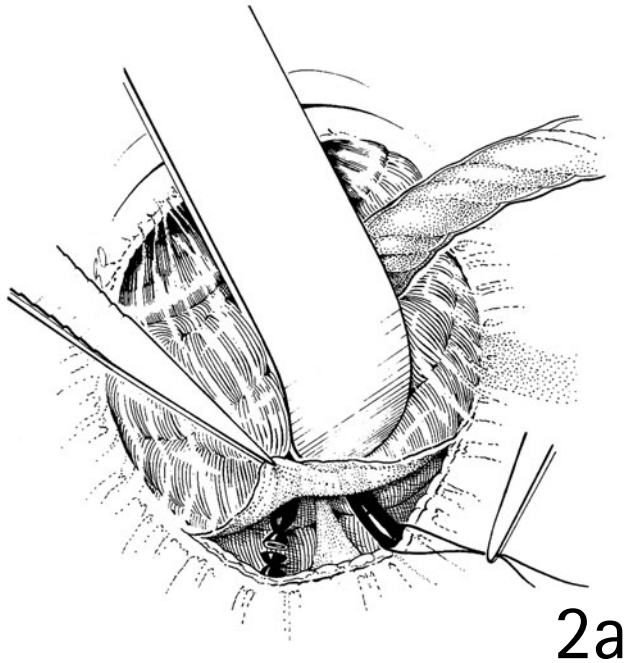
OPERATIONS

Small to moderate-sized omphalocele

1 A small ('hernia into cord') or moderate omphalocele, which may contain a small portion of liver, has the umbilical cord inserted into the top of the sac.

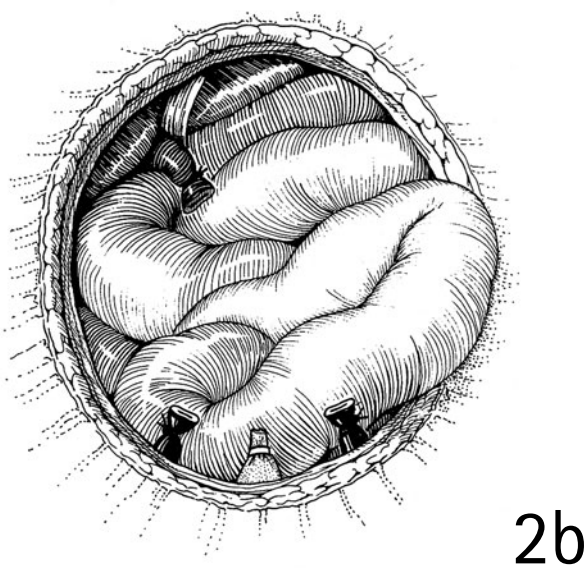


1



2a

2a,b Although some surgeons advocate leaving the sac intact and repairing the fascia and skin over it, most surgeons favor excision of the sac to allow complete intra-abdominal exploration. The sac is sharply removed at the skin/fascia edge, with careful identification and ligation of the umbilical vessels.



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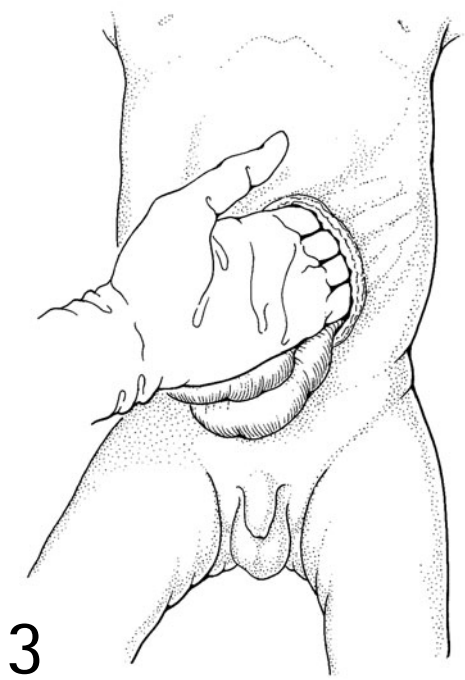
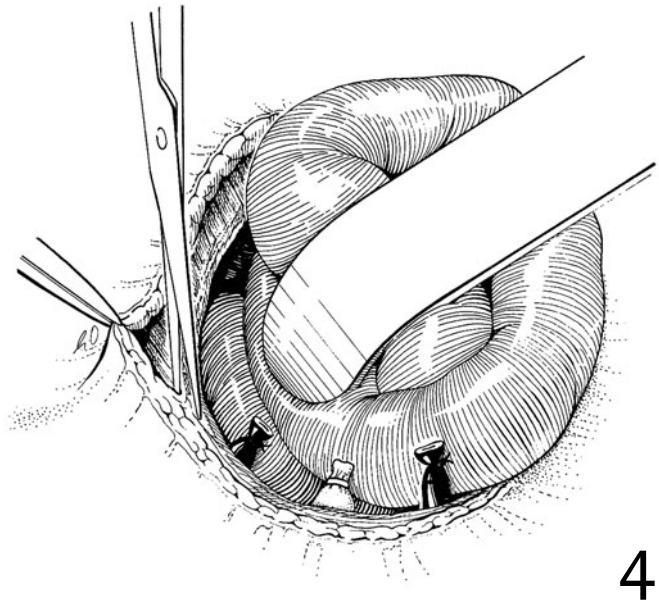


3

3 The abdominal cavity can be enlarged by manual stretching.

4 The skin is carefully 'undermined', separating it from the deep fascia layers.



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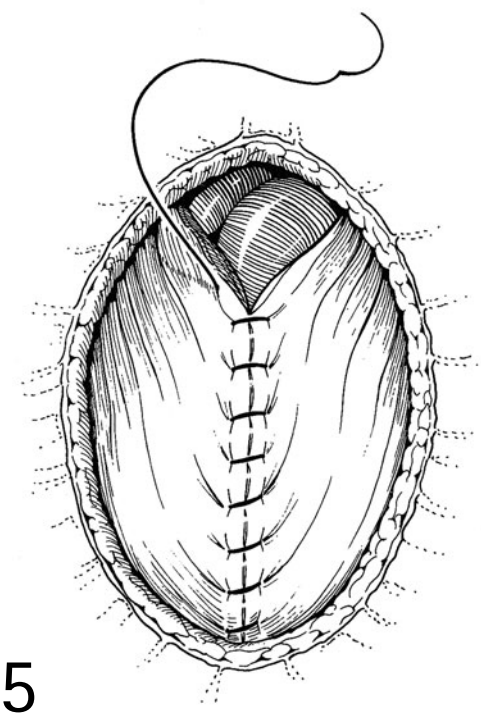
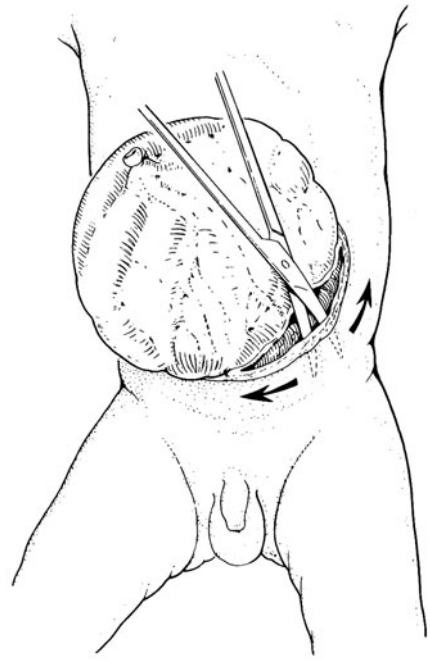


5

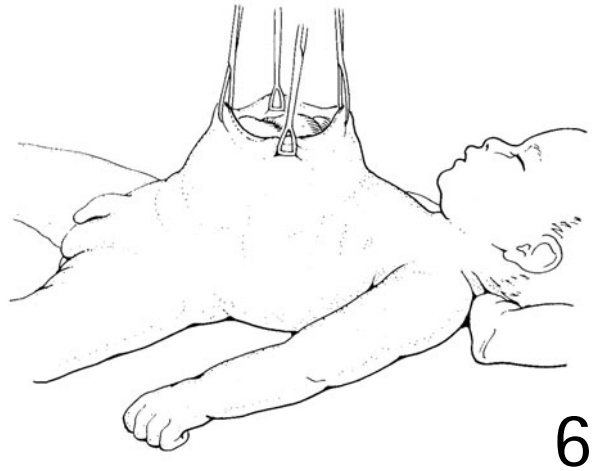
5 The fascia is closed with running or interrupted absorbable sutures (polyglactin or polydioxanone) and the umbilicus reconstructed.



6a

Large omphalocele: staged repair

6a–c Large omphaloceles, frequently containing most of the liver, are usually not fully reducible at the first operation, and staged repair is necessary. After undermining the skin, the skin is closed over the abdominal viscera, producing a ventral hernia that can be repaired 6–12 months later.



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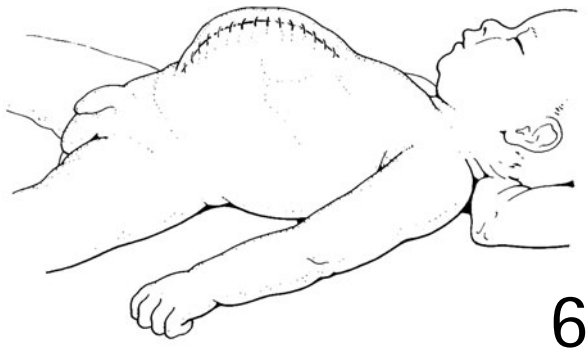
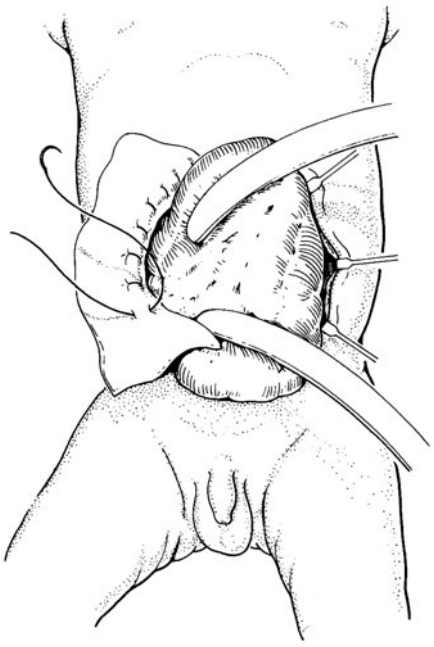
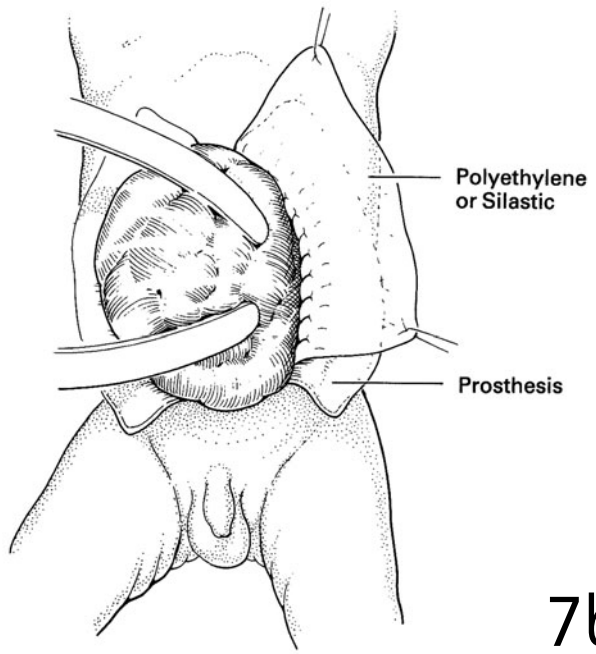


6c



7a

7a,b An alternative approach utilizes prosthetic closure of the fascia defect over polyethylene or Silastic sheeting to prevent adhesion of the viscera to the prosthetic material. The skin is closed over the fascia prosthesis.



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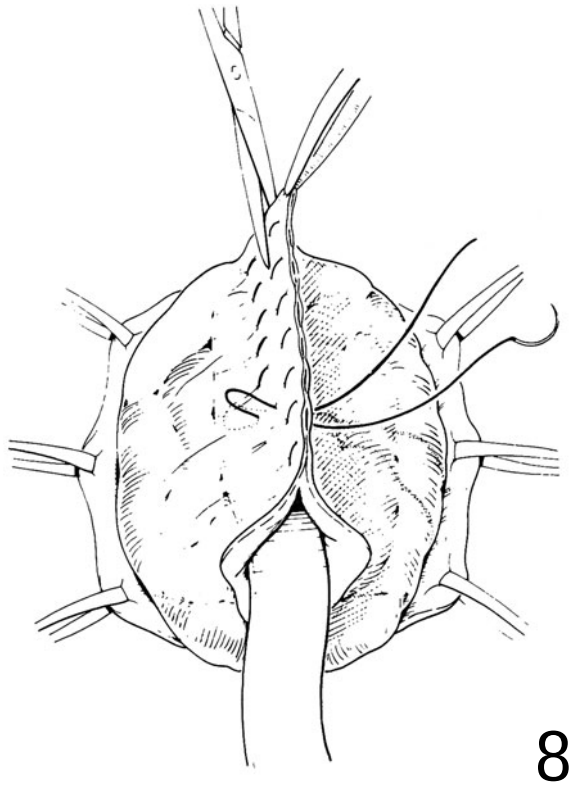

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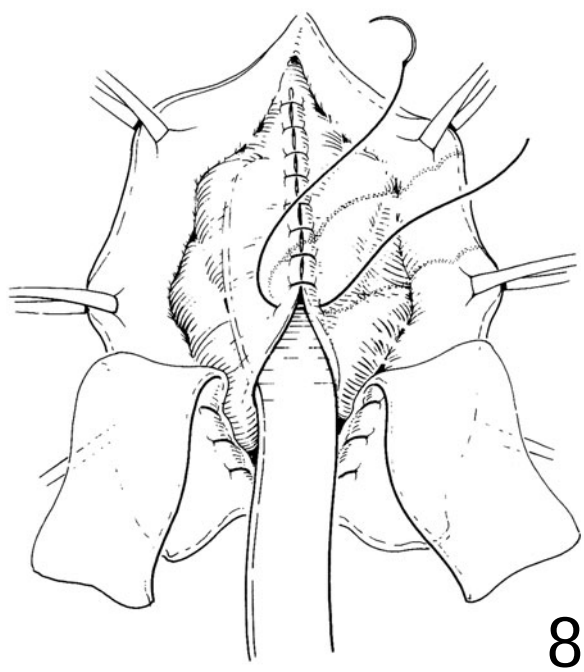
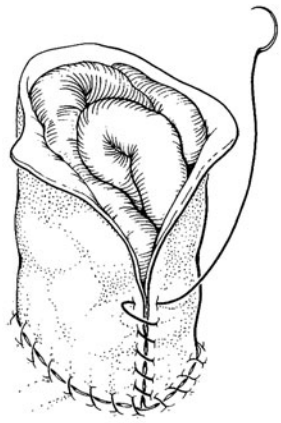


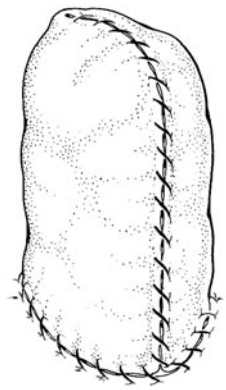
8b

8a,b At 4–6-week intervals the wound can be reopened and the skin dissected from the prosthesis. The central portion of the prosthesis and sheeting are resected and resutured to pull the fascia together. Eventually the fascia can be closed without the prosthesis.

9a–d In cases of ruptured omphalocele, an alternative method of management is necessary if the viscera cannot be reduced primarily and there is insufficient skin for coverage. A Dacron-reinforced Silastic sheet is attached to the abdominal wall with a running, non-absorbable suture and fashioned into a 'pouch' using the same suture. A preformed 'pouch', with a spring-loaded base that fits into the abdominal cavity to hold the 'pouch' in place and therefore does not require suturing, is also commercially available.



9a



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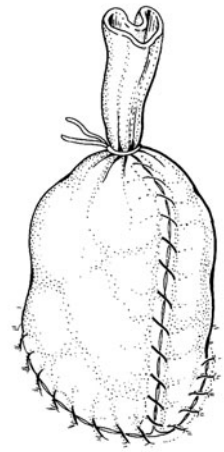

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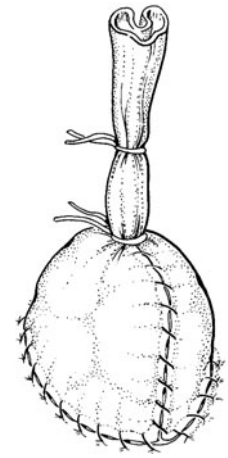
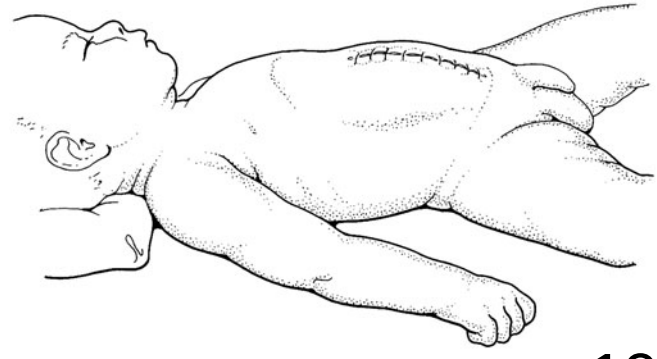


9d

The viscera are gradually reduced into the abdominal cavity, using gentle squeezing pressure on top of the 'pouch', which is then occluded by umbilical tape tie or suture to maintain reduction. This is usually performed without anesthesia every other day, over a 7–10-day period, until the gut is fully reduced.

10 The infant is then returned to the operating room for removal of the 'pouch', with fascia and/or skin closure.



10

POSTOPERATIVE CARE

All infants should be maintained on systemic antibiotics for 5–7 days or until the prostheses are removed. Intravenous nutrition should be initiated as soon as possible and continued until bowel function returns. In infants with omphalocele, this may be a period of several days.

Respiratory compromise is common after primary repair, or during staged reduction of omphalocele, and endotracheal intubation, ventilators, sedation, and, occasionally, muscle relaxants are common interventions in these infants in the early postoperative period.

OUTCOME

The outcome in these infants depends on the presence and degree of prematurity, associated anomalies, and the loss of bowel length due to atresia or gut infarction from mesenteric vascular compromise.

Neonates with omphalocele have a high (50–60 percent) incidence of associated anomalies and chromosome abnormalities that precludes long-term survival in 20–30 percent of cases. Severe cardiac defects are particularly troublesome in these infants. Modern neonatal intensive care, improved ventilator management, and intravenous nutrition have undoubtedly been responsible for the continued improvement in survival for these critically ill infants.

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31

Gastroschisis

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HISTORY

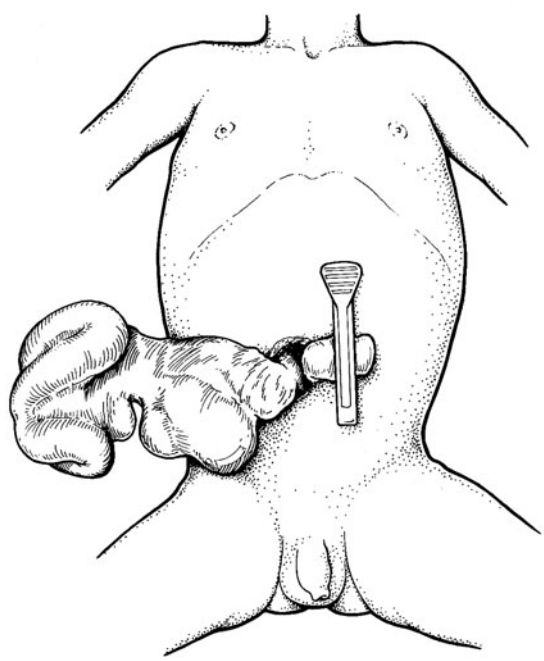
The first successful surgical repair of gastroschisis was performed by Watkins in 1943. Although there were improvements in the perioperative management and surgical procedures for gastroschisis, the mortality remained significant and was reported to be as high as 90 percent. Two major advances occurred in the late 1960s that led to a dramatic improvement in the survival of infants with gastroschisis. In 1967, Schuster et al. described a technique of staged reduction of the herniated bowel and abdominal closure for patients in whom primary closure was not possible. The second major advance was the evolution of intravenous nutrition, which, remarkably, allowed for growth and development during the prolonged period that these infants would not tolerate enteral feeding.

Over the past three decades, the outcome for infants with gastroschisis has dramatically improved. The survival rate is now greater than 90 percent.

PRINCIPLES AND JUSTIFICATION

Gastroschisis is an anterior abdominal wall defect that occurs in utero, through which there is herniation of intra-abdominal viscera into the amniotic sac. It is thought to result from a defect at the site of involution of the second (right) umbilical vein. This anomaly is accompanied by nonrotation of the bowel and an increased incidence of intestinal abnormalities, including atresia, perforation, and infarction, resulting from midgut volvulus or vascular thrombosis. The incidence of gastroschisis is approximately 1 in 4000–6000 live births.

Most infants with gastroschisis are born prematurely (35–37 weeks' gestation), weighing 2000–2500 g. The defect is almost always to the right of the umbilicus and generally measures 2–3 cm in diameter. All or a portion of the midgut is usually herniated through the defect. In addition, the stomach, urinary bladder, and, in females, the fallopian tubes and ovaries may also be extracelomic.



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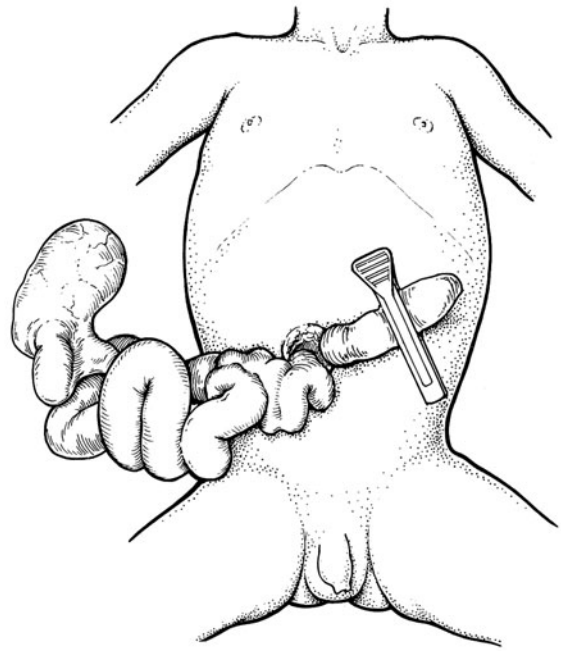
2 The intestine is foreshortened, edematous, and generally has a fibrin coating. Atresia of the small and large intestine occurs more often (approximately 14 percent) than in patients with omphalocele (1 percent). The most striking difference in appearance between omphalocele and gastroschisis is the absence of a sac or membrane covering the herniated contents in gastroschisis. Continuous contact of the herniated contents with amniotic fluid has been proposed as the reason for the thickened, foreshortened, and edematous bowel. There is also increasing evidence that the fascial defect begins to decrease in diameter near the end of the third trimester, which could lead to venous congestion of the midgut and contribute to the abnormal appearance of the bowel at birth. Therefore, serial fetal ultrasonography in the third trimester is warranted to follow the appearance of the bowel. Evidence of bowel wall thickening and dilatation can be an indication for early delivery. However, routine Cesarian section is not recommended for infants with gastroschisis.



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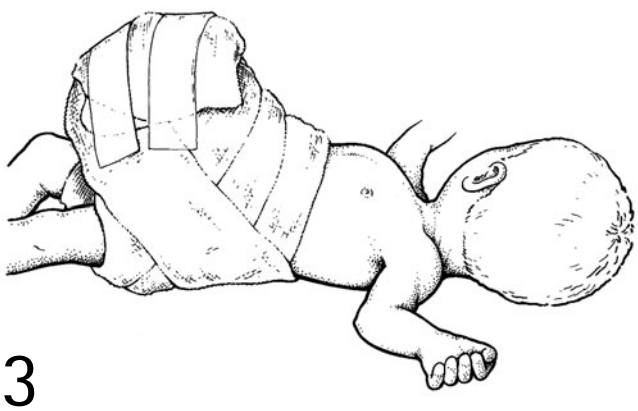
PREOPERATIVE ASSESSMENT AND PREPARATION

Gastroschisis requires prompt surgical intervention. Delays in surgical management should only be incurred as a result of transport of the infant to a pediatric surgical center or the need for prolonged preoperative stabilization. The evaluation of other life-threatening anomalies, which are rare, may delay surgery. Appropriate preoperative preparation is essential to ensure a good outcome.



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3 Maintenance of the infant's temperature within the normal range is critical because heat loss from the exposed herniated contents can be significant. There are numerous methods to minimize this heat loss depending on what supplies are available in the delivery room or intensive care nursery. Damp, warm gauze sponges or a damp, warm roll of gauze wrapped around the intestine followed by one or two dry gauze rolls wrapped around the patient's abdomen create an appropriate environment. The rolls of gauze help to stabilize the bowel and, therefore, diminish the risk of compromising its blood supply at the fascial ring. Thin plastic wrap or a plastic bag containing the baby from the chest to the feet has also been used to maintain normal body temperature and diminish evaporative fluid loss. The infant should be in a warming isolette or under an overhead radiant warmer to help maintain normothermia.

Most infants with gastroschisis are dehydrated at birth and require at least 125 percent of normal maintenance fluids to regain normovolemia. Eventually, almost all infants with gastroschisis will require central venous access. However, attempts are made to delay this procedure until 2–3 days after surgery to decrease the risk of catheter contamination by bacterial translocation or other sources. It is appropriate to give broad-spectrum antibiotics during the perioperative period because of exposure of the bowel and peritoneal cavity to bacterial contamination at the time of birth. A nasogastric tube placed at the time of birth is necessary for gastrointestinal decompression because of the bowel inflammation and resulting ileus.

Thus, before surgery, the infant with gastroschisis should be normothermic, hemodynamically stable, and have normal serum electrolytes following adequate fluid resuscitation.

Anesthesia

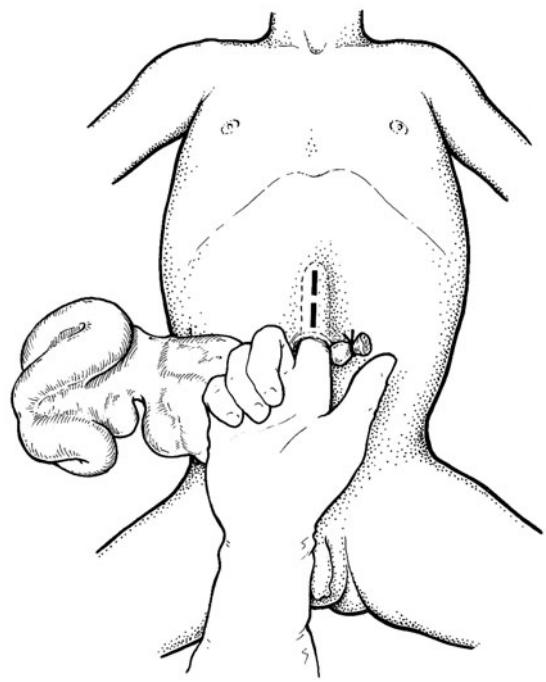
General anesthesia is required for appropriate operative management of gastroschisis. The choice of anesthetic agents should be made by the anesthetist, but two points should be emphasized: first, muscle paralysis is useful in optimizing the chances for complete reduction of the herniated bowel and primary abdominal wall closure, and second, nitrous oxide should not be used as it diffuses into the lumen of the bowel causing distension and compromising the likely success of primary abdominal wall repair.

OPERATION

The operation should be performed under conditions that maintain normothermia. Several methods exist to accomplish this goal. An overhead radiant warmer, warming lights, or a warming blanket should be used to maintain the infant's temperature in the normal range during the procedure. Raising the temperature in the operating room may also be necessary. After the induction of general anesthesia, the dressing previously placed over the herniated contents should be removed. The bowel should be handled with sterile gloves. The umbilical cord, which has usually been left long, should be clamped 2–3 cm above the abdominal wall and the excess cord then removed. Holding the bowel and clamp on the umbilical cord in one hand, the bowel should be prepared using gauze sponges soaked in a 50:50 mixture of povidone-iodine solution and saline. The antiseptic solution must be warm to the touch in an effort to minimize heat loss. After gently washing the bowel and the anterior and lateral abdominal wall, drapes are appropriately placed and the herniated contents are laid on the drapes. The surgeon should then scrub and put on gown and gloves. Next the herniated intestine should be carefully inspected for areas of perforation or sites of atresia, although no effort should be made to dissect matted loops of intestine.

4 It is nearly always necessary to extend the abdominal wall defect to facilitate reduction of the herniated bowel. This is generally done by extending the defect superiorly in the midline by 1–3 cm. Extending the incision caudally is not recommended because the urinary bladder is in close proximity to the inferior aspect of the abdominal wall defect. The length of this incision depends on the size of the original defect and the bulkiness of the herniated bowel.

The herniated intestine is reduced as much as possible, distributing the bowel to all quadrants of the peritoneal cavity. Two techniques have been described to facilitate complete bowel reduction and abdominal wall closure: (1) stretching of the anterior abdominal wall, and (2) 'milking' the intestinal contents into the stomach where they can be aspirated through the nasogastric tube. Although gentle stretching of the anterior abdominal wall can be useful, the authors are opposed to vigorous stretching. This maneuver can lead to rectus muscle hemorrhage and abdominal wall edema, producing a non-compliant, firm anterior abdominal wall, resulting in ventilation difficulties and wound-related problems. Nor do the authors advocate manipulating the intestine to 'milk' the intestinal contents into the stomach, believing that this can cause further damage to the bowel wall resulting in increased bowel wall thickening and additional delay in bowel recovery.



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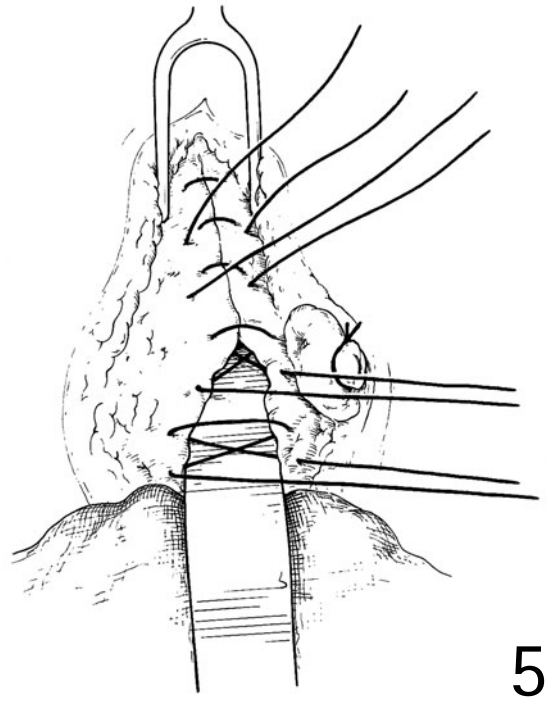
5 If reduction of the herniated intestine is successful, the abdominal wall is assessed for primary closure. If it can be closed without undue tension, 3/0 absorbable, monofilament sutures are used. These sutures are placed in a figure-of-eight, as this results in fewer knots. When all the sutures have been placed, they are tied in sequence with a thin, malleable retractor initially underneath the fascia to prevent a loop of intestine from becoming entrapped under the sutures.



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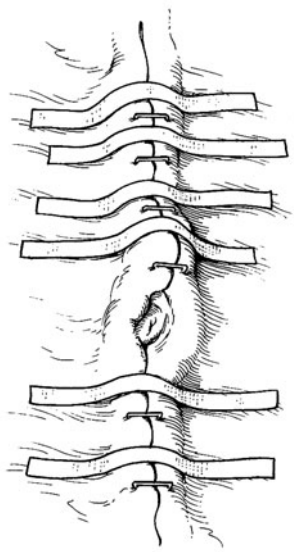

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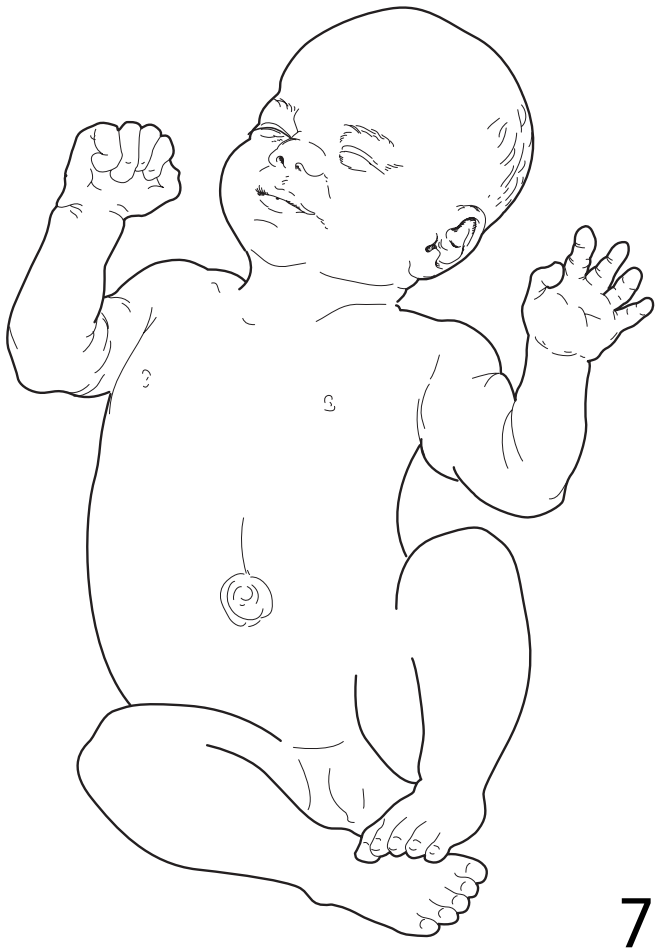
6 When the fascia has been closed, the skin edges are approximated using a few skin staples and sufficient sterile skin closure strips, allowing distribution of skin tension over a wider surface area and thus reducing the likelihood of skin disruption.



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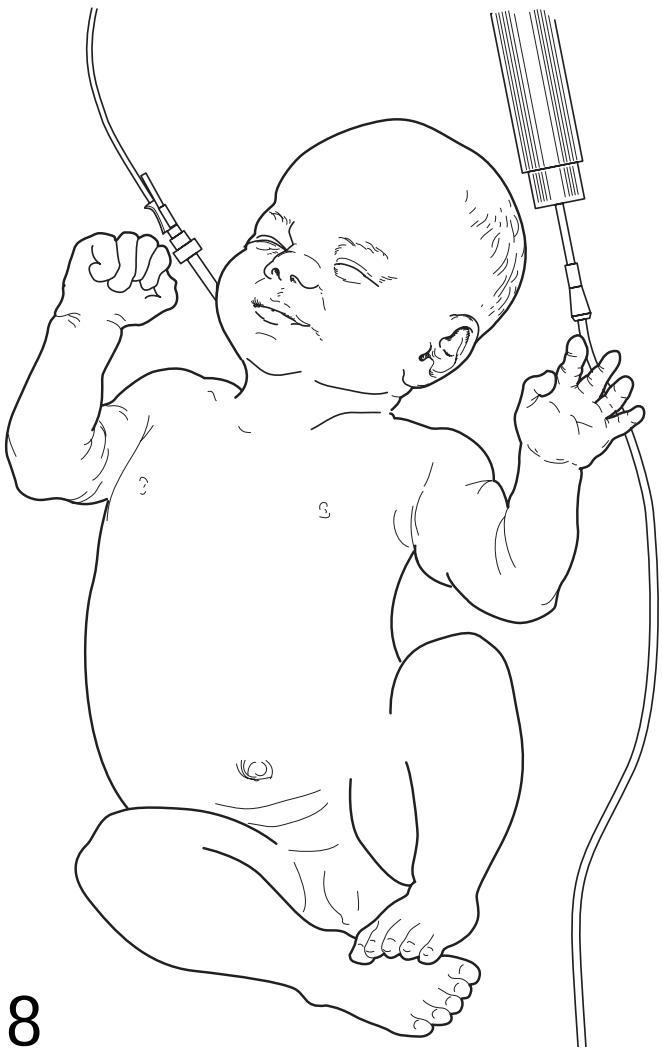
7 Appearance 1 week after primary closure. Note the umbilicus was retained.



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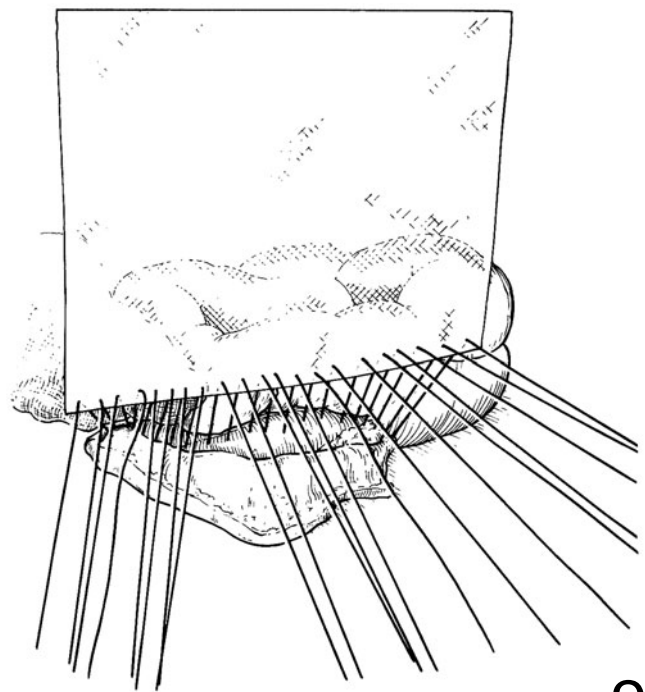
8 Appearance 3 weeks after primary closure.

About 60–70 percent of infants with gastroschisis can be operatively treated in this way without creating undue intra-abdominal pressure or tension in the abdominal wall closure. It is best to avoid high intra-abdominal pressure and excessive suture line tension. This can result in abdominal compartment syndrome, possibly leading to intestinal necrosis, renal hypoperfusion, and difficulty in ventilation, as well as wound disruption. Intragastric and bladder pressure monitoring has been used by some pediatric surgeons to determine intra-abdominal pressure. These two measurements are used as a guide to monitor intra-abdominal pressure during primary or staged closure of gastroschisis. The goal of therapy, to maintain intra-abdominal pressure below 20 mmHg, is based on prior studies showing that higher pressures compromise intra-abdominal organ perfusion.

9 For patients in whom complete reduction of the herniated bowel and abdominal wall closure are not possible or appropriate, the staged reduction technique described by Schuster in 1967 has proved to be very useful. Reinforced Silastic sheeting (0.8–1.0 mm thick) is sutured to the fascial edges. This is accomplished with interrupted 3/0 silk mattress sutures.



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10

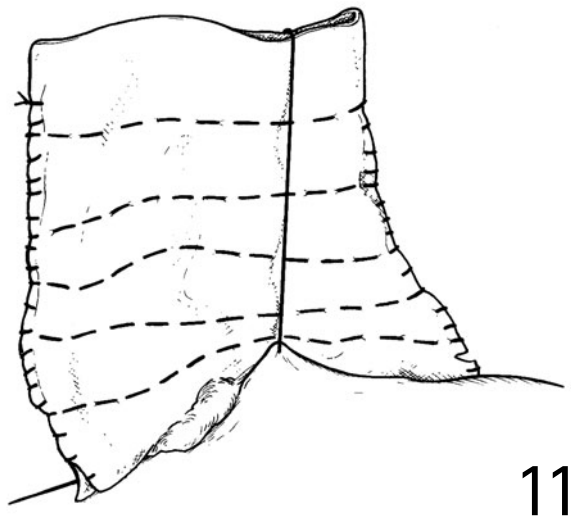
10 The cephalad and caudad vertical edges of the silo are constructed with running 3/0 monofilament sutures. Before closing the top of the silo, as much of the bowel as possible is reduced into the peritoneal cavity by manual compression within the sac while avoiding excessive intra-abdominal pressure. The top of the sac is oversewn with a 3/0 monofilament suture placed in a running horizontal mattress fashion.

The Silastic sac is covered with povidone-iodine ointment followed by dry roll gauze to act as a protective dressing and provide support to the Silastic sac at the fascial level.

POSTOPERATIVE CARE

Most patients with gastroschisis require parenteral nutrition to provide the necessary calories intravenously while awaiting bowel reduction and recovery of bowel function. Until recently, this was accomplished via a cuffed Silastic central venous catheter. An alternative is the placement of a small Silastic catheter inserted via a peripheral vein and threaded centrally (commonly referred to as a PICC line). Parenteral nutrition is typically required for 2–6 weeks after the operation while awaiting the return of intestinal function. Nasogastric decompression is necessary until there is evidence of bowel function. Broad-spectrum antibiotics are generally continued for a minimum of 5 days. Those infants who undergo the staged approach require a longer period of antibiotic treatment (usually until 1–2 days after the sac has been removed).

11 The staged reduction technique requires daily reduction of the herniated intestine within the silo. The target for completely reducing the bowel, removing the Silastic sac, and closing the abdominal wall is within 1 week of age. Any delay beyond 1 week substantially increases the risk of fascial infection, tearing away of the Silastic sheeting from the anterior abdominal wall, and failure of the technique. This is also true when using the spring-loaded, preformed silo. Daily reduction of the intestinal contents within the sac can be accomplished in the neonatal intensive care unit using sedation and sterile technique. Each time the procedure is performed, the sac and anterior abdominal wall are prepared with warm povidone-iodine solution before the reduction, and povidone-iodine ointment is applied followed by roll gauze after the procedure. Some pediatric surgeons do not use this approach when using the spring-loaded silo, but simply leave it exposed. General anesthesia is not necessary. When the herniated bowel has been successfully reduced into the peritoneal cavity and the fascial edges brought to within 1 cm of each other, the infant is ready for removal of the sac and primary abdominal wall closure in the operating room under general anesthesia. An alternative 'gentle touch' technique has recently been described. This method involves bedside placement of a spring-loaded silo, followed by passive reduction of the herniated contents into the abdomen via gravity. The infant is kept paralyzed or sedated and intubated with assisted ventilation during the passive reduction. The author states that this process typically takes 4–5 days. Subsequently, closure of the abdominal wall defect is performed.



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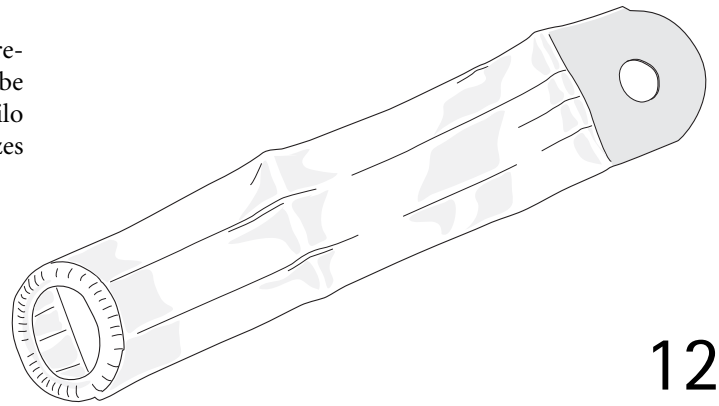
12 An alternative method is the placement of a preformed, spring-loaded silo at the bedside. This can be accomplished without general anesthesia. The preformed silo comes in different diameters to accommodate different sizes of defect and bulkiness of the herniated contents.



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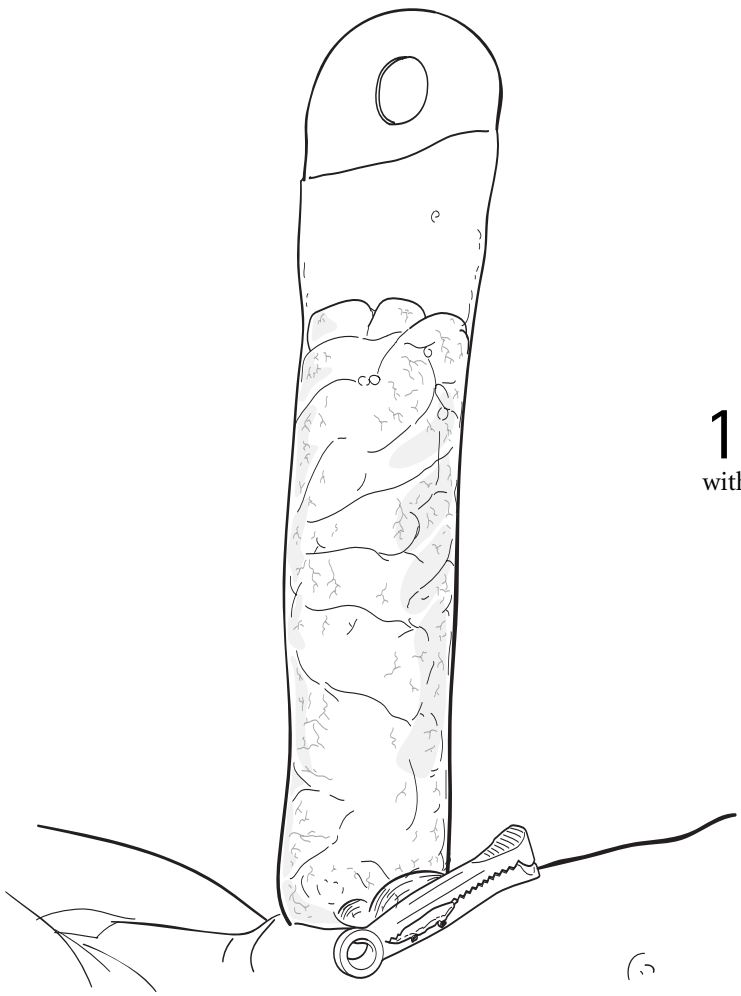

13 The spring-loaded silo is placed underneath the fascial defect after the herniated bowel is placed within it.

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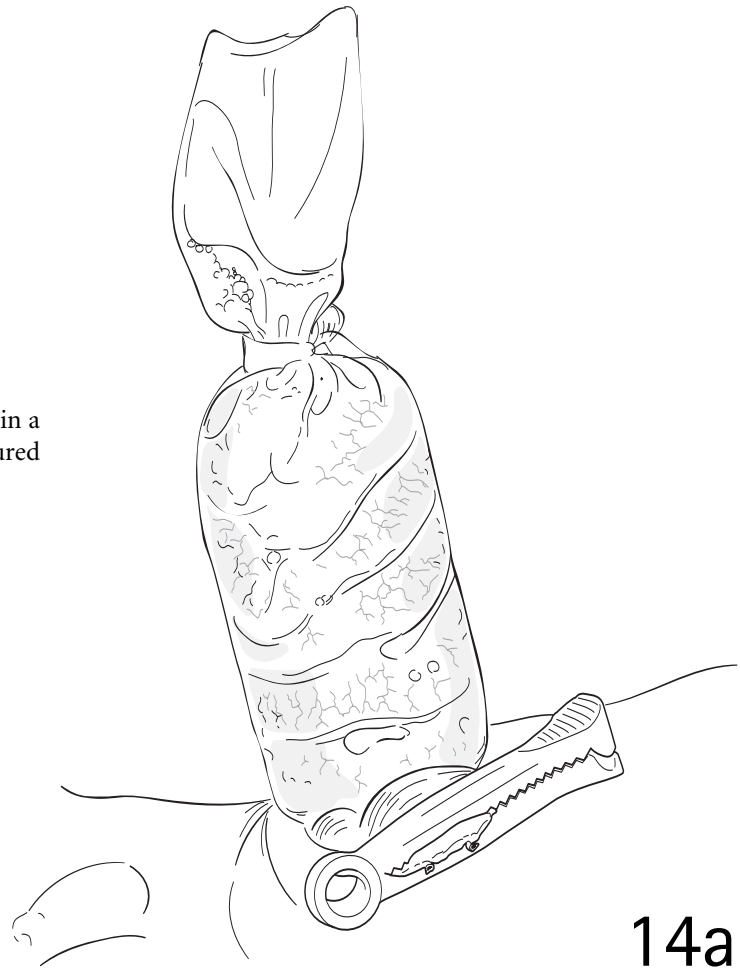
14a,b Reduction of the bowel is accomplished in a similar fashion to that used for the sutured silo.



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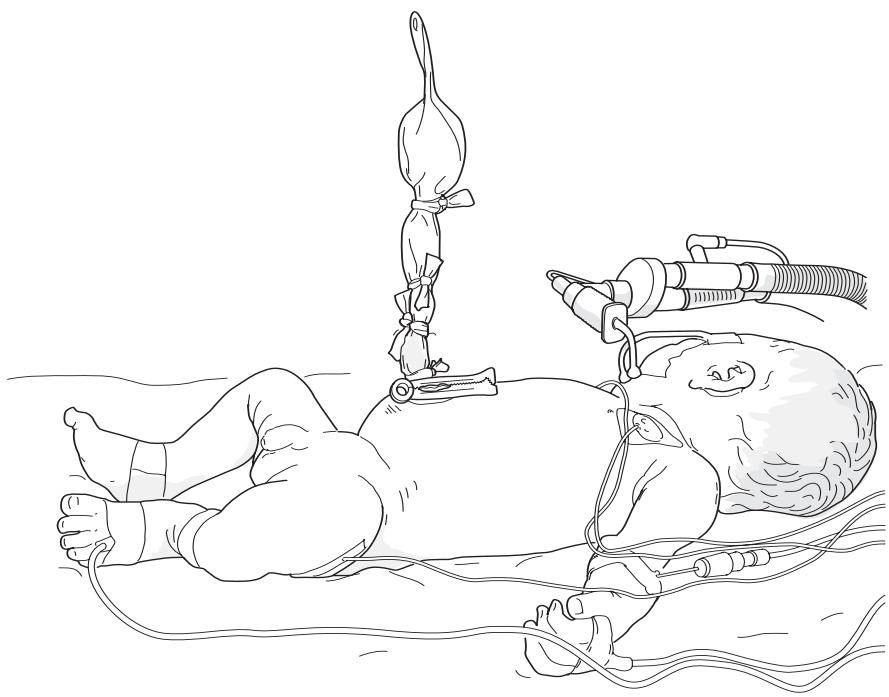
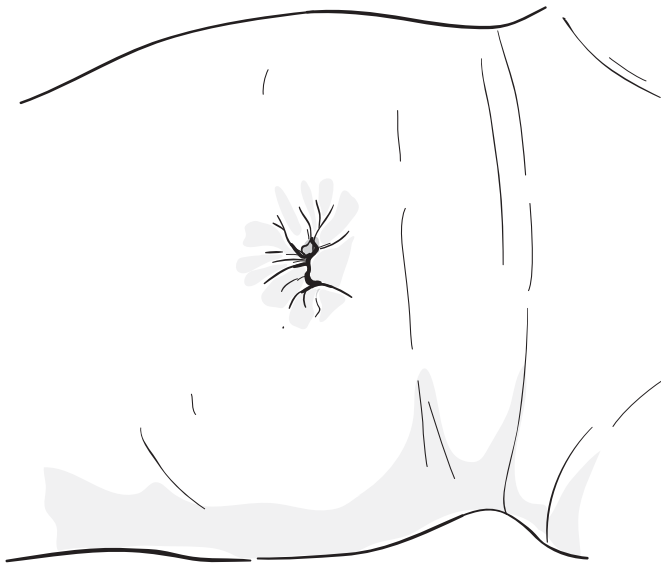


14b

15 Appearance after fascial closure and purse-string technique of skin closure. Umbilicus was removed.

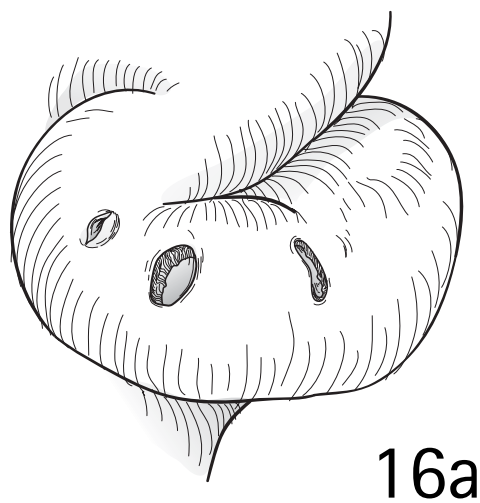


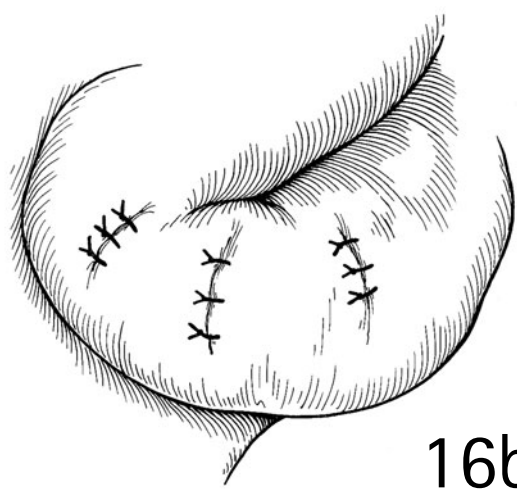
Once there is evidence of gastrointestinal function, enteral feeding can be introduced and gradually progressed using breast-milk or a low-residue elemental-type formula with appropriate caloric intake. In the past, enteral feeding in these patients was generally delayed for at least 4–6 weeks after surgery, as it was thought that early feeding could lead to an increased risk of developing complications. However, this approach has not been supported with clinical evidence and, more recently, enteral feedings have been started as early as 10–14 days postoperatively, with no increase in adverse outcomes.

COMPLICATIONS

16a,b Complications in infants with gastroschisis are generally related to the gastrointestinal tract or the abdominal wall closure. As noted earlier, in utero complications from intestinal atresia or perforation can occur. Intestinal perforation can be managed in one of several ways, depending on the specific circumstances. The options at the time of birth include suture closure, resection of the site of perforation with oversewing of the two ends of the bowel (i.e., creating 'intestinal atresia'), or creation of a stoma if primary abdominal wall closure can be accomplished.

It is generally not recommended to attempt a bowel resection or anastomosis because of the marked thickening and inflammation of the intestinal wall. Intestinal atresia can be managed by the creation of a stoma if primary abdominal wall closure is possible or by leaving the atresia in situ if staged reduction is undertaken. A stoma can be created at the time of removal of the Silastic sac and primary abdominal wall closure. A devastating complication can be partial or complete necrosis of the midgut as a result of excessive intra-abdominal pressure or kinking of the blood supply to the bowel at the time of reduction of the herniated bowel. This complication may lead to the death of the patient or to short bowel syndrome.





Additional complications associated with the abdominal wall closure are wound dehiscence and intestinal–cutaneous fistula formation. These complications are also often associated with excessive intra-abdominal pressure. It is preferable to use the staged reduction approach when primary abdominal wall closure might result in excessive intra-abdominal pressure.

A delayed complication is the development of necrotizing enterocolitis. The incidence of necrotizing enterocolitis in patients with gastroschisis has been reported to be as high as 20 percent. It generally has a delayed onset, usually 3–6 weeks after birth. The causes remain unknown, but associations have been made with total parenteral nutrition (TPN)-induced cholestasis and delay in feeding. Necrotizing enterocolitis associated with gastroschisis can be mild or severe and can involve a significant portion of the bowel resulting in a high mortality.

Finally, sepsis, resulting from intra-abdominal or wound infections, and central line infections are additional causes of morbidity in the gastroschisis patient.

OUTCOME

The availability of neonatal intensive care units, parenteral nutrition, and the technique of staged reduction have resulted in significant improvement in the outcome for infants with gastroschisis over the past four decades. The survival of infants with gastroschisis has exceeded 90 percent. Morbidity should be relatively low if attention is paid to the details of the surgical correction. In the authors' experience, infants successfully treated for gastroschisis do not have significant complications during later infancy and childhood.

In addition to the marked improvement in survival, the lengths of time to initiation of feedings and hospital discharge have been significantly shortened. Whereas hospitalization usually averaged approximately 6 weeks, the average is now around 4 weeks.

FURTHER READING

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32

Abdominal surgery: general principles of access

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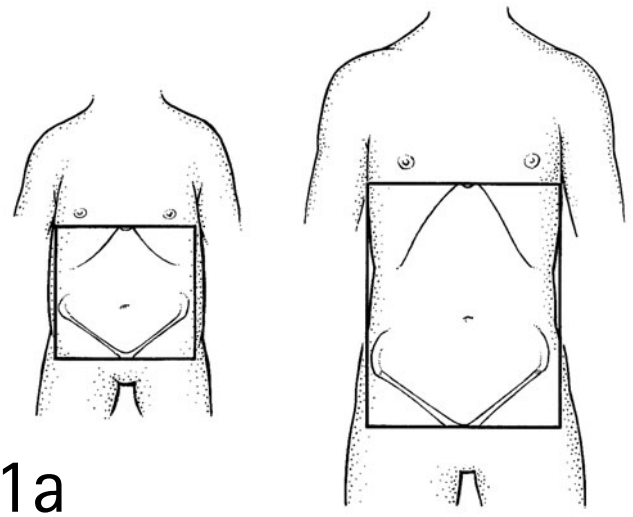
PRINCIPLES AND JUSTIFICATION

Anatomic considerations

1a,b There are important anatomic differences between the abdomen of the newborn infant and that of the older child or adult. The following are characteristics of the infant.

- The shape of the abdomen is a square compared with the rectangular shape of the older child. A transverse mid-abdominal incision in an infant will therefore provide access to the whole peritoneal cavity, with the possible exception of the pelvis.
- The compliant ribcage and wide subcostal angle facilitate access to the upper abdominal organs and diaphragm.
- The rectus muscle is wider and extends further laterally.
- The liver is relatively large and extends from below the costal margin on the left down to the right lower quadrant.
- The umbilicus is relatively low and nearer the pubic symphysis, and the bladder extends up to the umbilicus. There is limited space for an incision below the umbilicus in the neonate.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1b

Site and size of incision

The incision must be planned to provide optimal exposure for the surgeon whilst minimizing abdominal muscle damage. In a modern environment it is unusual to operate on a patient who is not stable, and the surgeon is able to place greater emphasis on the cosmetic aspects of the incision than was possible in the past. An incision that has been correctly sited does not need to be unduly long. Nonetheless, common reasons for a surgeon to experience difficulty are inadequate exposure due to an incision that is too small, poor retraction by the assistant, poor lighting, and inadequate muscle relaxation.

PREOPERATIVE

Anesthesia

General anesthesia with muscle relaxation and endotracheal intubation are required for abdominal operations on children.

Skin cleansing

A variety of topical preparations are in use for removing potential pathogens from the skin in the operative field. Alcohol-based solutions evaporate rapidly, promoting heat

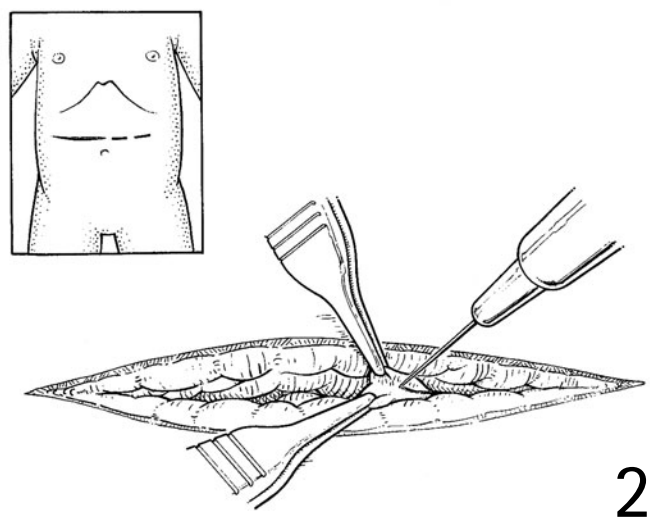
OPERATIONS

Transverse abdominal incision

The muscle-cutting transverse upper abdominal incision is suitable for most operations in infants, except when access is required to the distal colon and rectum. The incision may be limited to one side of the abdomen or can be extended across the midline, dividing both rectus muscles. For some procedures, such as reduction of an intussusception, a transverse incision lateral to the right rectus muscle will suffice.

POSITION OF PATIENT AND INCISION

2 The patient lies supine. The skin incision starts in the midline, 1–2 cm above the umbilicus, and extends laterally across the rectus muscle. The subcutaneous fat is lifted with two pairs of fine-toothed forceps (one held by the surgeon and one by the assistant) and cut with diathermy to minimize blood loss, particularly in the small infant. Bleeding from small vessels in the skin edge will stop spontaneously with compression; larger vessels are touch-coagulated with needle-point diathermy or picked up accurately with fine-toothed forceps and coagulated, taking care not to damage the skin.



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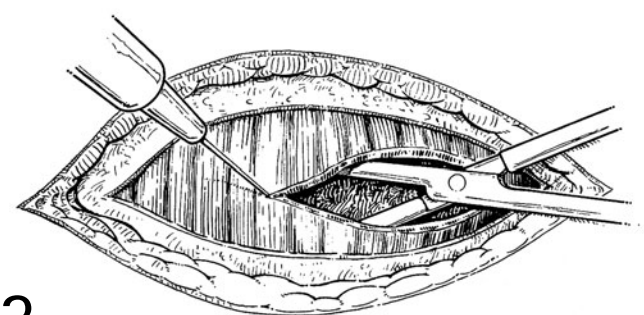


3

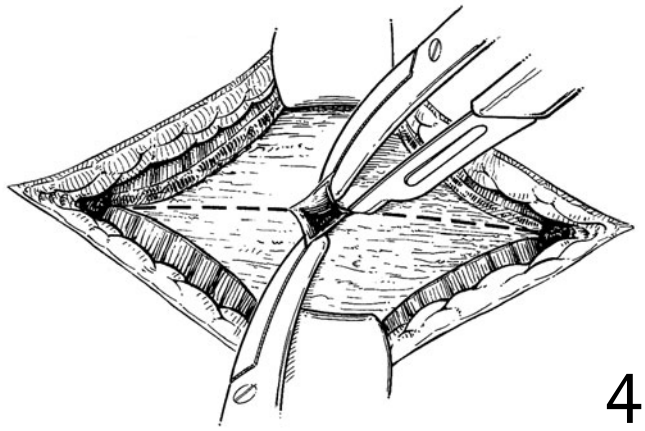
3 This exposes the anterior rectus sheath, which is incised transversely. A pair of artery forceps inserted deep to the rectus muscle is used to lift the muscle off the underlying fascia while cutting it with the diathermy. The vessels are identified and cauterized before being cut.

loss, and iodine-based preparations may irritate the skin. Aqueous chlorhexidine (Hibitane) has none of these potential disadvantages and has effective antibacterial activity. In newborn infants, the solution should be warmed. Excess fluid must not be used, as this may run under the patient, resulting in chemical or electrical burns, as well as promoting heat loss.

Draping

Sterile towels or drapes are used to provide a sterile environment around the incision. These are covered with a large, sterile, adhesive plastic sheet, which stabilizes the towels and also helps to keep the infant warm by reducing heat loss from the skin and by keeping the infant dry.

4 The posterior rectus sheath is picked up with two pairs of artery forceps placed about 1 cm apart, and a small incision is made between them, taking care not to damage the underlying bowel. Once air has entered the peritoneal cavity, the bowel falls away, unless it is distended or there are adhesions, and the incision can be completed safely using scissors. In the neonate, the transversalis muscle is well developed and vascular and may be divided using diathermy.



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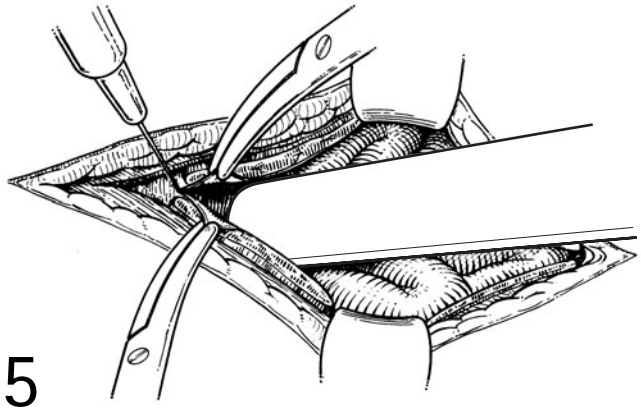
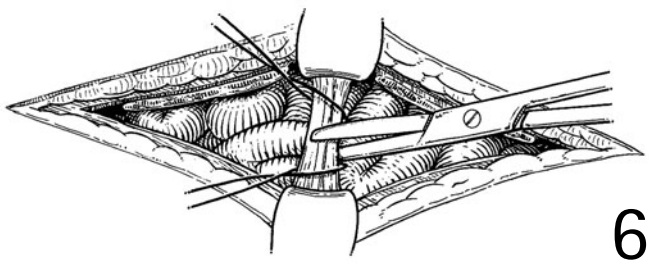


5

5 The incision is extended laterally by lifting the abdominal wall muscles with artery forceps applied to the upper and lower edges of the posterior rectus sheath, and cutting the external oblique, internal oblique, and transversalis muscle layers with the diathermy, while protecting the underlying bowel.

If the bowel is distended, it should be protected with a flat retractor or a swab. Adhesions must be carefully dissected off the peritoneum; when a pre-existing incision is being re-opened, it is advisable to begin the incision beyond the end of the scar so that the peritoneum is opened where it is 'normal' and underlying adhesions are unlikely to be present.

6 The incision may also be extended medially across the midline. The falciform ligament is cut with scissors and the ligamentum teres is ligated with absorbable ligatures and divided.



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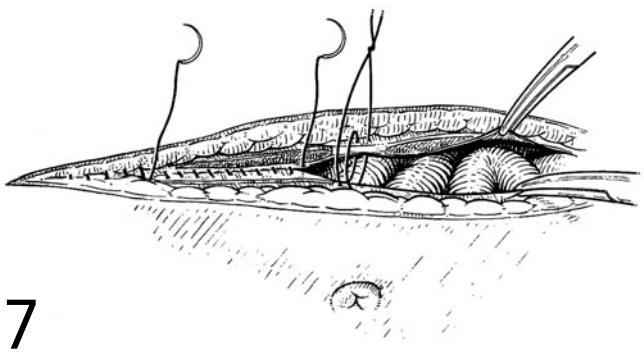


7

WOUND CLOSURE

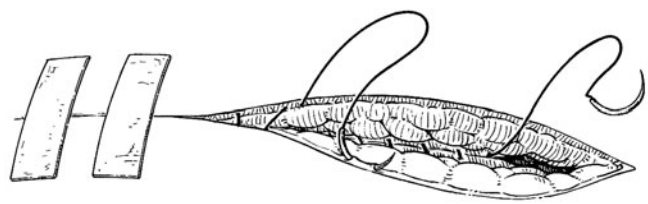
7 The abdomen is closed in layers using absorbable sutures (4/0 or 3/0 sutures on a round-bodied needle for infants and 2/0 for children).

The margins of the peritoneum and transversalis muscle or fascia are grasped with artery forceps, elevated, and approximated with a continuous absorbable suture. In the older child, if the incision divides both rectus sheaths, closure of the midline (linea alba) is reinforced with a single figure-of-eight suture. No sutures are placed in the rectus muscle, which is adherent to the rectus sheath and does not retract.

The anterior rectus sheath is also repaired using a continuous suture. Lateral to the rectus sheath, the internal and external oblique muscles are repaired in separate layers.

When there is doubt about the ability of the abdominal wall muscles to retain sutures, as in very premature or malnourished infants, the abdomen may be closed in a single layer with sutures incorporating all the muscles and the peritoneum. The skin is closed as a separate layer. The same technique may be used when a pre-existing incision is closed.

8 Before closing the skin, local anesthetic is infiltrated into the layers of the abdominal wall surrounding the incision. In most infants, the subcutaneous fat falls together without the need for sutures, and the skin is approximated with adhesive strips. In older children, the deep fascia is repaired with 4/0 or 3/0 absorbable sutures. This takes the tension off the skin, which is approximated with adhesive strips or a subcuticular 5/0 absorbable suture. The incision is covered with a dressing, mainly to allay the anxiety of the child.



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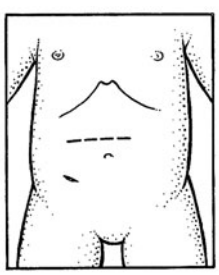

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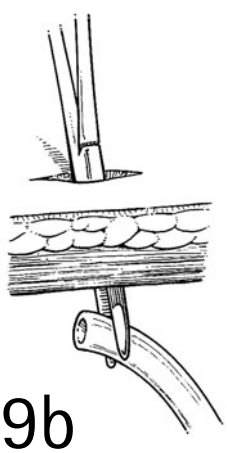

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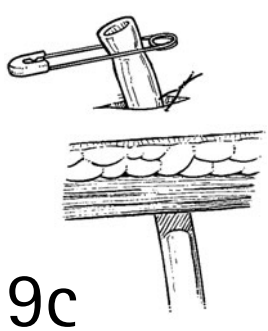


9c

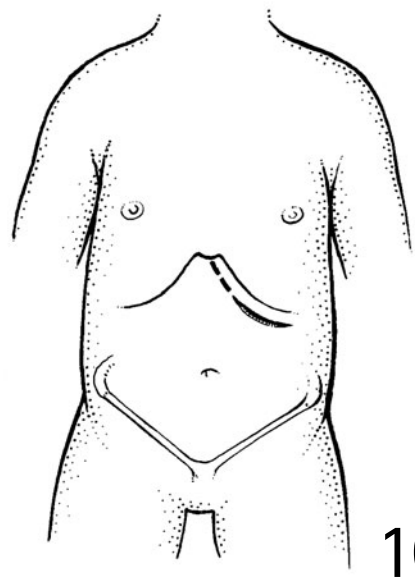
9a–c If an abdominal drain is required, it must be placed through the abdominal wall before closing the incision. At a suitable site, depending on the area to be drained, a short transverse incision is made through the skin and external oblique muscle.

With a hand in the peritoneal cavity to protect the bowel, an artery forceps is pushed through the abdominal wall into the peritoneal cavity to grasp the drain. The type of drain will depend on the specific situation; a Penrose drain suffices for most situations.

The drain is pulled out through the abdominal wall and sutured to the skin. A safety pin is placed through the drain to prevent it from slipping into the peritoneal cavity.

Subcostal incision

The left subcostal incision is useful for access to the diaphragm (congenital diaphragmatic hernia), esophagus (fundoplication), or spleen. On the right, the incision is used for operations on the gallbladder and bile ducts; if the liver is to be exposed, the incision is extended to the left subcostal region.



10

10 Depending on the age of the patient, the skin incision is made 1.5–3 cm below and parallel to the costal margin. It should not overlie the costal margin when sutured. In the midline, the incision may be extended cranially to the xiphisternum for better access to the esophagus or diaphragm.

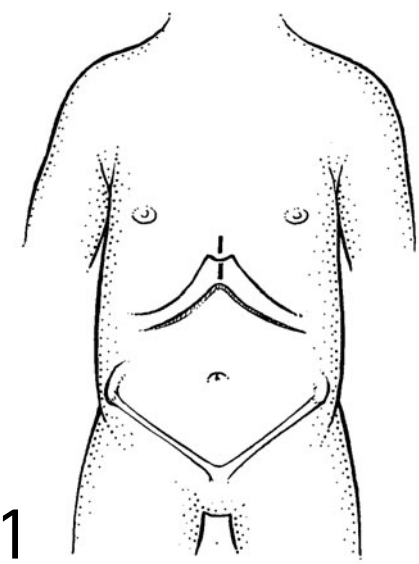
The layers to be divided are the same as for a transverse incision, but in an oblique direction.

WOUND CLOSURE

Closure is as for a transverse incision. If a gastrostomy tube has been inserted, ideally it should be brought out through a separate incision. On occasion, the most direct route is through the main incision; in this case the incision is closed in two halves, beginning on either side of the tube. On each side of the gastrostomy the stomach must be securely anchored to the abdominal wall with a non-absorbable suture.

Bilateral subcostal (rooftop) incision

This is the preferred exposure for surgery of the liver and portal structures.



11

11 For initial exploration of the liver, a right subcostal incision is made; this is then extended to the left with a curve across the midline. If necessary, a further extension may be made cranially in the midline to enter the mediastinum.

WOUND CLOSURE

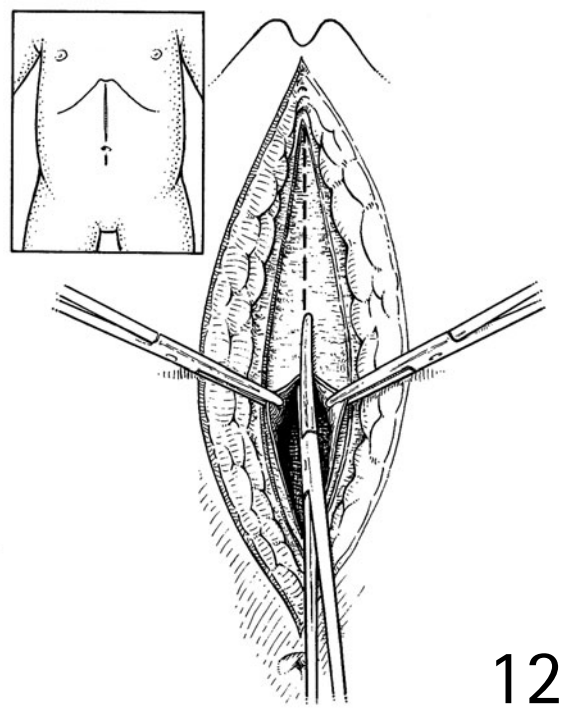
Closure is as for subcostal and midline incisions. A single reinforcing suture is placed in the midline prior to closing the peritoneum.

Midline abdominal incision

The access offered by the upper abdominal midline incision in the infant is restricted by the relatively large liver, but this disadvantage is offset by the wide costal angle and the cosmetic scar. The incision is useful for pyloromyotomy, gastrostomy, and fundoplication. In the older child, this is the incision of choice for blunt abdominal trauma.

12 The skin is incised from xiphisternum to umbilicus. (For pyloromyotomy, a shorter incision is adequate.) If it is necessary to extend the incision caudally, it should be taken straight through the umbilicus and not around it. This gives a superior cosmetic result, and the risk of infection is not increased if the umbilicus has been properly cleaned. The subcutaneous tissues are cut with the diathermy down to the fascia.

A short incision is made in the linea alba using a scalpel, and the falciform ligament is entered. The edges of the incision are grasped with artery forceps and elevated. A plane is developed deep to the linea alba, which is incised with scissors. Near the umbilicus, the peritoneum fuses with the linea alba, and the peritoneal cavity will be entered as the incision is extended caudally. The left or right fold of the faciform ligament is incised, depending on the exposure required; if necessary, the ligamentum teres is ligated and divided.



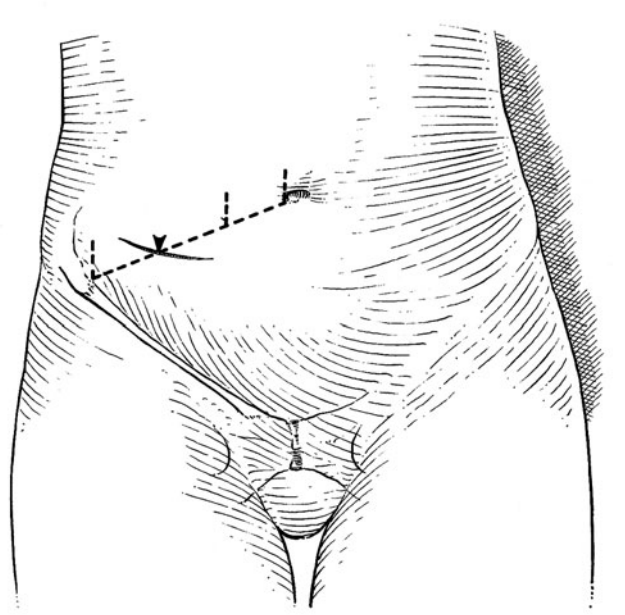
12

WOUND CLOSURE

The falciform ligament/peritoneum may be repaired, but this is not essential. The linea alba is approximated with a continuous strong suture of slowly absorbable material such as polydioxanone (PDS), or with a nylon suture (3/0 for infants, 2/0 or 0 gauge for children). The knot at each end should be buried to avoid an unsightly nodule. A subcutaneous suture may be required. The skin is closed with adhesive strips or a 5/0 continuous subcuticular absorbable suture.

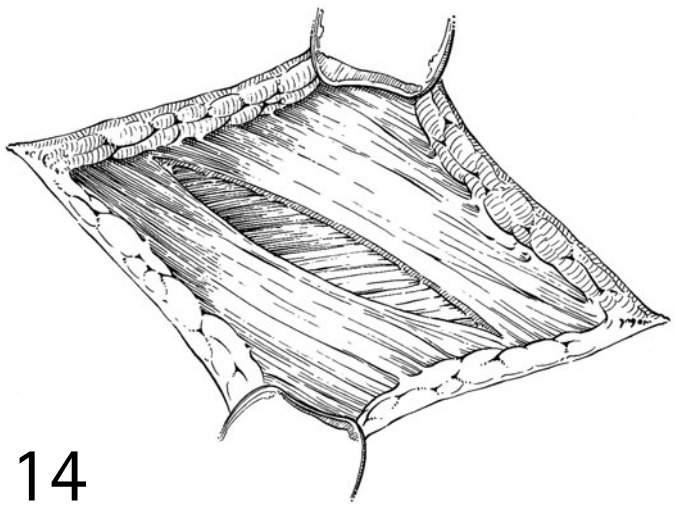
Grid-iron incision

The modified McBurney incision is the ideal incision for acute appendectomy in childhood.



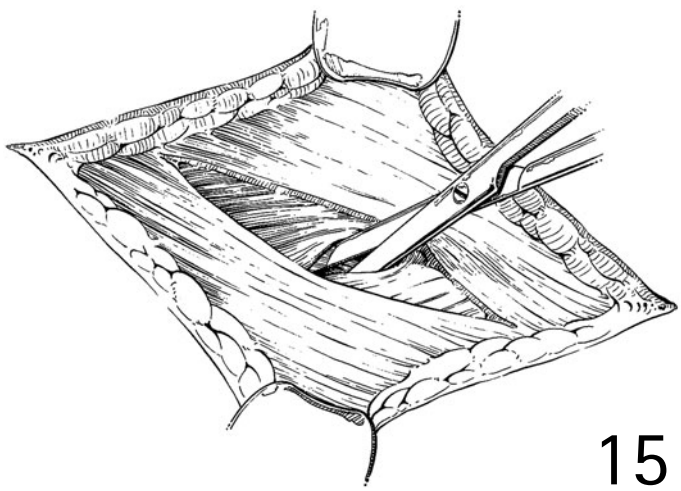
13

13 The traditional incision is centered over McBurney's point, which is two-thirds of the distance from the umbilicus to the right anterior superior iliac spine, and is aligned with the skin creases, which lie in a slightly oblique direction. It should be lateral to the rectus muscle, which is relatively broad in a child. A lower incision may be preferred for cosmetic reasons, but exposure of the appendix may be difficult if the incision is too low or too medial. If a mass can be palpated when the child is under anesthesia, this may influence the siting of the incision.



14

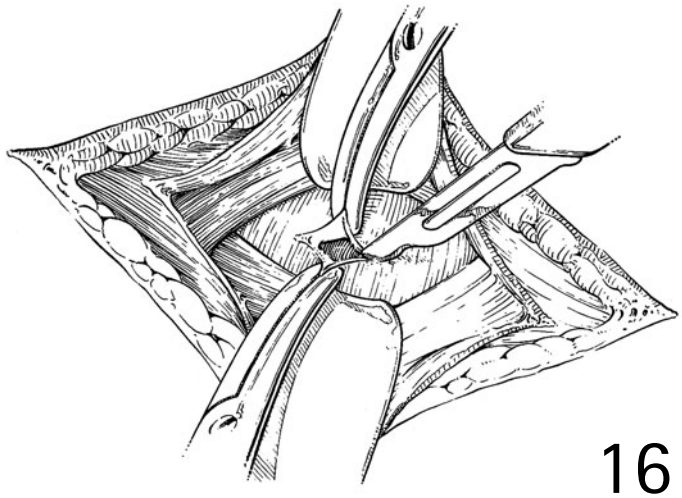
14 The subcutaneous tissues are divided with diathermy and swept aside with a swab to expose clearly the external oblique aponeurosis; this facilitates subsequent closure. The external oblique muscle is incised and then divided along the line of its fibers and separated from the underlying muscle.



15

15 The internal oblique muscle is opened by passing blunt scissors or artery forceps into the muscle and spreading it at right-angles to the direction of the fibers. Two Langenbeck retractors are inserted into the space and used to separate the fibers widely. The underlying transversus abdominis muscle and the fatty layer covering the peritoneum are opened in a similar fashion.

16 The peritoneum is grasped with two pairs of artery forceps, taking care to avoid the underlying bowel. The forceps are lifted and the peritoneum is incised transversely with a scalpel; the opening is enlarged using scissors.



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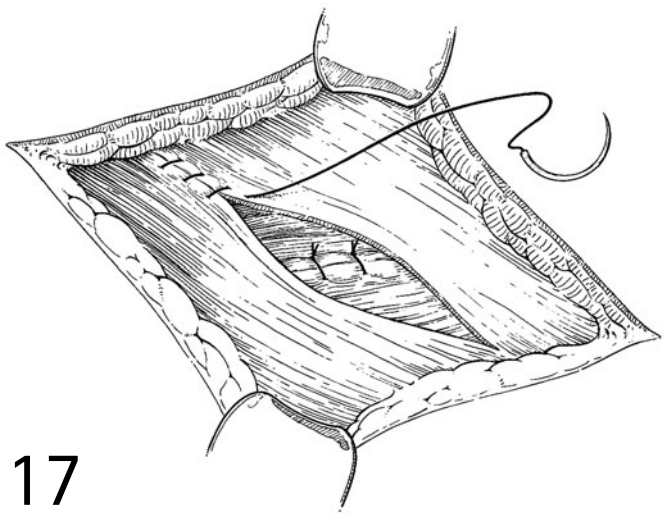


17

WOUND CLOSURE

17 The edges of the peritoneum are grasped with artery forceps and closed with a continuous absorbable suture.

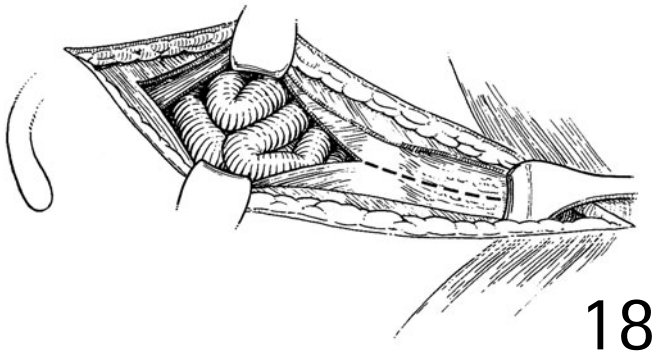
The fibers of the transversus and internal oblique muscles are closed as a single layer, using two or three interrupted sutures, which are tied loosely to avoid muscle ischemia. The external oblique muscle is closed with a continuous absorbable suture. The subcutaneous fat seldom requires sutures.

For skin closure, if adhesive strips alone are not adequate, a subcuticular suture of 5/0 absorbable material is used. In children, the skin is always closed after appendectomy, regardless of the degree of contamination; appropriate prophylactic antibiotic cover must be given.

EXTENDING THE INCISION

Circumstances may require the incision to be extended laterally; this is done by dividing the abdominal muscles in layers using the diathermy, as shown in Figure 5.

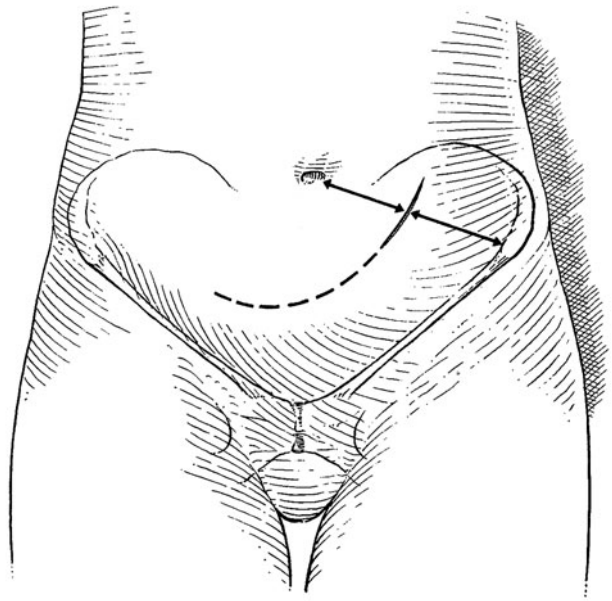
18 To extend the incision medially, the incision in the external oblique muscle is carried onto the anterior rectus sheath. The rectus muscle is retracted medially. The internal oblique and transversalis muscles are divided medially and this incision is extended to open the posterior rectus sheath and peritoneum. If necessary, the rectus muscle is also divided. The incision is closed in layers, as for a transverse abdominal incision.



18

Oblique muscle-cutting (Lanz) incision

In the left iliac fossa, this incision is used for colostomy formation. It may be extended medially as the 'hockey-stick' incision to provide access to the pelvis.



19

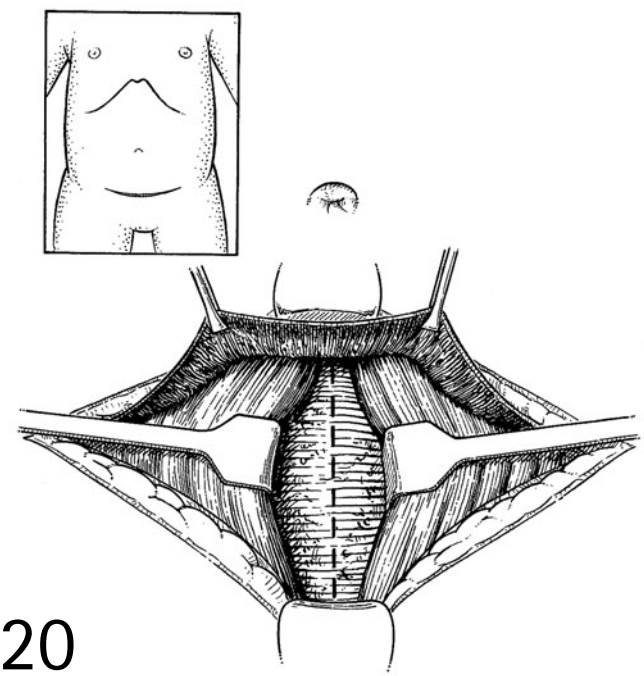
19 The skin is incised in an oblique direction at the midpoint of a line from the umbilicus to the anterior superior iliac spine. The external oblique muscle is incised along the line of its fibers, as for a grid-iron incision. The internal oblique and transverse muscles are cut obliquely in the same direction as the external oblique muscle, using the diathermy. For the 'hockey-stick' extension, the incision is continued medially, parallel to the skin creases. The rectus muscles and peritoneum are cut transversely.

WOUND CLOSURE

The incision is closed in layers, as for a transverse abdominal incision.

Pfannenstiel incision

This lower abdominal incision provides access to the pelvic organs, in particular the bladder, uterus, and ovaries, without dividing the rectus muscles.



20

20 The skin and subcutaneous tissues are incised transversely between the lateral borders of the two rectus muscles. The incision is slightly curved to follow the skin creases, and is centered about 2 cm above the pubic symphysis. The anterior rectus sheaths are divided transversely and dissected off the rectus muscles by blunt and sharp dissection, extending cranially well up to the umbilicus and caudally to the pubic symphysis. The rectus muscles are separated vertically in the midline and retracted laterally. The transversalis fascia and peritoneum are then opened vertically, taking care to avoid the bladder.

WOUND CLOSURE

The incision is closed in layers, as for a transverse upper abdominal incision. The rectus muscles are approximated in the midline with interrupted sutures.

COMPLICATIONS

The intestine is at risk of injury while the incision is being made, by being cauterized by the diathermy while incising the muscle layers, particularly in the newborn infant, or by being crushed by artery forceps or incised with the scalpel when the peritoneum is opened. During wound closure, the intestine is also at risk of injury from artery forceps applied to the edges of the incision or the peritoneal suture.

The risk of wound infection is reduced by minimising trauma such as from retractors, meticulous hemostasis and prophylactic antibiotics where indicated. Wound dehiscence has become uncommon with the use of slowly degrading, absorbable sutures.

33

The ergonomics of laparoscopic surgery

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Ergonomics is the study of people at work or in structured activities. It relates to the design of equipment, and how the equipment affects the surrounding environment. It is also a study of the interaction between the user and the equipment. Laparoscopic surgery, unlike conventional surgery, depends on interaction with very sophisticated equipment, and is not unlike flying an aircraft, which can only be flown by a pilot who understands the equipment and its limitations.

Many of the difficulties encountered in laparoscopic surgery – such a mastering eye–hand coordination, working with reduced freedom of movement, coping with a two-dimensional image, and reduced tactile feedback – are real. Most of these limitations of laparoscopic surgery can be overcome if the surgeon has a good understanding of ergonomics. Much of what works in laparoscopic surgery is counter-intuitive, and blind adoption of sound open surgical practices may actually be inappropriate.

Mastering laparoscopic skills is about:

- understanding ergonomics,
- knowing the limitations of equipment/user interphase,
- working within these limitations,
- training.

All laparoscopic instruments have to work around a fulcrum, the axis of which is the point of entry into the abdomen or chest. This means that all laparoscopic movements are completely paradoxical. This is first-order paradox and it is no different from rowing a boat. Most surgeons understand this principle but do not appreciate the fact that first-order paradox conditions only exist in laparoscopy when the surgeon, the video monitor, and the endoscopic camera are strictly in-line, and only when the camera is directed away from the surgeon. Any deviation from this will result in loss of first-order paradox. In the extreme example, if the camera is pointed toward the surgeon, a new order of paradox is introduced, even though the monitor is in-line. In this instance the horizontal movements are no longer paradoxical, but the vertical movements remain so. This is known as second-order paradox and has been investigated by Kohler using reversing prisms to introduce second-order paradox into daily activities. He found it impossible to perform simple tasks without having to think deliberately just to perform the task (Fig. 33.1).

Whereas most surgeons appreciate this and intuitively place themselves in an ergonomic position, they do not appreciate that intuitively positioning the assistant and operating room nurse across the table, which is the accepted convention in open surgery, places the assistant and operating room nurse in a position where they have to work with second-order paradox, and therefore reduces their effectiveness (Fig. 33.2).

It is not unusual to find nurses and assistants intuitively placing themselves in positions that are correct for open surgery, but that, in the case of laparoscopic surgery, severely disadvantages them (Fig. 33.3).

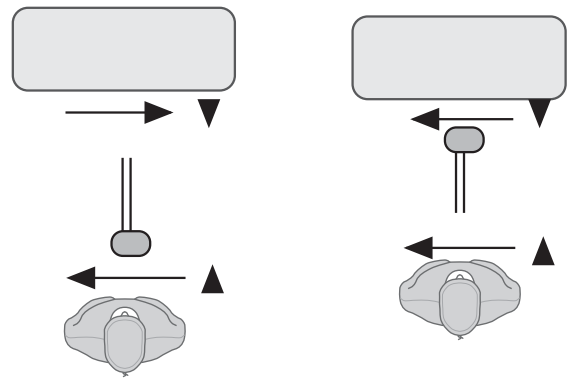

Fig. 33.1 (a) First-order paradox. Note that all movements are paradoxical. (b) Second-order paradox. Note if the camera is pointed directly at the surgeon, horizontal movements are no longer paradoxical but vertical movements remain paradoxical.

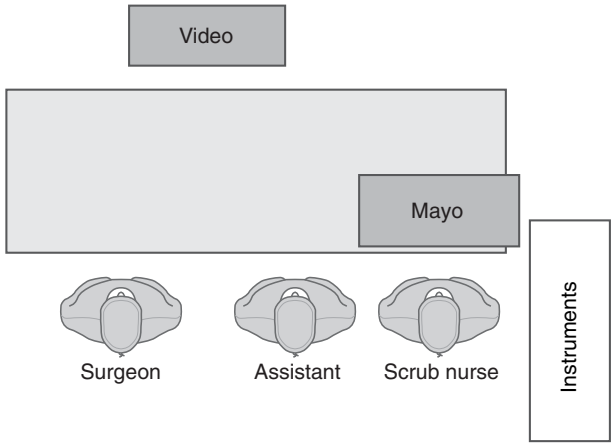

Fig. 33.2 Example of an ergonomic layout in which all personnel are working in first-order paradox.

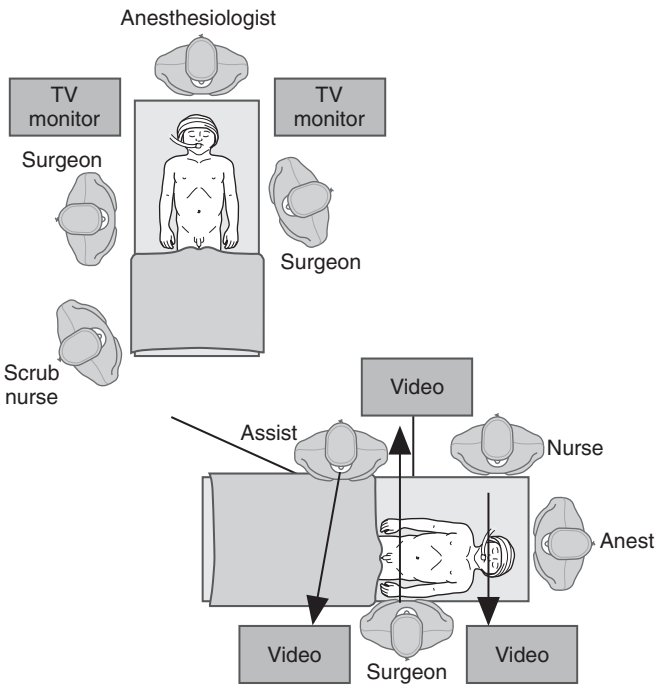

Fig. 33.3 Examples of non-ergonomic theater set-ups.

ERGONOMIC RULE

Whereas it can be tempting to use multiple monitors, one must be mindful of the fact that this may severely disadvantage the assistants if it places them in an environment where they have to work with second-order paradox. With few exceptions, it is therefore better for the assistant surgeon and the scrub nurse to be on the same side as the surgeon.

MANIPULATING INSTRUMENTS

A common problem encountered by the laparoscopic surgeon is finding the way back to the target when an instrument is removed and replaced with a new instrument. There are two easy ways of overcoming this problem, the first being to use the principle of triangulation, which is used every day to pinpoint the exact position of aircraft. Basically, if two reference points converge, it is easy to locate exact spatial position. In laparoscopic surgery, there are always at least three reference points, i.e., the camera and two instruments. This means that if the camera and another instrument are held steady during interchange of the third instrument, the surgeon should be able to locate (with practice) very accurately the internal location of the target by using the external landmarks of the two remaining instruments as reference points. It is useful to practice this exercise, which then becomes an automatic task.

The second way of returning instruments to the exact internal location is by using Gestalt. Gestalt involves having your own mental picture of the space around you and it allows one to navigate in total darkness in familiar surroundings. The surgeon learns to maintain a mental picture of the spatial position during interchange, so that the new instrument can be introduced in the same path. This requires practice, but once the skill is acquired, it becomes automatic. An important principle, however, is to make sure that you do not move at all during the interchange of instruments, as any change in your own position will result in loss of Gestalt.

OTHER ERGONOMIC CONSIDERATIONS

Surgeons intuitively place abdominal incisions as close to the operative site as possible, but this does not work in laparoscopy, because in laparoscopy one needs to have sufficient internal working space to manipulate the laparoscopic instruments. The volume of the cone formed from point of entry to surgical field determines the internal working space at laparoscopy. The ability to manipulate instruments on the inside is severely restricted if the instrument port is too close to the target organ, and leads to exaggerated external movements.

Likewise, placing instrument ports too far away simply amplifies external movements so that all internal movements, including tremor, are exaggerated.

The optimum port placement should be one that gives you enough space to manipulate instruments internally and which minimizes exaggeration of internal or external movements working space. In most instances, the ideal port position is where approximately half the working length of the instrument is on the inside. However, this rule can be broken to one's advantage, and the typical example of this is where it is necessary to perform microanastomosis. In this instance, it is to the surgeon's advantage either to use longer instruments or to place the port closer so that one needs to perform

exaggerated external movements to perform fine, accurate suturing.

COMFORT AND STABILITY OF SURGEON

It is important for the surgeon to be comfortable. The most comfortable position for a surgeon is for the arms to be hanging by his or her side, and not abducted. Placing instrument ports too close together will result in instrument clash, yet placing them too far apart will result in fatigue if the arms have to be abducted constantly.

Surgeons always set table height at their focal length in open surgery and it is common practice for laparoscopic surgeons to adopt the same practice. It should be noted that this is not ergonomic in laparoscopic surgery as it means that the surgeon needs to abduct his or her arms to manipulate instruments. Because there is no need for the table height to be at the same level as in open surgery, it is always better to adjust it so that it is low enough to allow one to work with arms by one's sides. This will reduce fatigue.

LAPAROSCOPIC SUTURING

Laparoscopic suturing is considered one of the most difficult tasks to perform. However, it can be made much easier if you understand the restriction on your freedom of movement incurred by working around a fulcrum. The first principle to understand is that it is impossible to suture if the needle holder is at right-angles to the intended suture line, in the same way as it is impossible to make an incision in the correct line if the scalpel is at right-angles to this line. The needle driver must therefore be in line with the anastomosis, and this is where one needs to plan port placement to optimize one's ability to suture.

The following points about laparoscopic suturing should be noted.

- Suturing is impossible if the needle driver is at right-angles.
- It is easier to suture toward a port.
- It is best to mount the needle at the halfway point.
- It is best to place the telescope as close as possible to where you wish to suture.
- It is best to form your loops over the loose end rather than bring the loop to the end.
- Use very short sutures (5–7 cm).

GENERAL PRINCIPLES OF ERGONOMICS

- Keep it simple.
- Keep equipment simple.
- Laparoscopic surgery is not intuitive, but an understanding of these guidelines allows you to plan every surgical procedure better, reduce fatigue and mistakes, and make laparoscopic surgery fun.

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34

Laparoscopy in infants and children

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HISTORY

The use of laparoscopy in infants and children was introduced by Gans and Austin in the early 1970s. Since then other investigators have confirmed their early results, and laparoscopy is now an established method of investigation and treatment for a wide variety of conditions in infants and children.

PRINCIPLES AND JUSTIFICATION

Indications

Laparoscopy is indicated for *diagnosis* only when more simple studies are inadequate and when exploratory laparotomy would otherwise be considered. It is indicated for *therapy* only when the procedure can be carried out safely with laparotomy, and either when the laparoscopic and open procedures are equally effective or when the laparoscopic approach has been shown to be superior.

Laparoscopy is most commonly used because of its ability to inspect the entire peritoneal cavity, as all four quadrants of the abdominal cavity can be inspected through one small puncture wound. Investigative procedures carried out by this technique include exploration for occult pain, contralateral hernias, intra-abdominal testis, trauma, biliary atresia, intersex, and a variety of other conditions.

Laparoscopy is also used for a number of operative procedures. These include appendectomy, cholecystectomy, adhesiolysis for partial small bowel obstruction, fundoplication for gastroesophageal reflux, splenectomy, inguinal hernia repair, pyloromyotomy, congenital diaphragmatic hernia repair, reduction of intussusception, Ladd's procedure for malrotation of the intestines, varicocelectomy, tumor staging, aspiration or excision of cysts of the mesentery or ovary, manipulation of the peritoneal end of ventriculoperitoneal shunts in patients who have had multiple previous procedures, intestinal resection such as that required for a Meckel's diverticulum, placement of a peritoneal dialysis catheter, orchidopexy for non-palpable undescended testes, and many other procedures.

LIVER BIOPSY

Many pediatric patients have hepatobiliary conditions, which, even after thorough study, may require a piece of liver tissue for accurate diagnosis. The simplest method of liver biopsy is using a percutaneous needle, but occasionally the results are unsatisfactory and open operation for liver biopsy is necessary. In the authors' experience, examination and biopsy using laparoscopy has distinct advantages over the open procedure.

The color, size, structure, and feel of the liver can be evaluated, and the presence of cysts, hemangiomas, nodules, tumors, or diffuse hepatic processes noted before the biopsy needle or forceps is directed into the most promising areas. Focal or nodular lesions may be missed by blind needling, and direct observation prevents penetration of vascular or other structures at risk of harm. Any bleeding or leakage of bile persisting after biopsy is readily observed and controlled by suture or electrocoagulation.

A better view of the liver and even of the spleen is obtained.

The risks of adhesion formation and other surgical complications are reduced. The operating time and stay in hospital are shorter and there is no abdominal scar.

CHOLANGIOGRAPHY

Percutaneous transhepatic cholangiography may be performed in well-selected cases. It is not an easy maneuver but, when successful, it is definitive.

In a small number of older infants or children with jaundice, laparoscopy is considered after the usual investigations. The appearance of the liver and gall bladder is significant, and

biopsy and percutaneous transhepatic cholangiography can be undertaken using this technique with acceptable safety, frequently avoiding open surgery. In some cases the findings on laparoscopy provide a definite indication for operation.

ASCITES

Removal of the ascitic fluid, replacing it with carbon dioxide, followed by laparoscopy, may reveal the etiology of the condition.

CYSTS AND TUMORS

A clear view and biopsy of abdominal cysts and tumors sometimes surpasses other procedures in providing necessary information for indications for surgery, or for the avoidance of open surgery when scattered metastases are seen. This is particularly true for tumors of the liver. In selected cases, second-look procedures may be appropriate.

PELVIC PROCEDURES

If the anatomic or hormonal status of the uterus, tubes, and ovaries is in doubt, laparoscopy will find its greatest application in three different age ranges: (1) the newborn with ambiguous genitalia; (2) the child with pain or precocious puberty; (3) the postpubertal adolescent with pain.

Ovarian tumors may be biopsied, ovarian cysts aspirated, and adhesions separated using electrocoagulation or the tine scissors.

OCCULT PAIN

Occult abdominal pain is categorized as recurrent bizarre or chronic abdominal pain for which a satisfactory diagnosis cannot be established by the usual methods and for which laparotomy is now being considered. Non-surgical causes of such pain, for example regional enteritis and salpingitis, may be identified using laparoscopy and the appropriate medical therapy given. Laparoscopy is also helpful, if no pathology is found, in providing reassurance that the organs are normal and that no disease is present.

VENTRICULOPERITONEAL SHUNT

Correction of malfunctions of ventriculoperitoneal shunts with the laparoscope is one of the most rewarding of its capabilities. Entrapment or encystations of the peritoneal catheter can be corrected by shifting its position to an appropriate site. Peritoneal fluid and tissue are easily obtained for culture. The entire peritoneal cavity is visualized and the region most suitable for repositioning of the shunt catheter is determined. Finally, the use of laparoscopy rather than laparotomy minimizes the formation of new adhesions within the abdominal cavity and may therefore reduce the risk of recurrent problems.

TRAUMA

Non-operative treatment for a ruptured spleen or liver is now preferred in abdominal trauma. When laparotomy is being considered, however (even when a considerable amount of blood is present in the peritoneal cavity), laparoscopy can be used to determine whether active bleeding is occurring, and which organ is bleeding. Furthermore, the infant or child with severe multiple organ system trauma, who is usually unconscious, requires rapid and accurate assessment, urgent respiratory and circulatory resuscitation and maintenance, and prompt therapeutic intervention. In a child who has sustained significant trauma to the head, chest, and extremities, the possibility of serious intra-abdominal injury must be ruled out before the priority of any particular regional or organ intervention is determined. Laparoscopy can be carried out quickly in the emergency room, intensive care unit, or in the operating room with portable equipment and will help provide the surgeons with the information to make decisions.

EMERGENCY LAPAROSCOPY

Indications for emergency laparoscopy in trauma are multiple organ system trauma, impaired sensorium, unexplained falling hemoglobin level, equivocal abdominal examination, or a stab wound with questionable abdominal wall penetration.

Contraindications

Laparoscopy is contraindicated in infants and children for whom general anesthesia is contraindicated, such as infants in shock. It is further contraindicated in a condition in which a coagulopathy may lead to hemorrhage that is difficult to control. Pregnancy in adolescents, peritonitis, adhesions, and intestinal obstruction with massive distension of the intestinal loops warrant caution and possibly some modification in technique, but are no longer considered contraindications to laparoscopy.

PREOPERATIVE

The stomach should be emptied if the upper organs are to be examined; the bladder should be emptied if the lower organs or pelvis are to be examined. A Credé maneuver is usually sufficient to empty the bladder, except when pelvic exploration is to be undertaken, in which case an indwelling Foley catheter should be inserted in older children.

Anesthesia

General anesthesia with endotracheal intubation is preferred because pneumoperitoneum significantly inhibits diaphragmatic movement.

INSTRUMENTS

A wide variety of instruments are suitable for laparoscopy in infants and children. The Storz laparoscopic pediatric instruments are the standard for infants; any other instruments used should be as delicate and useful as these to ensure the best possible results.

A high-flow insufflating device is used to introduce carbon dioxide and automatically to control the flow and pressure of the gas. A xenon or other similar high-intensity light source is also important.

The Veress needle with a spring-controlled stylet is used for closed access. A blunt trocar or Hasson cannula is used by many for open access.

Cannulas with trocars and instruments for grasping, retraction, biopsy, suction, electrocoagulation, palpation, cutting, and dissection complete the basic set. More advanced techniques require linear staplers, clip appliers, needle holders, a variety of advanced energy devices, and tissue extraction devices.

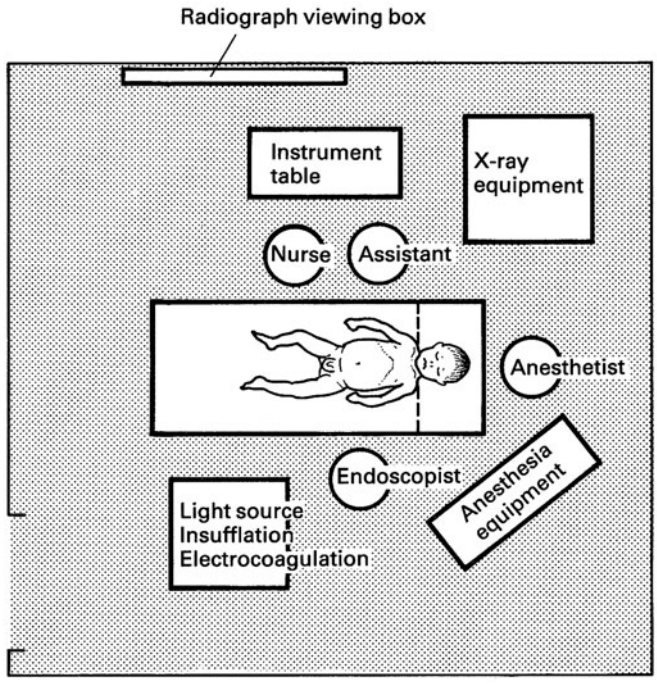
A high-definition video camera for projection onto multiple monitors is essential. Video recorders, printers, and other recording devices are desirable accessories for documentation and for teaching.

Carbon dioxide insufflation

While some prefer lower pressures in infants, unless a patient's condition mandates a lower pressure because of difficulty maintaining adequate ventilation, we generally use a pressure of 15 torr CO_2 for all of our laparoscopies. A note of caution should be mentioned when insufflation is initiated in tiny infants. Most insufflators today deliver their gas in volumes that exceed the intraperitoneal volume of the very small infant. In these cases, the intraperitoneal pressures can climb higher than desired very quickly. Knowing this beforehand will allow the surgeon to 'bleed off' some of the gas and enable the pressure to drop to a sufficiently low level to proceed safely with the operation. Computer algorithms in more modern insufflators permit smaller volumes of gas to be insufflated so that tiny infants can be safely subjected to laparoscopy.

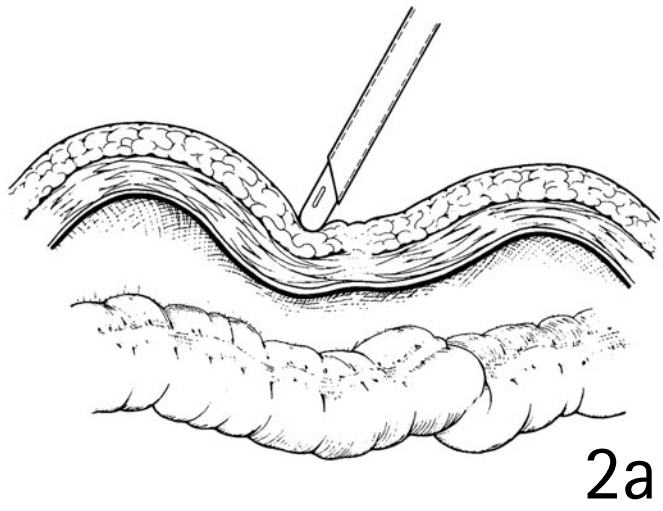
PATIENT POSITION

1 The patient is placed in the supine position on the operating table under endotracheal anesthesia and the operating team and equipment are appropriately positioned. The skin of the abdomen is prepared and draped.



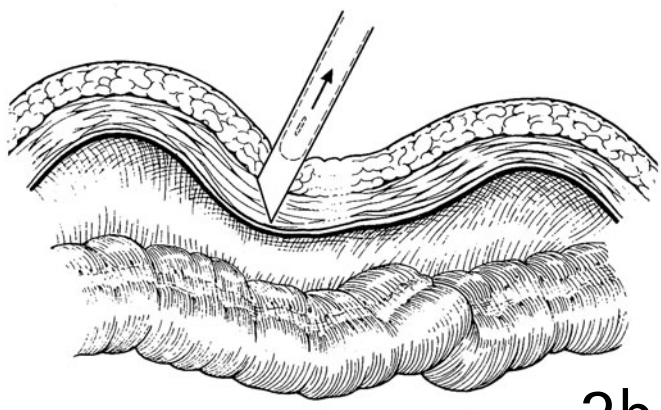
1

A stab wound is made in the skin with a knife blade. This puncture is usually made in the rim of the umbilicus where the abdominal wall is thin; this central location permits examination of the entire peritoneal cavity and leaves an almost invisible scar.



2a

2 The abdominal wall is tented upwards by the operator and assistant by grasping it above and below using a sponge to maintain traction, and the Veress needle with the spring-controlled blunt stylet is introduced into the peritoneal cavity. As the needle pierces the peritoneum, the blunt stylet springs out, thus protecting the abdominal contents from injury.



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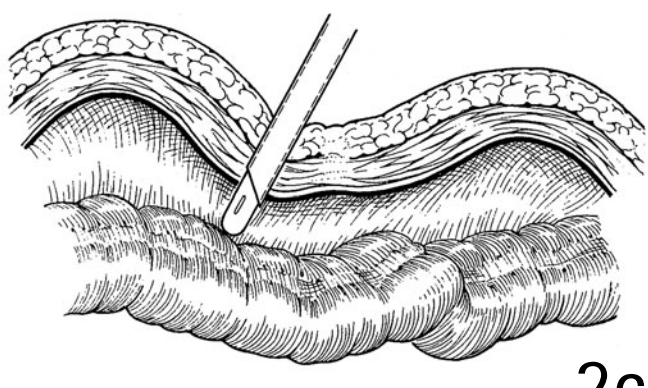
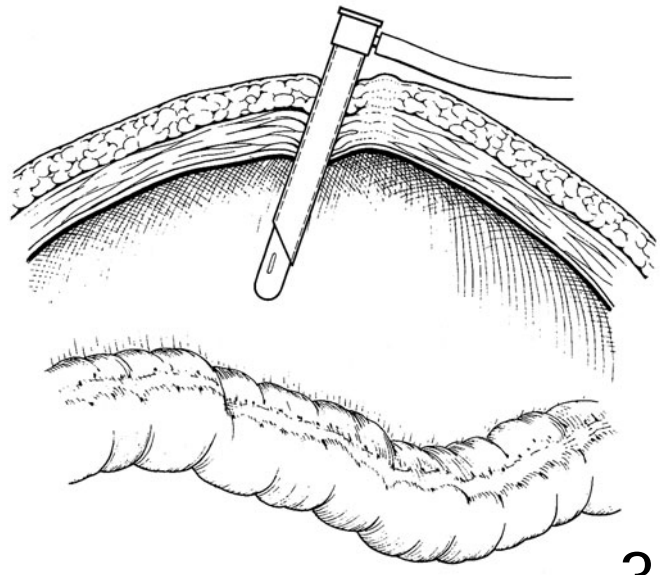


2c

A 10 mL syringe is connected to the needle, and aspiration is carried out to ensure that no bowel contents are present in the needle. Saline solution (5–10 mL) is then injected into the abdomen to demonstrate free flow. The meniscus of fluid in the hub of the Luer lock should descend when the abdominal wall is elevated.

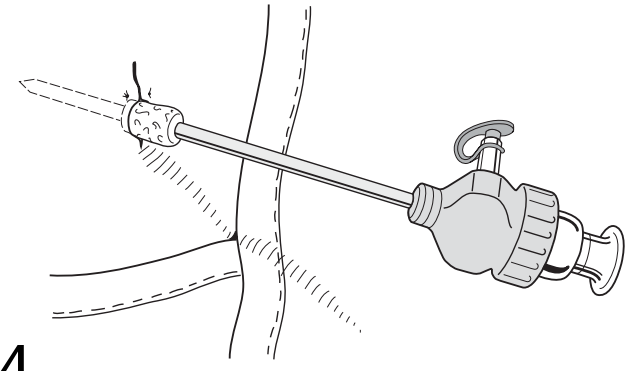
3 Pneumoperitoneum is accomplished by connecting the needle to a CO_2 cylinder through the insufflating device so that flow and pressure can be controlled as desired or set automatically. Abdominal pressure should not exceed 8–10 mmHg in an infant, or 10–15 mmHg in the older child.



3

During insufflation, the abdomen is gently percussed and palpated, and the needle is removed once the pneumoperitoneum is considered adequate. The skin puncture is enlarged by spreading it with a hemostat until it admits the trocar and cannula snugly.

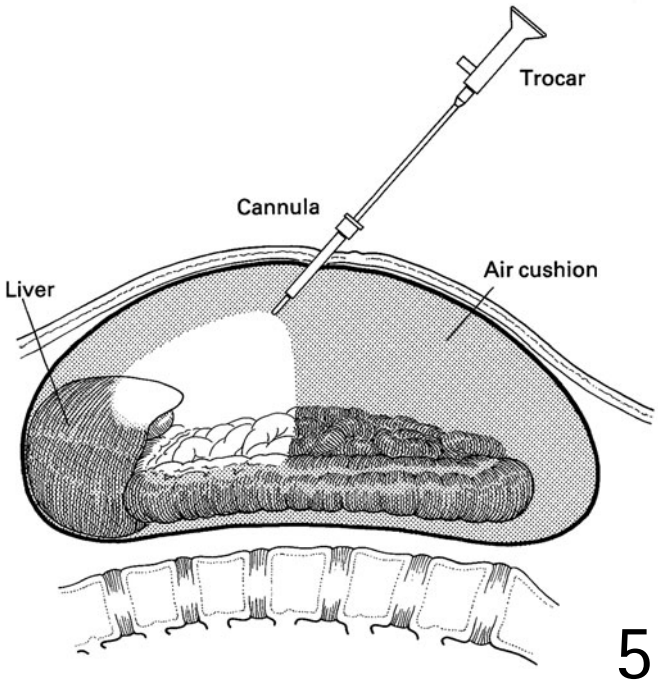
Open access may be achieved using a blunt trocar/cannula system. After the umbilical incision is made, the wound is spread and the midline fascia grasped with two hemostats, one on either side of the midline. Scissors are then used to incise the midline, and the peritoneum is opened sufficiently to introduce the trocar.



4

4 We find that a 1 cm sleeve of rubber tubing of an appropriate size placed on the outside of the cannula at the level of the skin is useful to keep the cannula from sliding during the procedure. This can be used as a retaining ring to suture the cannula to the skin to prevent it from dislodging.

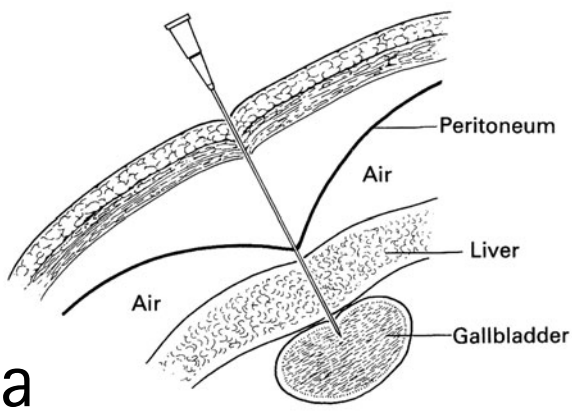
5 Once the pneumoperitoneum has been established with a Veress needle, the abdominal wall is again tented up by grasping it above and below the puncture, and the cannula with a pointed trocar is directed with a twisting motion through the abdominal wall and into the intra-abdominal air cushion. The trocar is then replaced by the appropriate telescope. We prefer to use a 30° telescope because it offers a wider field of vision. The viscera underlying the puncture after a Veress needle has been used should be inspected before proceeding, to ensure that no injury has occurred.



5

Inspection of the peritoneal contents may then begin. Any fogging of the telescope may be cleared simply and easily by gently touching the bowel wall with the telescope end. Needles for biopsy or injection may be inserted directly through the abdominal wall under direct vision through the telescope.

For more involved manipulations, other appropriately sized cannulas are introduced separately through the abdominal wall, observing their introduction through the telescope. All the accessory instruments listed earlier may be introduced through these cannulas under direct vision.



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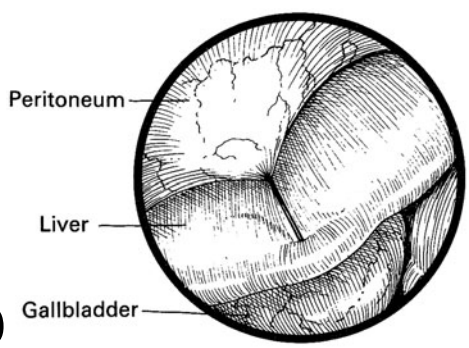
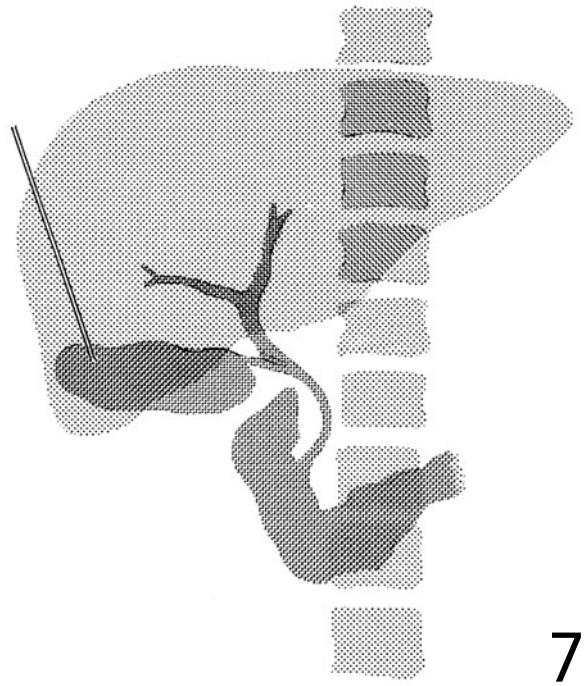


6b

OPERATIONS

Cholangiography

6a,b A plastic needle with a trocar is introduced through the abdominal wall and peritoneum, then through the liver and gallbladder bed into the gallbladder. The liver acts to tamponade any hemorrhage and to prevent bile leakage. Under direct vision, radio-opaque dye is injected and films are made under fluoroscopic guidance.

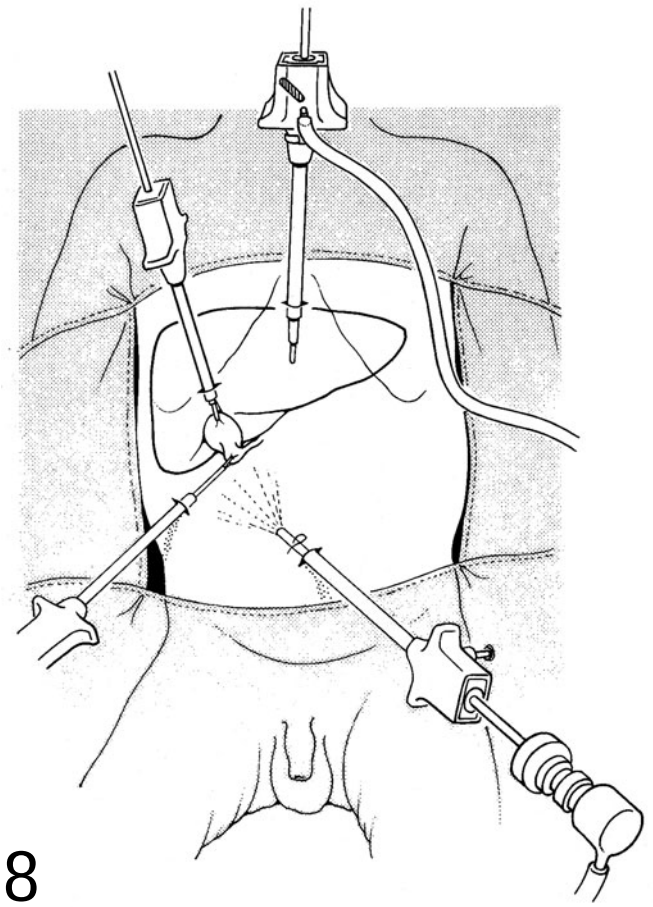


7

7 This method is useful in distinguishing neonatal hepatitis from biliary atresia and from bile duct hypoplasia. If the gallbladder is found to be absent on inspection, this is sufficient evidence to indicate exploration for biliary anomalies. If the gallbladder is present, cholangiography will demonstrate either a normal or an obstructed biliary duct system, which could be an indication for open exploration.

Cholecystectomy

Laparoscopic cholecystectomy has become the procedure of choice for cholecystitis and cholelithiasis. The procedure is particularly useful in children with sickle-cell disease because it is associated with fewer postoperative pulmonary complications.



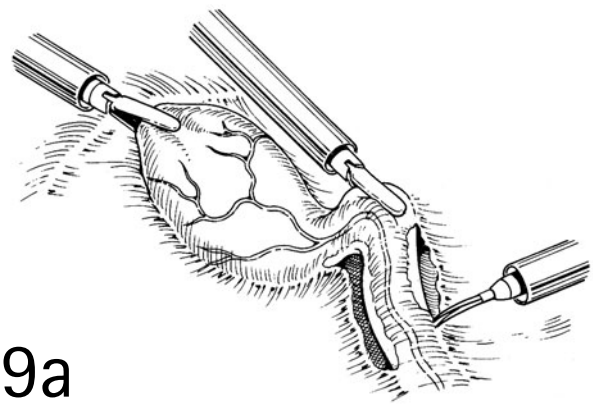
8

8 Most surgeons use four cannulas: a 10 mm umbilical cannula is used for the telescope and gallbladder extraction, and three 5 mm cannulas are also used. A subxiphoid cannula that can be moved to the patient's left is used for dissection and for clip application. A subcostal, midclavicular line cannula is used for retraction of the neck of the gallbladder, and an anterior axillary line cannula is used for retraction of the fundus.

The gallbladder is grasped using the two lateral cannulas for access and the cystic artery and duct are dissected free.

Most surgeons today believe that an operative cholangiogram should be performed before dividing the cystic duct to identify the ductal anatomy if this is not clear, if there is evidence of stones, or if there is a large common bile duct. This is performed either by placing a clip on the cystic duct at its junction with the gallbladder, making a small nick in the cystic duct with a pair of scissors and inserting a cholangiogram catheter through a separate port, or (if the anatomy is not clear) by performing a contrast study through the gallbladder itself as one might do in a case of biliary atresia.

9a–d Once the anatomy is clearly identified, the cystic duct and artery are divided between clips and the gallbladder is dissected.



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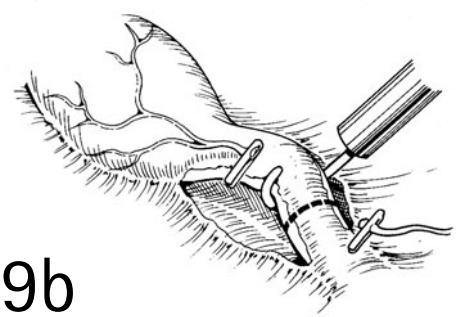

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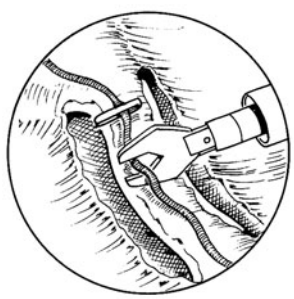


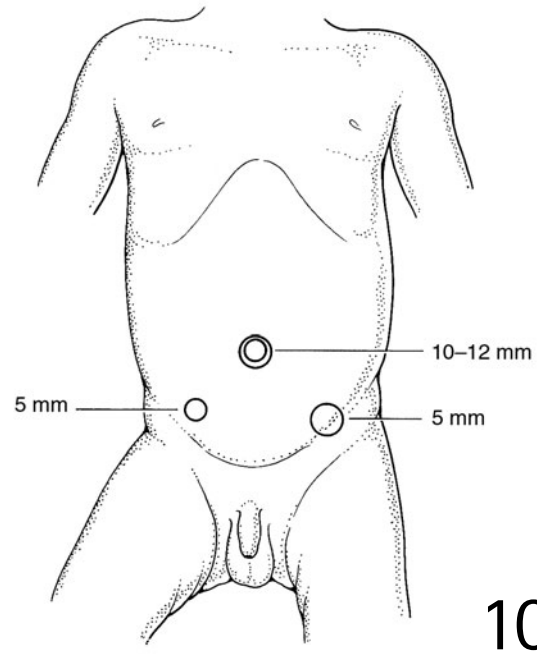
9c

9d

Once hemostasis of the liver bed is assured, the gallbladder is removed, usually through the umbilical cannula. If stones are present and prevent easy removal, they can be removed from inside the gallbladder, the neck of which has been exteriorized through the umbilical port. This will facilitate extraction.

Appendectomy

10 Patients should have their bladder catheterized and their stomach should be emptied. Three cannulas are sufficient for this procedure; an umbilical cannula (10–12 mm) is used for the telescope initially. This is later moved to a left lower quadrant port (5 mm). A midline suprapubic port (5 mm) and the umbilical port are used for instrumentation and removal of the inflamed appendix. We usually use an endoscopic stapler with a vascular load to remove the appendix because it is quick and simple. Alternatively, the mesoappendix can be divided with an ultrasonic scalpel or similar energy device and endoscopic loops can be used to ligate the appendix before it is transected between the loops.



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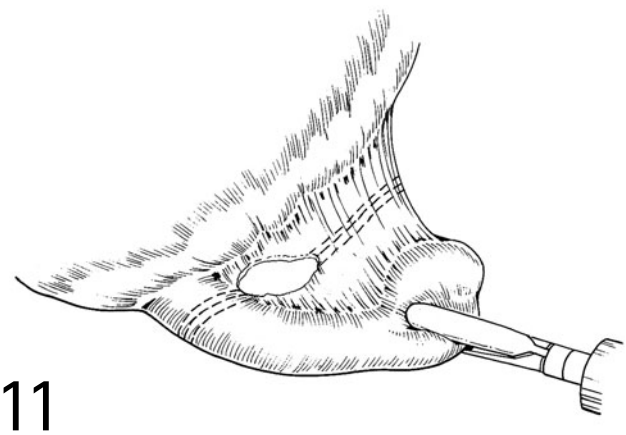


11

11 Once the appendix has been grasped from the suprapubic port, a dissector from the umbilical port is used to create a window between the appendix and the mesoappendix at the junction of the appendix with the cecum.

A linear stapler is then used to divide the appendix and the mesoappendix in turn. The appendix is then removed through the umbilical trocar.

Sometimes there is bleeding from the stump of mesoappendix, in which case an endoscopic loop can be used to reinforce it.

Alternatively, the appendix is grasped and the mesoappendix divided using an energy device or surgical clips to skeletonize the appendix. The appendix is then freed to its junction with the cecum.

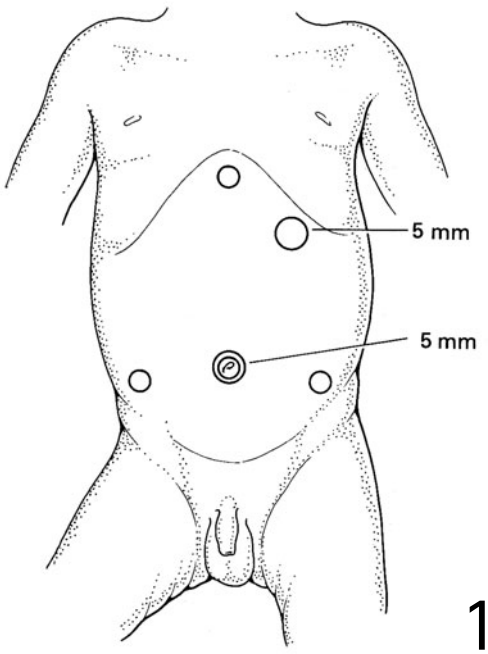
Endosurgical loops are applied to the base of the appendix and the appendix is divided and removed through the larger cannula.

In the case of ruptured appendicitis, an irrigation/aspiration cannula is used to evacuate any pus, minimizing the risks of abscess formation.

Fundoplication

The indications for a laparoscopic fundoplication are the same as those for the open procedure.

12 The authors use a four-cannula to five-cannula technique: an umbilical cannula is used for a 5 mm 30° telescope; 5 mm cannulas are placed in the right and left upper quadrants at the midclavicular line between the costal margin and the umbilicus. Another cannula is placed in the subxiphoid region. Here, we currently use a 5 mm disposable Kittner dissector held in place by a self-retaining retractor to elevate the liver and expose the hiatus. Lastly, it is sometimes helpful to place an additional cannula in the left lower quadrant to manipulate the stomach during the procedure to facilitate exposure.

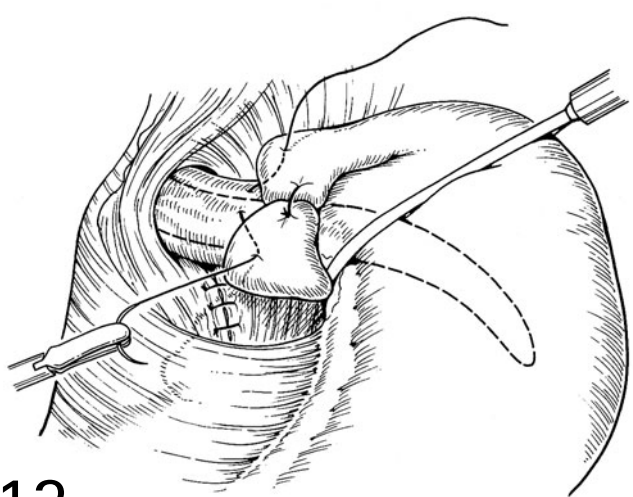


12

A bougie of appropriate size is inserted into the esophagus and used to judge the size of the wrap. We do not leave this in place during the dissection as it may interfere with manipulation of the stomach and esophagus.

We begin the dissection by dividing the short gastric vessels. We then carry the dissection cephalad to expose the left crus of the esophageal hiatus. Then the dissection moves anterior and toward the patient's right across the anterior of the esophagus.

The gastrohepatic ligament is divided next to expose the right crus. The peritoneum over this is divided to free the esophagus and display the esophageal hiatus.



13

13 The crura are then sutured closed and the fundus of the stomach is brought around the esophagus from behind. The wrap is then sutured into place in the same manner as for an open procedure.

Facility with endoscopic suturing is essential to the success of this procedure. A gastrostomy may be inserted if necessary under laparoscopic observation.

Splenectomy

Laparoscopic splenectomy is useful for a small or moderately sized spleen, but may be difficult for the enlarged spleen. The procedure is most often carried out for hereditary spherocytosis, idiopathic thrombocytopenic purpura, thalassemia major, and sickle-cell disease with life-threatening sequestration crises. See Chapter 67 for details of laparoscopic splenectomy.

POSTOPERATIVE CARE

At the conclusion of laparoscopy, the CO_2 is allowed to escape from the peritoneal cavity through the open cannulas. Postoperative care consists of the administration of appropriate analgesics and antiemetic medications as necessary.

Following fundoplication, patients are started on oral intake on the day of their procedure and are discharged from hospital within 24 hours. If patients have a gastrostomy, their discharge depends on the tolerance of gastrostomy feeds. After splenectomy, patients are started on oral intake 4–6 hours after surgery and are ready for discharge within 24 hours.

COMPLICATIONS

Two serious complications occur rarely: (1) perforation of the bowel during blind puncture of the abdominal wall or when using manipulating instruments; and (2) uncontrollable bleeding from a biopsy site or vascular injury. Both of these complications are readily identified through the laparoscope at the time of occurrence, and appropriate treatment by laparotomy should be instituted immediately.

The incidence of these complications is not accurately known, but must be very low, as they are absent in several large series.

Other complications include pneumothorax, gas embolism, and thermal burns from energy devices (recognition of this complication may be delayed). An understanding of the concepts of use of thermal energy in conjunction with intracavitary surgery helps to avoid such complications.

FURTHER READING

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35

Gastroesophageal reflux: Nissen fundoplication

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Gastroesophageal reflux is more common in infancy and childhood than is generally recognized. Although in the majority of cases (> 90 percent) the reflux resolves spontaneously within the first year of life as the lower esophageal sphincter matures, a small but significant proportion of cases develop complications requiring prolonged medical or surgical treatment.

HISTORY

In 1956, Rudolph Nissen of Basel, Switzerland, described the fundoplication procedure that he had been using for the previous 20 years to minimize postoperative reflux after resection of a peptic ulcer in the region of the cardia of the stomach. The technique did not involve division of the short gastric vessels, and the wrap extended for 4–6 cm on the esophagus. Subsequently, modifications have been made by Rossetti, Dor, Toupet, Donahue, and DeMeester.

PRINCIPLES AND JUSTIFICATION

Clinical presentation

EARLY INFANCY

The child presents with recurrent vomiting, which may be regurgitant or projectile. The vomitus may contain altered blood (the 'coffee-grounds' appearance). The infant fails to thrive and may be constipated.

OLDER CHILD

Vomiting is a major feature. In addition, the child complains of heartburn and dysphagia and may occasionally present with iron-deficient anemia secondary to chronic blood loss.

MENTAL RETARDATION

Recurrent vomiting occurs in 10–15 percent of neurologically impaired children. It is often regarded as part of the neurologic problem, but around 75 percent of these children have significant gastroesophageal reflux. It is notable that there is a high failure rate of medical treatment in this group.

ANATOMIC ANOMALY

Gastroesophageal reflux is more common in infants with esophageal atresia, diaphragmatic hernia, anterior abdominal wall defects, and malrotation.

ASPIRATION SYNDROMES

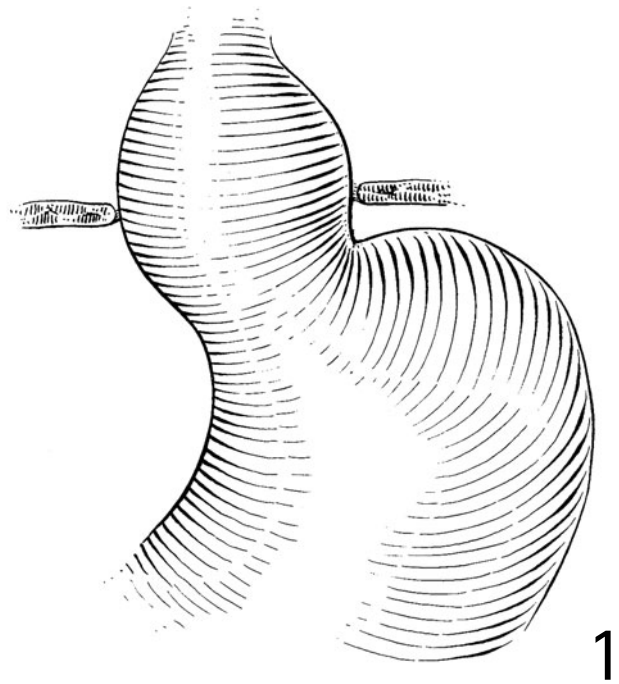
There is a small but definite association of aspiration symptoms (asthma, pneumonitis, cyanosis, apneic episodes) and gastroesophageal reflux.

OTHER PRESENTATIONS

Other presentations include rumination, Sandifer's syndrome (torsion spasms of the neck), protein-losing enteropathy, irritability, and hyperactivity.

Pathologic anatomy

Reflux may, or may not, be accompanied by an associated hiatus hernia. Two types of hiatus hernia are recognized.



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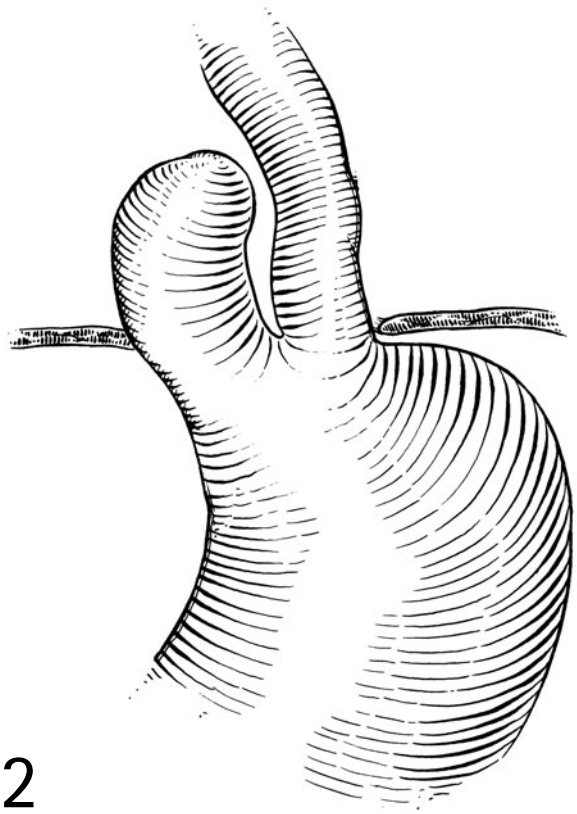
1 Sliding hiatus hernia, characterized by ascent of the cardia into the mediastinum.



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2 Paraesophageal or rolling hernia, in which the gastroesophageal junction remains in the abdomen while part of the gastric fundus prolapses through the esophageal hiatus into the mediastinum.

The sliding hernia is often associated with reflux, while gastric stasis in the paraesophageal hernia predisposes to peptic ulceration, perforation, or hemorrhage.

NORMAL MECHANISMS PREVENTING REFLUX

Physiologic control of reflux depends on the following factors.

- Anatomic:
  - length and pressure of the lower esophageal sphincter;
  - the intra-abdominal segment of the esophagus;
  - the gastroesophageal angle (angle of His);
  - the lower esophageal mucosal rosette;
  - the phrenoesophageal membrane;
  - the diaphragmatic hiatal pinchcock effect.
- Physiologic:
  - coordinated effective peristaltic clearance of the distal esophagus;
  - normal gastric emptying.

PATHOPHYSIOLOGY OF REFLUX

The squamous epithelium of the esophagus is unable to resist the irritant effect of gastric juices. The acid pepsin causes a chemical inflammation with erythema of the mucosa. With continued reflux, the mucosa becomes friable and bleeds easily on contact. Later, frank ulceration develops, which, with repeated attempts at repair and relapse, eventually leads to stricture formation. This process is summarized in Figure 35.1.

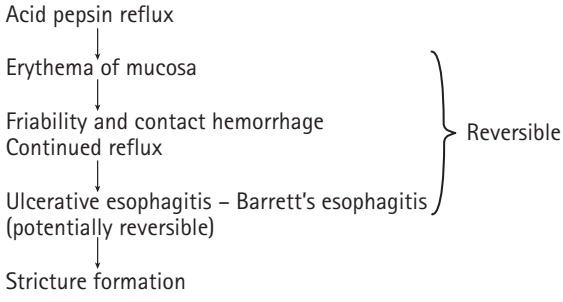


Fig. 35.1 Pathophysiology of reflux.

INDICATIONS FOR ANTIREFLUX SURGERY

Antireflux surgery should be undertaken in the presence of an established esophageal stricture or when conservative measures of treatment have failed. Surgery may also be considered at an early stage (1) in the presence of an anatomic anomaly, e.g., esophageal atresia, malrotation, exomphalos, and (2) in the presence of associated neurologic impairment, where the response to conservative measures is notoriously poor. Surgery may also be necessary if the patient is suffering from apneic episodes and repeated respiratory infections due to aspiration of refluxed material, or if the infant fails to thrive despite adequate therapy.

PREOPERATIVE

Investigations

A number of preoperative investigations should be performed.

- *Barium esophagogram*, with particular attention to the anatomy of the esophagus (presence of strictures, ulcerative esophagitis, abnormal narrowing or displacement); presence of a hiatus hernia; peristaltic activity of the esophagus and rate of clearance of contrast material; the degree of gastroesophageal reflux (grade I: distal esophagus; grade II: proximal/thoracic esophagus; grade III: cervical esophagus; grade IV: continuous reflux; grade V: aspiration into tracheobronchial tree); and evidence of gastric outlet obstruction.
- *Esophageal pH monitoring*. Continuous 24-hour monitoring of the pH in the distal esophagus is the most accurate method of documenting reflux. A pH of less than 4 is regarded as significant. During the 24-hour recording, the following parameters should be examined:
  - the number of episodes during which the pH falls below 4;
  - the duration of each reflux episode;
  - the number of episodes lasting more than 5 minutes;
  - the total duration of reflux, expressed as a percentage of recording time.
- *Esophageal manometry*. Pressure recordings are made with continuously perfused, open-tipped catheters or solid-state pressure transducers. A high-pressure zone is normally present in the distal esophagus. Individual pressure values are unreliable diagnostic indicators of reflux, but may be useful in predicting cases that will eventually require surgical treatment.
- *Endoscopy and biopsy*. Endoscopy will determine the degree of esophagitis; histology of the biopsy will provide pathologic grading of inflammatory cell infiltration. Four grades of esophagitis are recognized at endoscopy:
  - grade I: erythema of mucosa;
  - grade II: friability of mucosa;
  - grade III: ulcerative esophagitis;
  - grade IV: stricture.
- *Scintiscanning*. Technetium (^{99}Tc) sulfur colloid scans may be useful in documenting pulmonary aspiration.

MEDICAL MANAGEMENT

Small, frequent, thickened feeds should be given to infants with reflux. A 30° head-elevated, prone position is the most suitable posture for young infants. Antacids-alkalis with or without alginic acid (Gaviscon) should be administered. Histamine receptor antagonists (cimetidine, ranitidine) will suppress acid secretion and allow severe esophagitis to heal. Omeprasol (a proton pump inhibitor) is even more efficient.

Metoclopramide and bethanechol increase lower esophageal pressure and stimulate gastric emptying.

ANESTHESIA

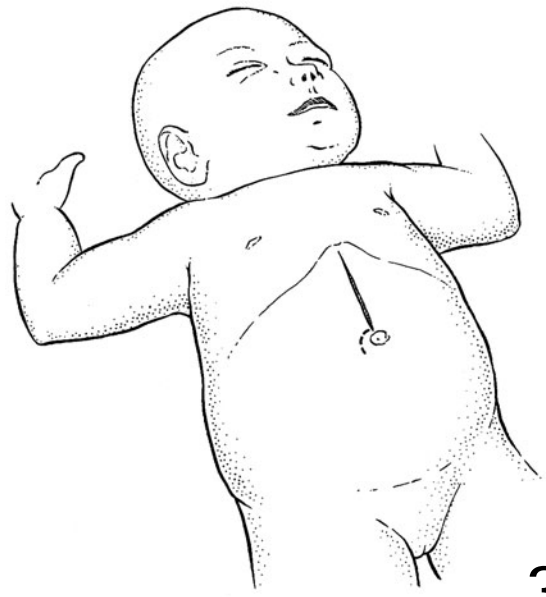
General endotracheal anesthesia is administered, with the patient supine. A single dose of prophylactic broad-spectrum antibiotics should be given after induction of anesthesia. The addition of epidural analgesia is extremely valuable in reducing postoperative pain and preventing respiratory complications.

OPERATION

Some surgeons insist on inserting a large-caliber bougie in the esophagus during the construction of the fundoplication to ensure that the wrap is not too tight. The author prefers a regular-size nasogastric tube and constructs a very loose wrap.

Incision

3 In the majority of cases the ideal approach is via a midline upper abdominal incision extending from the xiphisternum to the umbilicus. Alternatively, a left subcostal muscle-cutting incision may be used.



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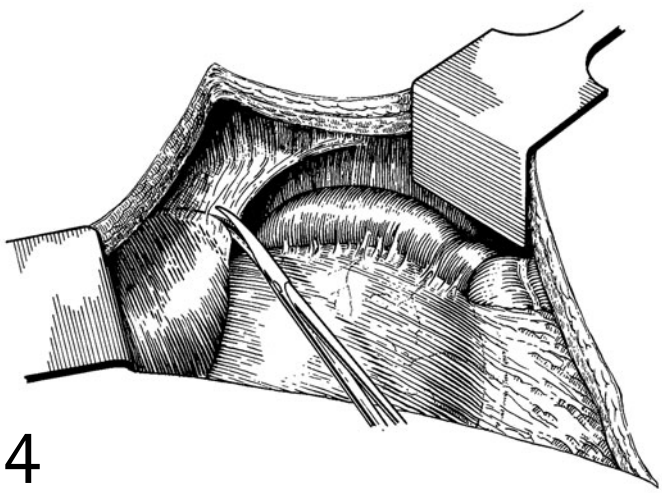


4

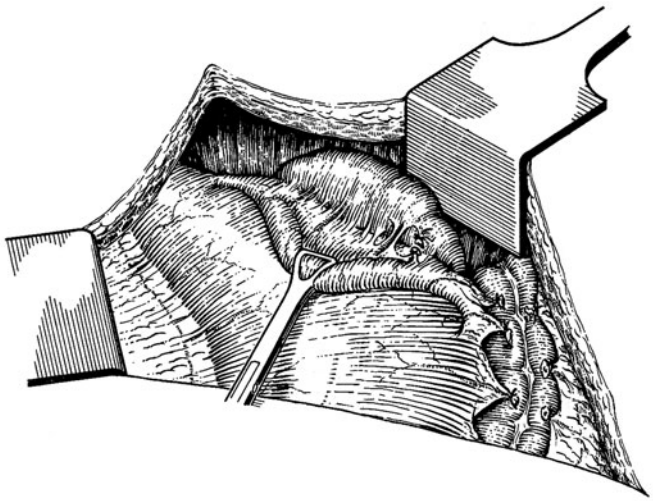
Exposure

4 Adequate exposure of the gastroesophageal junction will usually be obtained by retracting the left lobe of the liver anterosuperiorly. Additional exposure may be attained, if necessary, by dividing the left triangular ligament in the avascular plane and then retracting the left lobe of the liver to the right. In older children, especially children with kyphoscoliosis or obese children, the use of the self-retaining retractor is invaluable in obtaining adequate exposure of the operative field.

Mobilization of the fundus of the stomach

5a,b The proximal one-third to one-half of the greater curvature of the stomach is liberated from its attachment to the spleen by ligating and dividing the upper short gastric vessels in the gastrosplenic ligament. This is accomplished most safely by passing a right-angled clamp around each vessel in turn and ligating or coagulating with bipolar diathermy the vessel on the gastric and splenic side before dividing it.

When the vessels in the gastrosplenic ligament have been divided, the spleen should be allowed to fall back into the posterior peritoneum, thereby avoiding inadvertent trauma. Splenectomy should rarely be necessary in this procedure, even in revision fundoplication operations. The fundus is now sufficiently free to allow for a loose ('floppy') fundoplication.



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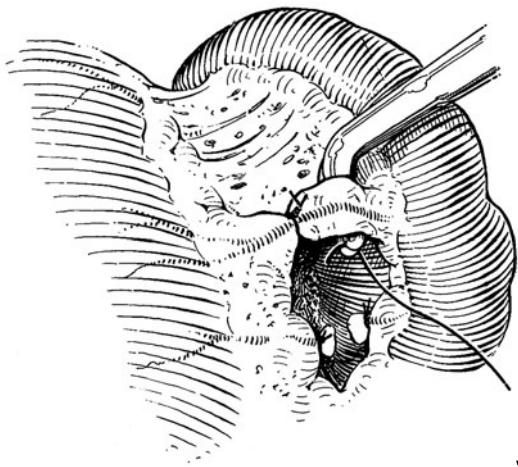

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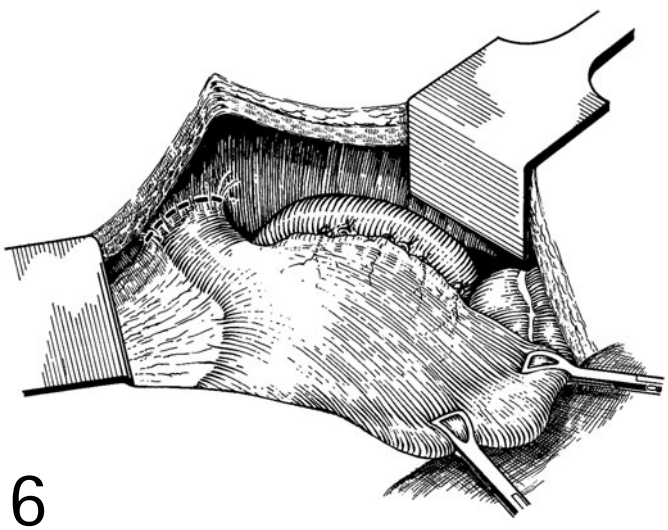


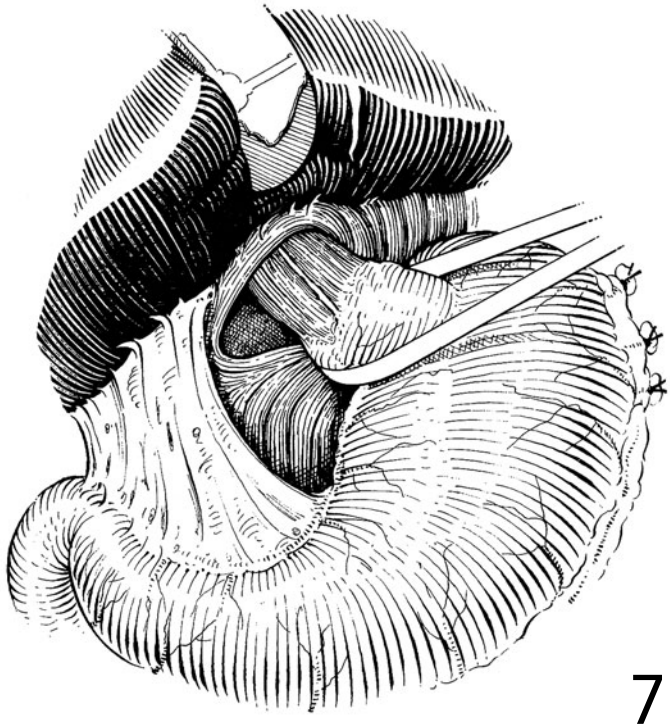
6

Exposure of the esophageal hiatus

6 The phrenoesophageal membrane is placed on stretch by downward traction on the stomach while the diaphragmatic muscle is retracted superiorly. The avascular membrane marked by a 'white line' is incised with scissors and the musculature of the esophagus displayed. The anterior vagus nerve will be seen coursing on the surface of the esophagus. It should be carefully protected and preserved.

Mobilization of the distal esophagus

7 Using a combination of sharp and blunt dissection, the lower end of the esophagus is encircled, avoiding injury to the posterior vagus nerve. A rubber sling is placed around the esophagus incorporating the posterior vagus nerve, which will be included in the fundoplication. The lower 2 cm or 3 cm of esophagus hiatus is completely exposed by dividing the upper part of the gastrohepatic omentum above the left gastric vessels.



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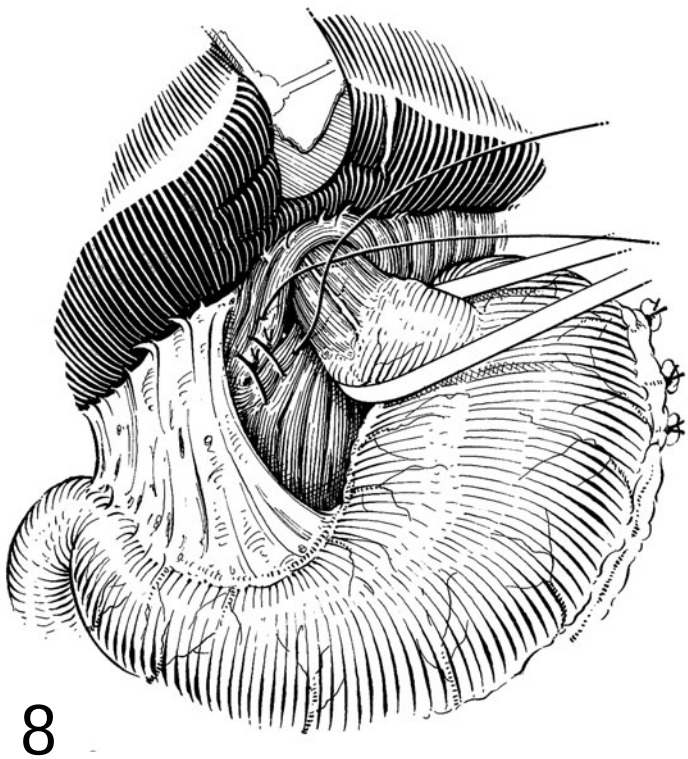


8

Narrowing of the hiatus

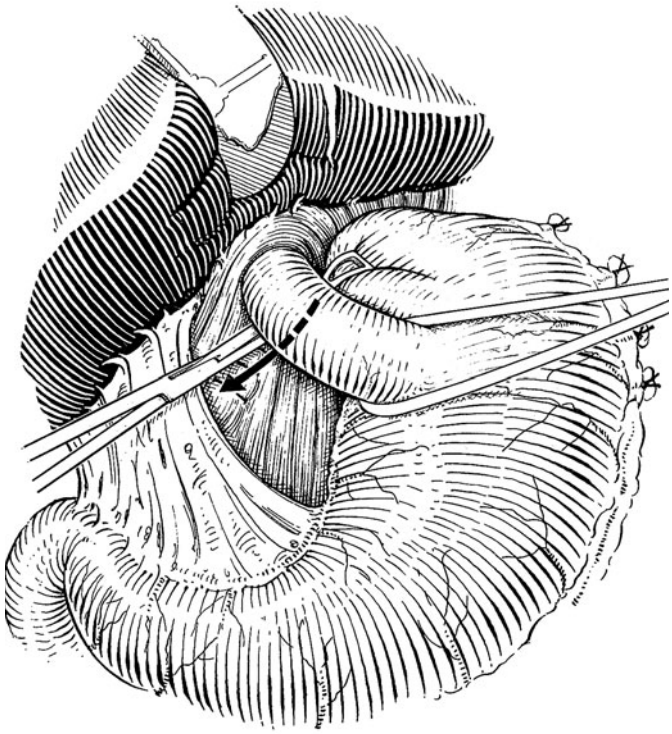
8 The esophageal hiatus is narrowed posterior to the esophagus by placing deep sutures through the crura of the diaphragm. The sutures are tied loosely to prevent them cutting through, but leaving sufficient space alongside the esophagus to allow the tip of a finger to pass. Two or three or more sutures may be required for this purpose.

Construction of the fundoplication

9a–c The mobilized fundus of the stomach is folded behind the esophagus so that the invaginated part of the stomach appears on the right side of the esophagus. It is important not to twist the stomach during this maneuver and to ensure that the stomach has been sufficiently mobilized to be able to fashion a loose wrap.

The length of the wrap varies from 1.0 cm in the infant to 2–2.5 cm in the older child. Commencing at the level of the gastroesophageal junction, three to four sutures of non-absorbable material (3/0 or 4/0) are placed through the stomach and esophageal muscle. Each suture passes from left to right through the anterior wall of the stomach, through the esophageal muscle (taking care not to enter the lumen of the esophagus), and through the wall of the mobilized portion of the fundus of the stomach, which has been folded behind the esophagus. Traction on the first (untied) suture brings the rest of the operating field clearly into view and facilitates the insertion of the remaining two or three proximal parallel sutures through the anterior wall of the stomach, esophageal muscle, and 'prolapsed' fundus. When all these sutures are in place, they are tied serially without tension on the wrap.

A second layer of non-absorbable sutures including the seromuscular surface of the stomach only may be placed superficial to the primary sutures to prevent disruption of the wrap.



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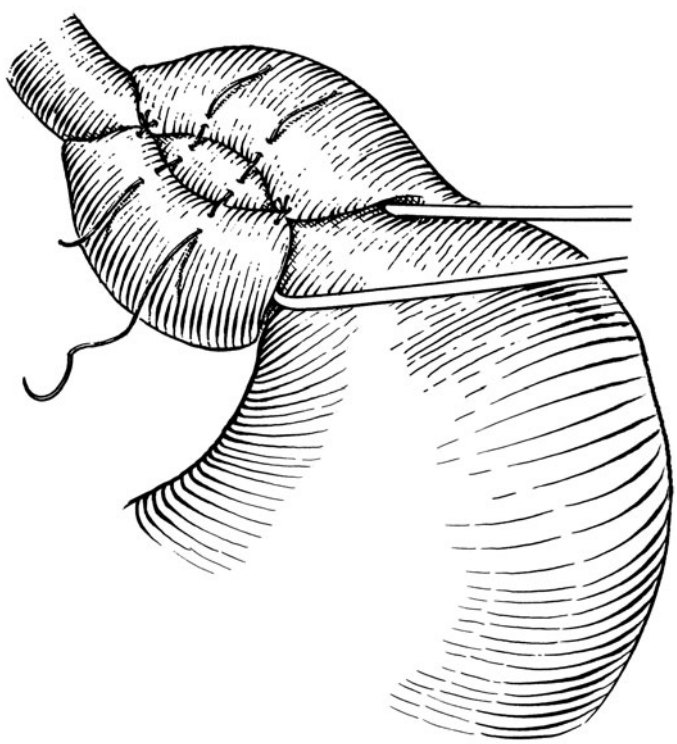

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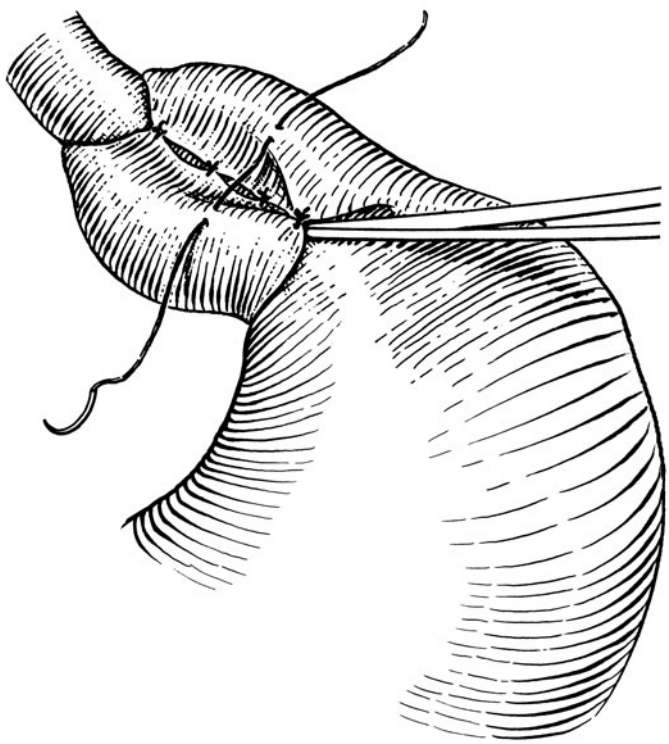
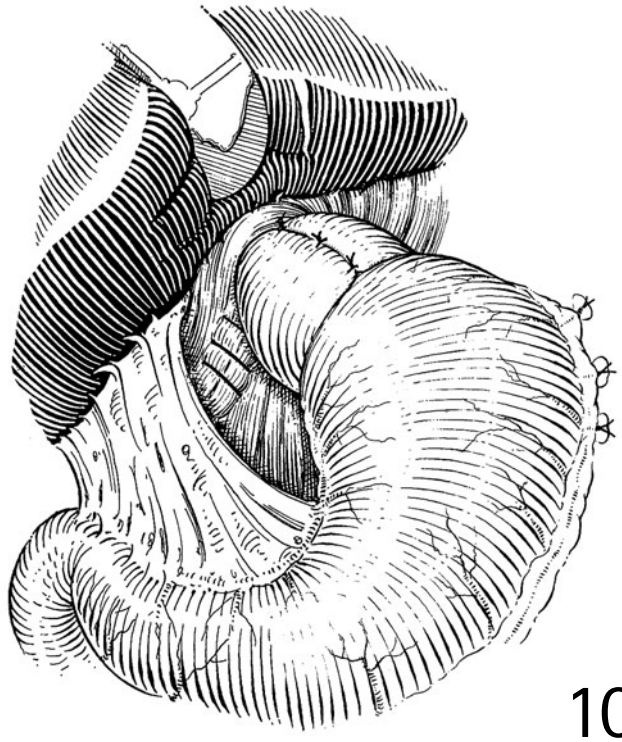


9c

10 The proximal suture of the second layer should be passed through the anterior wall of the esophageal hiatus in the diaphragm. An additional two or three sutures may be placed between the hiatus and the fundoplication to prevent the wrap migrating into the posterior mediastinum.



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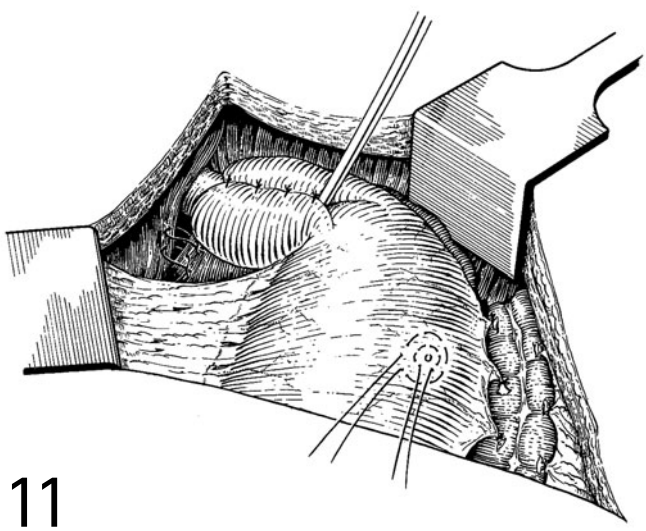

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Gastrostomy

11 A feeding gastrostomy should be constructed in neurologically impaired children who are unable to eat normally. A suitable area on the anterior wall of the stomach is selected to permit the gastrostomy site to be anchored to the anterior abdominal wall without exerting traction on the fundoplication.

Two rows of circumferentially placed non-absorbable sutures are placed through the seromuscular layer of the stomach. A suitable-size Malecot catheter is inserted through a centrally placed gastrotomy into the stomach and the sutures are tied, invaginating the gastrotomy site. The tube is brought out through a separate stab incision in the left hypochondrium, and the stomach wall is sutured to the anterior abdominal wall at the exit site to prevent leakage.

Closure

The wound is closed either in layers or with interrupted en-masse sutures. A subcuticular suture approximates the skin edges.

POSTOPERATIVE CARE

Nasogastric decompression and intravenous fluids are continued until postoperative ileus has resolved (usually 48–72 hours).

Complications

Death following this procedure is extremely uncommon. In severely retarded children there is a small but not insignificant risk of mortality, related mainly to the underlying disease.

Wound infection occasionally occurs.

Resiratory complications such as pneumonia or atelectasis particularly affect severely retarded patients and patients undergoing fundoplication for chronic respiratory complications secondary to aspiration.

Dysphagia may result from a wrap that is either too long or too tight.

The gastroesophageal reflux may recur because of either disruption of the fundoplication or herniation of the fundoplication into the posterior mediastinum.

Paraesophageal hernia occurs following inadequate approximation or disruption of the crural repair.

Gas bloat, hiccup, retching, and dumping symptoms are usually transient.

Adhesion intestinal obstruction is particularly common if an additional intra-abdominal procedure such as gastrostomy, incidental appendectomy, or correction of malrotation is performed. *Note*. It is important to alert the parents to the danger of intestinal obstruction, as the inability to vomit may result in inordinate delay in establishing the diagnosis.

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36

Laparoscopic fundoplication

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HISTORY

Laparoscopic fundoplication has become the gold standard for the correction of gastroesophageal reflux and is one of the commonest elective pediatric laparoscopic procedures performed today. The indications for laparoscopic fundoplication are the same as those for open fundoplication, and include failure of medical treatment, recurrent aspiration, esophagitis, and, in some centers, proven gastroesophageal reflux disease in patients with recurrent respiratory illness or asthma. The main indication for fundoplication in children today is reflux in neurologically impaired children, a legacy of complications from being very low-birth-weight infants.

It is important to appreciate that gastroesophageal reflux disease in these neurologically impaired infants is usually associated with pharyngeal incoordination, poor swallowing, and other motility disorders that may result in a less than ideal outcome, and this must be brought to the notice of the parents, who often expect a fundoplication to be the answer to many of their children's difficulties.

PRINCIPLE OF FUNDOPLICATION

The principle of fundoplication is to produce an antireflux mechanism by wrapping the fundus of the stomach around the esophagus, producing an inkwell effect. Variations on fundoplication include a partial wrap or an anterior fundoplication, but by far the commonest procedure performed in children is the Nissen fundoplication, a complete 360° wrap.

DIAGNOSIS

The diagnosis of gastroesophageal reflux disease is made on barium swallow or esophageal pH study, documenting the frequency and severity of reflux episodes into the lower esophagus over a 24-hour period. A palatogram or videofluoroscopy may be useful in neurologically impaired patients to determine the extent to which swallowing incoordination contributes to aspiration episodes.

SURGICAL TREATMENT

There are few contraindications to laparoscopic fundoplication. Patients with scoliosis and fixed flexion deformities of the hip present special difficulties and may make it impossible to manipulate laparoscopic instruments within a severely distorted abdominal cavity, or to maneuver instruments around permanently flexed limbs.

Previous surgery, such as laparotomy for necrotizing enterocolitis, is not necessarily a contraindication, as the lack of visceral adhesions in these patients is often surprising. The presence and extent of visceroparietal adhesions can be assessed preoperatively by using ultrasound to check for the presence of 'visceral slide'.

Preoperative preparation

It is important to exclude severe constipation in neurologically impaired patients. A distended transverse colon will severely impede surgical access. In these patients, a plain abdominal film is helpful in evaluating the need to perform a bowel washout prior to surgery.

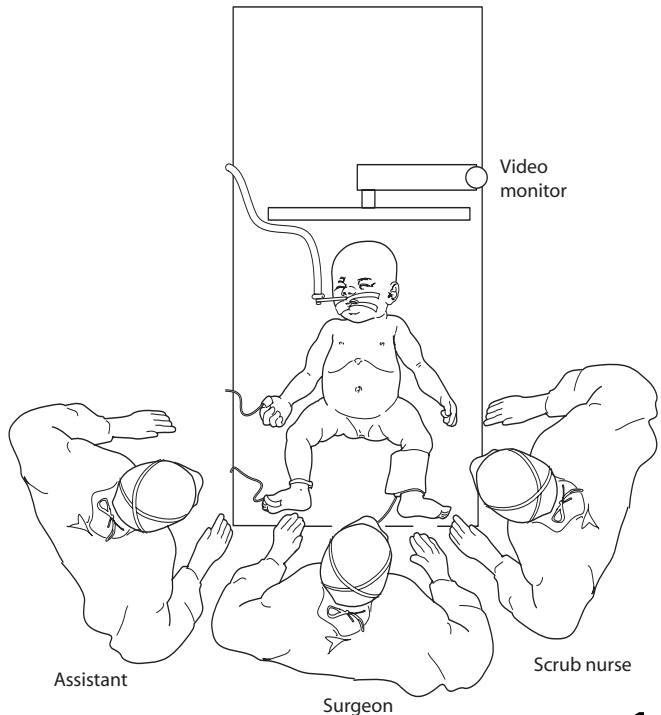
Anesthesia

General anesthesia with full relaxation and endotracheal intubation is mandatory in children undergoing any form of laparoscopic surgery. The small attendant risk of breeching the left pleural cavity during esophageal mobilization and causing a tension pneumothorax is an additional reason for having complete airway control.

A large nasogastric tube is inserted to ensure that the stomach is empty for the duration of the operation.

Patient positioning

1 The patient is positioned at the foot of the bed as per floor plan and the video monitor is placed at the head of the table. The table should be tilted head-up about 20–30°.



1

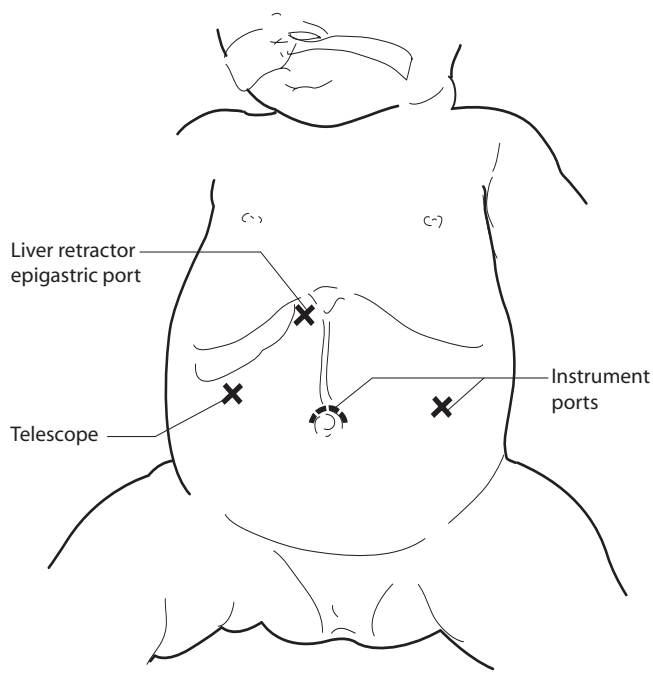
Laparoscopic instrumentation

The following are the instruments required for laparoscopic fundoplication.

- 30° 5 mm telescope.
- 7 mm Hasson cannula.
- Liver retractor (tooth ratcheted grasper).
- Tissue graspers (Reddick–Olsen).
- Needle holder.
- Scissors.
- Ultrasonic scalpel and/or bipolar scissors.

Port placements

2 The operation is performed with four ports. A primary port for the telescope (7 mm Hasson) is usually placed in the umbilicus. An instrument port is sited in the epigastrium for a liver retractor, and two additional ports, one in each upper quadrant, are required for the laparoscopic instruments. Some surgeons may use an additional port in the left paracolic gutter below the left instrument port to grasp the stomach, but with increasing experience this is not necessary.



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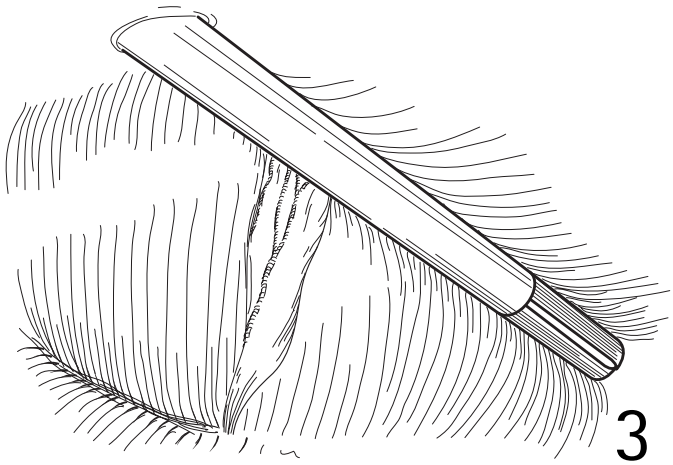
Liver retraction

3 As with the laparoscopic Heller's procedure, the left lobe of the liver can be easily retracted with a toothed, ratcheted grasper. The instrument is curled around the falciform ligament just at the point where the ligament attaches to the anterior edge of the liver, and the falciform ligament is lifted toward the anterior abdominal wall. This single action will lift the liver off the underlying viscera.



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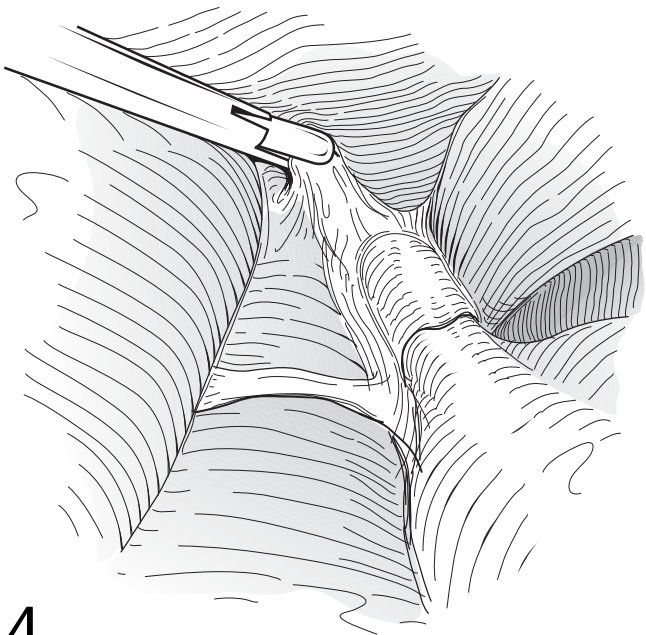

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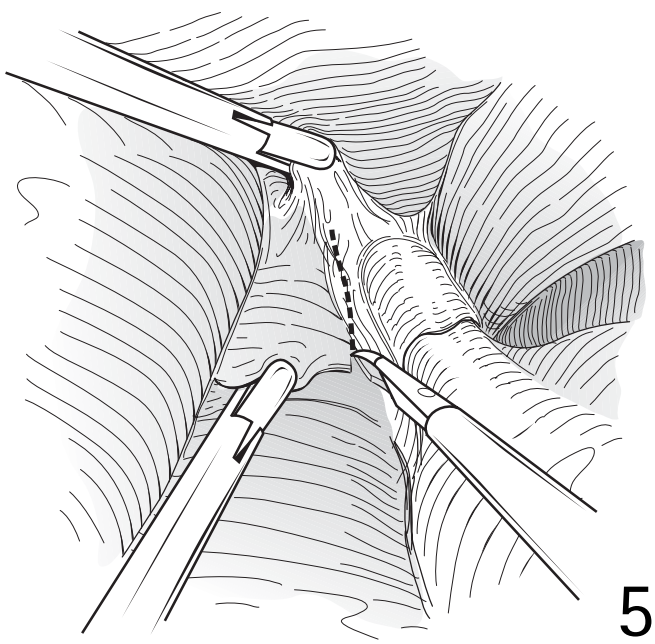
4 The instrument is directed toward the hiatus and fixed to the diaphragm by grasping the muscular diaphragm just above the hiatus. It can then be left in situ without any further attention paid to it during the operation. It may occasionally be necessary to introduce a second liver retractor in the left subcostal region if the liver proves to be very floppy.

Exposure of cardioesophageal junction

5 The key to this is to open the phrenoesophageal ligament and gastrohepatic omentum overlying the right crus. The right crus and esophagus are easily identified in this window created in the phrenoesophageal ligament.



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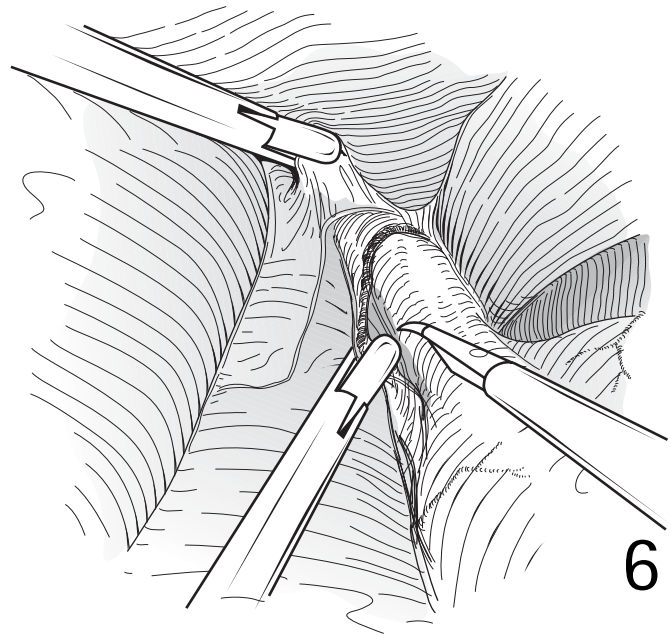
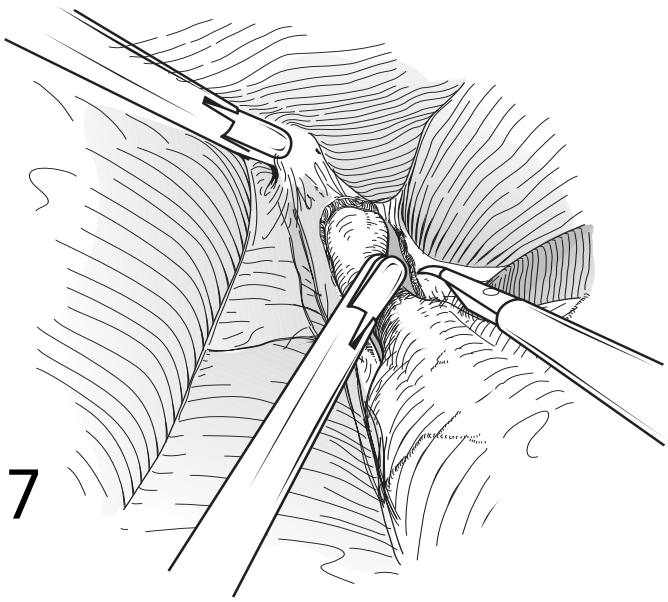
Crural exposure

6 The free edge of the right crus can then be separated from esophagus by a combination of sharp and blunt dissection. The entire intra-abdominal esophagus should be exposed and separated from its surrounding adventitia and visceral peritoneum.



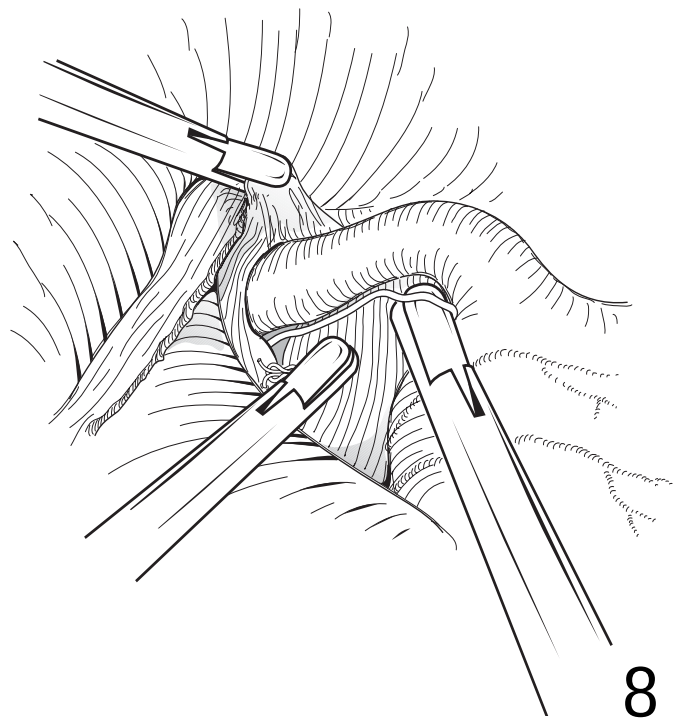


7 The left crus can be identified by following the edge of the right crus over the 'white line' where it merges with the left crus.

The edge of the left crus should be exposed and separated from the esophagus. Some loose adventitial tissue attaching the fundus to the diaphragm and left crus can be identified by gentle downward traction on the fundus. These attachments must be divided to create the posterior window for the fundus to be pulled through. There may be some bleeding to control.

Creation of posterior esophageal window

8 The posterior esophageal window is created by opening the gastrohepatic omentum. The esophagus is lifted anteriorly and the posterior vagus nerve identified. The posterior vagus nerve with the esophagus should be lifted anteriorly and a posterior window developed below the left crus. The left crus must be positively identified before creating the posterior window, otherwise there is a risk of breeching the left pleura, with resultant left tension pneumothorax, if a mistaken attempt is made to develop the posterior window above the left crus.



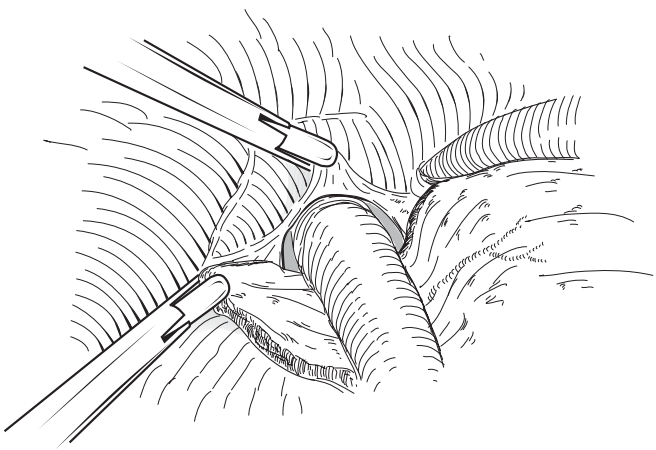
Danger zones

The two greatest dangers in laparoscopic fundoplication are perforating the esophagus and tension pneumothorax during this maneuver. The risk of esophageal perforation is minimized by identifying the posterior vagus and developing the window posterior to the vagus. If the patient develops a tension pneumothorax due to an inadvertent breech of the pleura, an intercostal drain is inserted in the left thoracic cavity to drain the pneumothorax and is left in position for the duration of the operation. The chest tube should be removed at the end of fundoplication, only *after completely desufflating the abdomen*.

The posterior crural gap is repaired with a single non-absorbable suture just where the right and left crura meet (see Illustration 8) and the posterior window is enlarged to allow the fundus to be pulled through unimpeded.

Fundal mobilization

The fundus can usually be identified through the posterior esophageal window created, and it should be grasped, and brought through the posterior window. This is considerably easier without a bougie in the esophagus, as recommended by some surgeons.



9

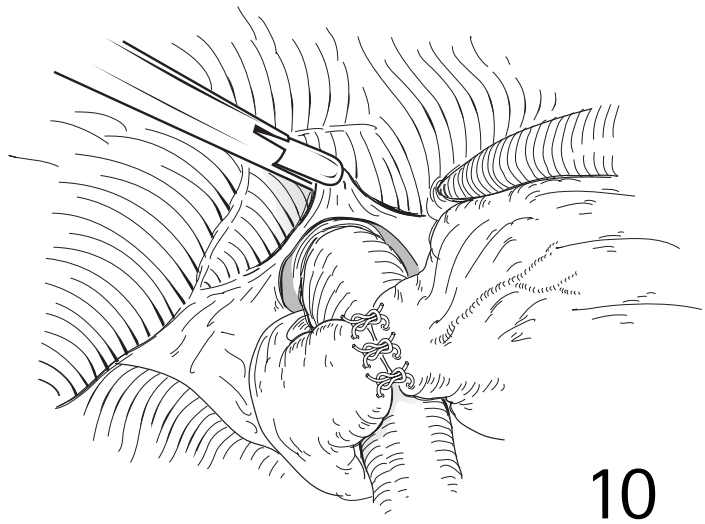
Shoe-shine maneuver

9 A visual evaluation with the aid of the shoe-shining technique – pulling on the fundus on each side of the window – will determine if there is sufficient fundus to perform a loose wrap.

Completing the wrap

A non-absorbable monofilament suture is then used to perform the wrap. Do not be concerned if this first suture appears too tight or if it appears to be in an unsatisfactory position. The first suture can be replaced after placing a second suture in a more suitable position. Do not anchor this first suture to the anterior wall of the esophagus for the same reason. This will then allow you to move the wrap along the esophagus to determine its optimal position. The wrap should be lifted away from the esophagus after this first suture has been placed, to determine if the wrap is sufficiently floppy. You should be able to lift the wrap off the esophagus with a clear 1 cm-gap window between the wrap and anterior wall of the esophagus. This is important in reducing the risk of postoperative dysphagia.

10 Once the correct position for the wrap has been determined, the wrap can be completed with three interrupted, non-absorbable sutures, anchoring at least one of the sutures to the anterior esophageal wall to prevent it from sliding. There should not be any tension on the fundus or the sutures.



10

The abdomen should then be desufflated and the fascia closed with absorbable sutures. The skin can be closed with Dermabond.

POSTOPERATIVE CARE

The nasogastric tube should be left in overnight and removed the following morning. Patients should remain on a sloppy, semi-liquid diet for the first few weeks after fundoplication, gradually increasing the consistency of the food.

OUTCOME

Laparoscopic fundoplication generally has a good outcome, except in patients with neurologic impairment in whom the incidence of wrap migration, disruption, and failure to correct reflux is higher.

The commonest immediate postoperative complication is dysphagia, which, if sufficiently symptomatic, can be relieved with an early single balloon dilatation. If symptoms persist and there is evidence of a tight wrap, it is generally easier to redo the fundoplication within the first week.

ACKNOWLEDGEMENT

Mr S Cascio FRCS for assistance with the further reading list.

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37a

Achalasia

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HISTORY

Achalasia was first described by Willis in 1672. He treated the patient by fashioning a rod of a whale bone with a sponge on the end with which the patient was able to force food into his stomach. In 1877 Zenker and von Ziemssen, and in 1884 Mackenzie, suggested that achalasia was due to diminished contractile power of the esophageal musculature. In 1888 Meltzer and Mikulicz independently postulated that spasmodic contraction of the cardiac sphincter was the etiologic factor. In the same year, Einhorn proposed that the condition was due to failure of relaxation of the cardia on swallowing.

PRINCIPLES AND JUSTIFICATION

Achalasia is a motility disorder of the esophagus characterized by an absence of peristalsis and a failure of relaxation of the lower esophageal sphincter. The cardinal symptoms in childhood are vomiting, dysphagia, chest pains and recurrent respiratory infections, and weight loss. The child learns to eat very slowly and to drink large quantities of fluid to encourage food to enter the stomach. At first there is only regurgitation of food, but later vomiting of undigested food eaten days earlier occurs. The child with achalasia is often first referred to a psychiatrist for treatment of food aversion or anorexia.

Histopathology

Strips of muscle from the distal esophagus reveal varying pathologies from complete absence of ganglion cells to chronic inflammatory changes through to normal ganglia. Histochemistry reveals a significant reduction in all neuropeptides, particularly vasoactive intestinal polypeptide, galanin, and neuropeptide Y.

Treatment

MEDICAL TREATMENT

Transient relief of symptoms can be achieved with nifedipine, a calcium antagonist that reduces the pressure at the lower esophageal sphincter.

FORCEFUL DILATATION

The aim of this treatment is to physically disrupt the muscle fibers of the lower esophageal sphincter by means of pneumatic or balloon dilatation. A fluid-filled (Plummer) or air-filled (Browne–McHardy, Rider–Moller, angioplasty catheter) bag of fixed diameter, or the balloon dilator, is radiologically positioned in the distal esophagus and gently inflated. Relief of symptoms in children is at best temporary but may occasionally last for prolonged periods. Recently, it has been shown that botulinum toxin injected into the lower esophageal sphincter musculature results in symptomatic relief, but the effect is short lived.

SURGICAL TREATMENT

The basis of all surgical procedures is the cardiomyotomy described in 1914 by Heller. Controversies concern the length of the myotomy, the extent to which the myotomy extends onto the stomach, and the necessity for an antireflux procedure.

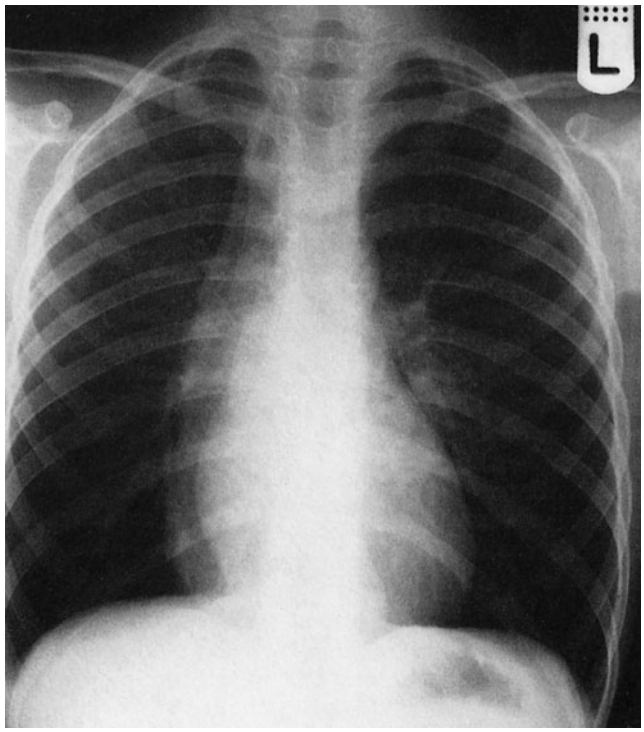
The principle of the procedure is to perform a myotomy over the distal 4–6 cm of esophagus, extending the incision for 1 cm onto the anterior wall of the stomach. The myotomy is covered by a short, floppy Nissen fundoplication to protect against subsequent gastroesophageal reflux.

PREOPERATIVE

Diagnosis

RADIOLOGIC FEATURES

1 A plain chest radiograph may show a dilated, food-filled esophagus with an air–fluid level. There may be radiologic signs of recurrent aspiration pneumonitis.



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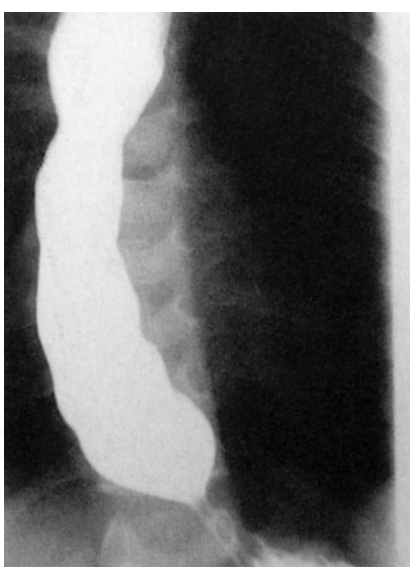


2

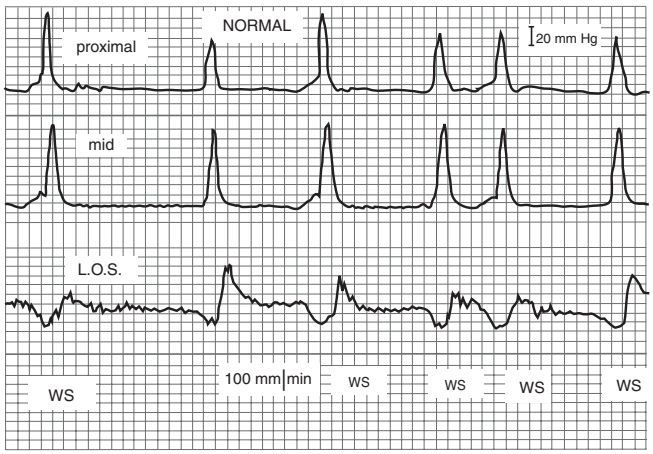
2 The diagnostic features of achalasia on barium swallow are a dilated esophagus, absence of stripping waves, incoordinated contraction, and obstruction at the gastroesophageal junction with prolonged retention of barium in the esophagus. Failure of relaxation of the lower esophageal sphincter gives rise to the classical 'rat-tail' deformity of funneling and narrowing of the distal esophagus.

ENDOSCOPY

The main value of esophagoscopy is to exclude an organic cause for the obstruction.

ESOPHAGEAL MANOMETRY

3a,b The criteria for diagnosis include: (1) a high-pressure (> 30 mmHg) lower esophageal sphincter zone; (2) failure of the lower esophagus to relax in response to swallowing; (3) absence of propulsive peristalsis; and (4) incoordinated tertiary contractions in the body of the esophagus.



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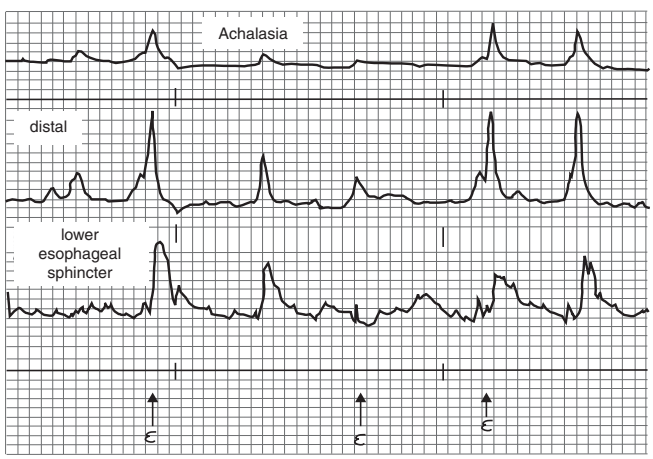


3b

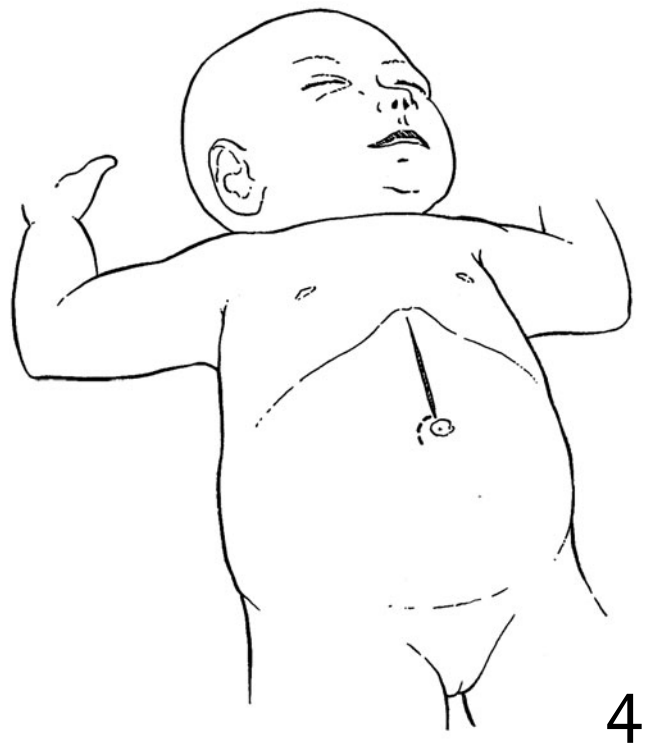
Anesthesia

General endotracheal anesthesia is administered, with the patient supine on the operating table. Measures must be taken to avoid aspiration of esophageal contents during the induction of anesthesia. Preoperative esophagoscopy is recommended to ensure complete evacuation of retained food and secretions from the esophagus. A medium-caliber nasogastric tube is passed into the stomach.

OPERATION

Incision

4 The approach is via an upper abdominal midline incision extending from the xiphisternum to the umbilicus.



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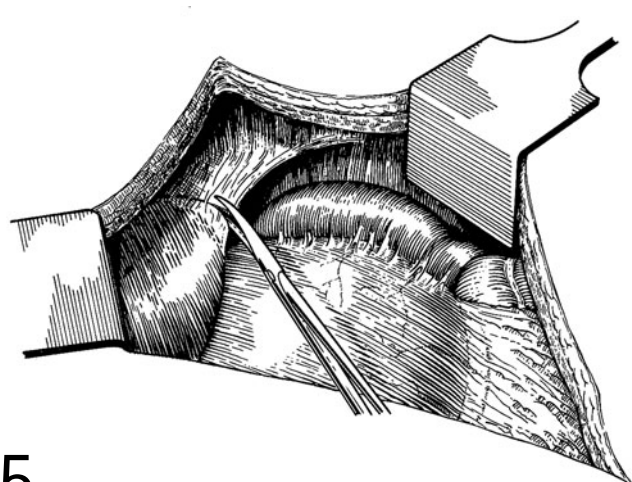
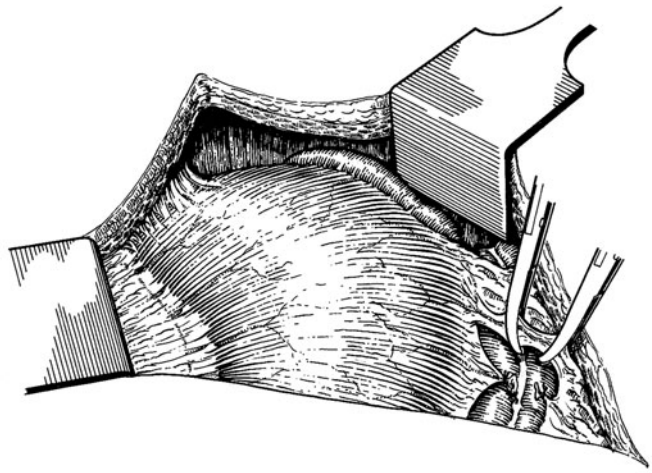


5

Exposure

5 In most cases, adequate exposure of the abdominal esophagus can be obtained by retracting the left lobe of the liver anterosuperiorly with a wide retractor. If necessary, additional exposure may be attained by dividing the left triangular ligament in the avascular plane and retracting the left lobe of the liver towards the midline.

Mobilization of fundus of stomach



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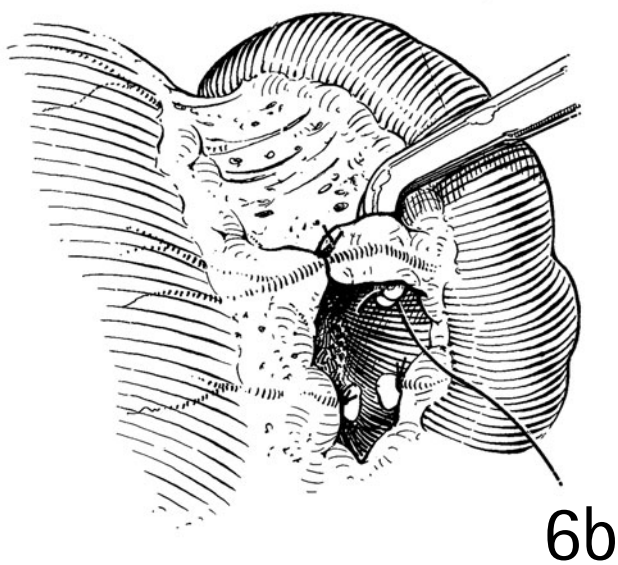


6b

6a,b As a Nissen fundoplication will be performed in addition to the extended gastroesophageal myotomy, the operative procedure for fundoplication should be followed at an early stage.

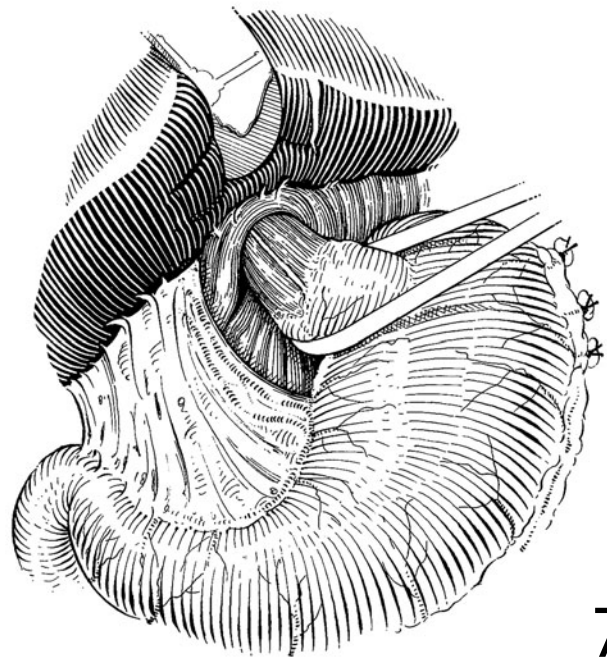
The proximal one-third of the greater curvature of the stomach is liberated from its attachment to the spleen by ligating or coagulating with bipolar diathermy and dividing the short gastric vessels in the gastrosplenic ligament. This is accomplished most safely using a right-angled forceps passed around each vessel in turn. When the vessels in the upper part of the gastrosplenic ligament have been divided, the spleen should be allowed to fall back into the posterior peritoneum, thereby avoiding inadvertent trauma. Splenectomy should never be necessary in this procedure. The fundus is now sufficiently free to allow for a loose (floppy) fundoplication. The esophageal hiatus is completely exposed by dividing the upper part of the gastrohepatic omentum above the left gastric vessels.

Exposure of esophageal hiatus

The phrenoesophageal membrane is placed on stretch by downward traction on the stomach while the diaphragmatic muscles are retracted superiorly. The avascular membrane is incised with scissors and the musculature of the esophagus displayed. The anterior vagal nerve will be seen coursing on the surface of the esophagus; it should be carefully protected and preserved.

Mobilization of the distal esophagus

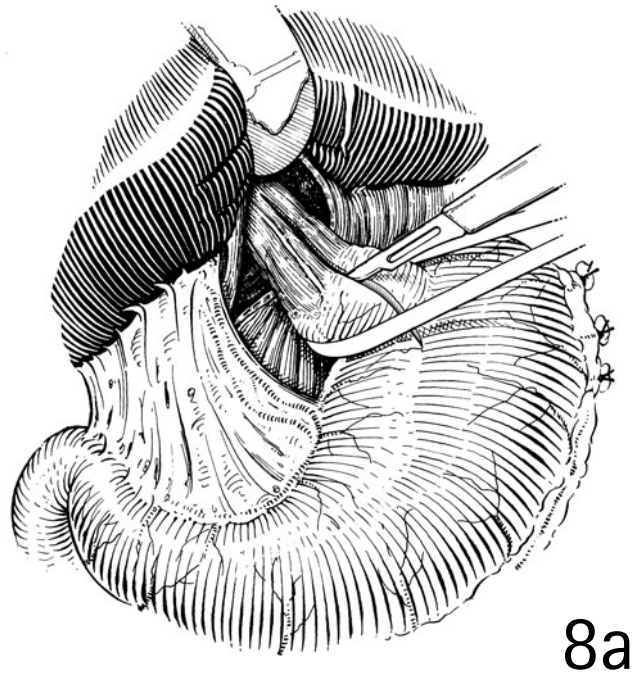
7 Using a combination of sharp and blunt dissection, the lower end of the esophagus is encircled, taking care not to injure the posterior vagal nerve. A rubber sling is placed around the esophagus. The lower 5–8 cm of esophagus is now exposed through the esophageal hiatus into the posterior mediastinum using blunt dissection with either a moist pledget or right-angled forceps.



7

Gastroesophageal myotomy

8a–c The myotomy is performed on the anterior wall of the esophagus, extending for 1 cm onto the fundus of the stomach. A superficial incision (1–2 mm in depth) is made in the musculature of the distal 4–6 cm of the esophagus. The divided muscle is gently parted with a blunt hemostat until the underlying mucosa of the esophagus is encountered. The thickness of the muscle of the lower esophagus varies from a few millimeters to 0.5 cm or more. Great care must be taken to avoid opening into the lumen of the esophagus.



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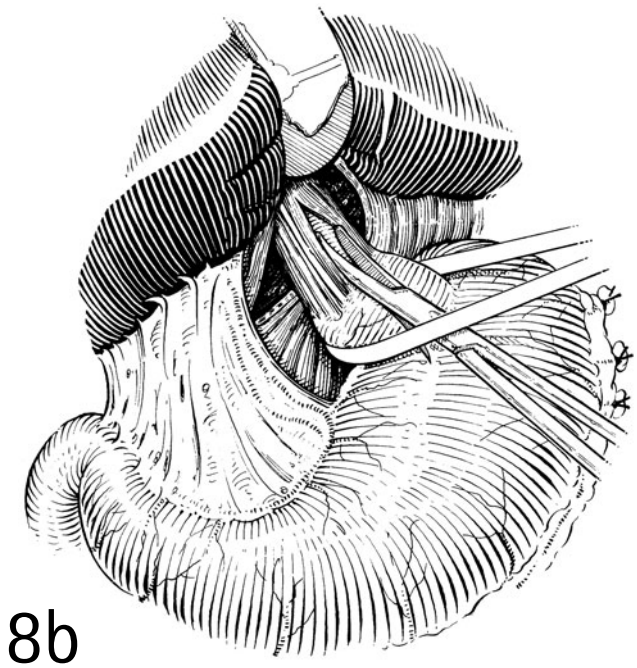

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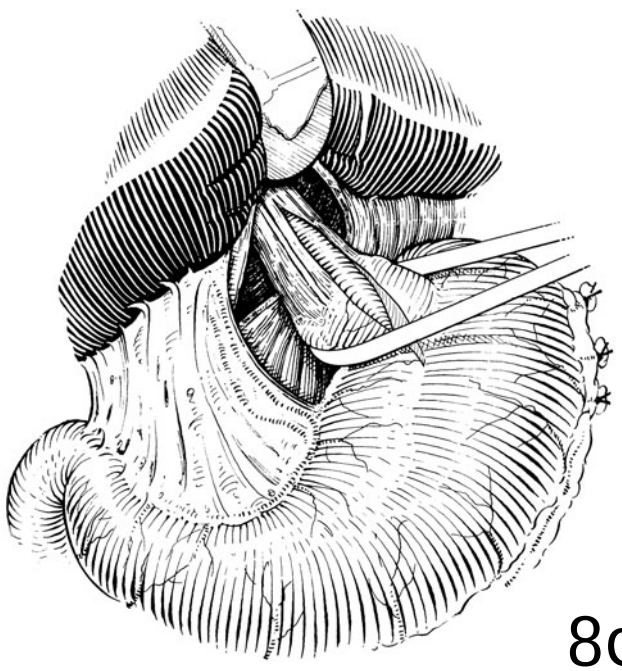

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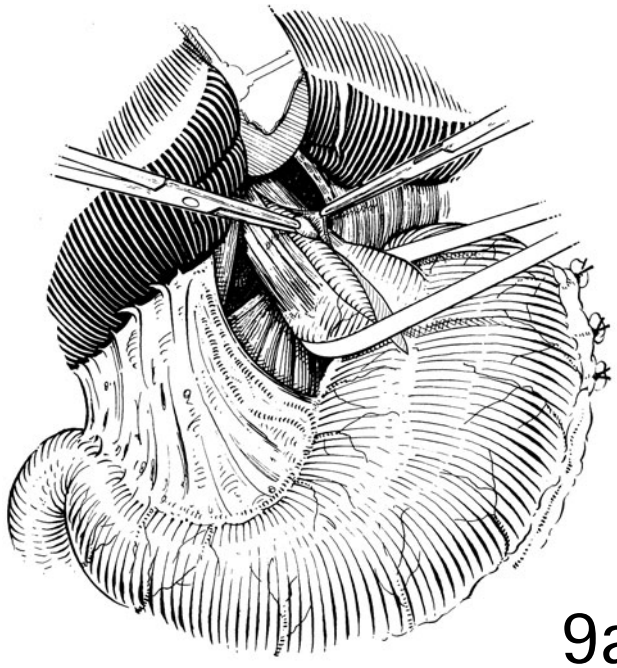

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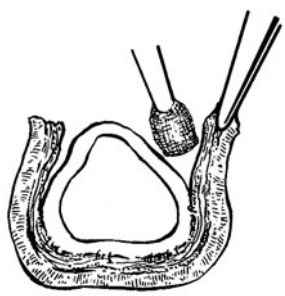


9b

9a,b The divided muscle is now separated from the underlying mucosa by blunt pledget dissection in the submucosal plane. The dissection is continued until at least 50 percent of the circumference of the esophagus is free of the overlying muscle.

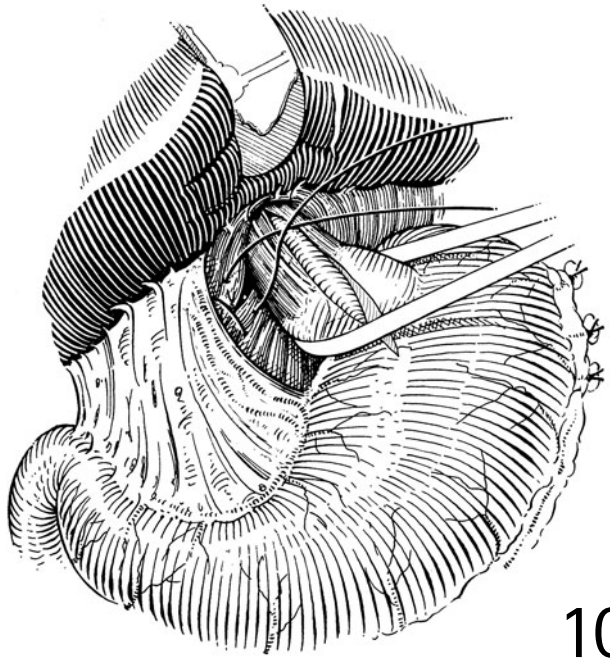
The myotomy is extended through the gastroesophageal junction for 1 cm onto the fundus of the stomach and the musculature is similarly elevated from the underlying mucosa.

Testing for esophageal perforation

The stomach and esophagus are distended with air introduced through the nasogastric tube, and the exposed mucosa is carefully inspected for perforation. A mucosal defect should be carefully closed with fine polyglycolic acid sutures.

Narrowing of hiatus

10 The esophageal hiatus is narrowed posteriorly to the esophagus by placing deep sutures through the crura of the diaphragm. The sutures are tied loosely to prevent them from cutting through, leaving sufficient space alongside the esophagus to allow passage of the tip of a finger. Two or three sutures may be required for this purpose.



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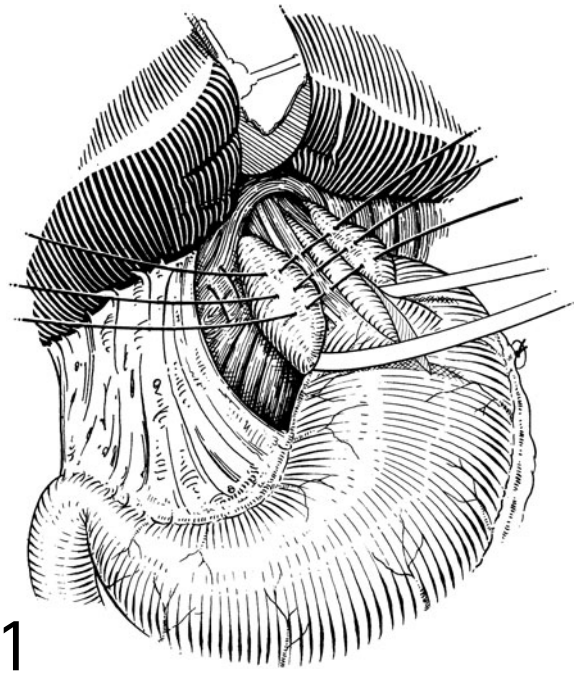

11

Fundoplication

11 A loose (floppy) Nissen fundoplication is now constructed over the distal 1–1.5 cm of the esophagus. The esophageal sutures are only placed through one side of the divided esophageal muscle in order to prevent re-approximation of the edges of the myotomy (see Chapter 35).

Wound closure

The wound is closed either in layers or with interrupted en masse sutures of 3/0 polyglycolic acid. A subcuticular suture approximates the skin edges.

POSTOPERATIVE CARE

Nasogastric decompression and intravenous fluids are continued until the postoperative ileus has resolved (mean of 3–4 days).

COMPLICATIONS

These can include mediastinitis due to failure to detect a mucosal perforation, and recurrence of symptoms if the muscle is not separated from the underlying mucosa for at least half the circumference of the esophagus. Gastroesophageal reflux is due to an inadequate fundoplication, and dysphagia for solids is due to too tight a fundoplication.

OUTCOME

After myotomy alone without an antireflux procedure, the long-term incidence of gastroesophageal reflux is around 15 percent. Relief of the dysphagia and respiratory problems is usually complete, but residual or recurrent pain may occur in 25 percent of patients and is due to diffuse esophageal spasm. The esophageal pain generally responds to pneumatic dilatation.

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37b

Laparoscopic Heller's cardiomyotomy

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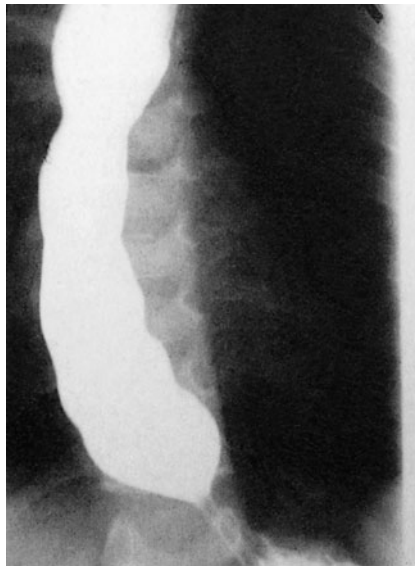
HISTORY

Achalasia is uncommon in children, with an incidence of 0.1 cases per year per 100 000 population under the age of 14 years. While it usually presents as a primary esophageal dysmotility disorder, it may be associated with the Triple A syndrome of achalasia, alacrima, and adrenocorticotrophic hormone insensitivity, or its variants.

The usual presenting symptoms are dysphagia, chest pains, regurgitation of undigested food (often mistaken for 'vomiting'), recurrent chest infections, and weight loss. These symptoms result from failure of relaxation of the lower esophageal sphincter leading to poor esophageal emptying and resultant progressive dilatation of the proximal esophagus. The symptoms can be mistaken for gastroeosophageal reflux disease, and some patients are in fact managed for this until the definitive diagnosis is established.

DIAGNOSIS AND ETIOLOGY

1 The diagnosis is made on a barium swallow with the classical appearance of a dilated proximal esophagus, with tapering of the lower esophagus into the so-called 'rat's tail' appearance. The diagnosis should be confirmed on esophageal manometric studies.



1

The etiology of achalasia is unknown, but patients with this condition have anomalies in the eosophageal musculature as well as in the innervation of the esophagus, of varying intensity to complete aganglionosis. A similar condition is encountered in Chagas' disease, a trypanosomiasis infestation that causes degeneration of the neurons of the enteric nervous system.

MEDICAL TREATMENT

Several forms of treatment are now available. Nifedipene, a calcium antagonist, can produce transient relief by lowering the lower esophageal sphincter pressure, but this does not provide long-term benefit.

Botox injection

Botulism toxin injection into the lower esophagus is effective for symptomatic relief, but the relief is usually temporary and multiple courses of injection are normally necessary. Botox injection is also associated with a significantly higher incidence of mucosal perforation should these patients require surgical intervention, mainly due to scarring and the resultant loss of tissue planes.

Radial balloon dilatation

Radial balloon dilatation has been used for the relief of symptoms with variable results. The best result is obtained with this technique when the lower eosophageal sphincter is overstretched by introducing two radial balloon dilators side by side. The lower esophageal sphincter is maximally dilated to the point of disrupting the lower esophageal musculature. In our experience, it is always necessary for patients to undergo a series of repeated dilatations to achieve significant symptomatic relief. Unlike Botox injection, however, repeated courses of dilatation do not appear to increase the incidence of mucosal perforation at subsequent surgery if dilatation fails.

SURGICAL TREATMENT

Cardiomyotomy remains the main surgical method for managing achalasia. The principle is to perform a 5 cm myotomy at the lower end of the esophagus, preserving the integrity of the mucosa.

Heller's cardiomyotomy may be performed via either a thoracoscopic or a laparoscopic approach. The thoracoscopic approach is associated with a high incidence of postoperative reflux, and it can be difficult to gain adequate endoscopic access to the intra-abdominal esophagus. The transabdominal laparoscopic approach is now widely accepted as the method of choice.

There is also debate about whether the myotomy should extend onto the stomach and also whether one should perform an antireflux procedure. The Dors anterior fundoplication is the commonest procedure performed in conjunction with Heller's cardiomyotomy, but a floppy Nissen's fundoplication has also been described.

Laparoscopic cardiomyotomy can be performed in situ, without detaching the esophagus from its abdominal attachment. An in-situ operation minimizes the risk of postoperative reflux if the attachments of the esophagus are left largely undisturbed.

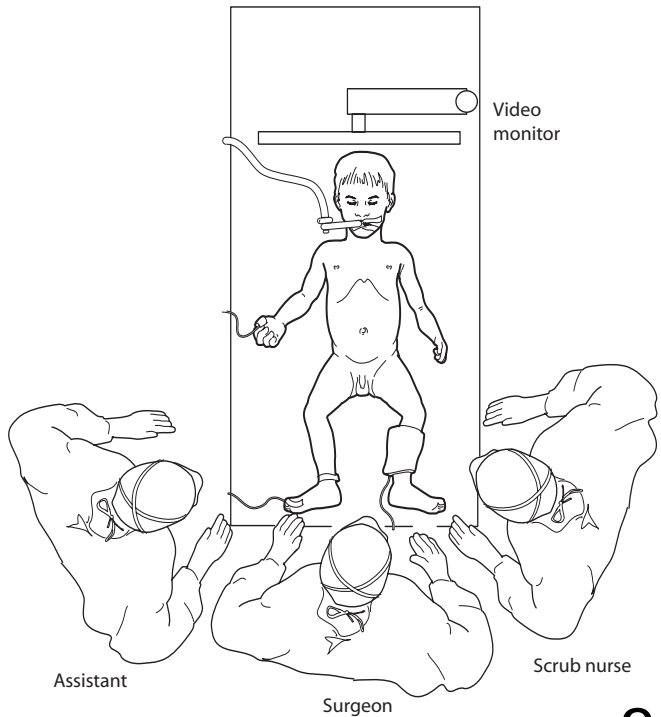
Anesthesia

General anesthesia with full relaxation and endotracheal intubation is mandatory. As with fundoplication, there is a small risk of breaching the left pleural cavity during dissection of the anterior wall of the mediastinal esophagus, and a tension pneumothorax may ensue.

A 10 Fr nasogastric tube is inserted to ensure that the stomach is empty for the duration of the operation. It is not necessary to perform a concurrent gastroscopy.

Patient positioning

2 The patient is positioned at the foot of the bed as per fundoplication and the video monitor is placed at the head of the table.



2

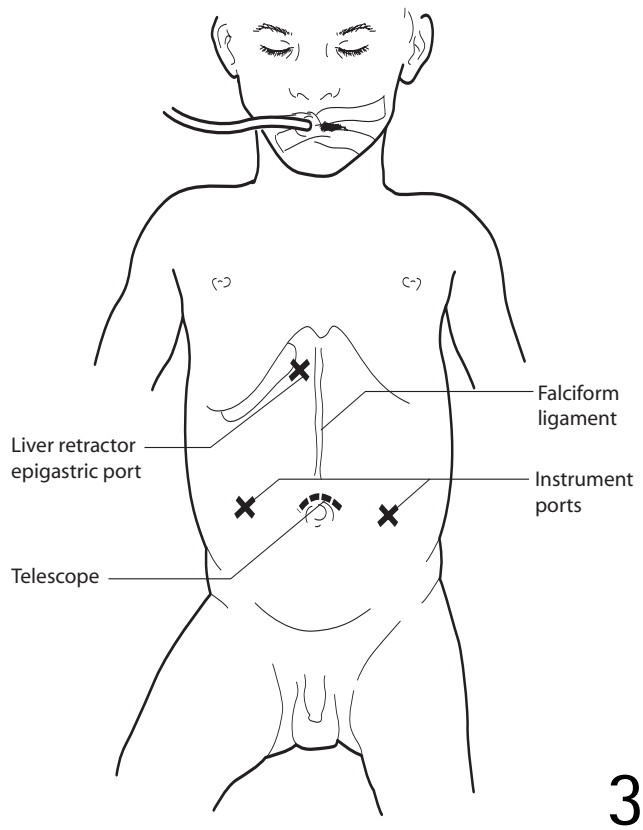
Laparoscopic instrumentation

The following instruments are required for laparoscopic Heller's cardiomyotomy.

- 30° telescope, 5 mm telescope.
- 7 mm Hasson trocar and cannula.
- 5 mm trocars and cannulae.
- Liver retractor (toothed, ratcheted grasper).
- Tissue forceps.
- Needle holder (standby).
- Bipolar scissors.
- Needlepoint bipolar diathermy forceps.

Port placements

3 The 7 mm Hasson port for the telescope is usually inserted in the umbilicus with an open laparoscopy technique. An instrument port is placed in the epigastrium one finger breadth below the subcostal margin just to the right of the falciform ligament for the liver retractor. Two other instrument ports are inserted, one in each upper quadrant.



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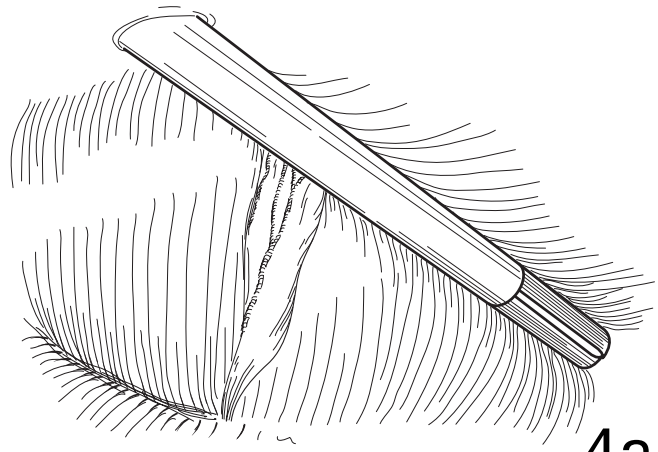
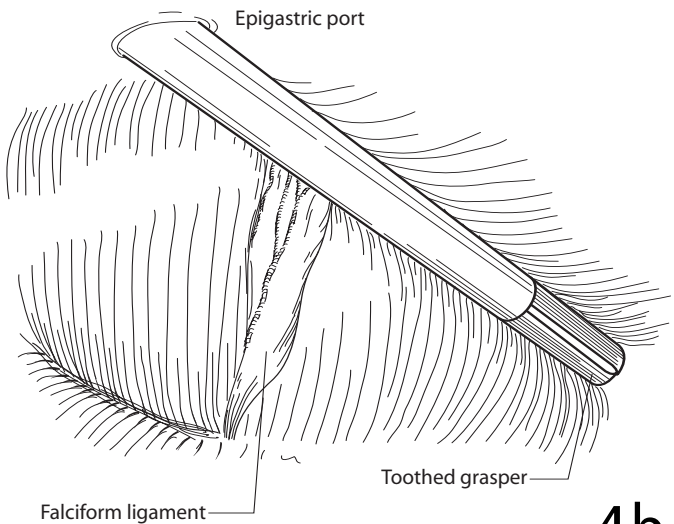


4a

Liver retraction

4a,b As with laparoscopic fundoplication, the left lobe of the liver can be easily retracted with a single toothed ratcheted grasper. The instrument is curled around the falciform ligament just at the point where it attaches to the anterior edge of the liver. This maneuver will lift the falciform ligament and the liver off the underlying viscera.



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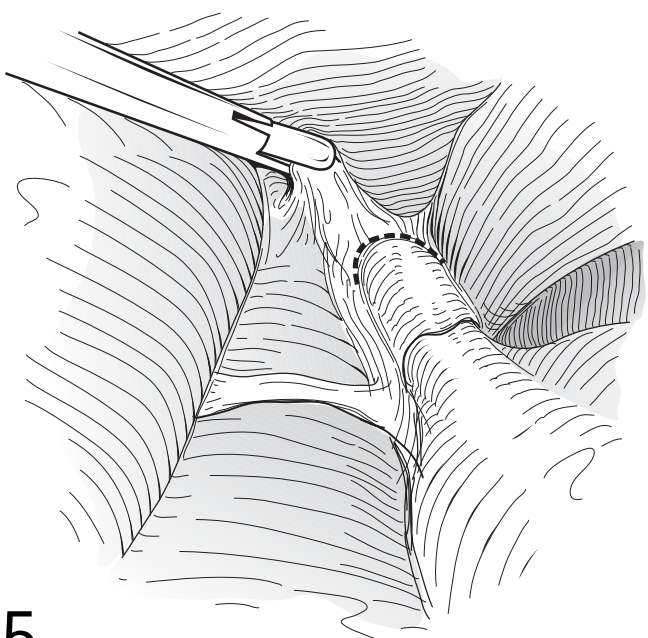
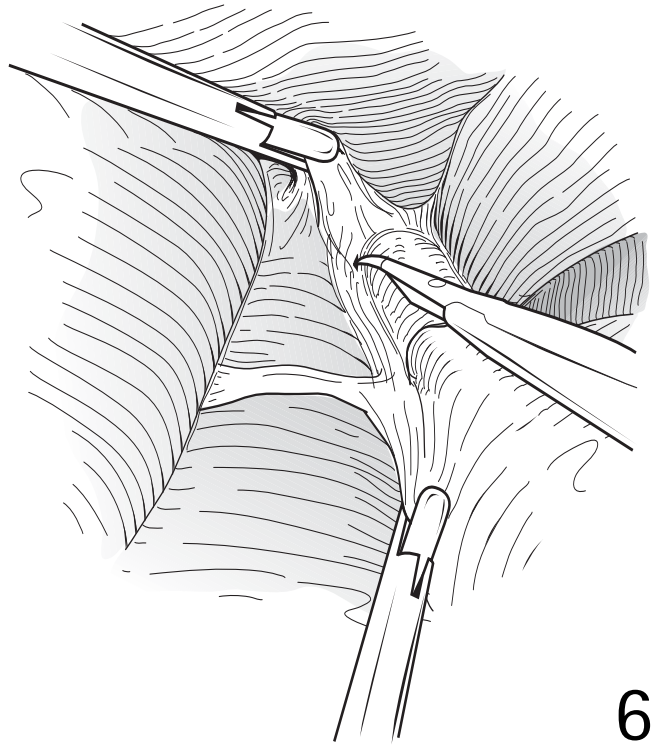


5

5 The grasper is fixed to the muscular diaphragm by grasping it just above the hiatus. It can then be left in situ without any further attention paid to it during the duration of the operation. It may occasionally be necessary to introduce a second liver retractor in the left subcostal region if the liver proves to be very floppy.

Exposure of intra-abdominal esophagus

6 The 'white line' (Illustration 5), which is the edge of the diaphragmatic crura, is easily identifiable by gentle downward traction on the intra-abdominal esophagus or stomach.



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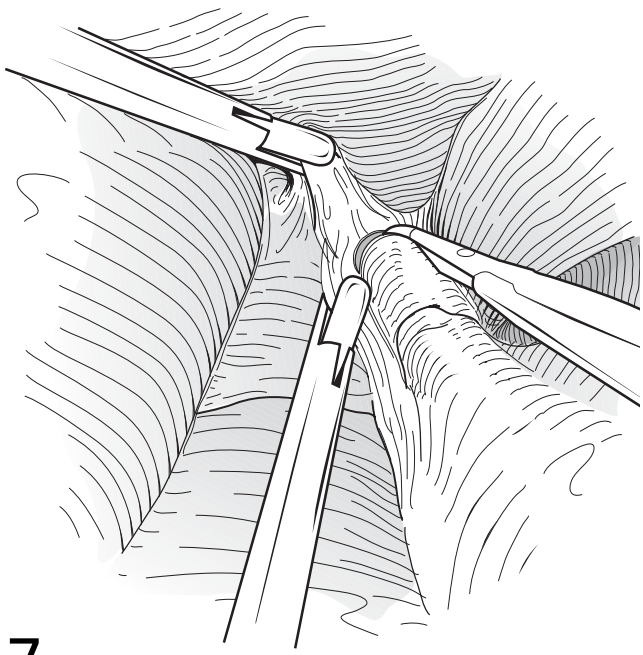

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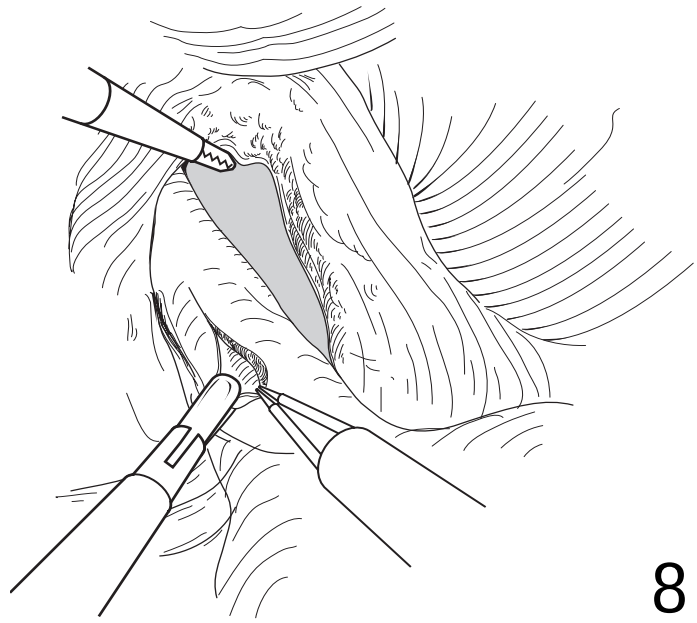
7 Unlike in fundoplication, the phrenoesophageal membrane should be left undisturbed and intact. The parietal peritoneum overlying the 'white line' should be incised from the 10 o'clock to the 2 o'clock position to expose the underlying esophagus. The anterior wall of the esophagus should be exposed from the crus down to the esophagogastric junction. Do not mobilize the lateral attachments of the abdominal esophagus, as this will result in reflux. Some larger vessels near the esophagogastric junction may bleed. The myotomy is performed in situ without mobilizing the abdominal esophagus. The gastrohepatic omentum should be left undisturbed.

Exposure to mediastinal esophagus

8 The crus is lifted away from the esophagus to gain entry into the mediastinal esophagus. An easy plane can be developed between the overarching crus and the esophagus, allowing the esophagus to be exposed in the mediastinum for about 5 cm. It is neither possible nor advisable to identify the inferior pulmonary vein. If necessary, the telescope can be introduced into the hiatus to view the mediastinal esophagus, but simply lifting the crus off the esophagus will usually provide sufficient exposure. It may be necessary occasionally to use an additional instrument for this part of the operation.



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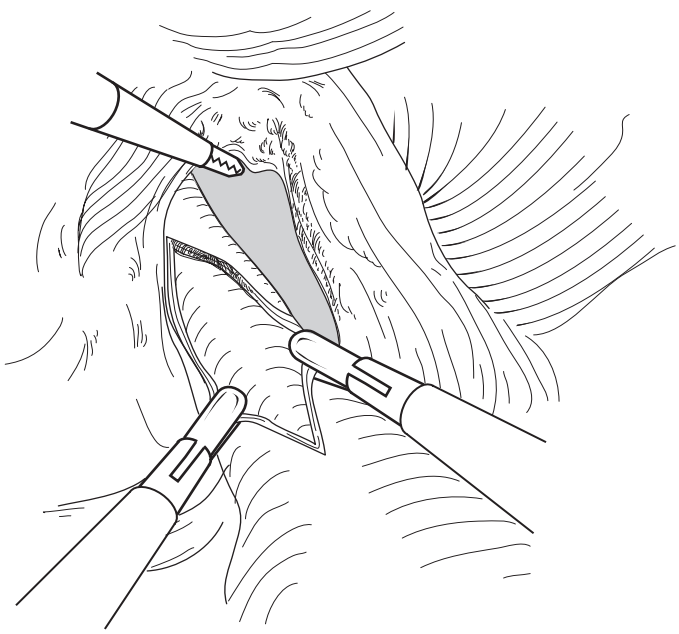
Once an adequate length of intrathoracic esophagus has been exposed, attention should be redirected to the abdominal esophagus.

Esophageal myotomy

An esophageal myotomy is best started on the anterior wall of the esophagus with needlepoint bipolar forceps (Tan bipolar forceps) about 1 cm proximal to the esophagogastric junction (Illustration 8). While it is possible to use monopolar diathermy or an ultrasonic scalpel to make this initial myotomy incision, both these instruments can produce deeper thermal damage, which may cause unrecognized damage of the underlying mucosa resulting in delayed perforation.



9

9 The myotomy can be extended with either bipolar scissors or the needlepoint bipolar forceps. The myotomy can be extended proximally by grasping the edges and gently tearing the muscle apart. The mucosa can be seen to bulge through the muscle defect, and this should be an easy tissue plane. In reality, a combination of bipolar diathermy and tearing is usually necessary. Beware of a tendency to spiral the myotomy off the anterior esophageal wall in the mediastinum.

The muscle should be spread further apart using blunt laparoscopic spreaders to allow the underlying mucosa to pout out as much as possible. The myotomy is extended only to the gastric junction, which can be identified when one sees the edge of circular gastric muscle fibers. Do not extend the myotomy onto the stomach, as this may result in reflux.

The mucosa is inspected for evidence of perforation and if there is concern, methylene blue should be instilled via the nasogastric tube to check for mucosal leak. Any mucosal leak can be repaired with a mucosal suture.

Fundoplication is not performed, because the entire esophagus is left undisturbed except for the small crural window created. The esophageal attachments are essentially undisturbed with this technique.

Patients can usually commence feeding immediately post-operatively, with immediate relief of symptoms.

ACKNOWLEDGEMENT

Mr Nicholas Chao FRCS for assistance with the further reading list.

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38

Gastrostomy

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HISTORY

Gastrostomy is one of the oldest abdominal operations in continuous use and its history is closely associated with the evolution of modern surgery.

Gastrostomies are important in the management of a wide variety of surgical and non-surgical conditions of childhood. Although pediatric surgeons have become more selective in the use of gastrostomies for congenital malformations, there has been a marked increase in the use of feeding stomas in infants and children without associated surgical pathology, mainly those with an inability to swallow secondary to central nervous system disorders. Refinements in traditional procedures and the introduction of newer and simpler endoscopically, radiologically, ultrasonographically, and laparoscopically aided gastrostomies have enhanced the safety and expanded the applicability of this operation. The use of softer, minimally irritating materials in the manufacture of gastrostomy catheters and the development of skin-level gastrostomy devices have greatly facilitated the long-term use of this type of enterostomy.

PRINCIPLES AND JUSTIFICATION

Gastrostomy is indicated in infants and children primarily for long-term feeding, decompression, or a combination of both. It is also commonly employed in conjunction with other interventions, such as antireflux procedures. Additional uses include gastric access for esophageal bougienage, placement of transpyloric jejunal feeding tubes, gastroscopy, and administration of medication.

TECHNIQUES

Three basic methods of constructing a gastrostomy are commonly used: (1) formation of a serosa-lined channel from the anterior gastric wall to the skin surface around a catheter; (2) formation of a tube from full-thickness gastric wall to the skin surface, a catheter being introduced intermittently for feeding; (3) percutaneous techniques, in which the introduced catheter holds the gastric and abdominal walls in apposition. With certain modifications, each of these interventions can be performed by minimally invasive techniques or in conjunction with laparoscopy.

Channel formation around a catheter

In the first group of techniques, the catheter may be placed parallel to (Witzel technique) or perpendicular to (Stamm technique) the stomach with a laparotomy (see below). The stomach is usually anchored to the abdominal wall with sutures. The essence of the Stamm-type gastrostomy is the use of concentric purse-string sutures around the gastrostomy tube, producing an invagination lined with serosa.

Gastric tube brought to the surface

The gastric tube is constructed and then brought to the abdominal wall either as a direct conduit (Depage, Beck–Jianu, Hirsch, and Janeway methods) or interposing a valve or torsion of the tube to prevent reflux (Watsudjii, Spivack techniques). This conduit is secured to the layers of the abdominal wall and/or the skin. The main appeal of the Janeway-type stoma is that the patient does not need a catheter between feedings. The use of automatic stapling devices, including those designed for laparoscopic use, has greatly facilitated the construction of the tube from the anterior gastric wall.

Percutaneous techniques

In this third group the catheter is placed with endoscopic, radiologic, ultrasonographic, or laparoscopic assistance without a laparotomy. The percutaneous endoscopic gastrostomy (PEG) is the most widely employed of these interventions. Depending on the method of introduction of the catheter, PEG may be performed using a pull technique (Gauderer–Ponsky – see below), a push technique (Sachs et al.) in which a semi-rigid catheter guide is advanced over a Seldinger-type wire instead of being pulled into place by a string-like guide from inside the stomach to the skin level, or the introducer technique (Russel et al.) in which a Foley catheter is advanced through a removable sheath from the skin level into the stomach. Percutaneous endoscopic gastrostomy was initially developed for children at high risk who were unable to swallow and was later adapted for use in adults.

Laparoscopic techniques

Laparoscopically assisted gastrostomies are modifications of the above basic types allowing surgeons numerous options either as single interventions or associated with other intracavitary procedures.

INDICATIONS

The type of gastrostomy, the preoperative work-up, the technique, and the choice of gastrostomy device depend mainly on the indications for the procedure, the child's age and underlying disease, and the familiarity of the surgeon with the different operations.

Feeding and administration of medications

Placement of a gastrostomy for enteral feeding has two prerequisites: the upper gastrointestinal tract must be functional, and the need for enteral feedings must be long term, at least several months. Children benefiting from gastrostomy fall into two broad categories: (1) those unable to swallow, and (2) those unable to consume adequate nutrients orally. The first group is the largest and composed primarily of patients with neurologic disturbances. The second group includes patients with a variety of conditions in which the central nervous system is intact: failure to thrive, complex bowel disorders (e.g., short gut syndrome, Crohn's disease, malabsorption), malignancy and other debilitating illnesses, and various congenital or acquired diseases interfering with growth.

In selected patients a gastrostomy is the most effective means of administering a non-palatable special diet (e.g., that used in chronic renal failure) or ensuring compliance with medication (e.g., administering cholestyramine in Alagille's syndrome).

All the gastrostomy types described above are suitable for this purpose (these are compared in Table 38.1). A comparison of devices used in all gastrostomy types (except gastric tube stomas) is given in Table 38.2.

Decompression with or without enteral feeding

When a gastrostomy is placed in conjunction with another intra-abdominal procedure (such as fundoplication or repair

Table 38.1 *Comparison of the most commonly used gastrostomies*

	Serosa-lined channel (Stamm)	Gastric tube (Janeway)	Percutaneous endoscopic technique	Laparoscopic technique
Catheter/stoma device continuously in situ	Yes	No	Yes	Yes
Laparotomy	Yes	Yes	No	No
Laparoscopically feasible	Possible	Yes	Yes	Yes
Need for gastric endoscopy	No	No	Yes	No
Need for abdominal relaxation during operation	Yes	Yes	No	Yes and insufflation
Procedure time	Short	Moderate	Very short	Short
Postoperative ileus	Yes	Yes	No	Some
Potential for bleeding	Yes	Yes	No	Some
Potential for wound dehiscence/hernia	Yes	Yes	No	No
Potential for early dislodgement of catheter	Yes	No	No	Some
Potential for gastric separation	Possible	Possible	Yes	Possible
Potential for infection	Yes	Yes	Yes	Yes
Potential for gastrocolic fistula	Possible	No	Yes	Possible
Incidence of external leakage	Moderate	Significant	Low	Low
'Permanent'	No	Yes	No	No
Suitable for passage of dilators for esophageal stricture	Yes	No	No	Possible
Interferes with gastric re-operation (fundoplication)	No	Yes	No	No
Suitable for infants	Yes	No	Yes	Yes

Table 38.2 *Comparison of commonly used gastrostomy devices*

	de Pezzer, Malecot, T-tube	Foley (balloon type)	Skin-level (button type)
Suitable for initial insertion	Yes	Yes	Yes
Suitable for decompression	Yes	Yes	Yes[a]
Tendency for accidental dislodgement or external migration	Moderate[b]	Moderate	Very low
Tendency for internal (distal) migration	Moderate	High	Unlikely
Tendency for peristomal leakage (particularly large tubes)	Moderate	Moderate	Low
Balloon deflation	No	Yes	Depending on type
Re-insertion	Easy to moderately difficult	Easy	Easy to moderately difficult
Long-term (particularly ambulatory patients)	Adequate	Adequate	Best suited

[a]With special adaptor.
[b]High with Malecot.

of duodenal atresia), the tube is used initially for decompression and then for intragastric feedings. If prolonged gastric or duodenal dysmotility is anticipated, a smaller, more flexible catheter is advanced into the lumen of the jejunum, exiting either along the gastrostomy tube or through a counter-incision.

A palliative decompressive gastrostomy can be of value in the management of patients with intestinal obstruction secondary to unresectable malignancy.

All gastrostomy types, except gastric tubes, are suitable for this purpose.

CONTRAINDICATIONS

Contraindications to PEG are inability to perform upper tract endoscopy safely or to identify transabdominal illumination and see an anterior gastric wall indentation clearly. Anatomic abnormalities such as malrotation or marked scoliosis, ascites, coagulopathy, and intra-abdominal infection, if severe, may render the procedure inadvisable. In such cases, laparoscopic or radiologic control, a fully laparoscopic procedure, or gastrostomy with a conventional laparotomy may be indicated.

PREOPERATIVE

Gastrostomy for feeding

Gastroesophageal reflux, as a manifestation of foregut dysmotility, is a common problem in neurologically impaired children both before and after the placement of a gastrostomy. Evaluation for the degree of reflux in these patients is therefore necessary. Patients with severe reflux are best managed with an antireflux procedure and a gastrostomy. Children with mild or no reflux are candidates for gastrostomy only.

Gastrostomy as an adjunct in children with surgical lesions

The addition of a stoma to the surgical correction of a congenital or acquired lesion should be considered only if it will substantially facilitate perioperative or long-term care. Examples in neonatal surgery include complex esophageal atresia, certain duodenal obstructions, select abdominal wall defects in which long-term ileus is anticipated, and short gut syndrome. Indications in older children include severe esophageal stricture, complex foregut trauma, intestinal pseudo-obstruction, malignancy, and complex adhesive bowel obstruction.

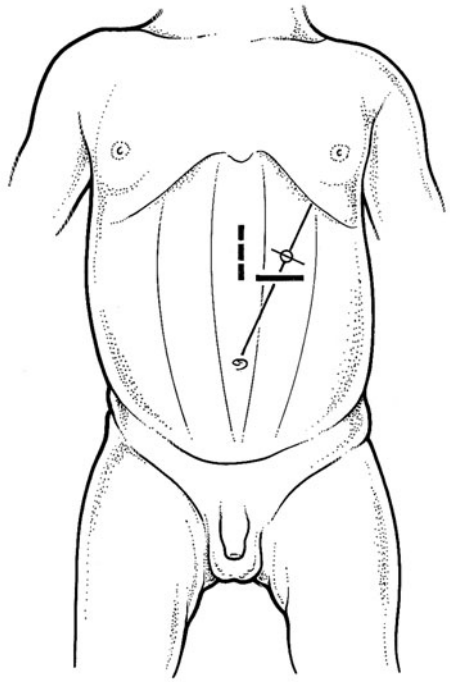
OPERATIONS

Stamm gastrostomy

This operation is performed using general endotracheal anesthesia. The child is positioned with a small roll behind the back to elevate the epigastrium, then prepared and draped. The author prefers to use a silicone mushroom-type catheter ranging in size from 12 Fr (full-term neonates) to 20 Fr for adolescents, and a 10 Fr T-tube or Malecot catheter for preterm infants or neonates with very small stomachs such as those found in children with esophageal atresia without distal fistula. The procedure may be modified slightly to accommodate the initial placement of a skin-level device.

Incision

1 The stomach is approached through a short, left, transverse, supraumbilical incision. Fascial layers are incised transversely and the muscle retracted or transected. The catheter exit site is approximately at the junction of the lower two-thirds and the upper one-third of a line from the umbilicus to the mid-portion of the left rib cage, over the mid-rectus muscle. A vertical incision may be useful in children with a high-lying stomach or a narrow costal angle.



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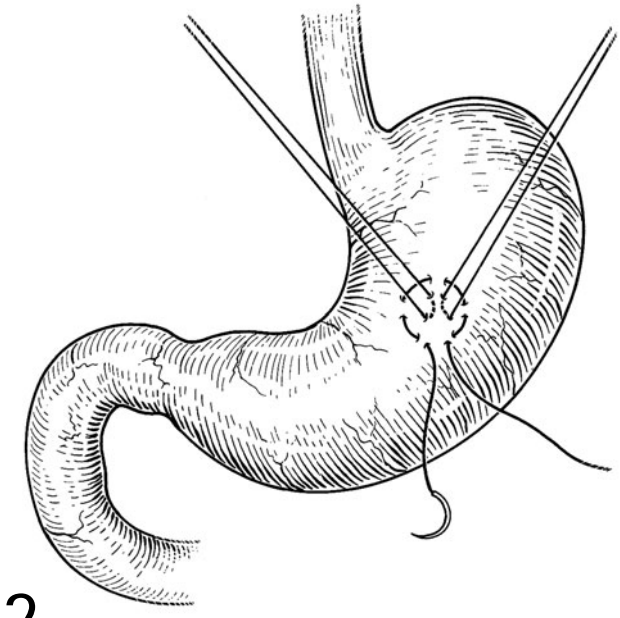

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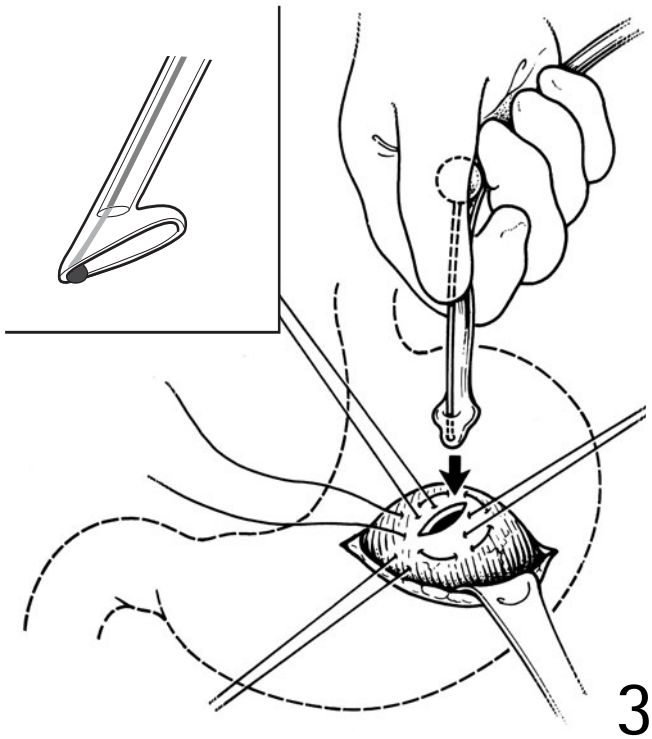
Production of a gastrotomy site on the anterior gastric wall

2 Traction guy sutures and a purse-string suture (synthetic, absorbable) are placed as shown. The opening should be away from the gastric pacemaker at the level of the splenic hilum; away from the greater curvature because that site may be needed for construction of a gastric tube for esophageal replacement; away from the fundus to allow for a possible fundoplication; and away from the antrum to prevent excessive leakage and pyloric obstruction by the catheter tip. If the catheter is to be placed cranially and close to the lesser curvature for a gastrostomy with antireflux properties, care must be taken to avoid the vagus nerve.

3 A lower guy suture pulls the stomach caudally, enhancing exposure and allowing better gastric access. The gastrotomy is performed with fine scissors or cautery while the upper guy sutures are lifted to prevent injuring the back wall. The de Pezzer catheter is introduced using a simple stylet while these sutures are elevated. The insert shows a 'tulip' or 'dome' PEG-type catheter being stretched in the same manner for insertion. These catheters have a reinforced 'notch' for the tip of the stylet.



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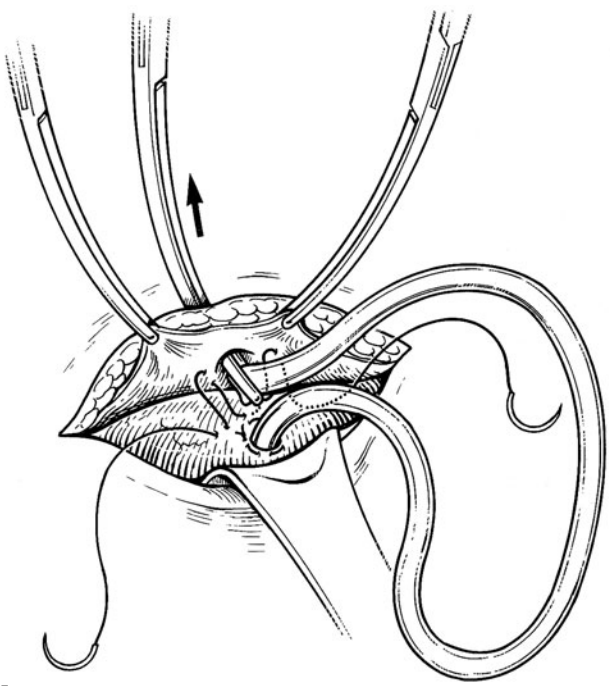
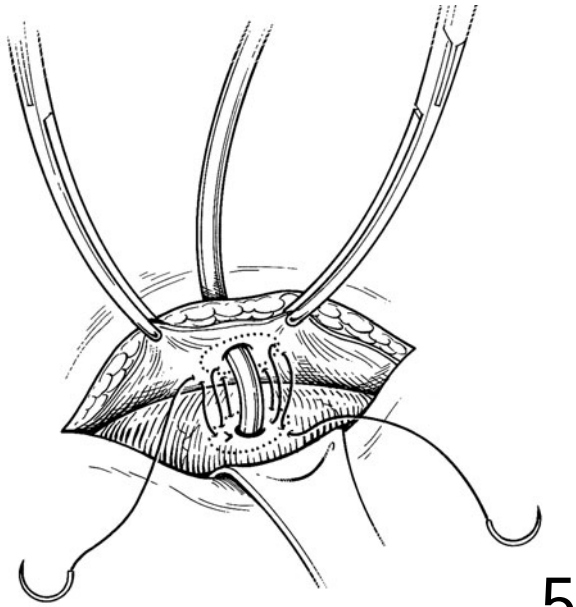


4

4 The purse-string suture is tied. A continuous, synthetic, absorbable, monofilament suture (polydioxanone) is used to anchor the stomach to the anterior abdominal wall. A Kelly clamp is placed through the counter-incision and the abdominal wall layers are pushed inwards. The posterior 180° of the anastomosis are completed, the peritoneum and fascia are incised, and the tip of the clamp is pushed through. The catheter end is grasped and the tube is brought out through the counter-incision.

5 Placement of the continuous monofilament suture is now completed. When tied, this suture provides a 360° fixation with a watertight seal. In most cases, this maneuver obviates the need for a second purse-string suture.



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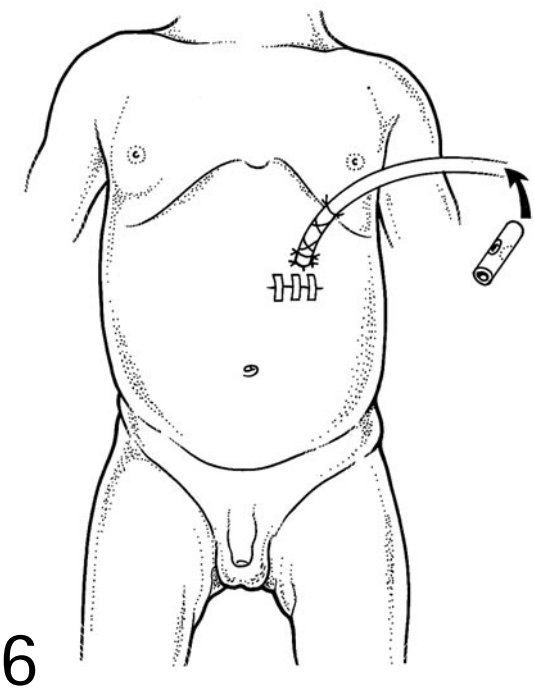

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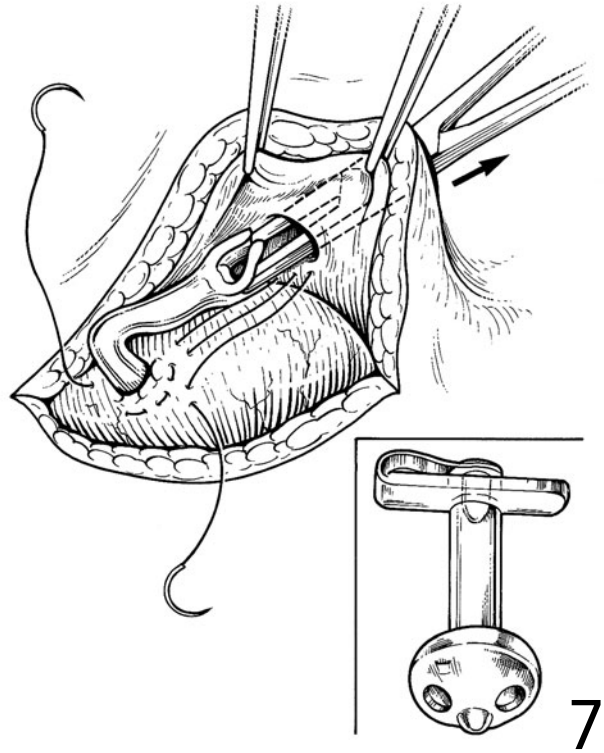
6 The abdominal wall layers are closed with synthetic absorbable sutures and the skin is approximated with subcuticular stitches and adhesive strips. The catheter is secured with synthetic monofilament sutures (polypropylene). These are removed 1–2 weeks after the operation, and a small crossbar is placed to prevent distal catheter migration.

7 This standard procedure may be modified to allow the insertion of a skin-level gastrostomy device, the original 'button' (inset), a balloon-type skin-level device, or a changeable skin-level port-valve. These devices are available in different shaft lengths and diameters. The shaft length should encompass the invaginated gastric wall, the abdominal wall, and an additional few millimeters of 'play' to allow for postoperative edema, ease of care, and subsequent growth and weight gain.



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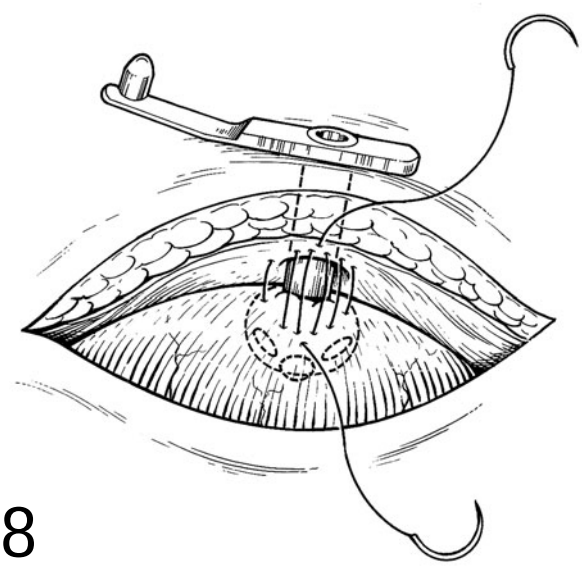

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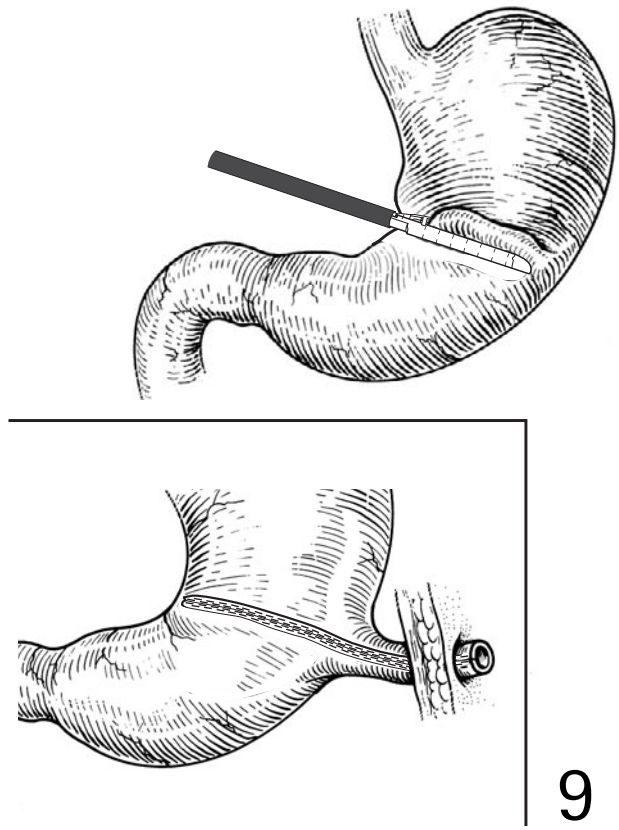
8 The stomach is vented with a nasogastric tube or a special decompression device, which, when inserted in the shaft, deactivates the one-way valve at the gastric end of the shaft.

Janeway gastrostomy

This procedure may be accomplished using either a conventional laparotomy or a gastrointestinal anastomosis stapler or employing an endoscopic stapler under laparoscopic control.

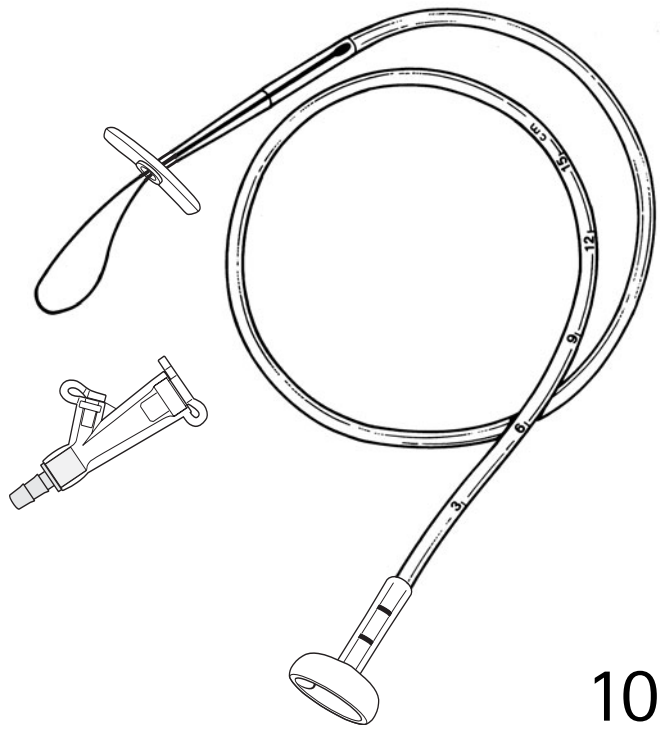


9 The stapler is employed to tubularize the gastric wall. The gastric tube is brought out away from the incision if the open technique is used, or through one of the port sites if it is performed laparoscopically.

Percutaneous endoscopic gastrostomy ('pull' technique)

The procedure is best performed in the operating room. Older children and those able to tolerate endoscopy without compromising the upper airway receive local anesthesia with sedation as needed. Younger children require general endotracheal anesthesia, primarily because of anticipated difficulties with the airway management. A single dose of a broad-spectrum intravenous antibiotic is given shortly before the procedure. For the endoscopy, the smallest available pediatric gastroscope is used. The catheter and retaining crossbar or catheter head should be soft and collapsible enough to glide atraumatically through the oropharynx and esophagus. The 16 Fr silicone rubber catheter depicted in Illustration 10 is suitable for most children. Hybrid catheters for primary implantation of skin-level devices are now also available. The operative field is prepared in the usual sterile manner.

10 A pediatric catheter should be used, with a gastric retainer, markings on the shaft, and dilating tapered end with steel wire loop. Also shown in this illustration are a skin-level retainer (external cross-bar) and a catheter adapter. The catheter is cut to an appropriate length after insertion.



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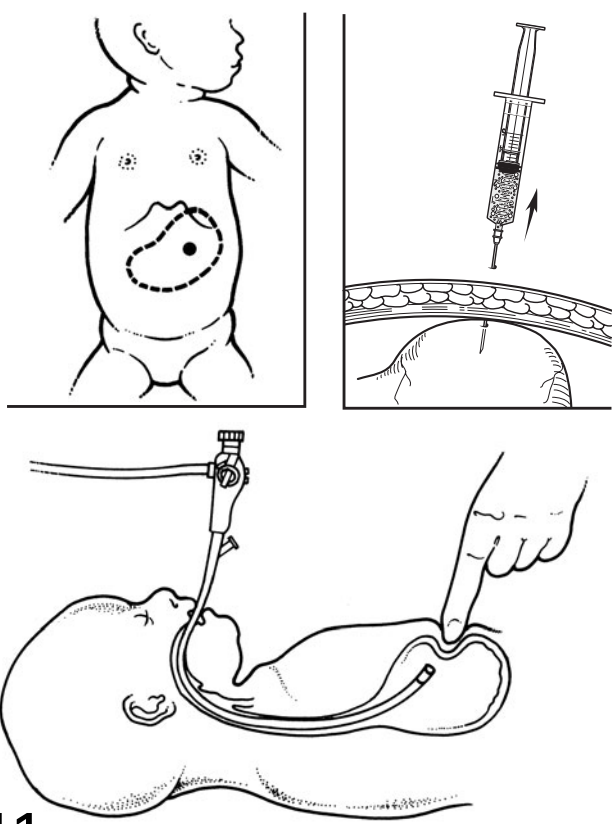
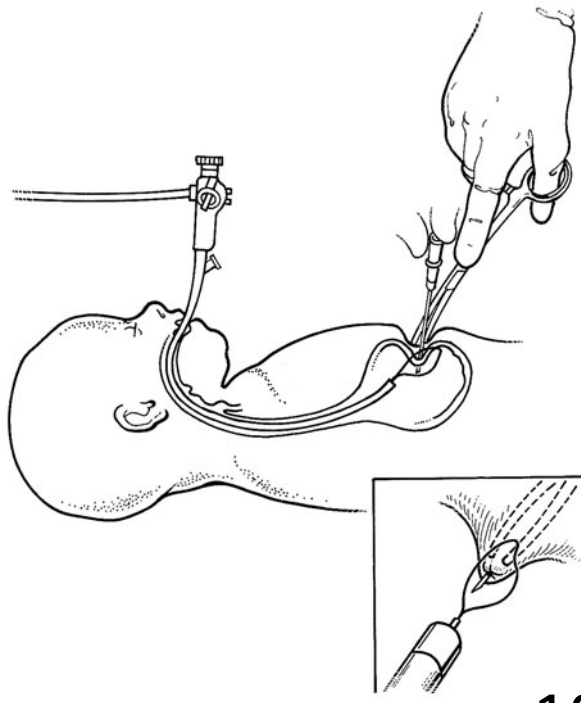


11

11a–c The gastroscope is inserted and the stomach insufflated. The stoma should be away from the ribcage to allow placement of an incision if a fundoplication becomes necessary in the future. Under-insufflation or over-insufflation should be avoided to minimize the possibility of accidentally piercing the transverse colon. Insufflation of the small intestine tends to push the transverse colon in front of the stomach and should thus be avoided. Digital pressure is applied to the proposed gastrostomy site, which usually corresponds to the area where transillumination is brightest. Transillumination and clear visualization of an anterior gastric wall indentation are key points. Without these, an open or laparoscopic technique should be employed.

Long-lasting local anesthetic is drawn into a syringe and the proposed PEG site injected. The needle is advanced further and continuous aspirating pressure is applied to the plunger. Air bubbles should be visible in the remaining fluid when the tip of the needle is seen by the endoscopist. If air bubbles are noticed before the needle tip is in the stomach, the colon or other intestinal loop may be interposed between the stomach and the abdominal wall (inset).

12 An incision of 8–10 mm is made in the skin and a Kelly-type hemostat applied to maintain the intragastric indentation. The endoscope is moved gently in small increments. The endoscopist then places the polypectomy snare around this 'mound'. The intravenous cannula is placed in the incision between the slightly spread prongs of the hemostat and then firmly thrust through the abdominal and gastric walls, exiting through the tip of the 'mound' into the loop of the polypectomy snare. The snare is partially closed, but not tightened around the cannula.



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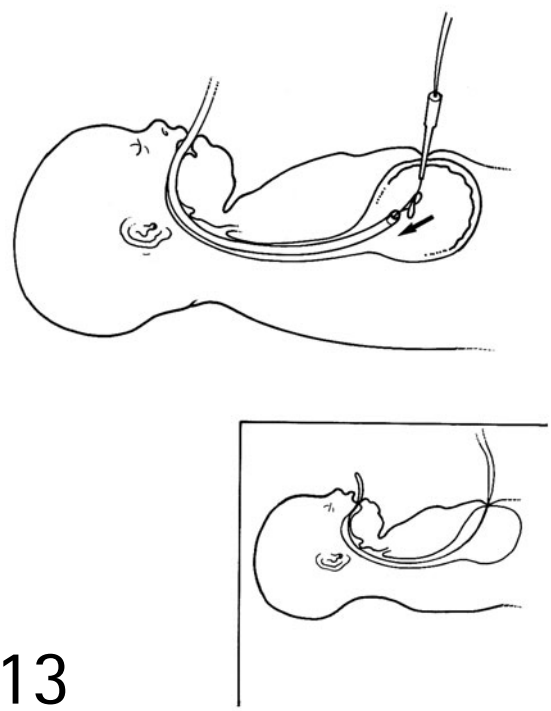
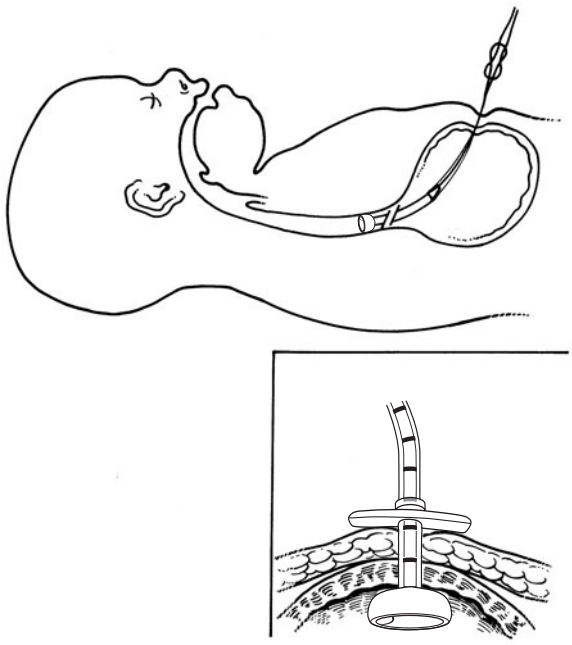


13 The needle is removed and the looped steel wire inserted through the cannula. The polypectomy snare is allowed to slide away from the cannula and is tightened around the wire. An alternative method is to retrieve the wire with alligator or biopsy forceps. The wire is then pulled back with the endoscope through the stomach and esophagus, exiting through the patient's mouth. A guiding tract is thus established.

13

14 The catheter is attached to the guidewire by interlocking the two steel wire loops. Traction is applied to the abdominal end of the wire, guiding the catheter through the esophagus and stomach and across the gastric and abdominal walls. The collapsed gastric retainer minimizes the risks of esophageal injury. (For diagrammatic purposes, a shortened catheter is shown.) The tapered end of the catheter exits through the abdominal wall before the gastric retainer enters the patient's mouth, allowing complete control of the catheter during placement. Traction is continued until the gastric and abdominal walls are in loose contact. The external crossbar is slipped over the catheter and guided to the skin level, avoiding pressure from the retaining crossbar on the mucosa or skin. The catheter is cut to the desired length and the feeding adapter attached. No sutures are used, and the catheter is connected to a small, clear plastic trap. A gauze pad and tape are applied.



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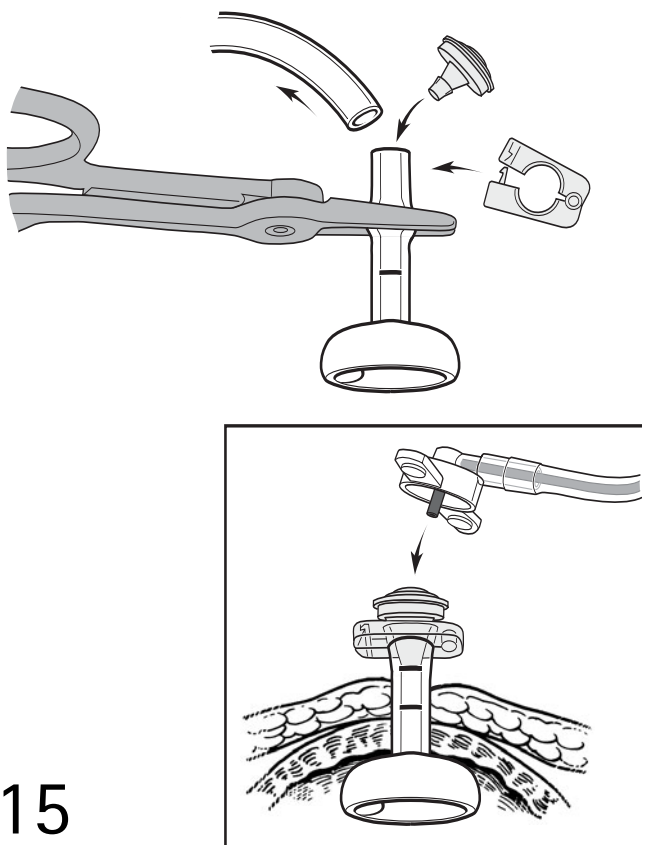
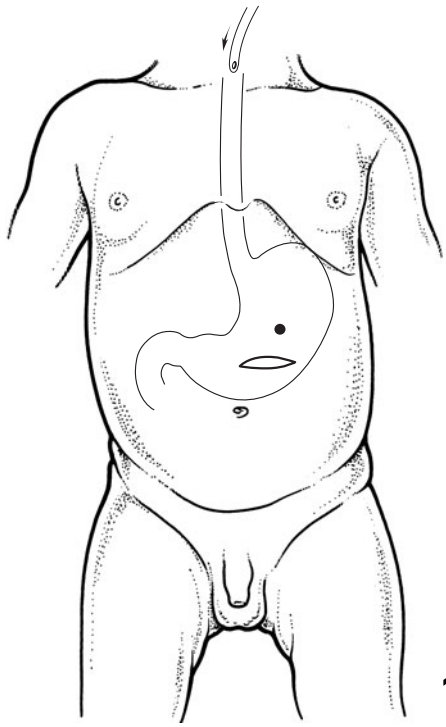


15

15 If desired, the PEG catheter can be converted to a skin-level device using the changeable skin-level port valve, either immediately after completion of the procedure or, preferably, a few days later when the edema has subsided and it is easier to determine the amount of 'play' necessary to avoid tissue compression. The main concern with this approach is cutting the catheter too short. To avoid this, one can use a 'temporary crossbar' between the skin and the external valve to allow a good apposition between the stomach and the abdominal wall for 7–10 days. This temporary crossbar is then simply cut off, allowing the necessary 'play'. The illustration depicts the atraumatic plastic clamp holding the tube, the cut-off long tubing, the external valve, and the securing cup. The inset shows the completed conversion and the feeding adapter.

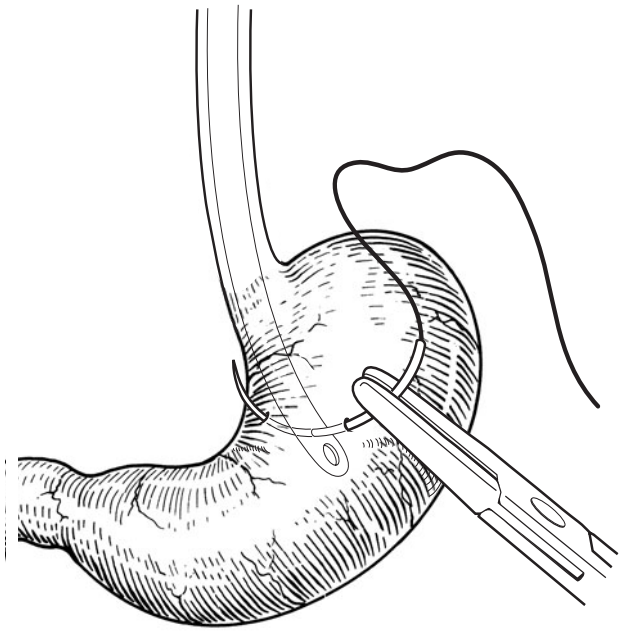
Hybrid gastrostomy

In children who have a markedly abnormal epigastric anatomy or dense adhesions because of previous abdominal operations or peritonitis, it may be impossible to perform a PEG or a laparoscopic gastrostomy safely. At times, even a Stamm gastrostomy may be technically challenging. For these cases, a hybrid procedure was developed that combines the 'open' technique employing a mini-laparotomy with the 'pull' PEG insertion of a gastrostomy catheter.

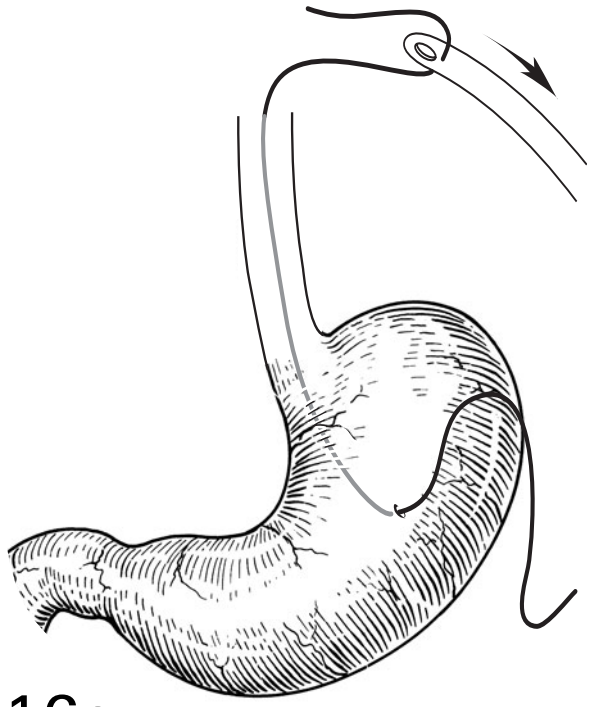


16a

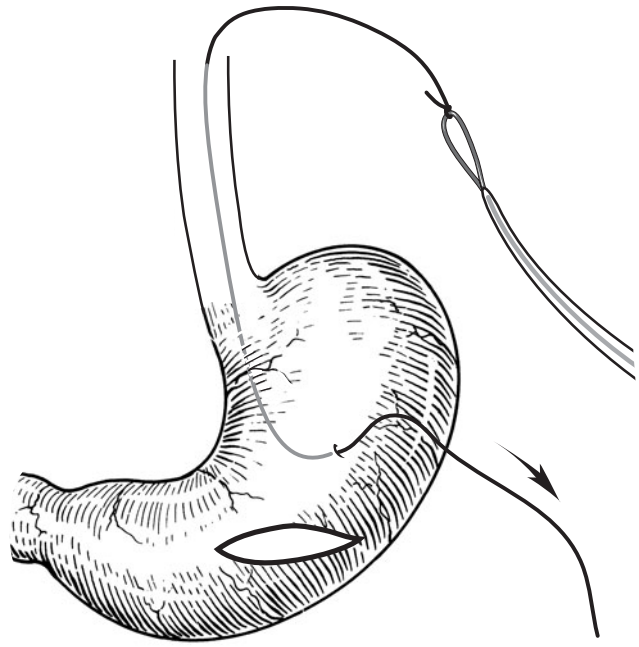
16a–e A mini-laparotomy is made in the left upper quadrant (16a). The anterior surface of the stomach is identified. A large (20–26 Fr) Nelaton-type catheter is inserted orally and advanced into the stomach, where it is clearly identified. A sturdy monofilament suture on a tapered needle is selected. The needle is pushed through the anterior gastric wall at the desired gastrostomy site, through the catheter and then out of the stomach (16b). The needle is then cut off. The catheter is pulled back with the suture imbedded in it (16c). The gastric end of the suture is brought out through a counter-incision (16d). The oral end of the suture is attached to a PEG-type catheter, which is then pulled back in a manner similar to the 'pull' PEG illustrated above. The inset (16e) demonstrates the completed procedure.



16b



16c



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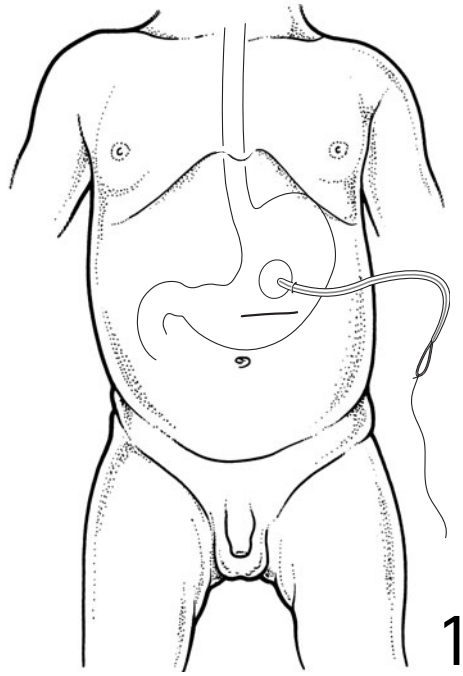


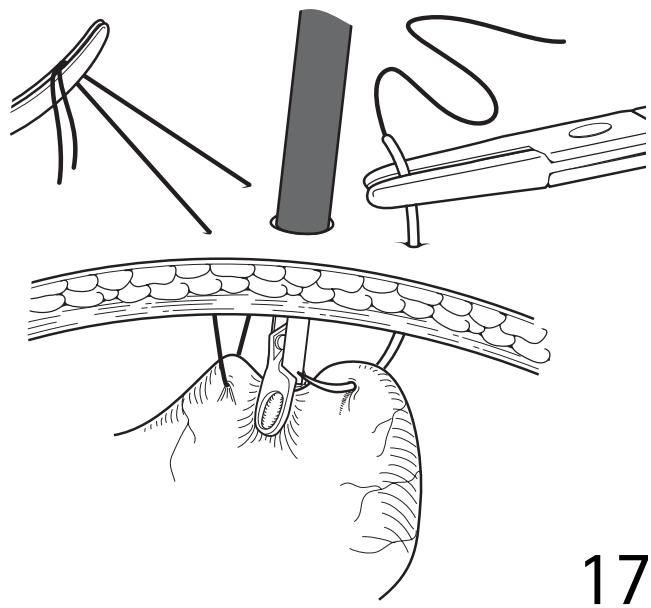
16e

Laparoscopic gastrostomy

Several methods for establishing a laparoscopic gastrostomy have been developed. In addition to the videoscopically controlled PEG, the two commonly employed methods are based on adaptations of the Stamm gastrostomy and modifications of the 'push' PEG using the Seldinger technique. Our preference is for the latter, because in order to place a purse-string suture through the exposed segment of the anterior gastric wall, the trochar site must be sufficiently enlarged. Bringing the gastrostomy catheter or skin-level device through this enlarged opening may predispose the site to leakage.

In order temporarily to anchor the stomach to the abdominal wall, different approaches may be employed: T-fasteners, U-stitches, or double-needle mechanisms. The U-stitch technique is illustrated here.

17 The most suitable site of a gastrostomy is selected in the left upper quadrant and marked. As in other types of gastrostomies, it should be away from the costal margin and the midline. A nasogastric tube is introduced. Pneumoperitoneum is established in the usual, age-appropriate manner and a 30° laparoscope introduced at the umbilicus. The left epigastric area is inspected. The previously marked site is infiltrated with a long-lasting local anesthetic. The needle is then pushed through the abdominal wall and the appropriate relation between the anterior gastric wall and the stoma site established. A small skin incision is made and a 5 mm trocar inserted. A grasper is introduced and the stoma site on the anterior gastric wall is lifted toward the parietal peritoneum. A U-stitch is passed through the abdominal wall, through the anterior gastric wall, and back out through the abdominal wall. A second U-stitch is passed parallel to the first one, 1–2 cm apart. The sutures are lifted, maintaining the stomach in contact with the abdominal wall. The grasper and the trocar are removed.



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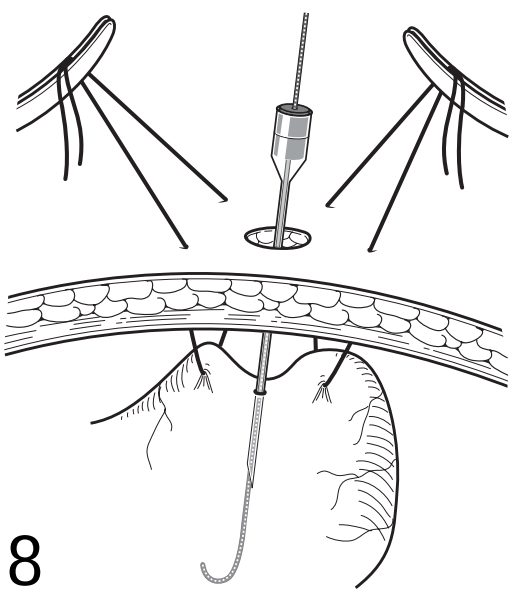
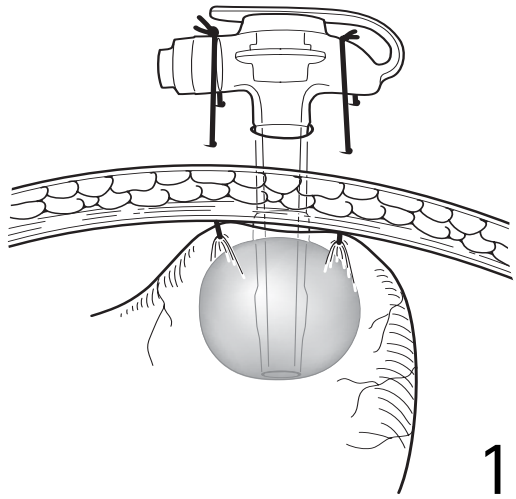


18

18 The stomach is insufflated with air through the nasogastric tube and a needle is inserted through the trocar site into the gastric lumen, between the two U-stitches. A Seldinger-type guidewire is passed through the needle into the stomach. The tract is dilated over the guidewire to the size required to insert either a Foley-type catheter or a balloon-type skin-level device. These are placed over the same guidewire. Stiffening of the catheter shaft with a thin metallic dilator is helpful during this insertion.

19 A balloon-type 'button' has been introduced. The previously placed U-stitches are tied over the wings of the 'button'. If a long tube is placed, a pair of bolsters is employed.



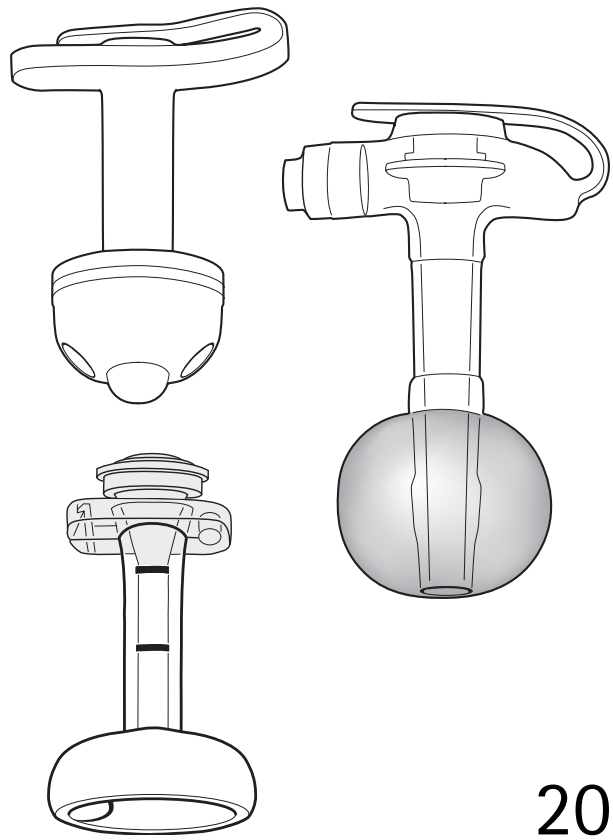
19

Percutaneous gastrostomy guided by fluoroscopy, computed tomography, or ultrasonography gastrostomy

These techniques are, in many ways, similar to the previously described methods of creating an approximation of the anterior gastric wall to the parietal peritoneum and inserting a self-retaining tube. For the fluoroscopic and computerized tomography-guided procedures, the stomach is insufflated with air. In the ultrasonographic-guided gastrostomy, the stomach is filled with saline. In some techniques, the stomach is approximated and held in place by T-fasteners prior to the insertion of the catheter in an 'introducer' PEG variation, whereas in others a pig-tail catheter is used to gain access and retention. In both cases, a Seldinger-type technique is employed, followed by dilatation of the tract and catheter insertion. A fluoroscopically guided 'pull' PEG has also been described. (see Chapter 99.)

POSTOPERATIVE CARE

Enteral feedings begin following open gastrostomies once the ileus has resolved, and on the day after the operation for the minimally invasive procedures. The dressing is removed after 24 hours, the wound is examined, and the tension on the external immobilizers adjusted in order to avoid excessive pressure that could lead to tissue damage. Our preference is to leave the stoma uncovered. We avoid harsh antiseptic solutions and, after a few days, simply use soap and water for cleaning. Granulation tissue tends to form after a couple of weeks and is controlled with gentle applications of silver nitrate. If granulation tissue becomes excessive, it leads to leakage and needs to be excised. We have observed good results with the application of a triamcinolone and antifungal combination to prevent the recurrence of granulation tissue. Once the tract becomes epithelialized, no medication should be used.



20 The three most commonly employed skin-level devices: the original 'button' (with a flapper-type one-way valve at the gastric end of the shaft), a balloon-type skin-level device (with a 'duck-bill' one-way valve in the upper shaft), and a changeable skin-level port valve (with an external slit valve).

20

COMPLICATIONS

Although generally considered a basic procedure, gastrostomy is associated with a long list of complications related to technique, care, and catheter used. Serious technique-related problems include separation of the stomach from the abdominal wall leading to peritonitis, wound separation, hemorrhage, infection, injury to the posterior gastric wall or other organs, and placement of the tube in an inappropriate gastric position. Separation of the stomach from the abdominal wall is usually due to inadvertent, premature dislodgement of the tube or a disruption during catheter change. It requires immediate attention. It is generally managed with a laparotomy, although in select cases a laparoscopic correction is possible. Most complications can be avoided by careful choice of the procedure and stoma device, considering it a major intervention, and using meticulous technique, approximating the stomach to the abdominal wall, exiting the catheter through a counter-incision, and avoiding tubes in the midline or too close to the ribcage.

Among the most serious long-term problems are the so-called 'buried-bumper syndrome' (or external catheter migration) and severe gastrostomy leakage. The first complication becomes apparent when there is difficulty with the administration of feedings that may also be associated with pain. This mishap can be avoided by always allowing sufficient 'play' between the skin-level device or external bumper and the skin. Severe leakage is initially managed using conservative measures. If these fail, the stoma may be relocated using a simple, non-endoscopic variation of the PEG. A new stoma site is selected and a small incision made. A large curved needle is placed through the leaking stoma, exiting through the new site. The suture is pulled through, establishing a tract. The catheter follows the tract, entering through the malfunctioning stoma and exiting through the new one. Once the catheter is in place, the leaking stoma is closed extraperitoneally.

FOLLOW-UP

All children with gastrostomies should be carefully followed up to prevent long-term catheter-related complications and monitored for manifestations of foregut dysmotility, particularly gastroesophageal reflux.

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Esophagogastroduodenoscopy

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HISTORY

Endoscopists in the nineteenth century used open, rigid tubes to visualize the upper gastrointestinal tract. The characteristics of rigid endoscopies were improved by the addition of conventional lenses that provided magnification and some increase in the viewing angle. When miniaturized for pediatric applications, the narrow viewing angle and the poor light transmission inherent in these devices limited their usefulness. A major advance in the field of endoscopy was the development of the rod-lens telescope, which first became available in 1966. Flexible fiberoptic endoscopes were first developed in 1958. Technical improvements combined with the development of specialized equipment have expanded the diagnostic and therapeutic potential of this technique. Flexible endoscopes for upper gastrointestinal endoscopy are now available in small sizes ideally suited to pediatric applications.

PRINCIPLES AND JUSTIFICATION

Indications

Endoscopy of the upper gastrointestinal tract is performed for both diagnostic and therapeutic reasons. The most common diagnoses requiring esophagogastroscopy in children include esophageal foreign bodies and reflux disease. Other diagnoses include caustic ingestion, food impaction, and tracheo-esophageal fistula evaluation. The most common symptoms requiring endoscopy include dysphagia, pain, bleeding, and food refusal. Additional procedures performed in conjunction with esophagogastroscopy include stricture dilatation and percutaneous gastrostomy insertion.

UPPER GASTROINTESTINAL TRACT BLEEDING

Flexible esophagogastroduodenoscopy diagnoses causes most upper gastrointestinal tract bleeding. Definitive or palliative treatment may be provided for specific lesions. Esophageal varices may be treated by endoscopic sclerotherapy or variceal banding. Ulcer bleeding may be controlled by the injection of sclerosants or coagulation using electric current, heat, or laser energy.

ESOPHAGEAL FOREIGN BODIES

Either flexible or rigid esophagoscopy is mandatory in the evaluation and treatment of suspected esophageal foreign bodies. When encountered, foreign bodies may be removed or advanced into the stomach. Most gastric foreign bodies will pass spontaneously. Those that fail to pass after a period of observation require extraction with a flexible scope.

ESOPHAGEAL STRICTURE

Endoscopy is beneficial in the diagnosis and treatment of strictures and achalasia. Dilatation of either process may be made easier and safer using endoscopy to visualize and assist in the passage of guidewires, strings, and dilators. Fluoroscopy to confirm proper guidewire position and to monitor the passage of each dilator is a useful adjunct to minimize the risk of esophageal perforation or damage.

CAUSTIC INGESTION/ESOPHAGEAL INJURY

Esophagogastroscopy is useful in the evaluation of infants following suspected ingestion of caustic substances. The degree of esophageal injury may be diagnosed and graded. Follow-

up evaluations can assess mucosal healing and early stricture formation.

ADJUNCTS TO FEEDING

Endoscopic techniques may be used to place gastrostomy tubes (percutaneous endoscopic gastrostomy) and transpyloric feeding tubes.

Choice of endoscope

The choice of rigid or flexible endoscopes depends on the procedure planned and the experience and training of the surgeon. The safety and ease of flexible endoscopy make it the procedure of choice for most pediatric upper gastrointestinal endoscopic procedures. The ability to insufflate air and thus distend the esophagus and allow complete visualization ahead of the advancing esophagoscope is the principal advantage of the fiberoptic endoscope. Removal of an impacted foreign body may be easier with the rigid esophagoscope. Endoscopists should be familiar with both techniques.

RIGID ESOPHAGOSCOPES

The Storz–Hopkins rod-lens telescope is utilized almost exclusively in rigid esophagoscopy. Unlike a conventional telescope that uses small lenses with long intervening air spaces, this system uses long glass rods with their ends shaped in the form of a lens and small intervening air space. The lens system allows transmission of a brilliant magnified image through a small-diameter tube. A wide variety of endoscopic instruments is available to perform such tasks as foreign body retrieval and sclerotherapy. General anesthesia with endotracheal intubation is required for most circumstances.

FLEXIBLE ENDOSCOPES

Flexible adult endoscopes are typically 11.0–12.6 mm in diameter. Although they may be passed into the esophagus of children, they are too large for maneuvers such as intragastric retroflexion for fundic visualization or transduodenal passage. Large endoscopes with two channels may have some advantages in emergency sclerotherapy for bleeding, as one channel can be used for suction and irrigation while the other is used to direct the sclerotherapy needle. Endoscopes of intermediate size, 9–10 mm, are versatile and may be used in both adults and older children. High-resolution 5 mm instruments are now available, allowing complete upper gastrointestinal endoscopy to be performed on the smallest of infants.

Image processing systems

Image processing systems that allow the endoscopic image to be projected onto a video monitor are invaluable for teaching and documentation (hard copy imaging). Endoscopes that utilize a video image processor located in the head of the endoscope offer incredibly sharp images.

PREOPERATIVE

Patients should be 'nil by mouth' for an appropriate period of time before sedation or anesthesia (e.g., 4 hours for infants and 6 hours for toddlers and young children). The mouth should be examined for loose and potentially dislodgeable teeth. Explanations appropriate to the age of the child should be given.

Anesthesia

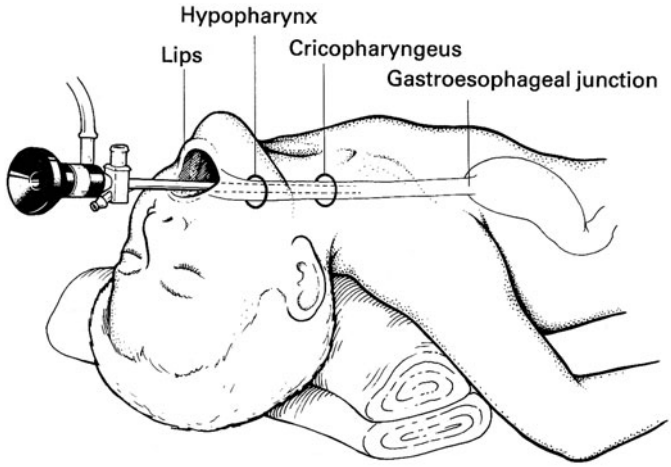
Upper gastrointestinal endoscopy should be performed either in the operating room or in an appropriately equipped, dedicated endoscopy suite. Equipment should be immediately available to provide airway and breathing support, including suction devices, supplemental oxygen, airways, bag/mask devices, and equipment for endotracheal intubation. Adjuncts to circulatory support, including intravenous therapy, fluids, intraosseous devices, and available resuscitation medications, are essential. The equipment and personnel to manage vomiting, seizures, anaphylaxis, and cardiopulmonary arrest must be available. Cardiorespiratory monitoring and pulse oximetry should be performed. An assistant for patient monitoring is essential. This may be an anesthetist, a nurse anesthetist, or a nurse assigned to the patient. It is preferable that this person is not the endoscopy assistant, whose attention is focused on the procedure and equipment. Guidelines for the monitoring and management of pediatric patients undergoing pharmacologic sedation for diagnostic and therapeutic procedures have recently been published.

OPERATION

The following endoscopic techniques are commonly used in pediatrics.

Rigid esophagoscopy

1 For this procedure the child is anesthetized and endotracheally intubated. Supporting towels are placed under the shoulders to maintain the head in full extension. The oropharynx is suctioned. Although the esophagoscope may be passed into the cervical esophagus by direct visualization, this maneuver is facilitated by lifting the tongue and epiglottis with a laryngoscope and directly visualizing the entry into the esophagus. It is critically important that the oroesophageal axis is straight ('sword-swallower's' position) during rigid esophagoscopy. The teeth, if present, are protected by a gauze pad or plastic guard.



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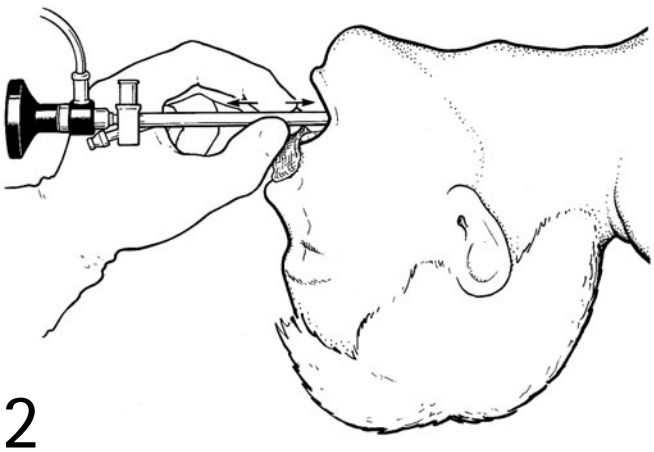


2

2 The esophagoscope is grasped with the supporting hand much as one would grasp a pencil, while the remaining fingers of that hand rest against the maxilla or upper teeth.

The esophagoscope is then advanced – in the words of Chevalier Jackson, '… the word insinuate is better than introduce, since it implies to introduce slowly, as through a winding and narrow passage … the esophagoscope is advanced … watching the folds as they unfold and recede'.

If any difficulty is encountered in negotiating the lumen of the esophagus, a small soft catheter may be used as a lumen finder. Again, Jackson advises: 'When no lumen is visible a search for a lumen is made by gentle palpation with the lumen finder. When the lumen is found the esophagoscope may be gently and safely advanced. The lumen finder is not, in any sense, a mandril for blind introduction…when it has found and entered deeply into the lumen, the esophagoscope is advanced'.

The narrowest points along the way are the cricopharyngeus and the gastroesophageal junction. Once the stomach has been entered, the esophagoscope is slowly withdrawn. It is during the removal of the esophagoscope that the best examination of both the stomach and the esophagus can be obtained.

Flexible esophagoscopy

Though conscious sedation can be used in children undergoing flexible esophagoscopy, general anesthesia with endotracheal control of the airway is preferred. The patient can be supine or positioned laterally with the left side down. The instrument is introduced by directly visualizing the oropharynx and the entry into the esophagus through the cricopharyngeus. The primary advantage of the flexible endoscope during advancement through the esophagus is the ability to insufflate air easily through the endoscope and with precise control in order to distend the esophageal lumen. Controlled air insufflation not only lessens the danger of esophageal injury during insertion of the endoscope, but also provides a valuable tool for examining the esophageal lumen since the distensibility of the esophageal wall may be altered in pathologic conditions. It must be emphasized that the same dangers found in using rigid instruments are present during flexible esophagoscopy. The esophagoscope should never be forcibly or blindly advanced. The esophagus

may be injured just as easily during manipulations with flexible instruments.

Air insufflation, although invaluable in visualizing the esophagus, must be controlled, especially in infants and small children. Gaseous distension of the abdomen can compromise ventilation. The lowest adjustment possible of the air insufflation rate is used, and the abdomen is left uncovered so that it may be continuously inspected. The child's temperature may need to be supported by elevating the room temperature, external warming lights, or a warming mattress.

After the stomach is entered, insufflation is used to allow a panoramic view. Excessive fluid is suctioned to allow complete visualization of the mucosa. A gastrostomy, if present, must be occluded to allow the stomach to fill. Orientation and endoscopic maneuvers are similar in the lateral and supine positions. Once the stomach is filled with air, the endoscope can be retroflexed, and the gastroesophageal junction visualized. The endoscope is straightened, and the remainder of the stomach is visualized. With the pylorus in view, the endoscope is advanced. Entering the pylorus may be facilitated by rotation of the endoscope. The best visualization of the pyloric channel may be obtained during the slow removal of the endoscope. In a patient with a pronounced angulation at the incisura (the 'J-shaped' stomach), a considerable length of the endoscope may need to be advanced before the tip will enter the pylorus. Changing the patient's position from supine to lateral or from lateral to supine may also be helpful. The ampulla of Vater is recognized by the drainage of bile. Cannulation of the ampulla for retrograde cholangiopancreatography is beyond the scope of this discussion. The duodenum, including the bulb and pylorus, is carefully inspected as the endoscope is slowly withdrawn.

Biopsy techniques

Small cup biopsy forceps that pass easily through the suction channel of the flexible endoscope or alongside the telescope within the rigid endoscope allow precisely directed biopsies to be performed.

Grading of esophagitis

Endoscopic grading of esophagitis is not uniform and is subject to both inter-user and intra-user variability. Nevertheless, description of the mucosal appearance should be part of the language of the endoscopist. The severity of esophagitis can be roughly graded on the basis of the visual appearance alone. Mild esophagitis is indicated by abnormal erythema and friability of the mucosa; moderate esophagitis describes linear erosions and superficial ulcers; and severe esophagitis refers to confluent erosions, deep ulcers and diffusely hemorrhagic mucosa.

Esophageal mucosal biopsies through the endoscope may help diagnose and manage such pathologic entities as gastroesophageal reflux, eosinophilic esophagitis, and Crohn's disease.

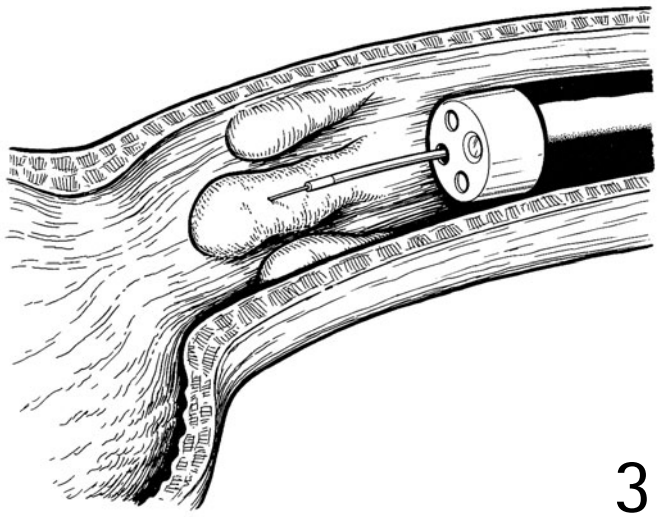
Grading of caustic injury

The severity of esophageal mucosal damage following caustic injury can be assessed by endoscopic visualization, preferably within 24 hours of injury. Mild injury is indicated by mucosal erythema and edema; moderate injury by superficial ulcerations with significant intact mucosa; and severe injury refers to circumferential ulceration, eschar formation, or very deep ulceration. As most children have mild injury after caustic ingestion, the role of mandatory esophagoscopy in all caustic injury patients can be debated.

Esophageal sclerotherapy and banding for variceal bleeding

Esophageal variceal bleeding usually stops spontaneously in children with conservative treatment and correction of coagulopathy. Emergency endoscopy to control bleeding is rarely required. Endoscopic variceal ligation has been proven to be safe and effective in children.

3 Under most circumstances, varices are present circumferentially in the distal esophagus and extend a variable distance proximally. Each sclerotherapy session is limited to 0.5 mL of sclerosant/kg body weight. Sodium morrhuate, 50 percent dextrose, and absolute alcohol have all been used for this purpose. The authors use a combination of perivariceal and intravariceal injection and preferentially obliterate an entire column of varices using injections at multiple levels rather than injecting circumferentially at one level. After injection of the varix, the endoscope is advanced and used to tamponade bleeding at the injection site.



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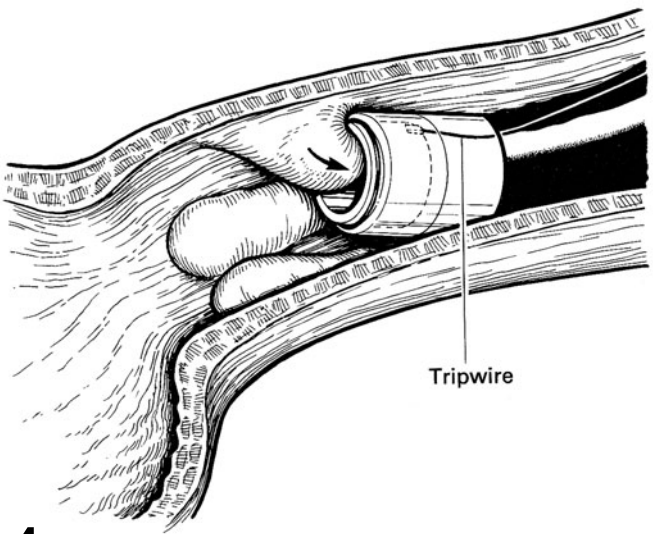

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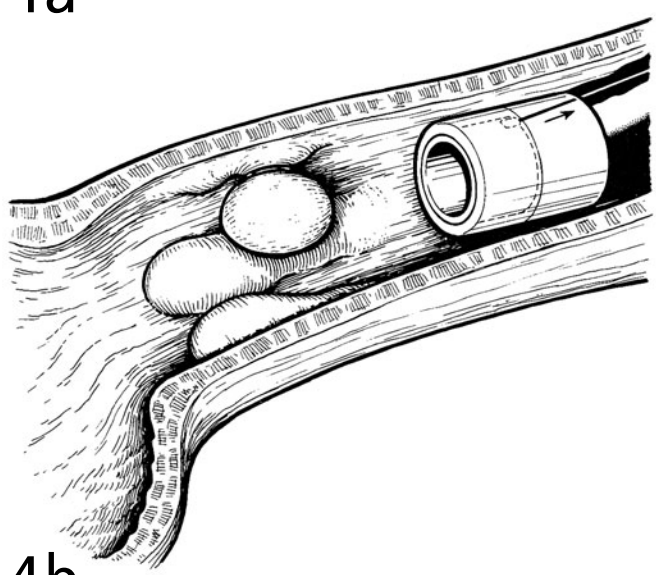


4b

4a,b With variceal banding, the varix is drawn into the end of the band applicator using suction. The band is released around the base of the varix, resulting in its thrombosis and subsequent obliteration. As many as six bands have been applied in one procedure.

Esophageal foreign body removal

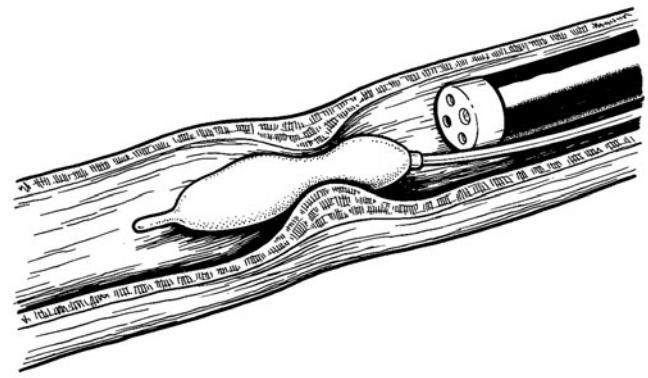
The most common esophageal foreign bodies in children are coins. Other objects include toy parts, hair beads, small bones, and food impaction. Although some advocate fluoroscopically-guided balloon extraction of coins as a safe and cost-effective alternative to endoscopic removal, the authors have not adopted this technique. The controlled environment of the operating room is preferred, where adjunctive equipment is readily available to provide airway control. In addition, when the foreign body is removed endoscopically, the esophagus can be directly assessed for damage. The most important technical consideration in foreign body removal is the availability of the proper grasping forceps. A 'coin forceps' with small teeth at its distal end, allowing it to grasp firmly the edge of a coin or small solid object, is essential. Removal of an irregular object such as impacted food material may be facilitated by the passage of a balloon catheter beyond the material, which is then withdrawn under direct vision while the airway is controlled by endotracheal intubation.

Unless the patient is known to have undergone prior esophageal surgery, a stricture is seldom the cause of an esophageal foreign body or food impaction.

Esophageal stricture

Stricture dilatation can be facilitated by the passage of a guidewire down the side channel of the endoscope and across the stricture. Proper position can then be confirmed with fluoroscopy before attempted passage of a dilator or a pneumatic dilating device. Very tight strictures can be initially dilated with sequential ureteral dilators before advancing to larger sizes.

5 When balloon dilatation is performed for esophageal strictures, the flexible endoscope is essential to position the balloon properly. It is worth noting that, since the operating channel of pediatric gastroscopes is small and passage of the larger dilating balloons may be difficult, the balloon may be directly inserted into the esophagus alongside the gastroscope and still be precisely positioned under direct vision. The dilating balloon is inflated to a preset pressure according to its diameter, as recommended by the manufacturer, and held at that pressure for 2 minutes. The application of radial forces using a balloon results in much less shear force to the esophageal lining than an equivalent dilatation using prograde or retrograde techniques.

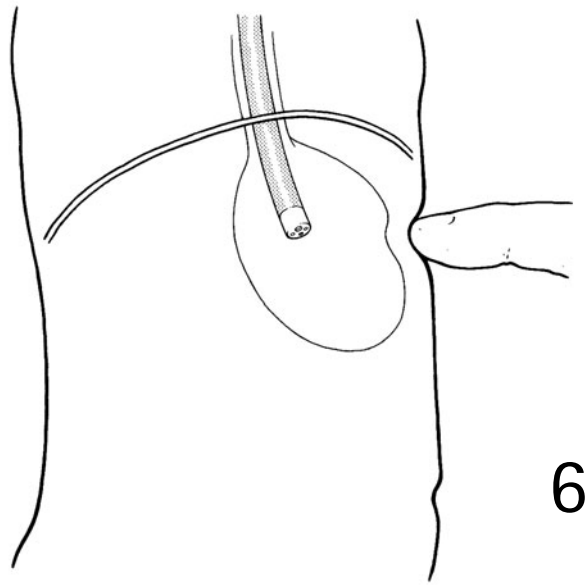


5

Percutaneous endoscopic gastrostomy

Percutaneous gastrostomy placement may be performed using either conscious sedation or general anesthesia in the endoscopy suite, intensive care unit, or operating theater.

6 The gastroscope is passed and used to identify the anterior stomach in contact with the anterior abdominal wall. Orientation is provided endoscopically visualizing the deformation of the anterior wall of the stomach caused by indenting the proposed gastrostomy site with a probe or finger.



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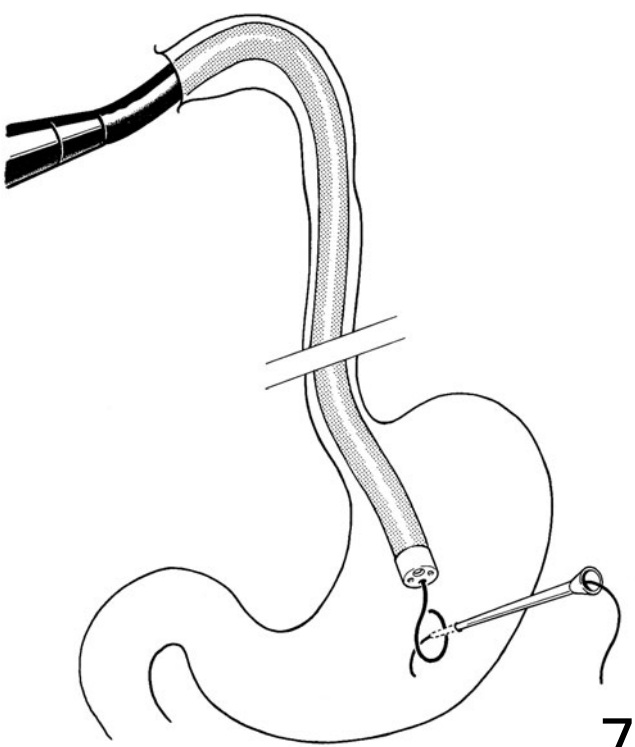
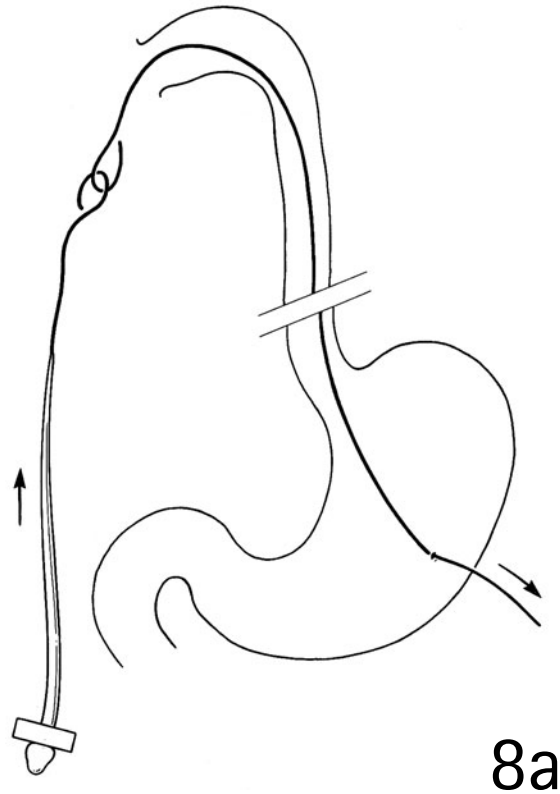


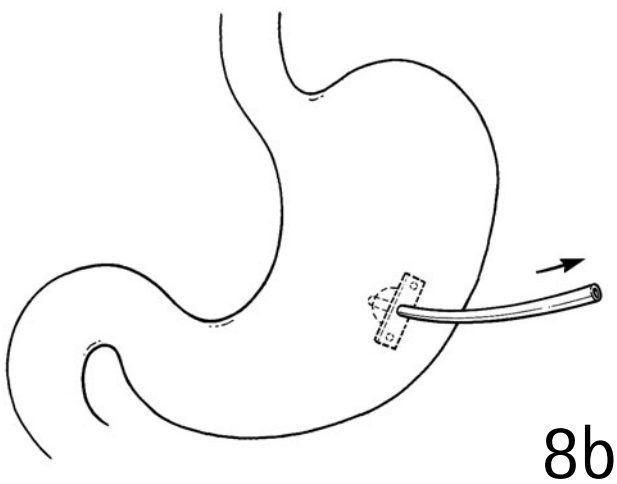
7

7 A needle is introduced through the gastrostomy site, full thickness of abdominal wall, and into the stomach, where its entry is visualized endoscopically. A wire passed through this needle is grasped with the endoscopic forceps or in a snare and withdrawn with the gastroscope out of the mouth.



8a

8a,b This wire is used to guide a dilator, on the end of which is a flanged feeding tube that will seat the stomach securely against the anterior abdominal wall when it is drawn out of the gastrostomy exit site. A small incision is made at the skin exit site to accommodate the gastrostomy tube.



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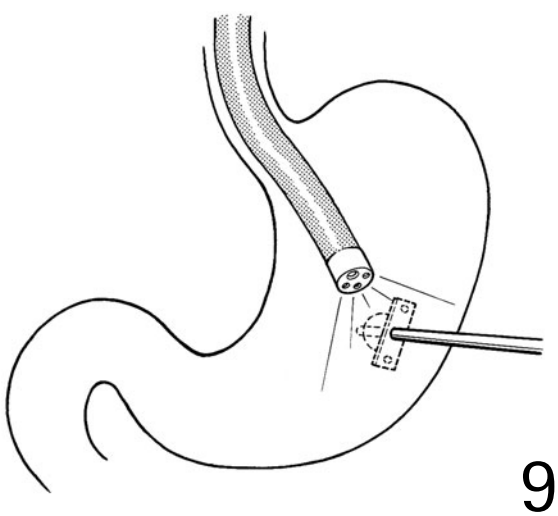

9

9 The gastroscope is again passed into the stomach and the proper positioning of the feeding tube is confirmed.

POSTOPERATIVE CARE

The endoscopic procedure itself may be performed on an outpatient basis. Concurrent or underlying disease may necessitate hospitalization. A chest radiograph is obtained whenever significant manipulation of the esophagus has occurred, such as during a dilatation procedure. The finding of mediastinal air or pneumothorax mandates esophagography with a water-soluble contrast medium to assess the degree of injury. Minor, self-contained perforations may be treated conservatively with antibiotics and hospitalization and possibly pleural drainage. Large perforations with significant pleural or mediastinal communication should be primarily repaired and drained.

Sclerotherapy may result in significant chest pain requiring narcotic analgesia. Esophageal ulceration and stricture may also result. Sucralfate is given by mouth for 1 week after sclerotherapy. Strictures following sclerotherapy usually respond to dilatation.

Percutaneously placed gastrostomy tubes may be used for feeding almost immediately, although the authors usually wait until the day after the procedure. Percutaneous gastrostomies require immediate operation for early tube dislodgement. Abdominal wall cellulitis will usually respond to antibiotic therapy. Failure to secure the gastric wall to the abdominal wall with resultant intraperitoneal leakage requires operative correction. Gastrocolic fistula may result from inclusion of a portion of the transverse colon in the path of the tube.

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Pyloromyotomy

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HISTORY

Sabricius Hildanus first described pyloric stenosis in 1646. Harald Hirschprung elaborated on the clinical presentation and pathology of the condition in 1888. At this stage the preferred treatment was medical, using a combination of gastric lavage, antispasmodic drugs, dietary manipulation, and the application of local heat, because the surgical mortality was almost 100 percent. In 1908 Fredet advocated longitudinal submucosal division of the thickened pyloric muscle, but recommended suturing the defect transversely. In 1912 Ramstedt simplified the Fredet procedure by omitting the transverse suturing, leaving the mucosa exposed in the longitudinal subserosal defect. This operation was successful and its essential elements have remained virtually unmodified ever since. Surgery has now completely replaced medical measures for the treatment of pyloric stenosis.

PRINCIPLES AND JUSTIFICATION

Incidence

The incidence of pyloric stenosis among whites is 2–3 per 1000 live births. Blacks are less frequently affected. The male-to-female ratio is 4:1. The disorder often occurs in first-born boys, and there is a strong familial pattern of inheritance. Vomiting due to pyloric stenosis has been noted in twins, whose symptoms began within hours of each other.

Diagnosis

Symptoms usually commence at 2–4 weeks of age, but can sometimes be seen in neonates or infants close to 2 months of age. Symptoms consist of projectile vomiting of non-bilious material, constipation, dehydration, lethargy or seizures, and failure to thrive. Hematemesis has been documented in a few cases.

Physical signs include variable degrees of dehydration, visible gastric peristalsis, and a palpable pyloric tumor.

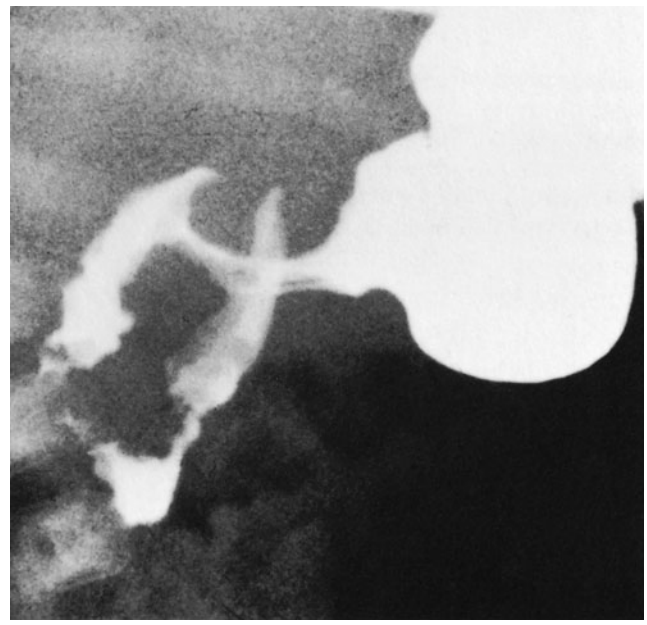
If one takes the time to perform a proper clinical examination, the diagnosis can nearly always be made without further investigations, but more often patients will be referred to the surgeons with diagnostic images suggestive of pyloric stenosis.

Special investigations

While diagnostic images may be helpful in difficult cases, none is necessary if clinical examination demonstrates a palpable 'tumor' or 'olive'. When physical examination fails to identify a pyloric mass, ultrasonographic images should be obtained. If ultrasound is equivocal, barium swallow can be helpful. Endoscopy should not be necessary to make the diagnosis of pyloric stenosis, but may be useful in symptomatic patients with other causes for gastric outlet obstruction.

Barium swallow

1 The following features are diagnostic: 'string sign' of the narrow elongated pyloric canal; 'double track' in the pyloric canal owing to infolding mucosa; delayed gastric emptying; gastric hyperperistalsis; the mushroom effect in the duodenal cap due to indentation by pyloric 'tumor'.



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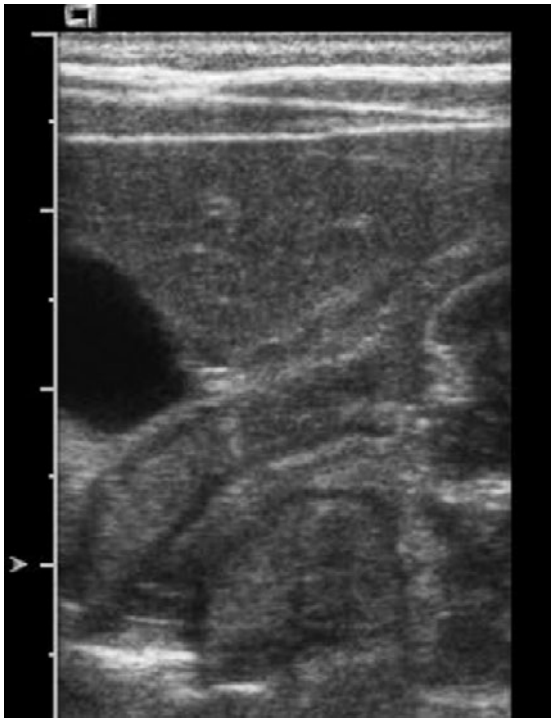


2

Ultrasonography

2 This typically shows a thickened pyloric musculature with a central sonolucent area representing the lumen. The thickened muscle is easy to recognize as a donut or bull's eye. Ultrasonographers also use the following measurement criteria to make the diagnosis: pyloric channel > 17 mm in length and pyloric thickness > 4 mm.

PREOPERATIVE

Preparation of patient

The operation of pyloromyotomy is never an emergency; correction of dehydration and acid–base imbalance takes precedence. Parenteral correction of metabolic abnormalities is the safest way to prepare a patient for surgery. An intravenous infusion of isotonic saline (20 mL/kg) is administered over 60 minutes. Once urinary output has been established, potassium chloride (20–30 mmol/L) is added to the infusate of 5 percent glucose in 0.9 percent saline. The infusion is given at a rate of 150–180 mL/kg body weight per 24 hours.

It may take as long as 48–72 hours for rehydration to be complete and for the infant to be ready for surgery. The goal is to correct the serum electrolytes to nearly normal. Accordingly, the serum potassium should be at least 3–4.5 mEq/L, the serum CO_2 should be < 27–30 mEq/L, and the serum sodium should be > 130 mEq/L before surgery is considered. The serum chloride will usually correct itself to > 100mEq/L with the above measures.

Evacuation of retained contents in the stomach by gastric lavage with warm isotonic saline is also recommended. Preoperative nasogastric suction should not be done because it will remove sodium, chloride, and potassium, thereby depleting the patient of the very electrolytes that need replacement.

Anesthesia

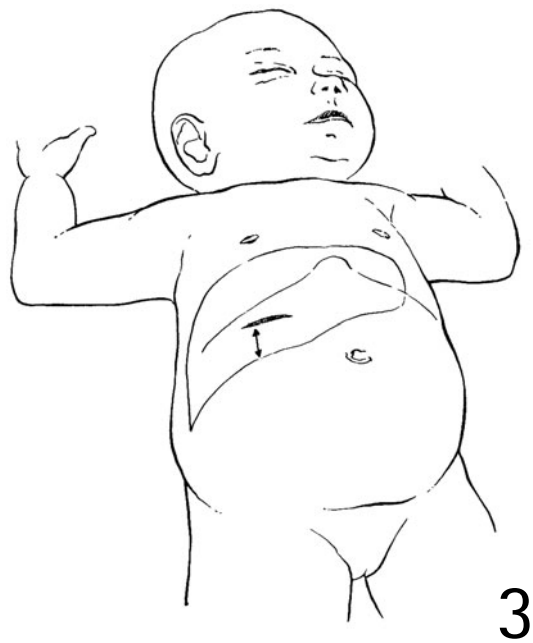
Although local anesthesia has been used successfully, general endotracheal anesthesia is now preferred, particularly if laparoscopy is performed. The stomach should be emptied immediately before induction to avoid vomiting and aspiration. The bladder is emptied using a Credé maneuver if laparoscopy is used. An intravenous infusion is set up for the administration of perioperative fluids and other drugs.

OPERATION

Open pyloromyotomy

POSITION OF PATIENT

The patient is placed supine on the operating table. A rolled towel under the level of the mid-thoracic vertebrae facilitates delivery of pyloric tumor into the incision. The stomach should be emptied of any contents with a suction catheter before starting the operation.



3

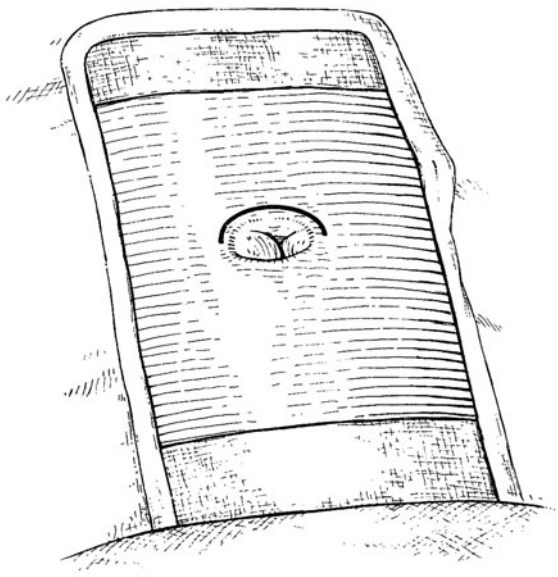
INCISION

3 A transverse incision approximately 2 cm long is made in the right upper quadrant of the abdomen, one finger breadth below the costal margin, starting immediately lateral to the border of the rectus abdominis muscle. The incision is deepened through the subcutaneous tissues and the underlying external oblique, internal oblique, and transverse abdominis muscles, which are divided in the line of the incision using needle-tipped electrocautery for hemostasis as needed. The peritoneum is opened transversely in the line of the incision.

An alternative approach is to enter the peritoneal cavity by splitting the muscles of the anterior abdominal wall in the direction of their fibers. This approach has the disadvantage of exposing unnecessary wide tissue planes, but is claimed to be associated with a lower incidence of dehiscence or later incisional herniation.

Some surgeons believe the transverse incision gives the best cosmetic result, but others prefer a vertical or paramedian incision.

4 More recently, an umbilical incision has been used for a better cosmetic result. A curvilinear skin incision is made one-half to two-thirds of the way around the superior circumference of the umbilicus in the umbilical fold. The subcutaneous tissues are spread with a hemostat to expose the midline fascia. The fascia is opened in the midline from the umbilical ring to as far cephalad as necessary to allow easy delivery of the pyloric mass.



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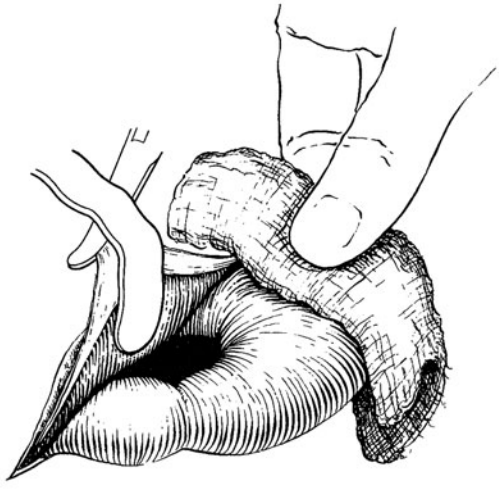


5

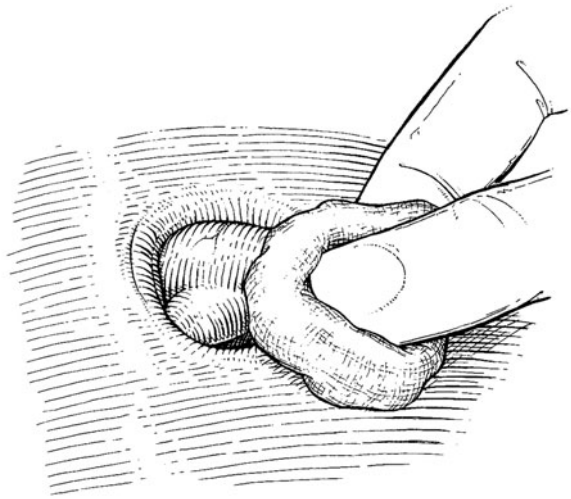
IDENTIFICATION OF THE STOMACH

5 With peritoneum opened, the liver covers the opening into the peritoneal cavity. The edge of the liver is gently retracted cranially using a malleable retractor protected by a moist gauze sponge.

No attempt should be made to grasp the pyloric tumor directly, as this leads to serosal tears and hemorrhage. The greater curvature of the stomach is identified and grasped in a moist gauze sponge. If the stomach is not readily found, traction on the transverse colon will draw the greater curvature of the stomach into wound.

DELIVERY OF THE PYLORIC TUMOR

6 With the greater curvature of the stomach firmly drawn across to the left and exerting traction on the antrum, the pyloric tumor is delivered out of the incision by applying a gentle to-and-fro rocking traction on the pylorus.

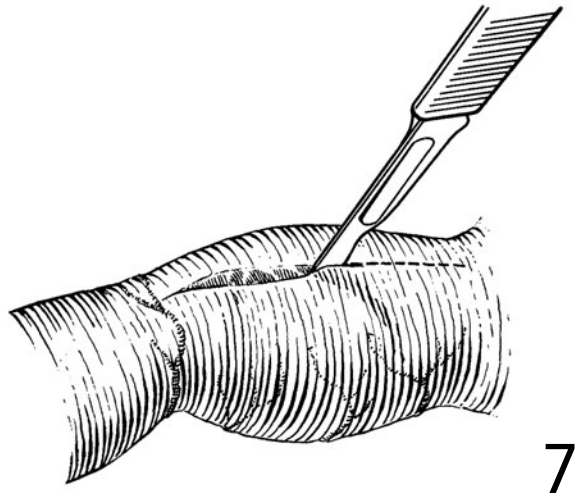


6

The pyloric vein of Mayo marks the distal extent of the tumor. Proximally, the tumor is less obvious where it merges with the hypertrophied stomach musculature. The tumor has a glistening, grayish appearance and is firm to palpation. There is a relatively avascular plane in the middle of the anterior surface where the vessels entering the pylorus superiorly and inferiorly merge.

INCISION OF PYLORUS

7 A serosal incision is then made in the avascular area on the anterior surface of the tumor. It is carried distally as far as the pyloric vein of Mayo, which marks the pyloroduodenal junction, while proximally it extends well onto the anterior surface of the antrum of the stomach. The length of the incision is 2–3 cm. Protrusion of the pylorus into the lumen creates a critical zone of folded duodenal mucosa in a very superficial position at the distal end of the stomach. It is in this area that perforation of the mucosa most often occurs.



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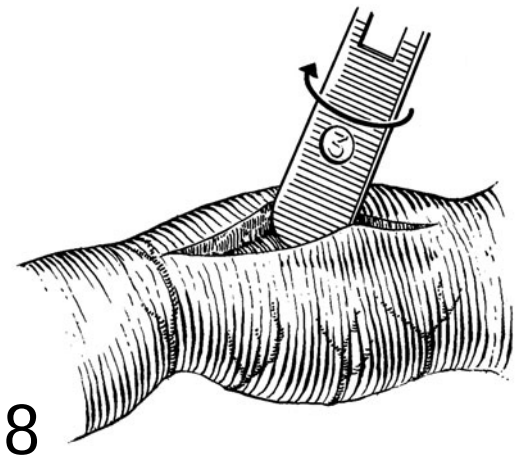
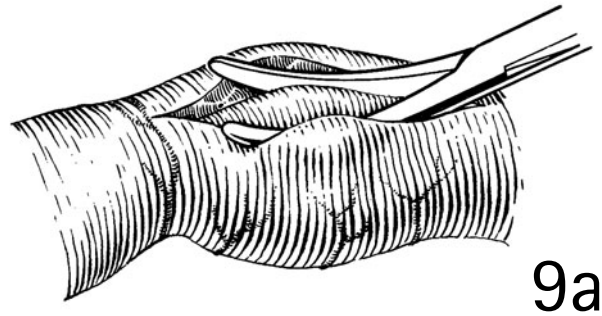


8

SPLITTING OF PYLORIC MUSCULATURE

8 Pressure with a blunt instrument, such as the back of the handle of a scalpel, into the incision with counter-pressure from a finger placed behind the tumor allows splitting of the hypertrophied muscle fibers down to the submucosa. This appears as a white, glistening membrane in the depth of the incision of the pylorus. A twisting movement on the blunt instrument produces an extension of the split proximally and distally and widens the incision. Alternatively, the blunt instrument is gently rubbed back and forth along the incision and over the muscle to split it.



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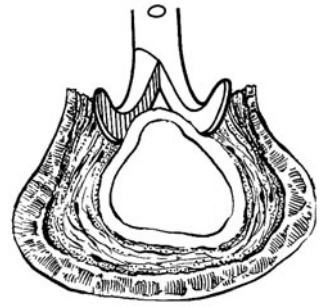


9b

9a,b To ensure that all muscle fibers have been divided throughout the length of the incision, the edges of the split muscle are spread apart with either a pair of blunt forceps (ensuring that the points are held away from the mucosa) or a pyloric spreader (Dennis Browne or Benson and Lloyd) so that the submucosa bulges into the incision. Special care must be taken at the pyloroduodenal junction to avoid entering the lumen of the duodenum, which is particularly vulnerable because of the protrusion of the pyloric tumor into duodenal lumen. The adequacy of the split is assured if the two halves of the muscle move independently of each other.

TESTING FOR PERFORATION

About 20–30 mL air are introduced into the stomach via the nasogastric tube and then gently milked through the pylorus into the duodenum, and a gauze sponge is dabbed on the incision to detect any bile staining. Any perforation of the mucosa will become obvious at this juncture and should be closed by direct suture with chromic gut or polyglycolic acid. Some surgeons advocate closing the pyloromyotomy completely and re-doing the myotomy on the opposite side of the pylorus. In either case, the important point is to recognize the perforation and to repair any leak found.

Slight hemorrhage from the edges of the pyloromyotomy will cease once the tumor has been replaced into the peritoneal cavity and hence venous congestion relieved.

WOUND CLOSURE

The wound is closed with a running suture of polyglycolic acid for peritoneum/muscle and fascia. The subcutaneous tissues are closed with continuous 4/0 polyglycolic acid suture and the skin is approximated with 5/0 polyglycolic acid subcuticular suture or surgical glue.

Laparoscopic pyloromyotomy

HISTORY

Alain et al. described laparoscopic extramucosal pyloromyotomy for the first time in 1991. Since then, several other groups have reported their experience with the technique for laparoscopic extramucosal pyloromyotomy. However, the application of laparoscopy to the treatment of hypertrophic pyloric stenosis occurred late compared to other procedures in the pediatric population, as it was difficult to justify using it rather than the open technique. Tan and Najmaldin reported their initial experience in 1993 and 1995, with good results.

In 1995, Castanon et al. described pyloric traumamyoplasty, an entirely new approach to split the hypertrophied pylorus. In this technique, two crushing applications of a laparoscopic Babcock clamp were used to rupture the thickened pyloric muscle, thus creating two grooves in the muscle and relieving the obstruction.

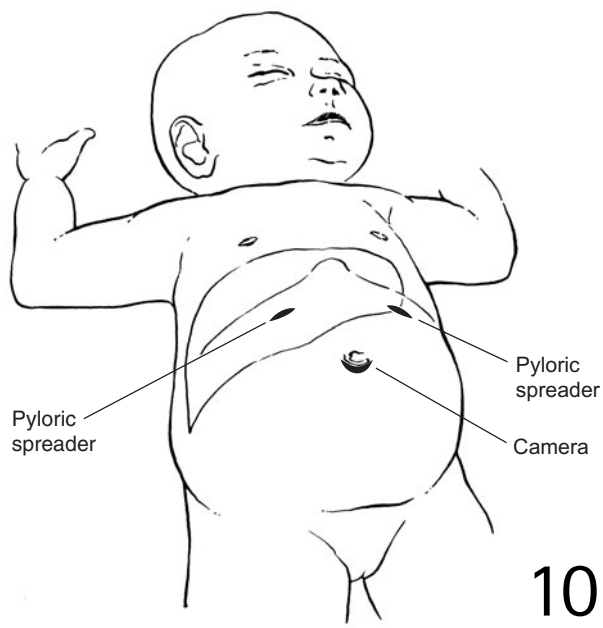
The further evolution of laparoscopic pyloromyotomy has been evident as many more authors have recently published their results as well as technical improvements in the procedure. Rothenberg described his slice and pull technique in 1997. In this procedure the duodenum is grasped with a laparoscopic Babcock clamp after a sheathed arthroscopy blade is used to perform the myotomy. Bufo et al. described a safer technique in which stomach rather than duodenum is grasped and pyloromyotomy is completed making the incision from the stomach toward the duodenum, similar to the incision made in the open approach.

Harris and Cywes presented a simple technique in which the only required instruments were an extended-tip bovie and two reusable pyloric spreaders.

OPERATIVE PROCEDURE

After the induction of anesthesia, the patient is placed in the supine position. The urinary bladder is emptied using the Credé maneuver. The abdomen is prepared and draped in the usual sterile fashion, with special attention being paid to meticulous preparation of the umbilicus. A preoperative prophylactic antibiotic should be given at the time of the operation to minimize the risk of postoperative wound infection.

10 The surgeon should stand on the patient's right side and the assistant on the left side. A stab incision is made in the infraumbilical skin crease or in the base of the umbilicus using a No. 11 knife blade, and a Veress needle is then placed through the incision and the abdomen is insufflated keeping the pressure between 8 mmHg and 12 mmHg. A small-caliber 0° telescope is placed through the umbilical port. A general abdominal inspection is done and the thickened pylorus is visualized. A small incision is made in the left upper quadrant just lateral to the midclavicular line, halfway between the umbilicus and the costal margin, and a small-caliber trocar is introduced for an atraumatic grasper with which to grasp the stomach just proximal to the thickened pylorus. Alternatively, some prefer to pass this grasper directly through a stab wound in the abdominal wall. Another small incision is made in the right upper quadrant lateral to the midclavicular line between the umbilical and costal margins, and a retractable, disposable arthroscopic knife is passed through the abdominal wall. The blade is extended to approximately 2–3 mm.



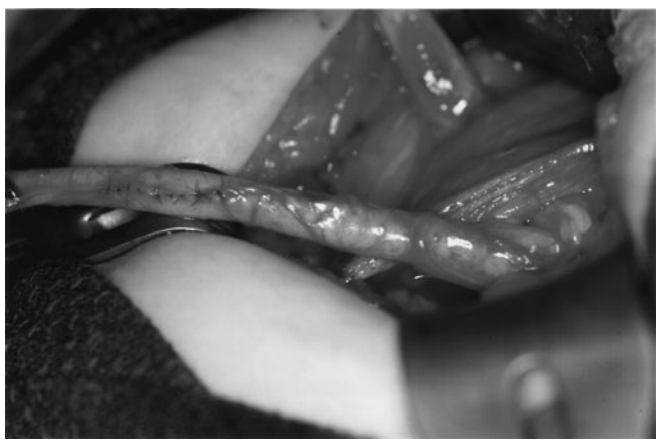
10

An incision is made in an area of avascularity over the pylorus. The incision should extend from the anterior gastric wall to just proximal to the duodenal bulb.

The pyloric incision is deepened bluntly using the retracted blade, as is done in the open technique. The arthroscopic knife is then withdrawn and a laparoscopic pyloric spreader is introduced.

11a,b The pyloromyotomy site is then slowly widened under direct vision and an adequate myotomy is assessed by holding the back edge of the myotomy using an atraumatic grasper and pyloric spreader and moving them independently to be certain that the two halves of the pyloric ring move independently.

11a



11b

The stomach should be insufflated using 60 mL air through a nasogastric/orogastric tube placed preoperatively to be certain that there are no leaks and that the air passes easily into the duodenum.

Following completion of the procedure, any ports should be removed and closed using adhesive strips or surgical glue, depending on the surgeon's preference.

Postoperative analgesia can be achieved by infiltrating the closed wounds with 0.25 percent bupivicane with epinephrine.

POSTOPERATIVE CARE

Patients are transferred from the recovery room to a regular hospital bed and intravenous fluids are continued. The patient is assessed after 4 hours to rule out peritonitis from an unrecognized perforation. Then feedings are begun and advanced as per surgeon preference. We find that, whereas in the past surgeons fed infants slowly, most of the time these infants can be advanced in their feedings quite rapidly once it is clear that they are tolerating oral intake. We start with an initial feeding of 30 mL (1 oz) of electrolytes in water solution. If that is tolerated, 2 hours later we give them 30–60 mL (1–2 oz) of formula and advance that as tolerated. It is not unusual to see some occasional postoperative emesis. If this persists, it is usually due to curds of milk retained in the stomach, in which case, we lavage the stomach with a 10 percent solution of sodium bicarbonate and begin the feeding regimen again.

COMPLICATIONS

Other than the aforementioned perforation and occasional postoperative emesis, the most frequently observed complication is wound infection. This is usually due to *Staphylococcus aureus* and is infrequently seen with the laparoscopic approach.

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Duodenoduodenostomy

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HISTORY

The surgical correction of congenital duodenal obstruction was made difficult by a lack of appropriate suture material and the lack of understanding of the perioperative care needs of neonates. By the middle of the twentieth century, enteroenterostomy, typically using a retrocolic, side-to-side duodenojejunostomy, became the standard operation for this problem. Improved perioperative care of the sick neonate resulted in many survivors. More recently, duodenoduodenostomy has been used to bypass congenital duodenal obstruction in an effort to hasten the return of intestinal function after surgery and to promote duodenal emptying. Duodenoduodenostomy may be performed in either a standard side-to-side fashion or in an eccentric fashion commonly known as the diamond duodenoduodenostomy. Video-endoscopic techniques for treating congenital duodenal obstruction have been reported.

PRINCIPLES AND JUSTIFICATION

The choice of surgical procedure for the treatment of duodenal obstruction is largely based on the preference of the surgeon. There may be unusual anatomic variants that make one procedure obviously preferred at the time of surgery. Each surgeon should be familiar with all options. The diamond duodenoduodenostomy may result in earlier postoperative feeding and shorter duration of hospitalization. This procedure is the choice of the authors and is described below. The duodenoduodenostomy is applicable to almost all cases of duodenal obstruction, whether caused by stenosis or atresia, with or without annular pancreas. It may also be used as a safe choice for the treatment of a duodenal web, since it does not risk damage to the pancreaticobiliary system, which may occur with web excision. In the hands of those experienced with the technique, almost any atresia is readily handled using the mobility afforded by the dilated proximal duodenum in combination with mobilization of the distal duodenum beneath the superior mesenteric vessels. A one-layer anastomotic technique is used most often. Although a decompressing gastrostomy was placed at the time of surgery in the past, this is no longer done routinely. In a child with multiple anomalies, in whom poor feeding is predictable, a gastrostomy may be added to the duodenoduodenostomy.

The only contraindication to surgery is the presence of multiple severe anomalies incompatible with life.

PREOPERATIVE

Duodenal obstruction may be diagnosed prenatally by fetal ultrasound or fetal magnetic resonance imaging. A dilated stomach and duodenum are seen on imaging studies. Neonates with congenital duodenal obstruction most often present with obvious symptoms on the first day of life. Feeding intolerance and vomiting, which is usually bilious, are noted from the outset. Vomiting may be non-bilious when pre-ampullary obstruction is present. Dehydration and electrolyte depletion rapidly ensue if the condition is not recognized and intravenous therapy begun. Secondary complications such as aspiration and respiratory failure may also be present. The presence of a 'double bubble' on a plain abdominal radiograph is essentially pathognomonic of duodenal atresia. Air may be seen distally in the gastrointestinal tract with an unusual double ampulla that opens both above and

below the stenosis. Contrast radiography is confirmatory and may be especially helpful in confirming the pathology when duodenal stenosis (incomplete obstruction) is present or unusual pancreaticobiliary anatomy allows air into the distal intestinal tract. Differentiating intrinsic duodenal obstruction from malrotation with volvulus may be difficult, and contrast radiography may also be helpful. Duodenal obstruction is treated less urgently than malrotation by some surgeons and therefore differentiating between the two entities is critical.

Trisomy 21, occurring in one-third of affected infants, and congenital heart disease must be suspected in all children with duodenal atresia. Concurrent malrotation and second intestinal atresias (most commonly in the duodenum itself) are gastrointestinal abnormalities that occur with increased frequency in patients with duodenal atresia. Rarely, duodenal atresia has occurred in association with esophageal atresia, hereditary multiple atresias, choledochal cyst, and biliary atresia. Duodenal atresia also occurs as part of the Feingold syndrome and Joubert syndrome.

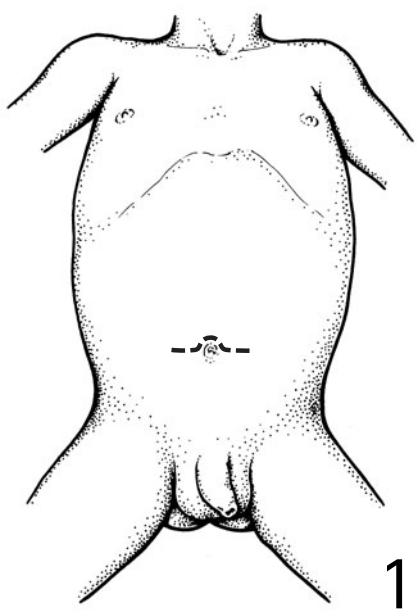
Anesthesia

General anesthesia with rapid sequence endotracheal intubation is required. Many pediatric anesthetists use epidural anesthetic supplementation for the operation as well as for postoperative analgesia. Prevention of hypothermia is accomplished by heating the operating room, warming the anesthetic gases, external warming lights, operating table warmers, warming of the intravenous fluids, and the use of adhesive plastic drapes for surgical draping.

OPERATION

Incision and initial evaluation

1 A small, transverse, right upper quadrant incision provides adequate exposure. Alternatively, a transumbilical laparotomy may be performed. The type and location of the atresia as well as any pancreatic abnormality or the presence of a rare preduodenal portal vein are noted. The patient is assessed for any abnormality of intestinal rotation; if present, a Ladd's procedure and appendectomy are performed. The presence of a normal gall bladder is noted.



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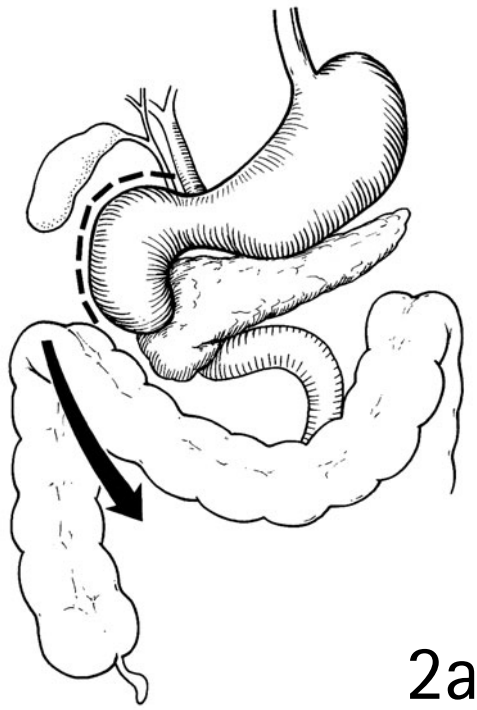
Mobilization and retraction

2a,b The hepatic flexure of the colon is mobilized sufficiently to expose the duodenum, and the proximal obstructed duodenum is freed from its retroperitoneal attachments. The requirements for distal mobilization vary according to the location of the atresia. If necessary, the entire distal duodenum may be mobilized from beneath the superior mesenteric artery. Rightward traction on the exposed distal duodenum allows these retroperitoneal attachments to be divided. A transpyloric tube may be passed to determine if a 'windsock' abnormality is present. Injection of air or saline into the distal segment is conveniently performed at this stage to rule out a second atresia. Either hand-held or fixed, table-mounted retractors may be used.



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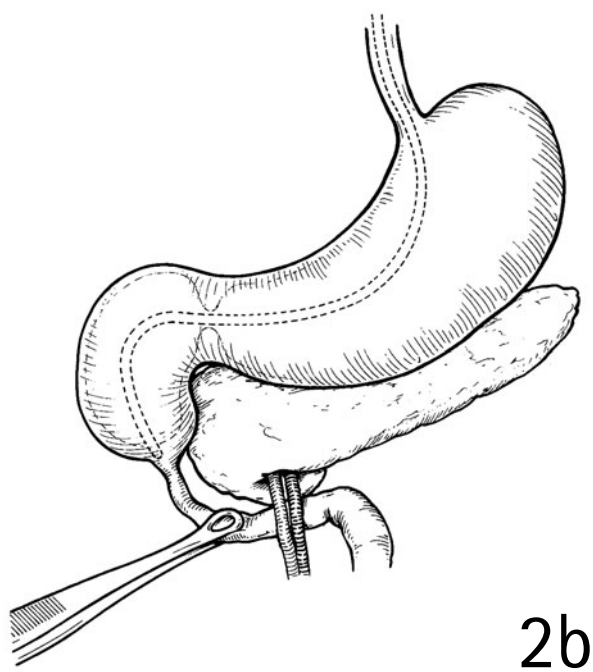
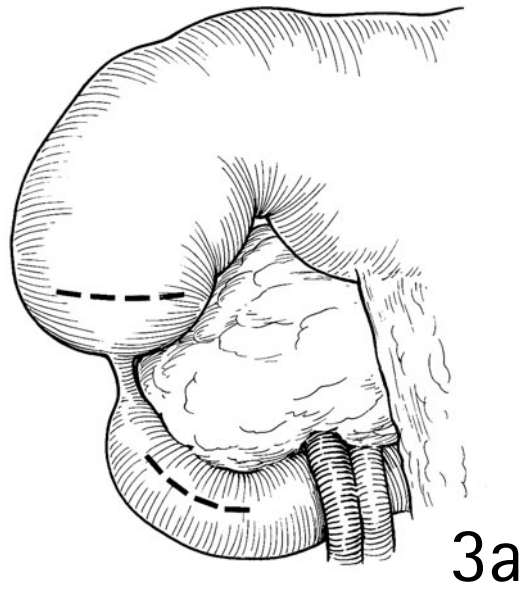


2b

Duodenoduodenostomy

3a–c A transverse duodenotomy is performed in the proximal segment. It is important that this incision be made 1 cm above the atresia to avoid any possibility of injury to the pancreaticobiliary system, which may enter anywhere in the vicinity of the duodenal web, stenosis, or atresia. Retrograde passage of a probe into the stomach confirms the absence of a duodenal web and eliminates the rare possibility of a concurrent gastric antral web. A longitudinal duodenotomy of the same length is created in the distal segment. Passage of a small tube distally confirms patency of the distal duodenum. A single layer of interrupted sutures with posterior knots tied inside and anterior knots tied outside ensures symmetry. The orientation of the sutures in the 'diamond' anastomosis is shown in b. The completed anastomosis is shown in c.



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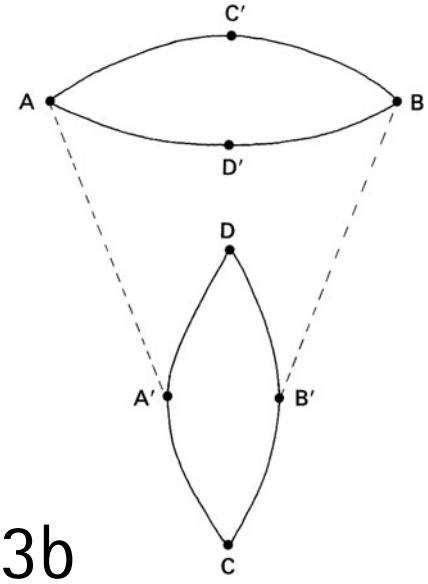

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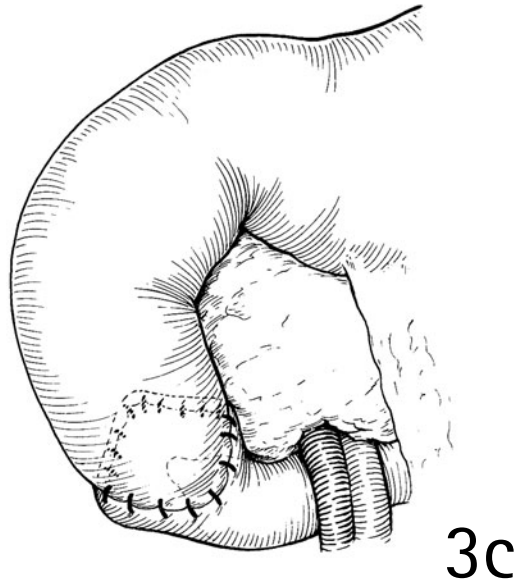

3c

If a long atretic segment is encountered or if it is difficult to mobilize the distal duodenum, a duodenojejunostomy may be performed. The first portion of the duodenum beyond the ligament of Treitz is passed through a small fenestration in the transverse colon mesentery for the duodenal–jejunal anastomosis. A gastrostomy may be performed if the need is anticipated.

Video-endoscopic approach

As with nearly all intracorporeal procedures, advances in instrumentation and surgical skill have made laparoscopic duodenoduodenostomy possible. Isolated reports have demonstrated feasibility. It is uncertain whether the video-endoscopic approach will be generally applicable, result in earlier return to feeds, and be free of technical problems. Advances in small articulating instruments will enhance the ability of pediatric surgeons to perform this procedure.

POSTOPERATIVE CARE

Postoperative care consists primarily of supportive measures to provide nutrition while awaiting the return of intestinal function. Immediate enteral feeding can be started if a transanastomotic tube is placed at the time of the initial operation. Transanastomotic feeding may reduce parenteral nutrition use and improve the time to oral feeding. The dis-

advantages of transanastomotic feeding include tube dislodgement and intestinal injury. Parenteral nutrition may be used to provide nutritional support postoperatively. To minimize the risks of parenteral nutrition (especially hepatotoxicity), total calories, protein and fat intake should be kept at the lowest levels possible to allow growth. Placement of a central venous catheter at the time of surgery is often convenient. Peripherally inserted central venous catheters are used as well. Unless the child was septic before surgery, only a prophylactic course of antibiotics is indicated. Ventilatory support is provided as needed. Nasogastric or gastrostomy drainage is maintained until gastric emptying begins, as heralded by a change in the quality of the gastric drainage from bilious green to clear or yellow and a decrease in gastric residuals.

Feeding is instituted slowly and may require a period of several weeks before full enteral nutrition is tolerated. Standard infant formulas or breast-milk are satisfactory, and expensive, partially digested formulas are usually unnecessary.

OUTCOME

The outcome depends almost entirely on the presence of other anomalies. Anastomotic leak, intra-abdominal sepsis, and wound complications occur rarely. Missed second atresias have been reported. Duodenal atony or paresis with a functional duodenal obstruction in the face of an anatomically patent anastomosis is a rare but frustrating problem. Plication of the duodenum or an alternative method of duodenal bypass is a surgical option if conservative observation is unsuccessful. Long-term complications are uncommon. Symptoms such as pain, vomiting, and feelings of fullness may be present in up to one-third of patients when studied as adults. The symptoms correlate poorly with objective findings on upper gastrointestinal radiographic studies and endoscopy. Late non-function may respond to duodenal plication.

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Malrotation

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The term malrotation refers to a condition in which the midgut – that part of the intestine supplied by the superior mesenteric vessels extending from the duodenojejunal flexure to the mid-transverse colon – remains unfixed and suspended on a narrow-based mesentery.

HISTORY

The first description of intestinal development was written by Mall in 1898. Frazer and Robins in 1915 expanded on the observations of Mall, and in 1923 Dott extended the embryologic observations to the problems encountered clinically. In 1936, William Ladd emphasized the importance of releasing the duodenum and placing the cecum in the left upper quadrant. The principles of the modern procedure are almost unchanged from those of Ladd.

PRINCIPLES AND JUSTIFICATION

Embryology

The alimentary canal initially develops as a straight tube extending down the midline of the embryo. As it lengthens, the intestine extends into the extra-embryonic celom of the umbilical cord, but later returns to the abdominal cavity. The foregut – stomach and duodenum – is supplied by the celiac artery, the midgut – small intestine and proximal colon – by the superior mesenteric artery, and the hindgut – mid-transverse colon to the rectum – by the inferior mesenteric artery. Three stages of development of the midgut are recognized.

STAGE I

The first stage occurs during the fourth to tenth weeks of gestation. Owing to rapid growth, the celomic cavity is unable to contain the midgut within its confines. The midgut is forced out into the physiologic hernia within the umbilical cord.

STAGE II

Stage II occurs at the tenth to twelfth weeks of gestation, during which the midgut migrates back into the abdomen. The small intestine returns first and lies mainly on the left side of the abdomen. The cecocolic loop returns last, entering the abdomen in the left lower quadrant but rapidly rotating 270° counterclockwise to attain its final position in the right iliac fossa. The duodenojejunal loop simultaneously undergoes a 270° counterclockwise rotation, coming to rest behind and to the left of the superior mesenteric vessels. The cecocolic loop lies in front of and to the right of these vessels.

STAGE III

During the twelfth week of gestation, various parts of the mesentery and the posterior parietal peritoneum fuse, notably the cecum and ascending colon, which become fixed in the right paracolic gutter.

CONSEQUENCES OF ERRORS OF NORMAL ROTATION

Errors may occur at any one of the three stages, with varying consequences.

Stage I

Failure of the intestine to return into the abdomen results in the formation of an exomphalos/omphalocele.

Stage II

During this stage, a number of errors could occur.

- Non-rotation: rotation may fail to occur following re-entry of the midgut into the abdomen.
- Incomplete rotation: counterclockwise rotation is arrested at 180°. The cecum lies in a subhepatic position in the right hypochondrium. The duodenojejunal rotation is similarly arrested and the duodenojejunal flexure lies to the right of the midline and superior mesenteric vessels. The base of the midgut mesentery is compressed and narrow and the entire midgut hangs suspended on the superior mesenteric vessels by a narrow stalk, which is prone to volvulus.
- Reversed rotation: the final 180° rotation occurs in a clockwise direction, with the colon coming to lie posterior to the duodenum and superior mesenteric vessels.
- Hyperrotation: rotation continues through 360° or more so that the cecum comes to rest in the region of the splenic flexure in the left hypochondrium.
- Encapsulated small intestine: the avascular sac that forms the lining of the extra-embryonic celom returns en masse into the abdomen with the intestine.

The most common error is incomplete rotation. Treatment of this condition forms the basis of the rest of the discussion.

Stage III

Rotation occurs normally but fixation is defective, resulting in a 'mobile cecum'. It is estimated that this situation is present in 10 percent of asymptomatic individuals, but it may predispose to cecal volvulus or to intussusception.

INCIDENCE

Malrotation may go undetected throughout life. Approximately 60 percent of clinical cases present in the first month of life, and more than two-thirds of these within the first week. Sporadic cases occur throughout life. It is generally accepted that once the diagnosis of malrotation has been established, surgical correction is mandatory to prevent the development of volvulus, which occurs in 40–50 percent of cases not treated surgically. Malrotation forms an integral part of exomphalos/omphalocele, congenital diaphragmatic hernia, and prune-belly syndrome. It has also been found in conjunction with intrinsic duodenal obstructions, esophageal atresia, Hirschsprung's disease, biliary atresia, and urinary tract anomalies. Associated anomalies are present in 30–45 percent of patients.

CLINICAL PRESENTATION

Neonatal period

Malrotation in early infancy may present either with acute strangulating obstruction or with recurrent episodes of subacute intestinal obstruction.

Acute, life-threatening, strangulating intestinal obstruction occurs as a result of midgut volvulus. The infant presents in a shocked and collapsed state with bilious vomiting (which often contains altered blood), abdominal tenderness with or (more commonly) without distension, and the passage of dark blood rectally. Edema and erythema of the abdominal wall develop as the volvulus becomes complicated by intestinal gangrene, perforation, and peritonitis.

Recurrent episodes of subacute intestinal obstruction are usually a forewarning of volvulus. The first, and often the only, sign may be *bile-stained vomiting*, which must be vigorously and intensively investigated.

Infants and children

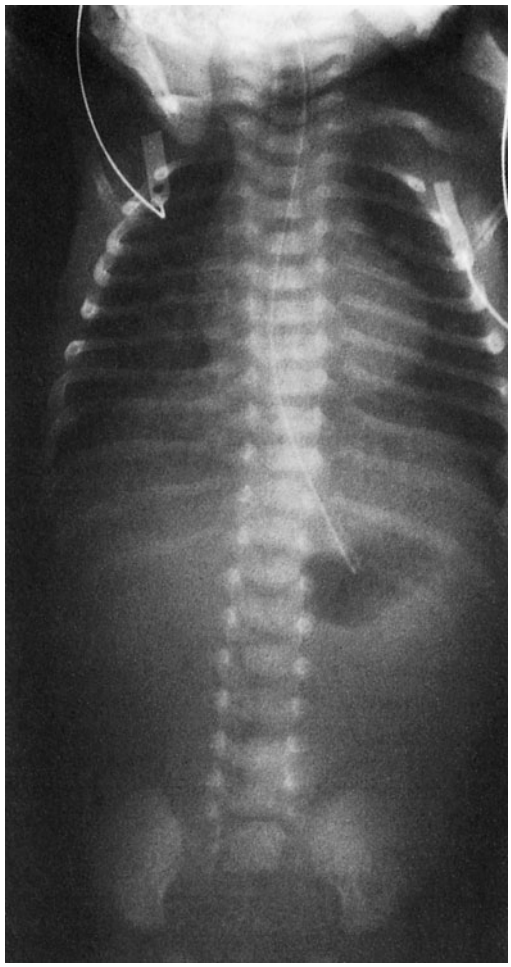
A wide spectrum of clinical symptoms has been ascribed to malrotation. The most common symptom is intermittent or cyclic vomiting, which is occasionally bile tinged. Failure to thrive and malnutrition may be a result of intestinal malabsorption secondary to lymphatic compression in the narrow-based mesentery of the small intestine. Older children may present with features of anorexia nervosa. Early satiety or pain associated with intake of food results in a reluctance to eat or food aversion.

PREOPERATIVE

Radiologic investigations

PLAIN ABDOMINAL RADIOGRAPHY

1 The features suggestive of a volvulus on the plain abdominal radiograph are air–fluid levels in the stomach and proximal duodenum (a 'double-bubble' appearance) and a paucity of gas in the rest of the intestine. In the infant presenting in shock with features of acute strangulating obstruction, further radiologic investigations only delay definitive treatment.



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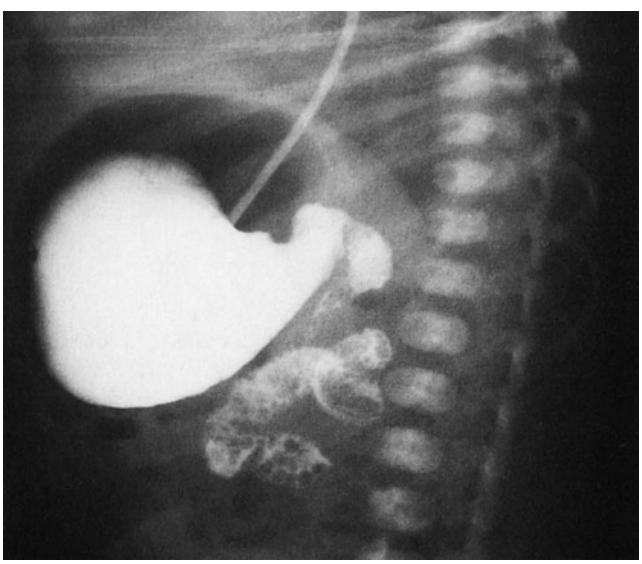


2

CONTRAST RADIOLOGY

2 The investigation of choice is an upper gastrointestinal contrast study. The features that should be elicited are an abnormal configuration of the duodenal C-loop, the identification of the duodenojejunal flexure to the right of the midline, and small bowel loops on the right side of the abdomen. A 'twisted ribbon' and 'corkscrew' appearance of the duodenum and upper jejunum indicates a midgut volvulus.

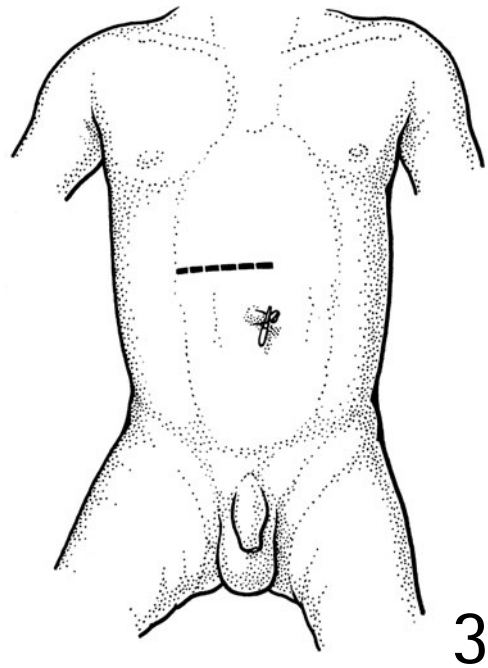
Contrast enema gives information only about the position of the cecum, which may occasionally be normally placed even in the presence of a volvulus.

Preoperative resuscitation

Patients presenting with acute strangulating obstruction require rapid resuscitation before proceeding to surgery. This comprises rapid intravenous volume replacement (plasma 20 mL/kg body weight), nasogastric decompression, correction of electrolyte and acid–base imbalance, and administration of broad-spectrum antibiotics. Attempts should be made to correct hypothermia. The period of intensive resuscitation should not extend for more than 1–2 hours before proceeding to surgery, as prolonging the time will expose the intestine to increased ischemia and may result in more extensive bowel necrosis.

OPERATIONS

Surgical correction of the anomaly should always be regarded as an emergency, even in patients presenting non-acutely. Volvulus may supervene at any stage and the operation should be scheduled as early as possible.

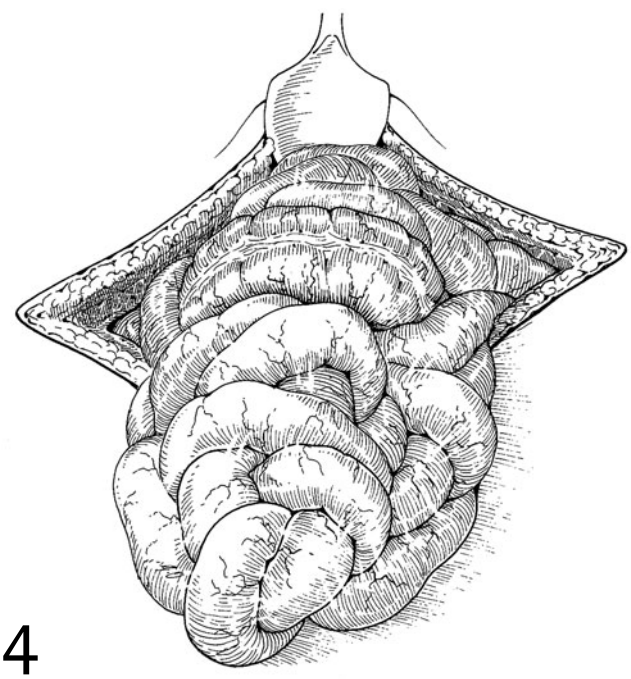


3

Laparotomy

INCISION

3 A laparotomy is performed via an upper abdominal, transverse, muscle-cutting incision, extending mainly to the right side. The obliterated umbilical vein in the free edge of the falciform ligament is ligated and divided. The entire bowel is delivered into the wound for careful examination. A small volume of yellowish, free peritoneal fluid is usually present in any early intestinal obstruction, but bloodstained fluid is indicative of intestinal necrosis.

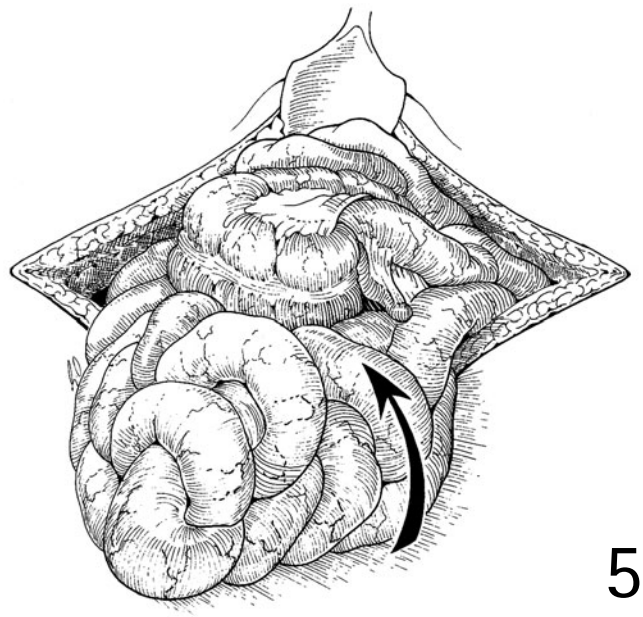


4

MIDGUT VOLVULUS

4 The volvulus occurs around the base of the narrow midgut mesentery. The twist usually occurs in a clockwise direction and is untwisted by as many counterclockwise rotations as required.

5 Moderately ischemic bowel, which appears congested and dusty, rapidly resumes a normal pinkish color on reduction of the volvulus. Frankly necrotic bowel is extremely friable and may disintegrate on handling. Bowel of questionable viability should be covered, after untwisting, with warm, moist swabs and left undisturbed for approximately 10 minutes before assessing the extent of ischemic damage. A Ladd's procedure for the malrotation is carried out (see below).



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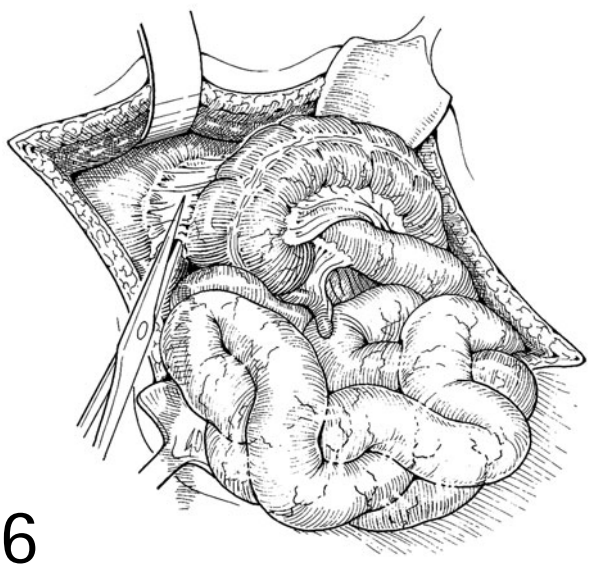
In patients with extensive intestinal gangrene, frankly necrotic bowel should be resected and the bowel ends either tied off or stomas fashioned with a view to performing a second-look laparotomy in 24–48 hours, when lines of demarcation will be clearly evident. At this stage, an end-to-end anastomosis may be feasible. In the intervening period, the patient is electively ventilated and resuscitative measures continued.

UNCOMPLICATED MALROTATION (LADD'S OPERATION)

The aim of this procedure is to restore intestinal anatomy to the non-rotated position with the duodenum and upper jejunum on the right side of the abdomen and the cecocolic loop in the left upper quadrant.



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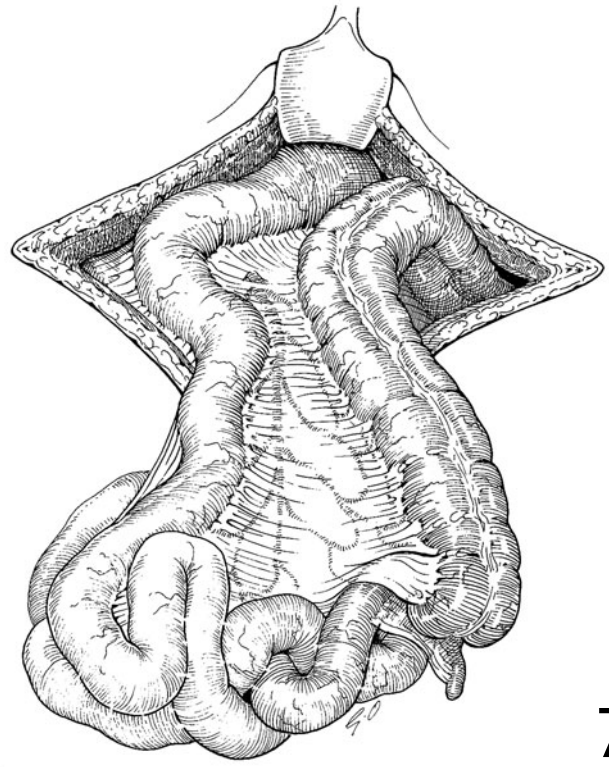
6 Folds of peritoneum extending from the cecum and ascending colon across the duodenum to the right paracolic gutter and to the liver and gallbladder are carefully divided. This maneuver leaves the cecocolic loop free laterally, but dense adhesions in the base of the mesentery must be divided before the cecum and ascending colon can be fully separated from the duodenojejunal loop. Separation is achieved by opening the serosa of the mesentery between the duodenum and cecum and exposing the anterior surface of the superior mesenteric vessels coursing in the narrow-based mesentery to the midgut.

The mesentery in this part is often thickened and edematous, especially if an associated midgut volvulus needs to be untwisted. Care should be taken to avoid trauma to the main vessels, and small branches may need to be ligated before being divided. Large, fleshy lymph and lymphatic channels that have been divided should be sealed by ligation or electrocoagulation to avoid postoperative chylous leakage.

7 The mesentery is widened peripherally to allow the right colon to be mobilized. Centrally the dissection is continued into the base of the mesentery until the superior mesenteric artery and vein are freed of any fibrous compression.



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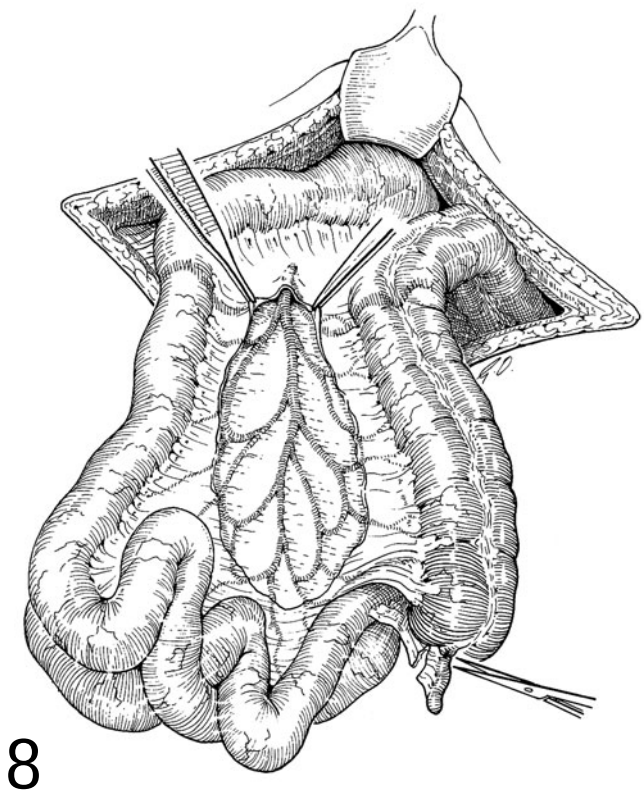

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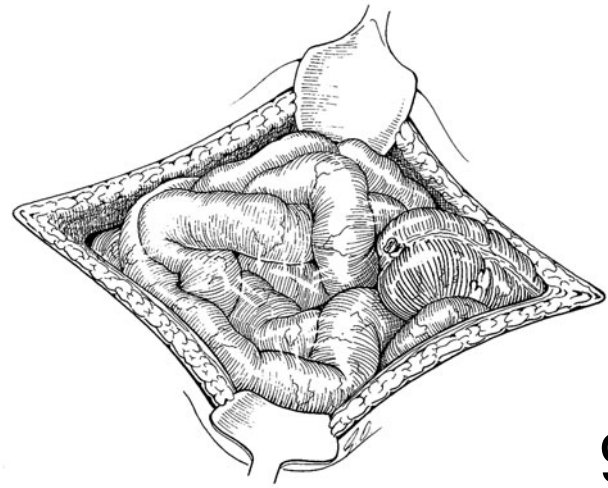
8 The duodenum is straightened by dividing the ligament of Treitz, following which the duodenum should be carefully inspected for the presence of any intrinsic obstruction. If there is any doubt, a balloon catheter should be passed perorally through the duodenum into the proximal jejunum, inflated, and carefully withdrawn into the stomach. Inability to pass the catheter through the duodenum or hold-up of the inflated balloon on withdrawal indicates an intrinsic obstruction.

An appendectomy should always be performed, as the cecum will be placed in the left upper quadrant of the abdomen and the diagnosis of subsequent appendicitis in later life could be extremely difficult to establish.

9 The intestine is replaced in the peritoneal cavity, commencing with the duodenum and proximal jejunum that lie on the *right* side and ending with the terminal ileum and cecum that are placed in the *left* hypochondrium. No attempt is made to fix the intestine in this position.



9

CLOSURE

The abdomen is closed in layers or en masse with continuous or interrupted sutures. The skin is closed with a continuous subcuticular suture.

Laparoscopy

Laparoscopy has two potential roles in the management of malrotation:

1. the treatment of symptomatic malrotation in the newborn period,
2. to determine in cases of doubtful malrotation whether or not surgical therapy is required.

TREATMENT OF SYMPTOMATIC MALROTATION IN THE NEWBORN PERIOD

It has been stated that volvulus is a contraindication for a laparoscopic approach. Volvulus is always present in the newborn with symptomatic malrotation and is amenable to laparoscopic correction. Strangulation with necrosis and/or perforation is a contraindication to laparoscopy.

The child is placed in a supine reversed Trendelenberg position on a short operating table (Fig. 42.1). The child's right side is 15° elevated. The sheet covering the operating table is enveloped over the legs to prevent slippage of the child when the table is in reversed the Trendelenberg position.

The surgeon is positioned at the bottom end of the table with the camera assistant to the left and the scrub nurse to the right. The laparoscopy column stands to the left of the child's head, the second screen to the right of the child's head (Fig. 42.2).

The first cannula is inserted in an open fashion through the inferior umbilical fold. A sleeve around the cannula prevents it from being pushed in, while a suture fixes the hub of the cannula to the skin and underlying fascia so that it cannot be pulled out (Fig. 42.1).

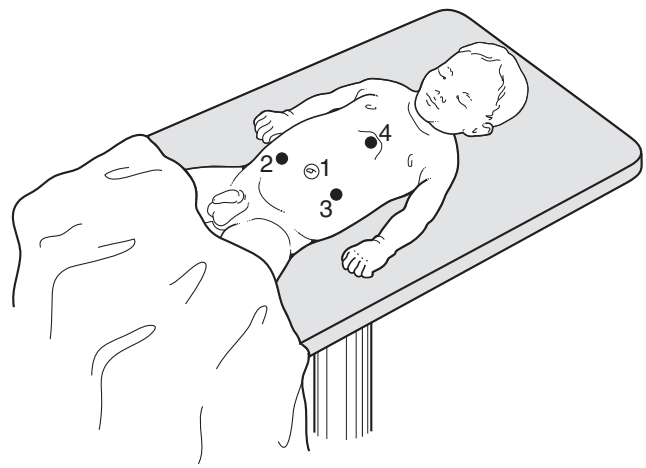


Fig. 42.1 Positioning of the patient for laparoscopic exploration and treatment.

Before starting CO_2 insufflation, the intraperitoneal position of the tip of the catheter is checked with the telescope. Pneumoperitoneum is established at a pressure of 8 mmHg using a flow of 0.5 L/minute. Optimal muscle relaxation helps to increase the working space.

Two working cannulae are inserted, one pararectally on the right at umbilical level and one in the left hypochondrium. An extra cannula is inserted subcostally on the left and is used for retraction (Fig 42.1).

The table is tilted to the left so that the bowel tends to move to the left by gravity. When the bowel is not critically ischemic, the volvulus should be left in place, as this keeps the bowel out of the way during the initial dissection of the proximal duodenum (Figs 42.3–42.6). Next, the volvulus, which is

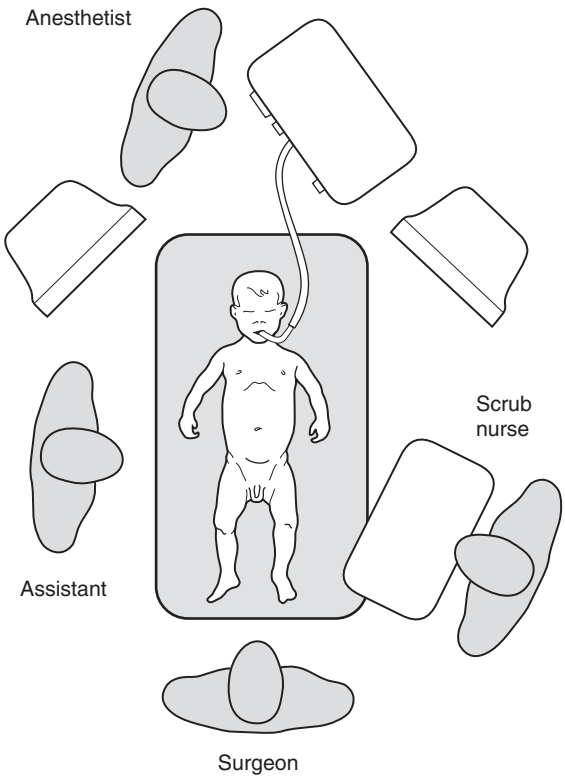


Fig. 42.2 Positioning of the team and equipment for laparoscopic exploration and treatment.

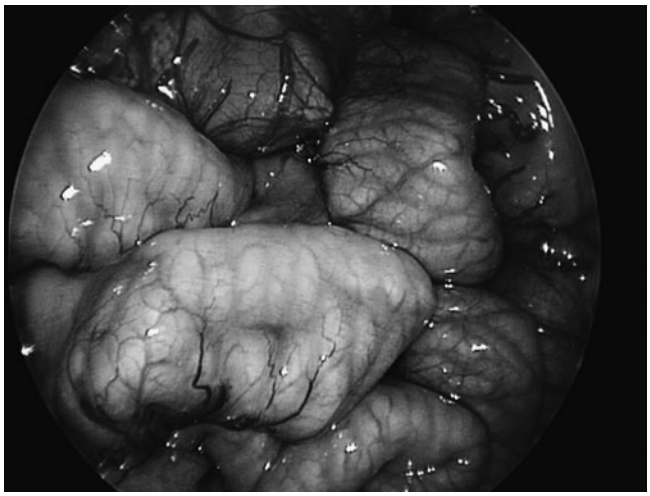


Fig. 42.3 Ischemic small bowel due to volvulus.

always in clockwise direction, is untwisted in counterclockwise direction. The cecum and ascending colon are dissected free and are displaced to the left. The kissing area between the ileocecal region and the duodenum should be undone by incising the anterior leaf of the mesentery (Figs 42.7–42.8). In

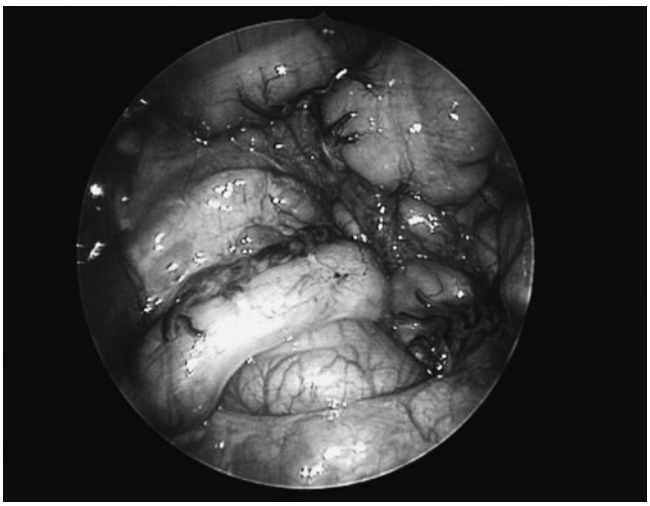


Fig. 42.4 Volvulus of small bowel around the mesenteric stalk.

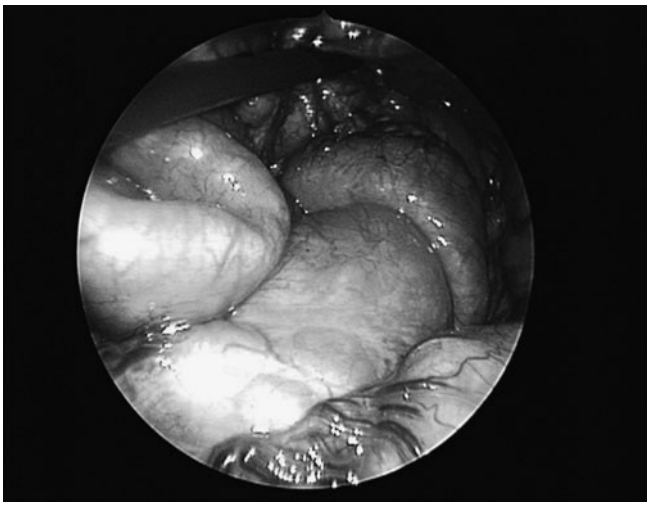


Fig. 42.5 Edematous mesenteric stalk due to volvulus.

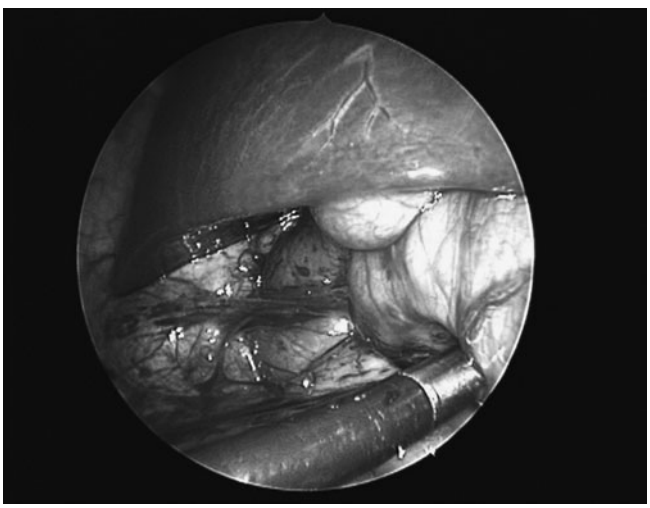


Fig. 42.6 The colon is displaced to the left. Next the duodenum is mobilized.

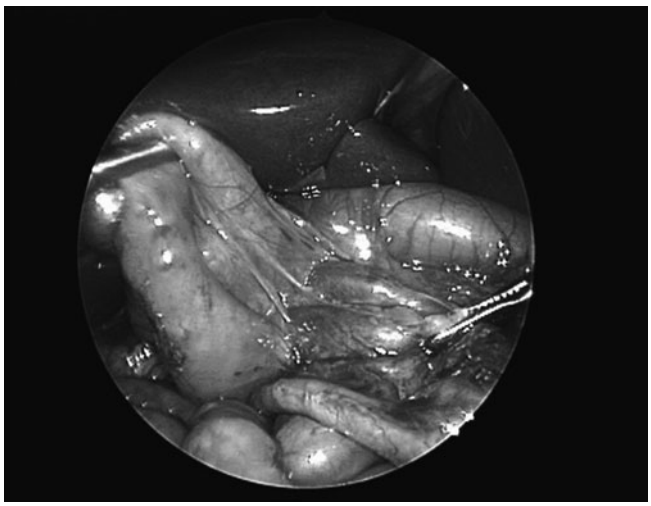


Fig. 42.7 Separation of the duodenum and ileocecal region. Note the engorged mesenteric veins.

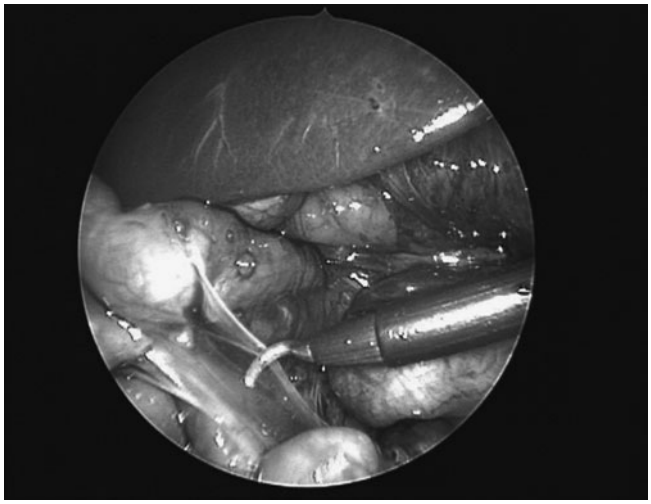


Fig. 42.8 Transection of bands between the duodenum and ileocecal region.

doing so, the mesenteric stalk is enlarged. The duodenum is now followed downwards and freed. The last band to be divided is the remnant of the duodenojejunal flexure, which is to the right of the vertebral column.

At the end of the procedure, the anesthetist is asked to inject air into the stomach so that its passage in the proximal small bowel can be checked, thus excluding a membranous occlusion. Removal of the appendix is optional. This can be done outside the body by exteriorizing the appendix through the porthole in the left hypochondrium (Fig. 42.9).

The cannulae are removed under endoscopic control. The fascia at the umbilicus is closed with one absorbable suture. Skin is approximated with adhesive strips.

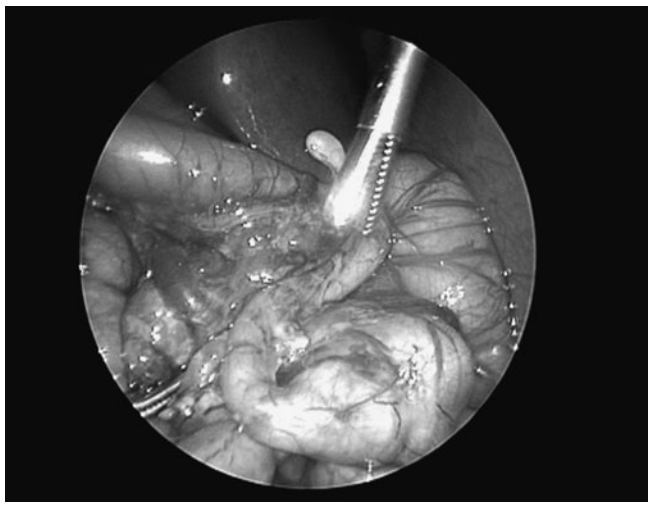


Fig. 42.9 Grasping and exteriorization of the appendix through the port in the left hypochondrium.

DOUBTFUL MALROTATION

The diagnosis of malrotation may be difficult to make. An inverted relationship between the superior mesenteric artery and vein may be present on ultrasound. More often, the suspicion of intestinal malrotation is raised during a barium meal examination. The critical question is not whether there is malrotation, but whether the mesenteric stalk is narrow and not fixed to the posterior abdominal wall. In other words, is there a high likelihood of volvulus? Laparoscopy is an ideal tool to look at the broadness and at the fixation of the mesenteric stalk.

The child is placed in a supine reversed Trendelenberg position on the operating table. The child's right side is 15° elevated. Small children are placed in a froglike position with the legs enveloped in the sheet, which covers the operating table (Fig. 42.1). The legs of larger children may be placed on abducted leg supports so that the surgeon stands in between them (Fig. 42.2).

A three-cannulae technique is used: one at the umbilicus, and two pararectally at umbilical level. A CO_2 pneumoperitoneum at a pressure of 8 mmHg suffices when there is optimal muscle relaxation.

The laparoscopic exploration should be systematic.

- Position and fixation of the cecal region to the lateral peritoneal wall (Figs 42.10–42.11).
- Position of the transverse colon and greater omentum (Fig. 42.12)
- Position of the ligament of Treitz (Fig. 42.12).
- Fixation of the mesenteric stalk.

More often than not, the diagnosis of 'dangerous malrotation' is excluded.

The cannulae are removed under endoscopic control, and the fascia defect at umbilical level is closed with absorbable suture. Skin is approximated with adhesive strips.

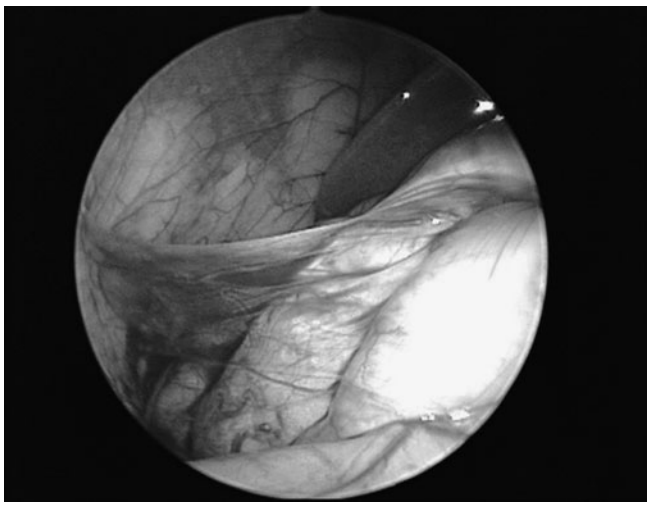


Fig. 42.10 The ascending colon is securely attached to the lateral peritoneum.

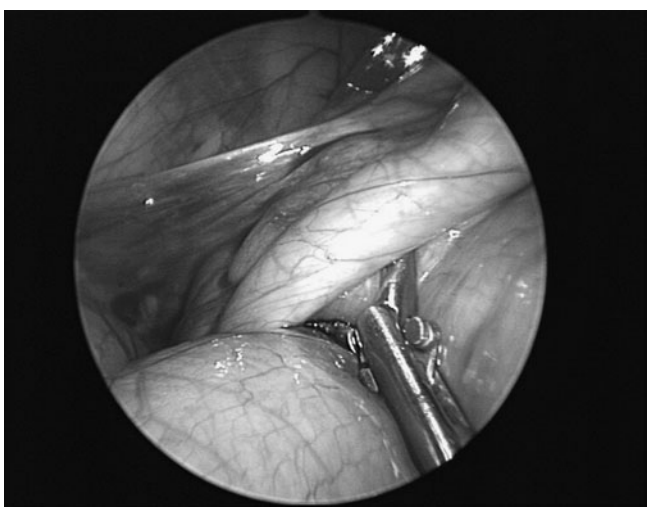


Fig. 42.11 The appendix is securely attached to the lateral peritoneum.

POSTOPERATIVE CARE

Nasogastric aspiration and intravenous fluid and electrolyte support continue until bowel function returns. The period of ileus generally lasts 48–72 hours, during which time intestinal fluid losses should be replaced with an equivalent volume of 0.45 percent sodium chloride in 5 percent dextrose solution containing 20 mEq/L potassium chloride.

OUTCOME

Recovery is generally prompt and uncomplicated. Infants who have suffered extensive bowel loss following midgut volvulus may experience problems of short bowel syndrome with a prolonged requirement for parenteral nutrition. Recurrent volvulus is rare, but adhesion intestinal obstruction is relatively common (3–5 percent of cases).

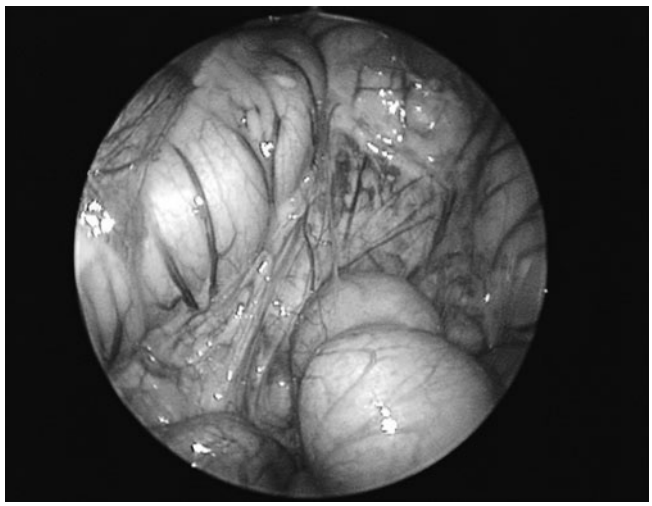


Fig. 42.12 Normal position of the transverse colon and greater omentum. The duodenojejunal flexure lies to the left of the vertebral column and is securely attached, as is the mesenteric stalk (not shown).

Acknowledgments

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FURTHER READING

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43

Congenital atresia and stenosis of the small intestine

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HISTORY

In 1911, Fockens of Rotterdam reported the first successfully treated case of small intestinal atresia. Up until 1952, however, the mortality rate of atresia of the small intestine remained very high, even at the best pediatric surgical centers. Late presentation, dysmotility of the proximal dilated atretic bowel, the blind loop syndrome, malnutrition, infections, prematurity, and associated congenital abnormalities contributed to the high mortality. In a comprehensive review of the world literature up to 1950, Evans could find reports of only 39 successfully treated cases of jejunoileal atresia.

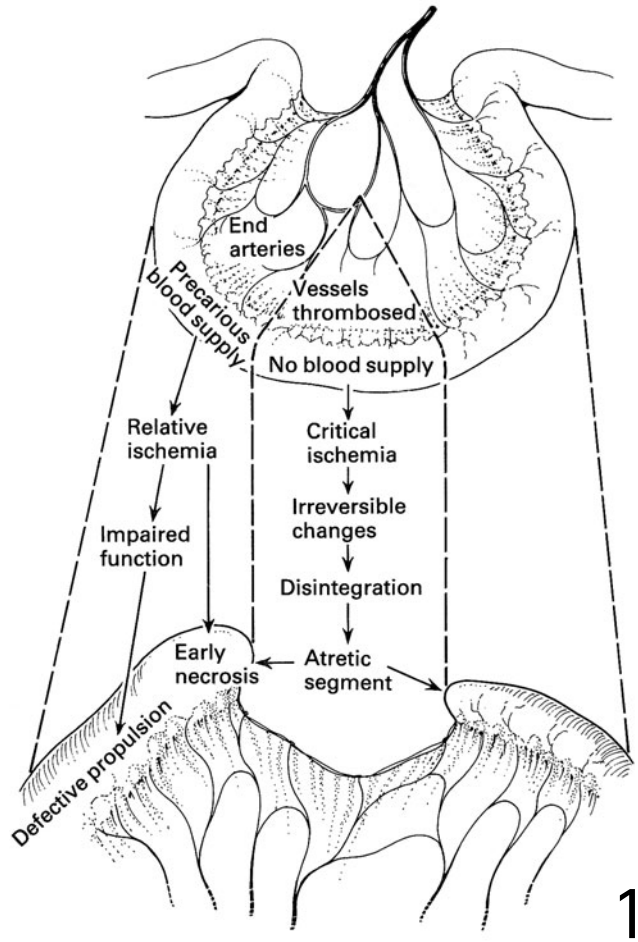
In 1952, Louw published results of an investigation of 79 patients treated at Great Ormond Street Hospital, London, and suggested that jejunoileal atresia was probably due to a vascular accident rather than being the result of inadequate recanalization, as had previously been commonly accepted. At his instigation, Barnard perfected an experimental model in pregnant mongrel bitches. This not only confirmed Louw's hypothesis, but also provided the opportunity to improve the technical aspect of bowel resection and primary anastomosis. These factors, along with advances in neonatal care, have achieved survival rates higher than 90 percent.

PRINCIPLES AND JUSTIFICATION

Pathogenesis

1 Although several mechanisms have been postulated, the most feasible theory is that of a localized intrauterine vascular incident resulting in ischemic necrosis, liquefaction, and absorption of the affected devascularized segment(s). The ischemic hypothesis is further supported by additional evidence of incarceration or snaring of bowel in an exomphalos or gastroschisis, fetal intussusception, midgut volvulus, transmesenteric hernia, and thromboembolic occlusions. The anomaly is usually not genetically determined.

The ischemic insult causes morphologic and functional abnormalities of the remaining proximal and distal bowel. The blind-ending proximal bowel becomes dilated and hypertrophied, resulting in functional abnormalities that include ineffective peristalsis. The viability may be at risk, and ischemia can lead to perforation. The distal bowel is unused and collapsed, with potential normal length and function. The discrepancy in luminal size between proximal and distal bowel may vary from two to twenty times, depending on the distance from the stomach and completeness of the obstruction.



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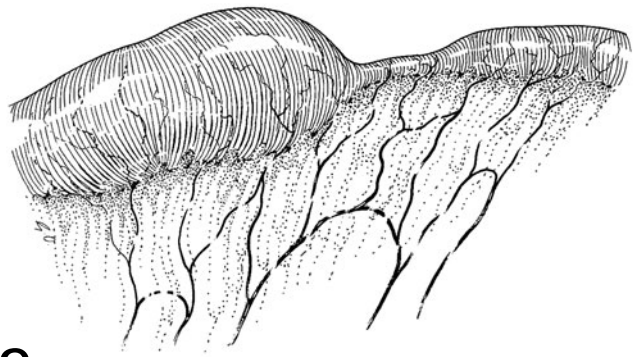
Classification

The most proximal atresia determines whether it is classified as jejunal or ileal atresia or stenosis.



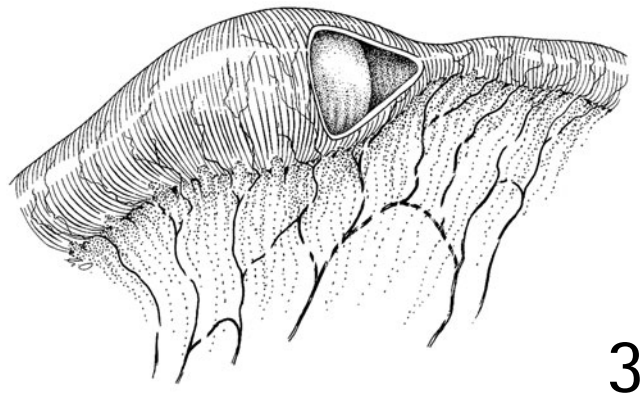
2

STENOSIS

2 The proximal dilated and distal collapsed segments of intestine are in continuity with an intact mesentery, but at the junction there is a short, narrow, somewhat rigid segment with a minute lumen which may mimic atresia type I. The small intestine is of normal length.

ATRESIA TYPE I (MEMBRANE)

3 The dilated proximal and collapsed distal segments of intestine are in continuity and the mesentery is intact. The pressure in the proximal intestine tends to bulge the membrane into the distal intestinal lumen, so that the transition from distended to collapsed intestine is conical in appearance: the 'windsock' effect. The distal intestine is completely collapsed. The small intestine is of normal length.



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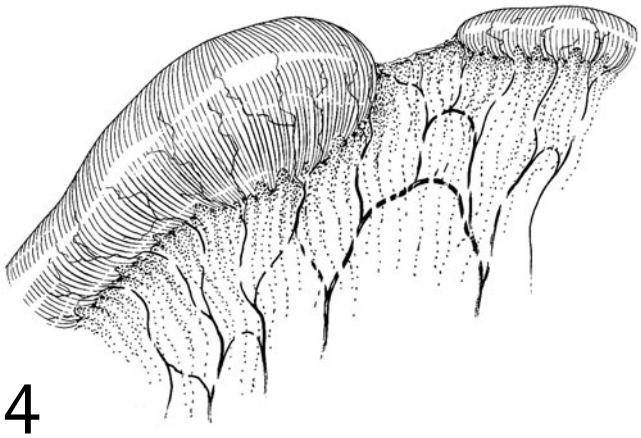

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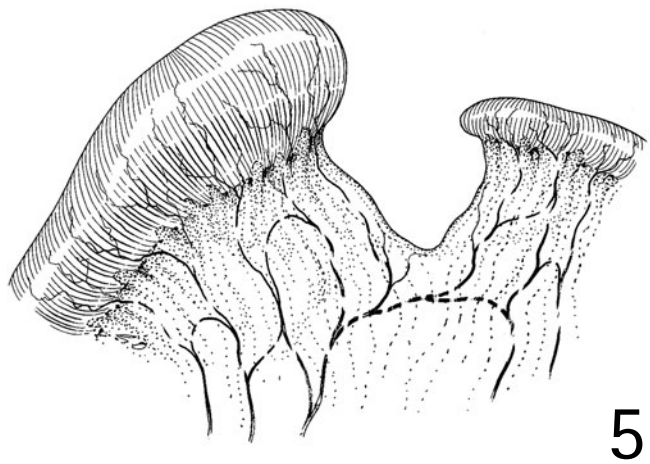
ATRESIA TYPE II (BLIND ENDS JOINED BY A FIBROUS CORD)

4 The proximal intestine terminates in a bulbous blind end that is grossly distended and hypertrophied for several centimeters but more proximally assumes a normal appearance. This blind end is often aperistaltic and poorly perfused, and may be necrotic with a perforation. The distal completely collapsed intestine commences as a blind end that is occasionally bulbous, owing to the remains of a fetal intussusception. The two blind ends are joined by a thin, fibrous band, with the corresponding intestinal mesentery intact. The small intestinal length is usually normal.

ATRESIA TYPE IIIA (DISCONNECTED BLIND ENDS)

5 The appearance is similar to that in type II, but the blind ends are completely separate. There is always a mesenteric defect of varying size, and the proximal intestine may, as a secondary event, undergo torsion or become over-distended with necrosis and perforation. The total length of intestine is reduced to a varying extent.



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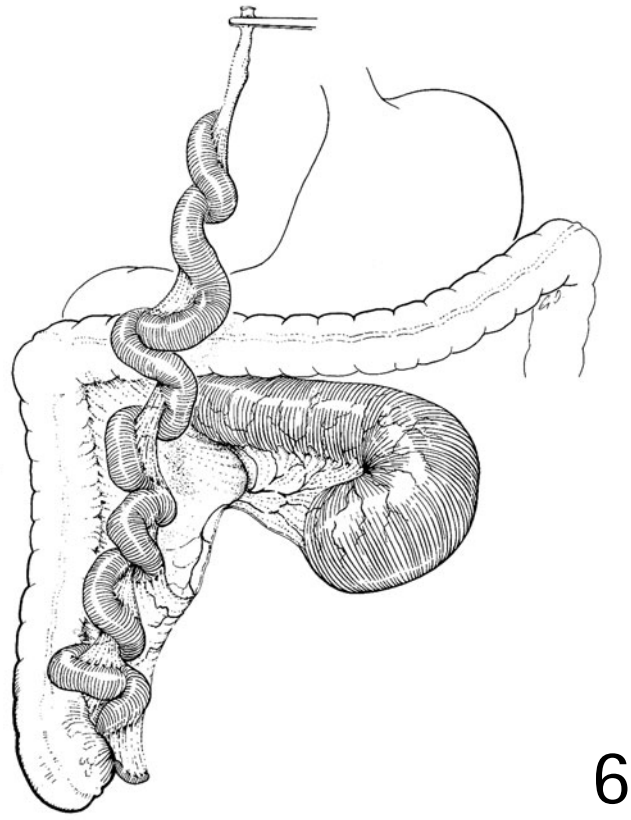

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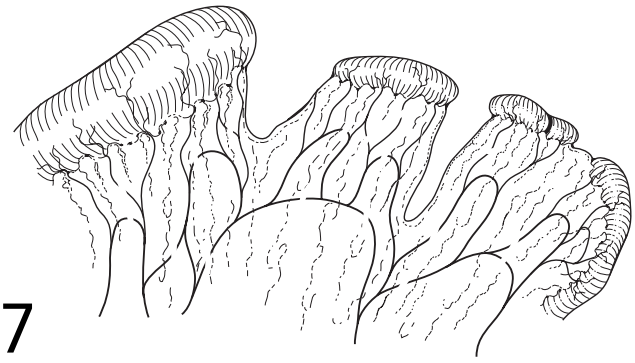
ATRESIA TYPE IIIB ('APPLE PEEL', 'CHRISTMAS TREE' ATRESIA)

6 As in type IIIa, the blind ends are unconnected and the mesenteric defect is large. The atresia is usually localized in the proximal jejunum near the ligament of Treitz, with absence of the superior mesenteric artery beyond the origin of the middle colic branch and absence of the dorsal mesentery. The distal intestine assumes a helical configuration around an attenuated, single perfusing vessel arising from the ileocolic or right colic arcade. Occasionally, atresia type I or II is found in the distal intestine, usually close to the blind end. Vascularity of the distal intestine may be impaired. There is always a significant reduction in intestinal length.



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ATRESIA TYPE IV (MULTIPLE ATRESIA)

7 Multiple atresias can be combinations of types I–III and often present morphologically as a string of sausages.

Prognostic factors

- Prematurity and congenital anomalies are associated with increased mortality.
- Delayed diagnosis may precipitate ischemia and necrosis of the proximal atretic segment.
- Type III atresia – has the highest mortality.
- Short bowel syndrome, which may be due to extensive intrauterine bowel loss, over-zealous bowel resection, ischemic injury to the bowel, or postoperative complications.

Complications

Although a survival rate of more than 90 percent can be expected, complications are not infrequent. These include anastomotic leaks and stricture formation, ischemia of the bowel due to the delicate blood supply, especially in type IIIb, adhesive bowel obstruction, the short bowel syndrome, and infections related to pneumonia and septicemia.

PREOPERATIVE ASSESSMENT AND PREPARATION

Clinical presentation

Many cases of intestinal atresia are now being diagnosed prenatally by ultrasonographic investigation of the fetus, showing dilated fetal intestine, suggesting obstruction, particularly in pregnancies complicated by third trimester polyhydramnios. Postnatally, atresia or severe stenosis of the small intestine presents as neonatal intestinal obstruction with persistent bilious (green) vomiting dating from the first or

second day of life, varying degrees of abdominal distension, and perhaps some abnormality in evacuating meconium. Tenderness, rigidity, and erythema of the abdominal wall may signify bowel ischemia or peritonitis.

Radiology

The diagnosis of atresia is confirmed by radiologic examination. Erect and supine abdominal radiographs done after 6–8 hours of birth will reveal distended, air-filled small intestinal loops proximal to the obstruction and a gasless distal abdomen.

In some cases, the first abdominal radiograph reveals a completely opaque abdomen due to a fluid-filled obstructed bowel. Emptying of the stomach by means of a Replogle nasogastric tube and the injection of a bolus of air will demonstrate the level of the obstruction. When intestinal stenosis is present, an abnormal differentiation in caliber of the proximal obstructed intestine and the distal collapsed intestine will be evident. The diagnosis may be difficult and is often delayed. When the radiograph suggests a complete low obstruction, a contrast enema is given to rule out associated colonic atresia or functional obstruction, e.g., total colonic aganglionosis or meconium ileus, which may be confused with atresia of the distal ileum. When an incomplete small intestinal obstruction is diagnosed, an upper gastrointestinal contrast study is indicated to demonstrate the site and nature of the obstruction.

Factors that can mimic jejunoileal atresia include midgut volvulus, meconium ileus, incarcerated hernia, Hirschsprung's disease, colonic atresia, birth trauma, prematurity, drugs, and hypothyroidism.

Preoperative preparation

- Decompression to prevent aspiration.
- Fluid management:
  - maintenance,
  - replacement of deficiency/ongoing losses.
- Plain abdominal radiograph (air contrast).
- Contrast enema.
- Correction of hematological and biochemical abnormalities.
- Prophylactic antibiotics.

Anesthesia

The major anesthetic considerations are related to prematurity, fluid and electrolyte disturbance, abdominal distension, the risk of aspiration, and additional congenital anomalies.

Abdominal distension may compromise both cardiac and ventilatory function. A tense abdomen may obstruct venous return, compounding the hypotension. Release of this tension at laparotomy may be associated with a fall in blood pressure, particularly if preoperative resuscitation is inadequate. Ventilatory function may be compromised by diaphragmatic splinting, which will improve significantly after the release of tension. These infants, particularly those who are premature, are also at risk of regurgitation and pulmonary aspiration, which may further compromise respiratory function. Bowel perforation may cause septicemia and further distension.

Prior to the induction of anesthesia, the nasogastric tube should be aspirated to reduce the risk of pulmonary aspiration, as a significant residual gastric volume may be present. Rapid sequence induction and cricoid pressure may further reduce the risk. Invasive monitoring is indicated in sick or unstable infants, and a central line may be required for intravenous feeding postoperatively.

The anesthetic management is dictated by the condition of the infant and the available facilities. Light general and epidural anesthesia may avoid the need for postoperative ventilation.

OPERATION

Principles of surgery

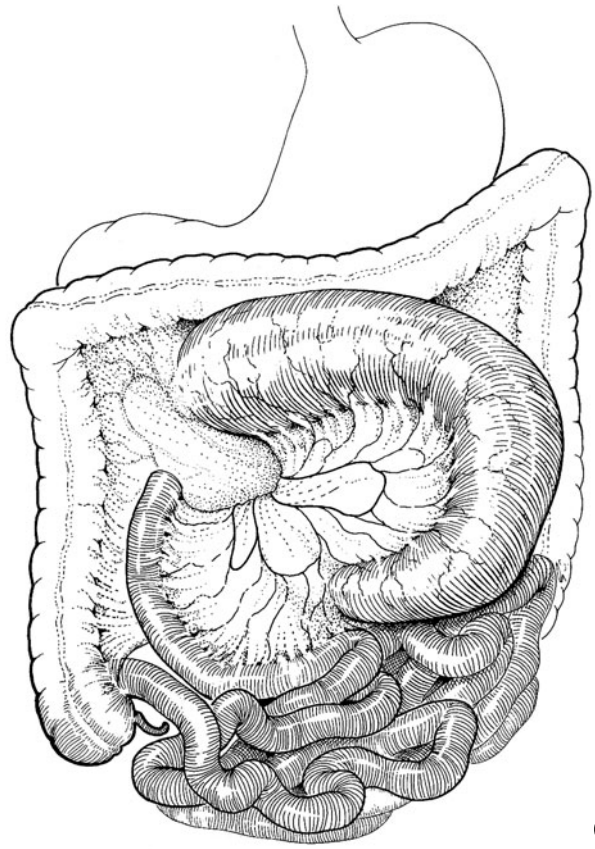
- The operative procedure depends on the type of atresia.
- The whole length of the small intestine should be inspected carefully to determine the site and type of atresia and the most likely pathogenesis.
- Patency of the distal small and large bowel must be confirmed.
- Back resection of the proximal dilated bowel with primary, single-layer, end-to-end (end-to-back) anastomosis with or without tapering or plication is the most common operative procedure.
- Residual bowel length must be documented.
- Ischemic or twisted bowel must be untwisted, and anastomosis may be delayed for 24 hours to assess viability.
- Every effort should be made to preserve bowel length in the presence of foreshortened bowel in type IV atresia.
- Proximal or distal stomas are rarely indicated.
- The bowel must be returned into the abdominal cavity, avoiding twisting or kinking of the anastomosis.
- Additional steps may include de-rotation of the proximal jejunum and duodenum in high intestinal atresia, with back resection of the dilated bowel followed by tapering or inversion plication of the megaduodenum.
- Gastric decompression is best achieved with a Replogle nasogastric tube on low continuous suction.
- Bowel-lengthening procedures should not be performed at the initial operation.

Incision

Adequate exposure is obtained through a long, supraumbilical, transverse incision transecting the rectus muscles 2 cm above the umbilicus.

Exploration

8 In uncomplicated cases, the intestine can be delivered through the wound by gentle exertion of pressure on the abdominal wall. If free gas escapes on opening the peritoneum, or if there is contamination of the peritoneal cavity, the perforation should be identified immediately and closed before further exploration. The intestine proximal to the obstruction is distended, whereas the intestine distal to the obstruction is collapsed, tiny, and worm-like, and may contain intraluminal contents. All of the intestine is exteriorized to determine the site and type of obstruction and to exclude other areas of atresia or stenosis as well as associated lesions, e.g., incomplete intestinal rotation or meconium ileus.



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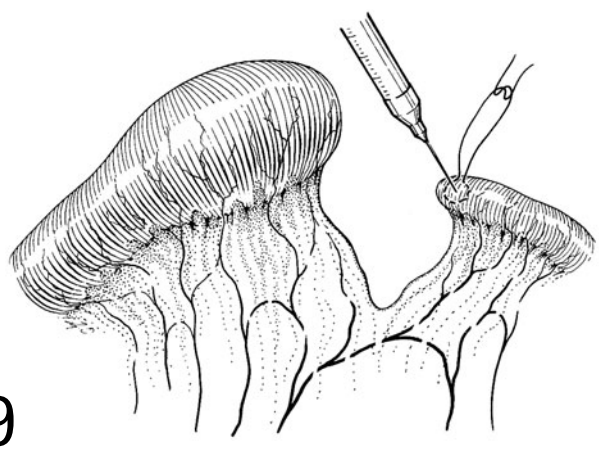


9

Detection of other atretic areas

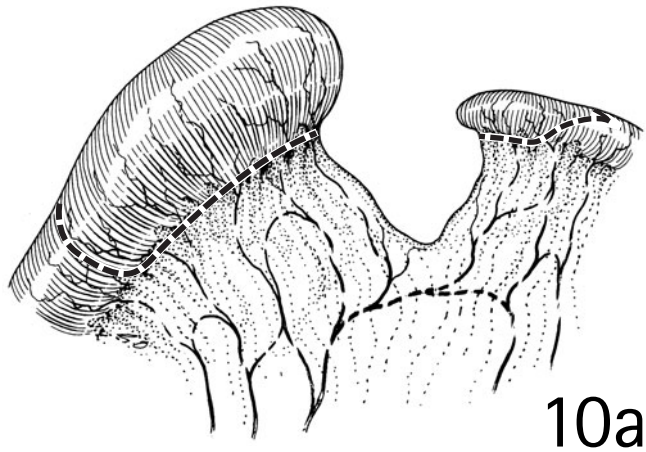
9 After the type and location of atresia have been determined, the distal intestine is carefully examined. If a volvulus is present, the bowel should be untwisted. Other atretic segments should be excluded, which can occur in 6–21 percent of cases. Intraluminal membranes are best detected and localized by injecting normal saline into the lumen of the collapsed intestine and following the advancing fluid column down to the cecum. Colonic atresia is excluded by a similar procedure through the cecum or by a previously performed contrast enema.

The total length of small intestine is measured accurately along the antimesenteric border. The normal length at birth is approximately 250 cm and in the preterm infant 115–170 cm. Residual bowel length in excess of 80 cm and the presence of an ileocecal valve usually preclude the short bowel syndrome.

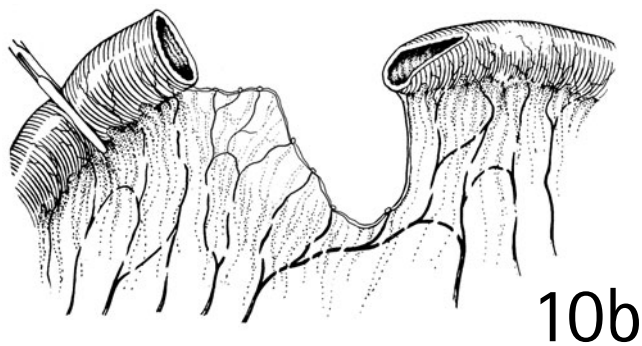
Resection

10a,b The atretic area and adjacent distended and collapsed loops of intestine are isolated by replacing the rest of the intestine into the abdomen and walling off the abdominal cavity with moist packs.

After milking the intestinal contents into the proximal bulbous end, or in high jejunal atresia into the stomach, from where it is aspirated, an atraumatic bowel clamp is applied across the bowel a few centimeters proximal to the site selected for transection. To ensure adequate postoperative function, the proximal distended and hypertrophied intestine must be liberally resected. We usually resect 10–15 cm even if it appears viable. The mesentery adjoining the portion to be resected is clamped, ligated, and divided. The proximal intestine is transected at right-angles. The blood supply at this point should be excellent. Some 2–3 cm of the distal intestine is then resected using a slightly oblique line of transection to create a 'fish mouth' that renders the opening about equal in size to that of the proximal intestine. A bacterial swab is taken for culture as the proximal gastrointestinal tract may have become colonized with bacteria.



10a



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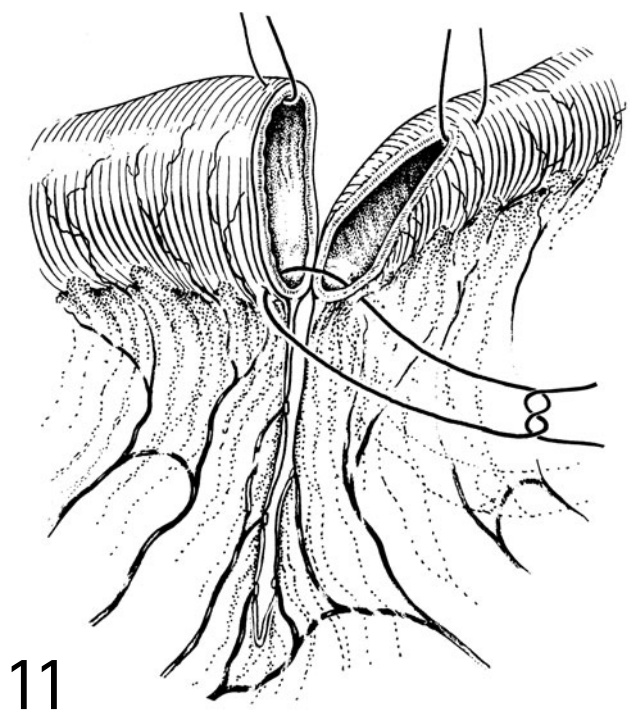
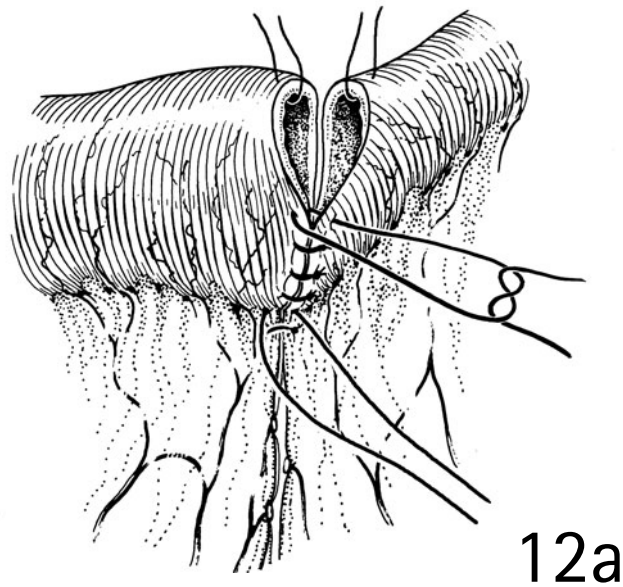


11

Uniting the mesenteric borders of bowel

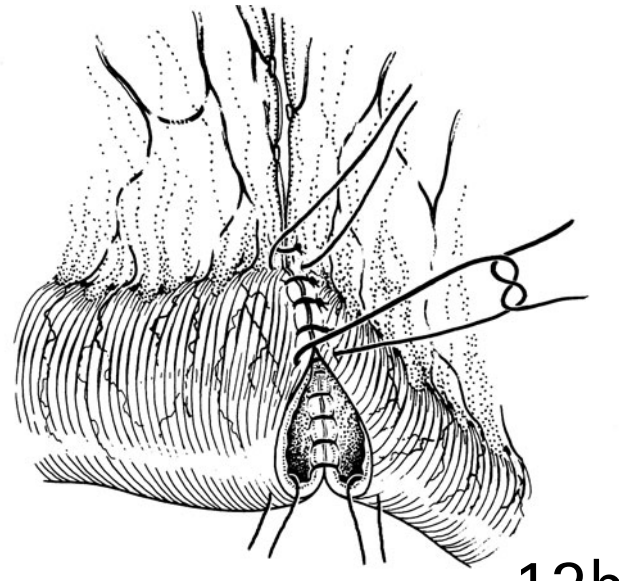
11 A 5/0 or 6/0 monofilament, synthetic, absorbable, full-thickness suture unites the mesenteric borders of the divided ends, and temporary stay sutures are placed at the antimesenteric angles to facilitate accurate approximation.



12a

Anastomosis

12a–c The 'anterior' bowel edges are joined by through-and-through interrupted sutures, starting from the mesenteric side, and tied on the serosal surface. Once the anterior layer has been completed, the bowel is rotated 180° to expose the back wall. If there is size discrepancy between the transected proximal and distal ends, a short cut back along the antimesenteric border can be performed on the distal bowel.



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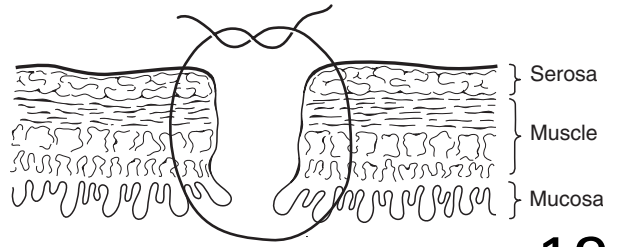
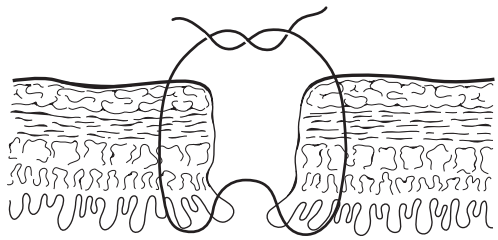


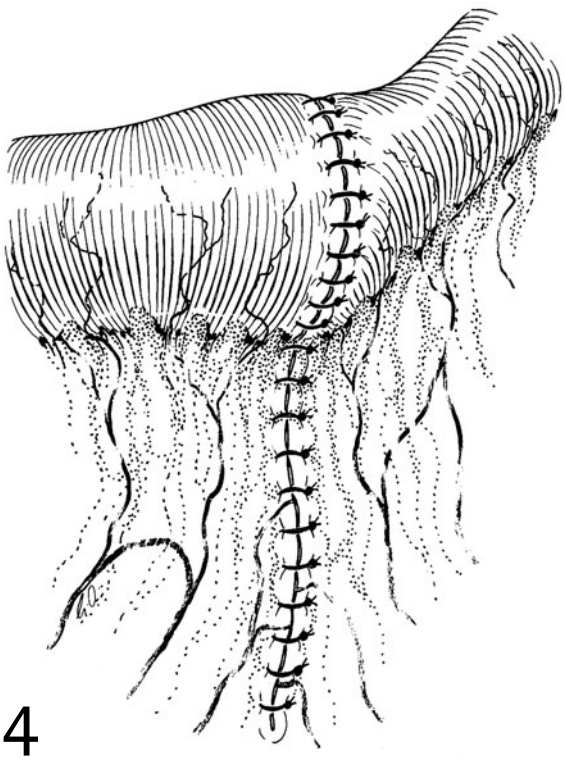
12c

13 Gambee interrupted inverting stitches may also be used instead of through-and-through stitches. Alternatively, the posterior bowel edges are united with interrupted through-and-through or inverting Gambee sutures, with the knots tied on the mucosal surface. The anterior bowel edges are then joined in a similar fashion, with the knots being tied on the serosal surface.



13

A similar technique is used for stenosis and intraluminal membranes. Procedures such as simple enteroplasties, excision of membranes, and bypassing techniques are not recommended because they fail to remove the abnormal segment of intestine. Side-to-side anastomosis is avoided because of the increased risk of creating blind loops.



14

Completion of anastomosis and closure of mesenteric gap

14 The completed anastomosis is not strictly end to end, but a modification of Denis Browne's 'end-to-back' method. The suture lines are inspected and additional stitches are placed if required to ensure a 'watertight' anastomosis. The defect in the mesentery is repaired by approximating (and overlapping if necessary) the divided edges with interrupted sutures, taking great care not to kink the anastomosis or to compromise the blood supply. Thereafter the intestines, well moistened with warm saline, are returned to the peritoneal cavity.

Closure of abdominal wound

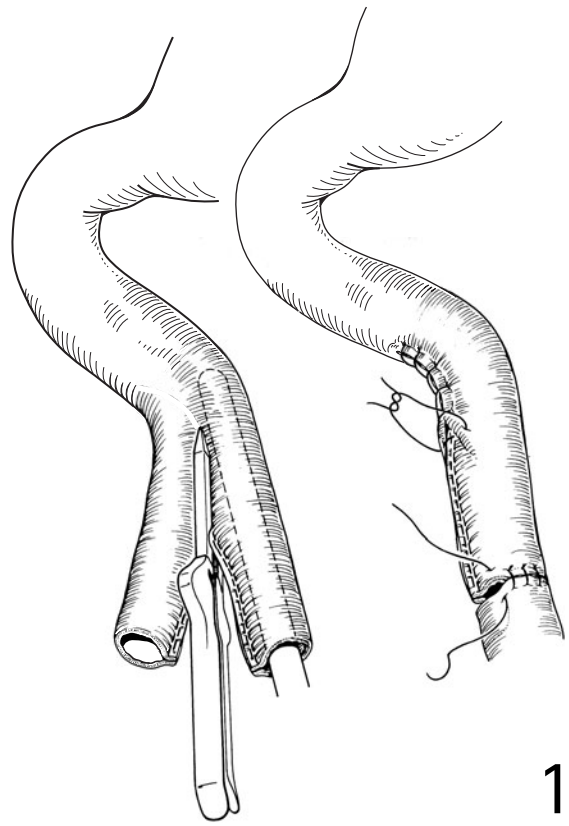
Before closure, the whole peritoneal cavity is irrigated with saline and all blood clots and particulate matter removed. The anterior abdominal wound is then closed in layers or, alternatively, with a single-layer mass closure utilizing a continuous through-and-through absorbable stitch, excluding Scarpa's fascia and skin. Scarpa's fascia and subcutaneous layers are approximated with absorbable sutures. The skin is sutured with a continuous, synthetic, absorbable suture or approximated with adhesive strips and then covered with a thin, sterile skin dressing. No drains are used.

BOWEL-SAVING PROCEDURES

Tapering duodenojejunoplasty or enteroplasty

15 This surgical procedure is indicated for bowel-length preservation, especially in type IIIb atresia and for high jejunal atresias. The bulbous, hypertrophied proximal bowel is de-rotated and back resected along the antimesenteric border into the third or second duodenal segments. The tapering is performed over a 22–24 Fr catheter to ensure adequate luminal size. An intestinal autostapling instrument may greatly facilitate this procedure. The linear anastomosis is reinforced with interrupted absorbable 5/0 or 6/0 sutures. Tapering can safely be done over an extended 10–20 cm length.

The tapered bowel is then anastomosed to the distal bowel and replaced in a non-rotated position. Tapering is also indicated for equalizing diameter size for more distal atresias and for correction of a failed inversion plication procedure.



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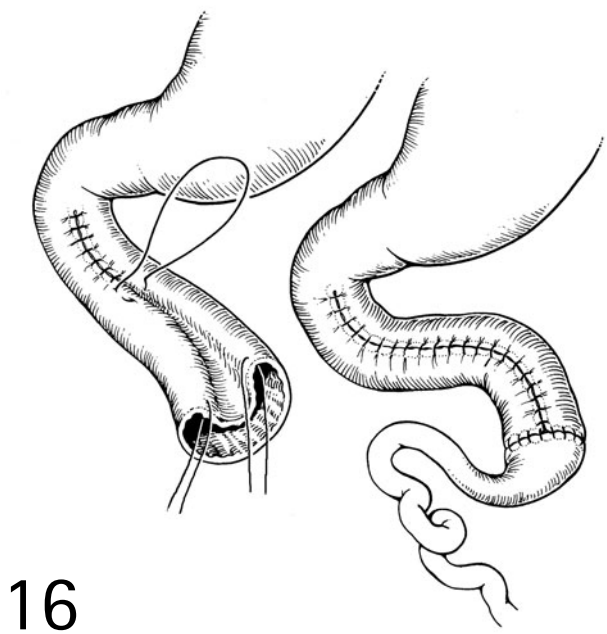


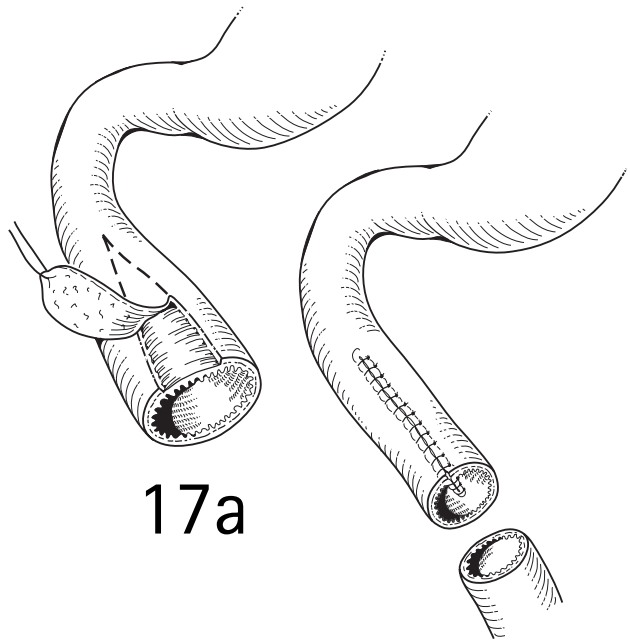
16

Plication or infolding enteroplasty

16 The same technique used for tapering enteroplasty is used, except that antimesenteric intestinal plication involves infolding of up to half or more of the intestinal circumference into the lumen over an extended length. The plication is performed by a running stitch up to 1 cm from the planned anastomotic site. The distal end is then completed by interrupted stitches to allow for additional surgical trimming if required. The keel created by the infolding should be sutured closed. Excessive infolding must be avoided and the patency of the lumen not compromised. The infolding is accomplished with non-absorbable sutures and the bowel is left in a position of non-rotation.

Antimesenteric seromuscular stripping and inversion plication

17a,b This technique tapers the dilated proximal bowel segment and prevents unraveling of a plication enteroplasty. An antimesenteric, elliptical, seromuscular segment of the dilated proximal bowel is resected by blunt and sharp dissection from the underlying submucosa. The muscular margins so created are approximated with interrupted sutures, with the underlying mucosa imbricated or inverted into the lumen of the bowel. The distal end of the created keel should be tapered and sutured closed. This technique prevents anastomotic leakage and enhances the establishment of prograde peristaltic activity.



17a

17b

SPECIAL SURGICAL CONSIDERATIONS

Multiple atresias

Multiple atresias are often localized to a short segment of intestine, and resection with one anastomosis is preferred if sufficient intestinal length remains. If the bowel length is critical, multiple anastomoses should be performed.

Bowel lengthening

Bowel-lengthening procedures have no place at the initial operation.

Exteriorization

Exteriorization of proximal and distal intestine may be required with established peritonitis or questionable vascularity of the remaining intestine. The authors do not favor the fashioning of stomas.

Intestinal atresia and gastroschisis

Primary anastomosis may be difficult and hazardous, and the favored option is reduction of the eviscerated intestine with the atresia left intact, closure of the abdominal wall defect, and delayed resection and primary anastomosis of the atresia at 14–21 days.

POSTOPERATIVE CARE

Nasogastric decompression is usually required for 4–6 days after the operation (longer for high jejunal atresias). Therapeutic antibiotics are continued for 5–7 days or longer, and an oral antifungal agent is given prophylactically. Oral intake is commenced when the neonate is alert, sucks well, and there is evidence of prograde gastrointestinal function, i.e., clear gastric effluent of low volume, a soft abdomen, or when flatus or feces has been passed. Surveillance for gastrointestinal dysfunction should continue until the infant has established normal gastrointestinal function.

If at any time there is suspicion of a leak at the anastomosis (suggested by sudden collapse, abdominal distension, and vomiting), a plain erect or decubitus radiograph of the abdomen should be taken. If this reveals free air in the abdomen more than 24 hours after operation, laparotomy should be performed immediately and the leaking site sutured, or the anastomosis redone.

OUTCOME

Before 1952, the mortality rate for congenital atresias of the small intestine in Cape Town was 90 percent. Between 1952 and 1955, 28 percent of the neonates survived. At that stage, most were treated by primary anastomosis without resection. With liberal back resection of the blind ends and end-to-end anastomosis, the survival rate increased to 78 percent during

the period 1955–58. During the 46-year period 1959–2005, 310 patients with jejunoileal atresia and stenosis were admitted to the Paediatric Surgical Service at the Red Cross Children's Hospital, of whom 31 have died, giving an overall mortality rate of 10 percent (Tables 43.1–43.3). However, in line with the improved preoperative and postoperative care of newborns, our last 15-year mortality rate dropped to 4.6 percent (5 mortalities out of 107 patients). Causes of death in these five patients were short bowel syndrome, moribund at presentation due to volvulus and necrotic bowel (IIIb), congenital syphilis, sepsis, and cot death at 2 months of age. Other causes of death prior to 1990 included five neonates who were moribund on admission with infarction of the proximal intestine due to volvulus of the bulbous end and established peritonitis (types I, II, IIIa, and two type IIIb). Five infants died from infection related to pneumonia and septicemia. Further medical care was withheld from two patients with less than 10 cm residual small intestine length. One infant died from bleeding due to vitamin K deficiency, and 13 died from the short gut syndrome (nine early and four late deaths).

There was a total of 18 complications in 107 patients seen during the last 14 years. These included short bowel syndrome ($n = 4$), anastomotic leak ($n = 4$), functional obstruction, wound dehiscence, wound sepsis, and generalized sepsis (2 each), one child with a kink at the anastomotic site, and one with anastomotic stenosis.

Table 43.1 *Jejunoileal atresia and stenosis: Red Cross Children's Hospital experience*

Type	Jejunum	Ileum	Total (%)
Stenosis	20	13	33 (10.6%)
Type I	57	15	72 (23.2%)
Type II	18	12	30 (9.6%)
Type IIIa	26	23	49 (15.4%)
Type IIIb	58	–	58 (18.7%)
Type IV	54	14	68 (21.9%)
Total	233	77	310

1959 to 2005 ($n = 310$).

Table 43.2 *Mortality related to type of atresia*

Type	Patients	Mortality	Percent
Stenosis	33	0	0
Type I	72	4	5.6
Type II	30	3	10
Type IIIa	49	8	16.3
Type IIIb	58	9	15.5
Type IV	68	7	10.3
Total	310	31	10

Table 43.3 *Jejunoileal atresia and stenosis: improvement in survival*

Authors	Years of study	*n*	Survival (%)
Evans	1950	1498	9.3
Gross	1940–1952	71	51
Benson and colleagues	1945–1959	38	55
De Lorimer	1957–1966	587	65
Nixon and Tawes	1956–1967	62	62
Louw	1959–1967	33	94
Martin and Zerella	1957–1975	59	64
Cywes and colleagues	1959–1978	84	88
Danismead and colleagues	1967–1981	101	77
Smith and Glasson	1961–1986	84	61
Vecchia and colleagues	1972–1997	128	84
Rode and colleagues	1990–2005	107	96.4

FURTHER READING

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Evans CH. Atresias of the gastrointestinal tract. *International Abstracts of Surgery* 1951; **92**: 1–8.

Kimura K, Perdzynski W, Soper RT. Elliptical resection of tapering the proximal dilated bowel in duodenal or jejunal atresia. *Journal of Pediatric Surgery* 1996; **31**(10): 1405–6.

Kling K, Applebaum H, Dunn J, Buchmiller T, Atkinson J. A novel technique for correction of intestinal atresia at the ligament of Treitz. *Journal of Pediatric Surgery* 2000; **35**(2): 353–6.

Louw JH, Barnard CN. Congenital intestinal atresia. *Lancet* 1955; **269**: 1065–7.

Smith GHH, Glasson M. Intestinal atresia: factors affecting survival. *The Australian and New Zealand Journal of Surgery* 1989; **59**: 151–6.

44

Meconium ileus

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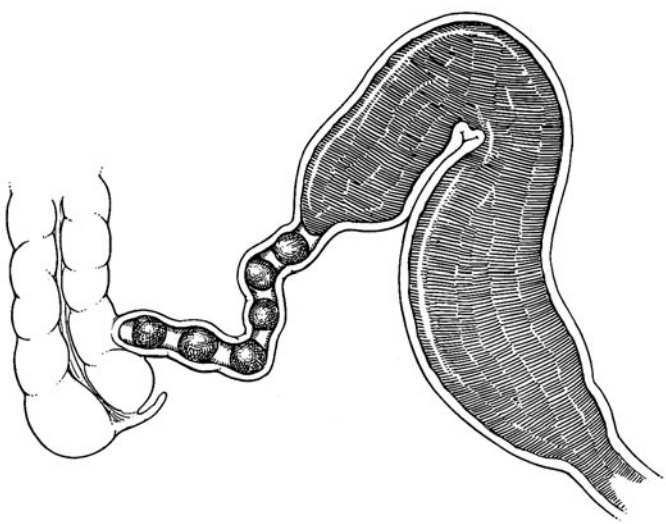
ETIOLOGY AND CLASSIFICATION

Cystic fibrosis is the most common serious inherited defect affecting the Caucasian population. Cystic fibrosis is transmitted as an autosomal recessive condition with a 5 percent carrier rate and an incidence of approximately 1:2500 live births. The cystic fibrosis transmembrane conductance regulator (*CFTR*) gene is located on the long arm of chromosome 7. According to the Cystic Fibrosis Genetic Analysis Consortium, 13 mutations occur at a frequency of greater than 1 percent and account for 87 percent of cystic fibrosis alleles. The delta F508 mutation is the most common and is present in 70 percent of CF alleles in the USA. There are great differences among populations, and among African Americans delta F508 only accounts for 43 percent of the alleles. The CFTR protein controls sodium and chloride transport and in cystic fibrosis this results in abnormal luminal secretions. Neonatal intestinal obstruction due to inspissated meconium has been identified since the early reports concerning cystic fibrosis and is referred to as meconium ileus. This presentation is observed in 10–15 percent of infants born with cystic fibrosis. The etiology of this abnormal meconium (mucoviscidosis) is due to deficient pancreatic and intestinal secretions as well as an abnormal concentration of the meconium within the duodenum and proximal jejunum. Instances of meconium ileus can be classified into uncomplicated and complicated cases.

Uncomplicated meconium ileus

CLINICAL PRESENTATION

In this condition the abnormal thickened meconium causes a simple obturator obstruction of the terminal ileum. The distal 15–30 cm of terminal ileum is filled with inspissated meconium pellets, which are adherent to the bowel wall. The ileum just proximal to the obstruction fills with thick, putty-like meconium and dilates to 3–4 cm in diameter. The colon is unused and small (microcolon), because meconium has not yet entered this segment of bowel.



1

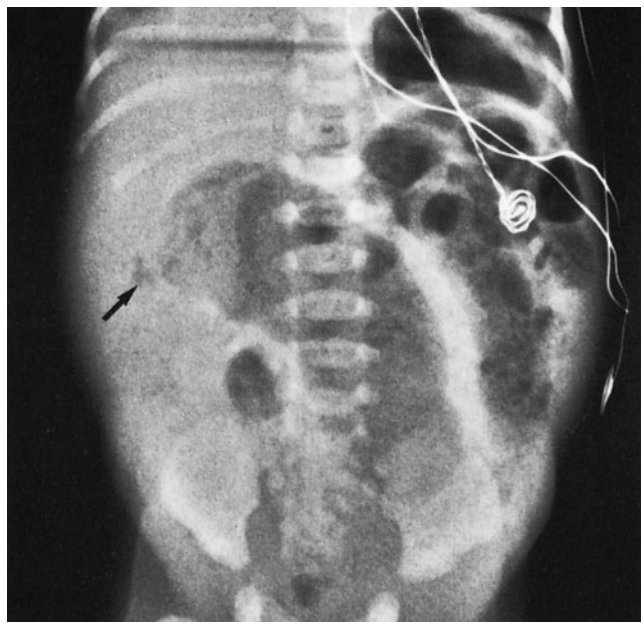
The typical neonate with meconium ileus may appear relatively normal for the first 12–18 hours of life and some tolerate several feeds. As the proximal bowel fills with swallowed air, however, abdominal distension and emesis (initially clear, later bilious) and failure to pass meconium are noted, heralding the presence of intestinal obstruction at 24–36 hours of age.

INITIAL MANAGEMENT

The neonate with suspected bowel obstruction should be treated with oral gastric tube decompression of the stomach and intravenous fluids to replace pre-existing fluid deficits and ongoing losses. Antibiotics are administered, as the differential diagnosis of a newborn with this presentation includes sepsis.

RADIOGRAPHIC EVALUATION

2 Plain abdominal radiographs and decubitus views usually demonstrate similar-sized dilated loops of intestine without air–fluid levels. A 'soap bubble' appearance is often noted in the right lower quadrant (arrow), a result of air mixing with the thick meconium.



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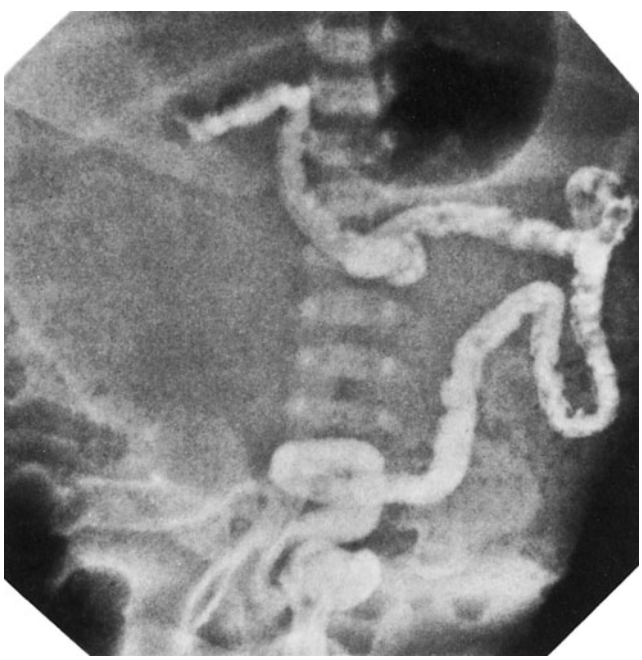


3

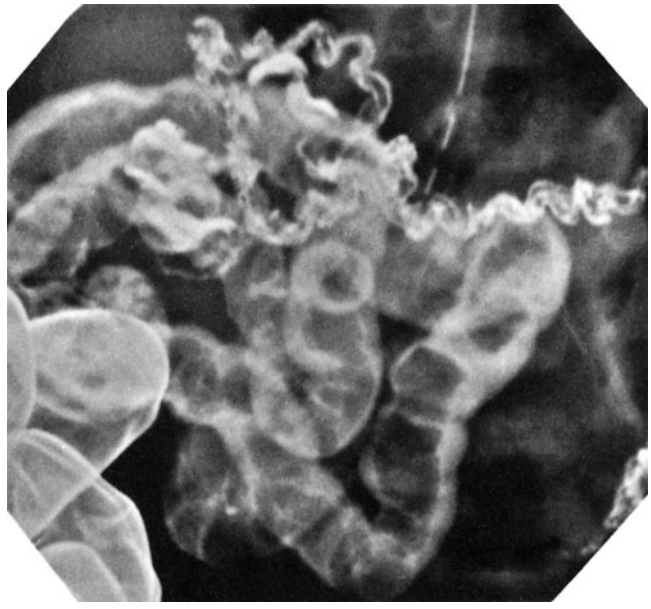
DIAGNOSTIC ENEMA

3 The initial diagnostic test is a contrast enema. If the diagnosis of meconium ileus is not apparent from the plain radiographs, a barium enema is the diagnostic procedure of choice. This will demonstrate a microcolon and may also document the presence of meconium pellets in the proximal ascending colon and terminal ileum.

This study will also exclude cases of colon atresia, small left colon syndrome, and meconium plug syndrome, and document the location of the cecum to rule out anomalies of rotation and fixation. Neonates with distal ileal atresia and total colonic aganglionosis may have similar appearance on a contrast enema examination, but these neonates usually have air–fluid levels in the dilated proximal small bowel and absence of pellets in the distal ileum and proximal colon. If the neonate is stable and there is no evidence of complicated meconium ileus (peritoneal calcifications, giant cystic structure, etc.), non-operative treatment with a hypertonic contrast material enema is recommended.

THERAPEUTIC ENEMA

The management of neonates with uncomplicated meconium ileus was significantly altered with the introduction of the diatrizoate meglumine (Gastrografin) enema by Noblett in 1969. The efficacy of this procedure is related to the hyperosmolar nature of Gastrografin (1100–1900 mOsm/L), which contains a wetting agent (Tween 80) and draws large volumes of fluid into the bowel lumen, thus washing out the obstructing meconium. Although initial reports used full-strength Gastrografin, most pediatric radiologists dilute the contrast material to approximately 3:1, and the Gastrografin currently used does not contain a wetting agent. Complications reported following Gastrografin enema include: perforation, necrotizing enterocolitis, shock, and the occasional death. Most of these events are probably related to the hyperosmolar nature of the contrast material causing fluid depletion, which results in decreased intestinal blood flow and perfusion. Some radiologists prefer using other agents, such as diatrizoate sodium (Hypaque) or iothalamate meglumine (Conray), alone or in combination with *N*-acetylcysteine, but the authors prefer to use dilute Gastrografin. It is essential that the radiologists and clinicians are aware of the osmolality of the solution.



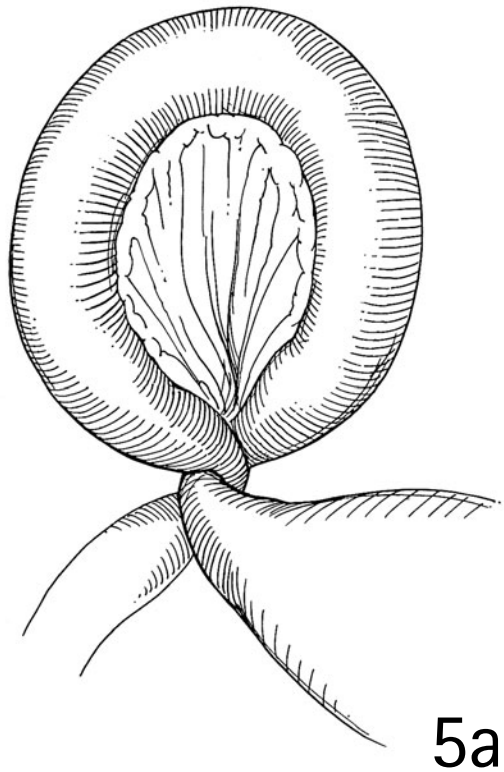
4 The enema is gently administered under fluoroscopic control and the contrast material flushed around the obstructing meconium pellets in the terminal ileum.

Before the enema, an intravenous route is established and the infant is appropriately resuscitated, and fluids are infused at a rate of 1.5 times maintenance during and after the procedure. The infant's pulse rate and urine output are carefully monitored in anticipation of fluid shifts into the bowel lumen. Meconium pellets, followed by loose meconium, generally pass through the rectum over the next 4–8 hours. If evidence of bowel obstruction persists and the infant remains clinically and hemodynamically stable, a second or third enema may be administered. *N*-acetylcysteine (2.5–5 percent, 5 mL every 6 hours) may be administered by oral gastric tube to aid in clearing the thickened meconium from above. As the clinical evidence of obstruction resolves, the oral gastric tube is removed and feeding advanced. The Gastrografin enema is successful in resolving the obstruction in approximately 55 percent of cases. Survival for these infants at 1 year is nearly 100 percent. If these non-operative efforts fail, surgical exploration is required.

Complicated meconium ileus

CLINICAL PRESENTATION AND INITIAL MANAGEMENT

5a–c Complicated cases include instances of volvulus, bowel perforation, intestinal atresia, and giant cystic meconium peritonitis. Volvulus usually occurs when the distended segment of ileum twists at the level of the narrow, pellet-filled, distal small intestine (a). In some cases, volvulus can result in bowel perforation, leading to meconium peritonitis (b), and in others, the bowel may become necrotic and liquefy, resulting in a pseudocyst. This latter condition is referred to as a giant cystic meconium peritonitis. Bowel atresias are thought to arise when the base of the volvulus becomes ischemic (c).



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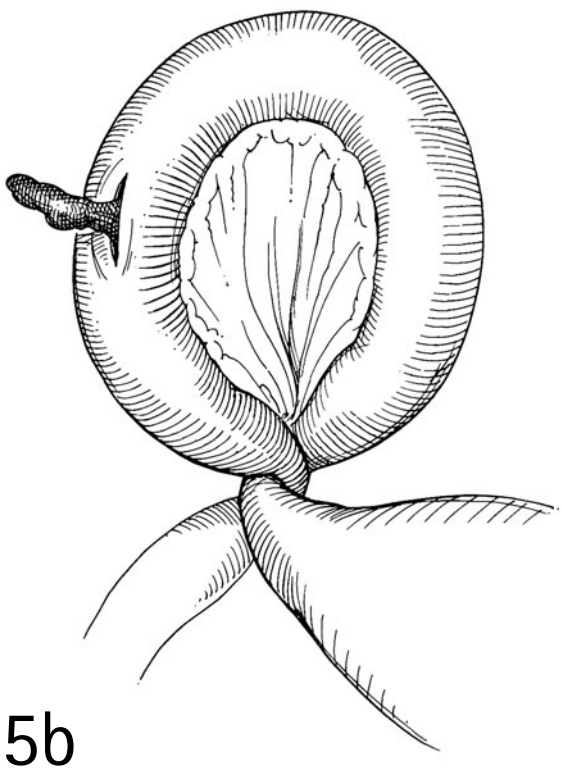

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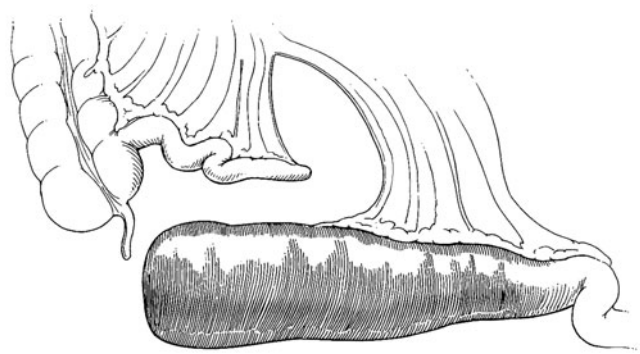


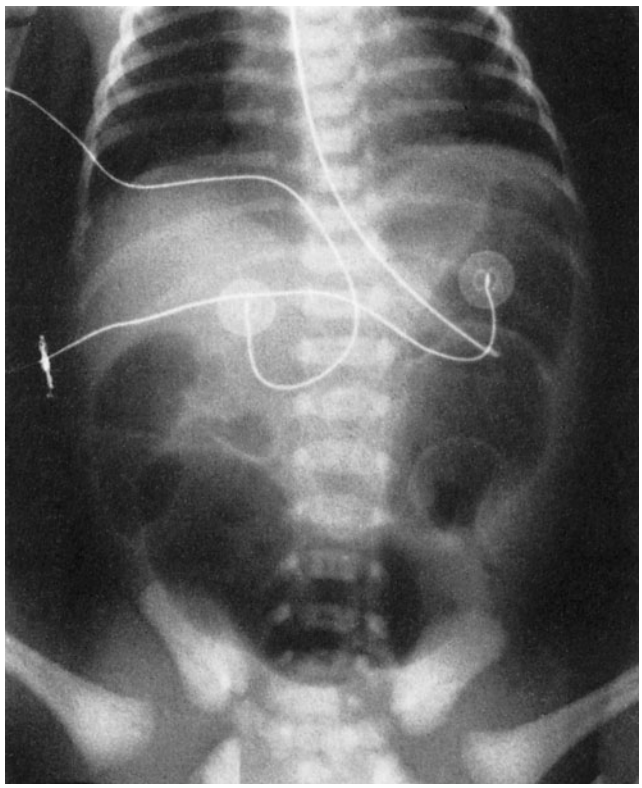
5c

Neonates with complicated meconium ileus usually present with abdominal distension at the time of, or shortly after, delivery. In addition, bile-stained fluid is usually noted in the stomach. On physical examination, an abdominal mass may be noted. Neonates with meconium peritonitis occasionally have meconium in the scrotal sac or vagina as a result of passage of this material through a patent processus vaginalis or the fimbriated ends of the fallopian tubes, respectively. In addition, in one unusual report a meconium pseudocyst appeared as a buttock mass. The early management of these neonates includes intravenous hydration, antibiotics, and oral gastric tube decompression of the stomach.

DIAGNOSIS

6 In contrast to neonates with uncomplicated meconium ileus, flat and erect or decubitus radiographs of the abdomen in complicated cases may demonstrate distended loops of small bowel of different size with air–fluid levels. Intraperitoneal calcifications from the extravasated meconium, characteristic of meconium peritonitis, may be noted. A mass effect or ascites may also be observed.

Neonates who can be identified as complicated cases by plain abdominal radiographs are taken to the operating room for prompt exploration. In uncertain cases, a barium enema may be useful to exclude other causes of distal obstruction.



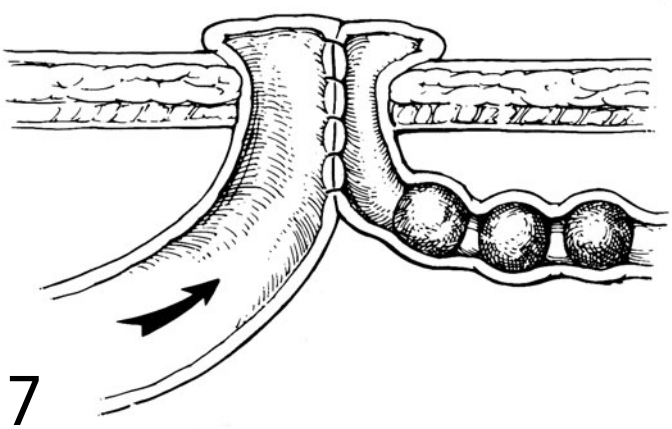
6

OPERATIONS

Uncomplicated meconium ileus

MIKULICZ PROCEDURE

Meconium ileus was often considered a fatal condition until 1948 when Hiatt and Wilson reported a number of survivors after enterotomy and irrigation. This technique was not widely utilized and, in 1953, Gross reported successful outcomes in infants with meconium ileus following bowel resection and use of Mikulicz enterostomy.



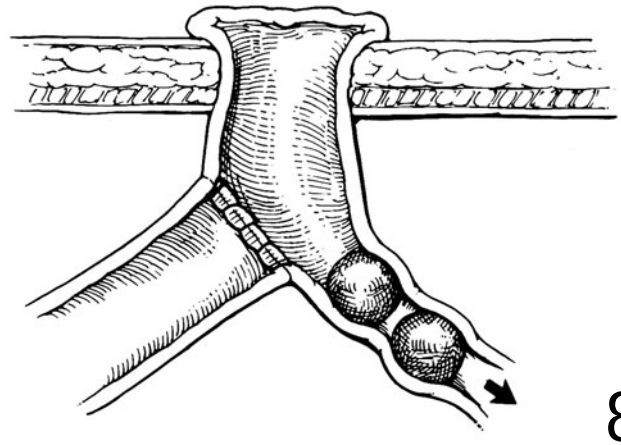
7

7 The dilated bowel loop filled with thickened meconium is brought out of the abdomen and the small bowel proximal and distal to this segment is sutured together in a side-to-side fashion by interrupted seromuscular sutures. Following closure of the abdomen, the exteriorized dilated bowel is resected, thus avoiding the risk of peritoneal contamination. This results in an enterostomy through which the distal bowel can be irrigated in the postoperative period to wash out the obstructing meconium pellets. A Mikulicz spur-crushing clamp is applied, resulting in a common lumen, and the ostomy is then closed at a later date.

The disadvantages of this procedure are the loss of fluids from the mid-small bowel ostomy, the need for a subsequent procedure to close the stoma, and some reduction of bowel length due to initial resection.

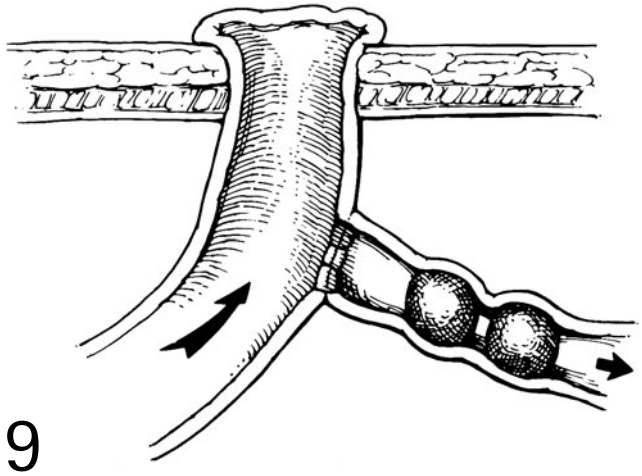
BISHOP–KOOP PROCEDURE

8 In 1957, Bishop and Koop reported resection of the large dilated loop followed by an anastomosis between the end of the proximal segment and the side of the distal segment. The end of the distal bowel is then brought out as an end ileostomy. A catheter is passed into the distal segment to allow postoperative irrigation. As the distal obstruction is relieved, the intestinal contents preferentially pass into the distal ileum and colon, thus decreasing loss of fluid and electrolytes from the stoma. The ostomy can be closed at a later date and, in some cases where it is trimmed beneath the skin, may close spontaneously.



8

The disadvantages of this technique include loss of bowel length at the time of the initial procedure, the need for an intraperitoneal anastomosis, and the need for a second operative procedure.



9

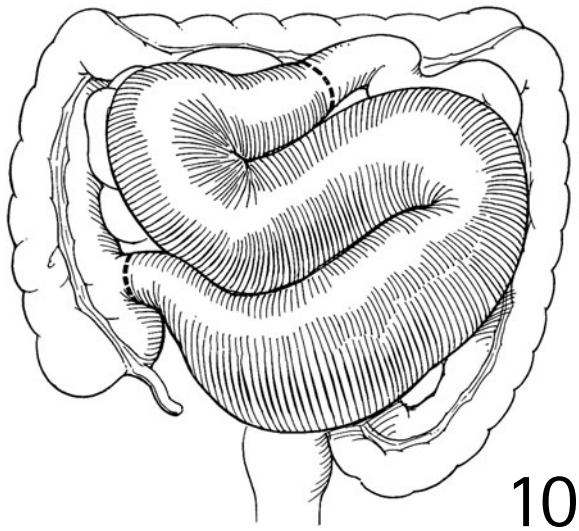
SANTULLI–BLANC ENTEROSTOMY

9 This modification of the Bishop–Koop procedure concept was reported in 1961. The operation involves resection of the distal dilated bowel segment followed by a side-to-end anastomosis with proximal enterostomy.

The disadvantages are similar to those noted for the Bishop–Koop procedure.

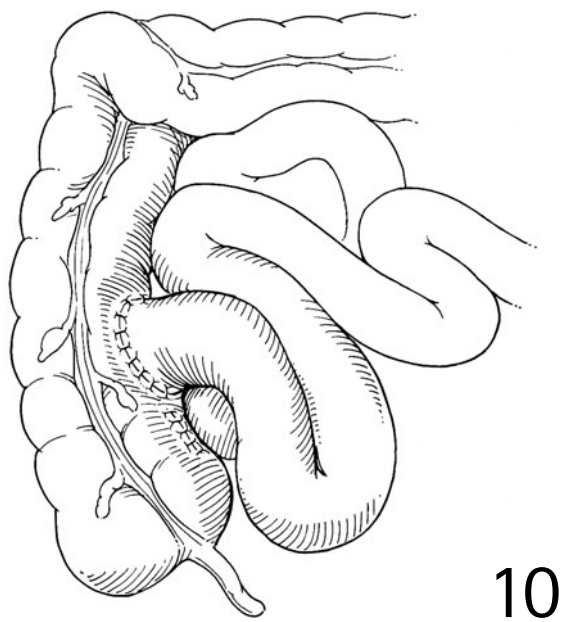
PRIMARY RESECTION AND ANASTOMOSIS

The use of resection with primary anastomosis in the management of meconium ileus was first reported by Swenson in 1962.



10a

10a,b After resection of the obstructed bowel segment, the remaining pellets in the distal bowel are irrigated clear and an ileocolonic anastomosis is performed.

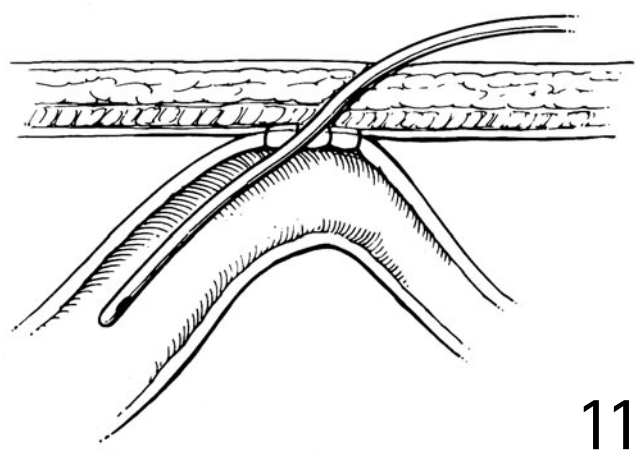


10b

The disadvantage associated with this procedure is resection of additional bowel, as the terminal ileum containing meconium pellets was usually resected along with the dilated segment of ileum. This, as well as concerns about an unvented intraperitoneal anastomosis, prevented wide acceptance of this procedure.

TUBE ENTEROSTOMY

11 In 1970, O'Neill and colleagues reported success with a simple procedure involving tube enterostomy with postoperative irrigation. Their initial report of five neonates was followed by a report by Harberg et al. concerning 9 of 11 neonates who had successful meconium washout using this technique. Harberg and colleagues modified the technique slightly by utilizing a T-tube. A follow-up study of this technique noted success in 20 out of 23 patients with T-tube irrigation utilizing pancreatic enzymes or 1 percent *N*-acetylcysteine, 5–12.5 mL per dose. Unfortunately, three patients required a second procedure in the neonatal period due to inadequate treatment with the T-tube.

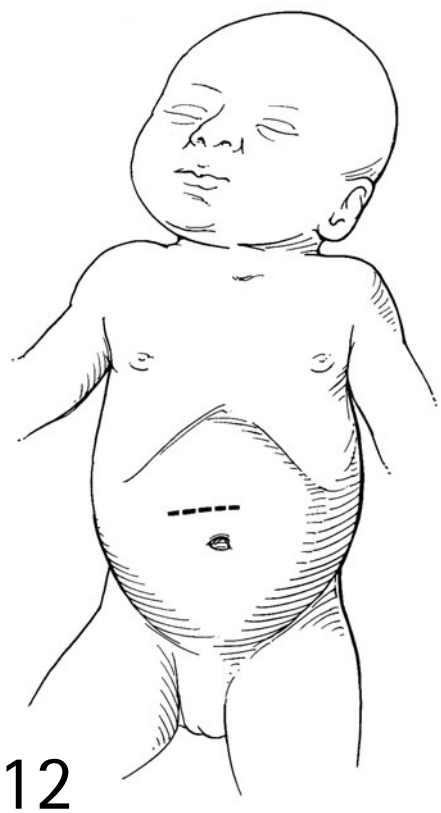


11

ENTEROTOMY AND IRRIGATION

The technique of enterotomy with irrigation has been one of the two procedures of choice at our institution since 1981. As previously noted, this procedure was originally described by Hiatt and Wilson in 1948. Reports by several other authors appeared in the literature between 1970 and 1990.

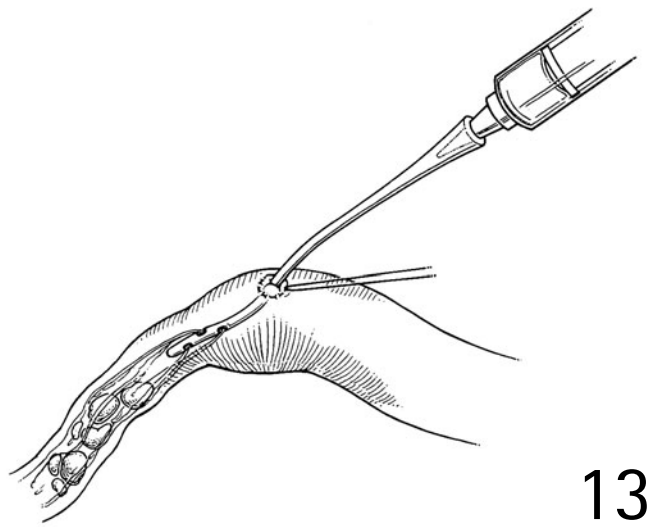
The infant is taken to the operating room and an endotracheal tube is placed before anesthesia to avoid aspiration.



12

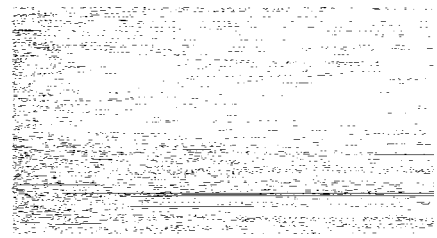
12 After induction of general anesthesia, the procedure is carried out through a right-sided supraumbilical transverse abdominal incision. The right rectus abdominis muscle, as well as a portion of the right external and internal oblique and transversus abdominis muscles, is divided using electrocautery. The peritoneal cavity is entered and explored. The distended meconium-filled loops and pellet-filled distal small bowel are identified and delivered into the wound.

13 A purse-string suture of 4/0 silk is placed on the antimesenteric border of the dilated bowel 6–8 cm proximal to the narrow region containing the obstructing pellets. An 8–10 Fr red rubber catheter is placed through a small enterotomy, the purse-string suture is snugged, and the bowel lumen is irrigated by gently instilling saline through a syringe attached to an adapter and three-way stopcock. Fluid is irrigated into the proximal thick meconium and around the distal pellets.

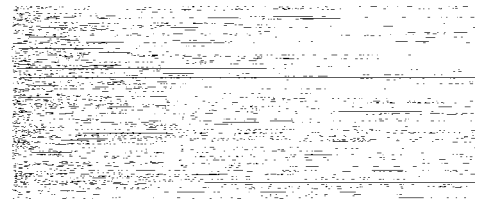


13

With irrigation and gentle manual manipulation, the thick meconium from the distended loop and the distal pellets can be removed through the enterotomy or washed out distally through the colon. Occasionally the enterotomy may need to be extended to a 1.5–2.0-cm opening to allow removal of the thick meconium and pellets. Numerous irrigations are usually required. Dilute Gastrografin or 2.5–5 percent *N*-acetylcysteine solution may also be used for irrigation. There have been occasional reports documenting hypernatremia with *N*-acetylcysteine. When the pellets are cleared, saline is irrigated through the distal ileum and into the colon to exclude the possibility of atresia and also to flush some of the distal pellet fragments into the colon. The enterotomy is then closed in two layers, using an inner full-thickness layer of absorbable 4/0 or 5/0 polyglactin (running or interrupted) and an outer seromuscular layer with interrupted 5/0 silk suture.



14a



14b

APPENDICAL IRRIGATION

14a,b In 1989, Fitzgerald and Conlon reported the use of the appendix to instill dilute Gastrografin around the thickened meconium and pellets and flush the material into the colon. This has become the other preferred technique at our institution. The catheter can usually be advanced from the cecum retrogradely into the ileum, and saline is gently irrigated into the terminal ileum (a); the surgeon gently compresses the pellets and meconium to allow smaller particles to mix with the saline. The catheter is then withdrawn into the appendix and the surgeon 'milks' the material into the colon (b). This process must usually be repeated several times and as the material is advanced into the ascending colon, it is gently flushed into the distal colon and out of the rectum. After the ileal obstruction is completely relieved, the appendix is removed in the standard fashion.

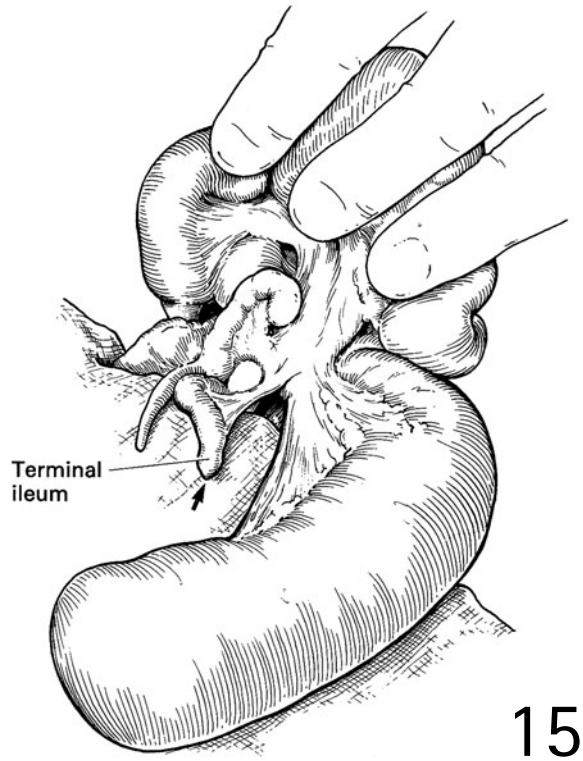
Complicated meconium ileus

The anesthetic management is similar to that described for uncomplicated cases. The abdomen is entered through a transverse supraumbilical incision. The operative findings and ease of procedure may vary significantly, from volvulus and bowel atresia to meconium peritonitis or giant cystic meconium peritonitis. These conditions are, therefore, be discussed separately.

VOLVULUS AND ATRESIA

In instances of meconium ileus associated with bowel volvulus or atresia, the pathology is usually easily identified at laparotomy, and a primary anastomosis is nearly always possible.

15a–c In cases of volvulus, the involved loop is resected and an end-to-oblique anastomosis is constructed between the dilated proximal bowel and smaller distal bowel. The proximal bowel is divided at a 90° angle with respect to the mesentery and the distal bowel at a 45° angle. The distal meconium pellets should be removed through the open bowel, and the distal segment should also be irrigated to facilitate return of bowel function and to avoid a postoperative obturator obstruction distal to the anastomosis. The anastomosis is constructed with one or two layers of interrupted 5/0 silk sutures.



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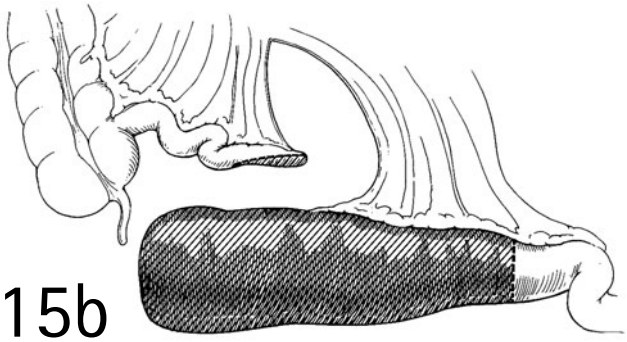

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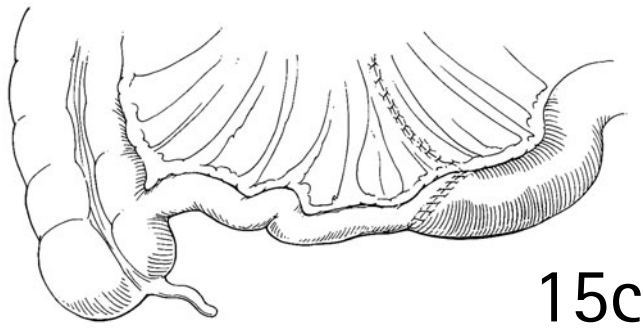


15c

In cases of atresia, if adequate bowel length is present, the proximal dilated segment (usually 10–15 cm) is resected, as it is frequently atonic, and an end-to-oblique anastomosis is fashioned.

MECONIUM PERITONITIS AND GIANT CYSTIC MECONIUM PERITONITIS

Neonates with these two disorders generally have numerous adhesions throughout the abdomen and may have significant blood and fluid losses during the operation. The abdomen is explored and the small bowel and colon carefully identified and dissected free from the numerous adhesions.

The necrotic dilated segment is resected and, if possible, an end-to-end or end-to-oblique anastomosis is constructed. In some cases this is not possible and a temporary enterostomy is required. The bowel opening may be found within the pseudocyst. Most of the pseudocyst wall should be resected, if possible. This may result in some blood loss requiring transfusion. The proximal end may be brought out as a temporary stoma through the corner of the wound or through a separate incision. The distal bowel can either be closed with sutures or staples and left in the abdomen, or brought out as a separate mucous fistula to allow irrigation as well as re-feeding of the effluent from the proximal ostomy into the distal bowel.

POSTOPERATIVE CARE

Uncomplicated meconium ileus

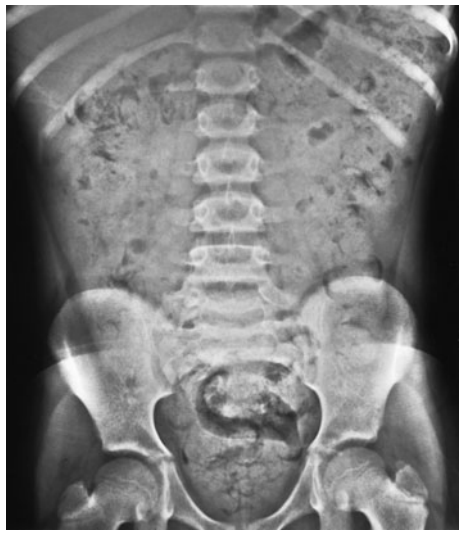
In the early postoperative period an oral gastric tube is left in place until bowel function returns. Use of *N*-acetylcysteine (2.5–5 percent, 5–10 mL in 6 hours) through an oral gastric tube may further aid passage of inspissated meconium. When bowel function returns, the tube is removed and enteral feedings are initiated along with pancreatic enzyme supplementa-

tion. The diagnosis of cystic fibrosis is confirmed by obtaining an elevated sweat chloride level on testing and the actual chromosomal defect is identified to assist in genetic counseling. The management of these patients requires a multidisciplinary team, including the pediatric respiratory physician, in order to optimize the pulmonary status, as pulmonary function deterioration is the major cause of morbidity and mortality.

LONG-TERM COMPLICATIONS

Gastrointestinal problems after the newborn period are relatively common in children with cystic fibrosis. These include intussusception, appendiceal distension with inspissated material and appendicitis, rectal prolapse, and gallbladder disease. In the early 1990s, colonic strictures were reported by several centers in association with high pancreatic enzyme replacement. One of the most common gastrointestinal disorders in children and adolescents with cystic fibrosis is the distal intestinal obstruction syndrome. The obturator obstruction often occurs after an intercurrent illness in which the child has a decreased oral intake and stops taking the pancreatic enzyme supplement.

16 The clinical presentation includes abdominal pain and decreased stool frequency. Plain radiographs frequently demonstrate large amounts of fecal material with the 'bubbly' granular appearance similar to meconium ileus (a). This may require treatment with Gastrografin enemas or administration of a balanced intestinal lavage solution orally or through a nasogastric tube.



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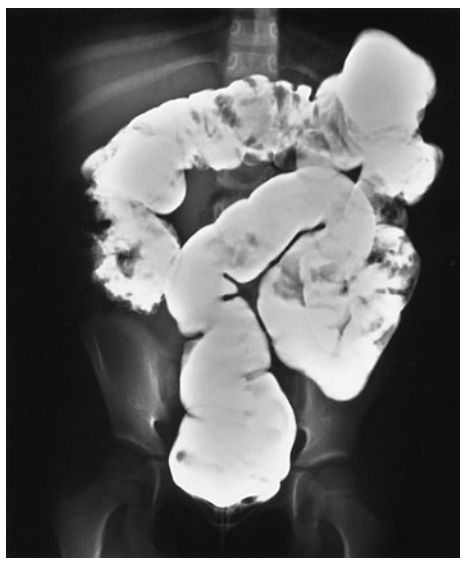

16b

The obstruction usually responds to non-operative management with a balanced salt solution such as GoLytley (20 mL/kg per hour for 3–4 hours). If dehydration is present, intravenous hydration will be required. In some more advanced cases or if the diagnosis is not clear, a water-soluble contrast enema with Gastrografin or other solution may be both diagnostic and therapeutic (b). The use of oral or nasogastric Gastrografin has also been reported with a dose of 50 mL followed by at least 200 mL of water in children less than 8 years of age, and 100 mL followed by at least 400 mL of water in children over 8 years of age.

Complicated meconium ileus

Neonates with complicated meconium ileus are managed in a similar manner to uncomplicated cases, although return of bowel function may be somewhat slower, particularly in cases of perforation. Some infants may require total parenteral nutrition if bowel function is slow to return, or if a proximal enterostomy does not provide an adequate absorptive surface to support the infant with enteral nutrition alone. Infants with both a proximal and distal stoma can often be managed with oral feeds combined with re-feeding of the proximal ostomy effluent into the distal stoma. Enterostomy closure is generally performed 5–6 weeks after the initial procedure.

OUTCOME

The results of the medical and surgical management of 60 neonates with meconium ileus treated between 1972 and 1991 at the James Whitcomb Riley Hospital for Children, Indianapolis, have been reviewed. The study included 20 girls and 40 boys. A family history of cystic fibrosis was present in six neonates.

Twenty-five neonates had uncomplicated meconium ileus due to intraluminal obstruction of the terminal ileum with concretions of abnormal meconium. The treatment of these patients can be divided into two time periods, 1972–1980 and 1981–1991. Ten infants presented during the first time period, and only two of them were successfully cleared with a diatrizoate meglumine (Gastrografin) enema. The eight remaining infants underwent resection, operative irrigation, and enterostomy formation. A Bishop–Koop stoma was constructed in two infants, and six had a double-barrel (side-by-side) enterostomy. Of the 15 neonates treated during the later time period, eight (53 percent) were successfully cleared with a Gastrografin enema. The remaining seven infants required laparotomy. Seven were treated with enterotomy and intraoperative irrigation with saline or dilute contrast agent (Hypaque or Gastrografin) and one with irrigation and double-barrel enterostomy.

Thirty-five neonates presented with 56 complications of meconium ileus, including volvulus (22), atresia (20), perforation (6), and giant cystic meconium peritonitis (8). Clinical presentation in these neonates included abdominal distension, bilious vomiting, and failure to pass meconium; these symptoms were usually noted earlier than in uncomplicated cases. Neonates with perforation and giant cystic meconium peritonitis often had abdominal distension at the time of delivery. Three were diagnosed by prenatal ultrasound.

Operative management of patients with atresia, volvulus, and perforation included resection and anastomosis in 15 and enterostomy in 12. The eight patients with giant cystic meconium peritonitis underwent excision of the pseudocyst and enterostomy.

The diagnosis of cystic fibrosis was confirmed in all cases by sweat chloride test. Pancreatic enzyme therapy was instituted, along with a routine formula feed. Enterostomy closure was usually accomplished between 4 weeks and 3 months of age. All patients have been followed by the Indiana University Cystic Fibrosis Clinic at James Whitcomb Riley Hospital for Children. Survival at 1 year was 92 percent (23/25) in patients with uncomplicated meconium ileus, and 89 percent (31/35) in complicated cases. The mortality in the uncomplicated cases was due to pulmonary problems, and both occurred during the early time period. Deaths in the complicated cases were the result of sepsis (2), renal failure (1), and severe cholestatic jaundice progressing to liver failure (1). Since this report, uncomplicated cases have been managed exclusively with either enterotomy and irrigation or irrigation through the appendix.

FURTHER READING

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45

Vitellointestinal (omphalomesenteric) duct anomalies

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INTRODUCTION

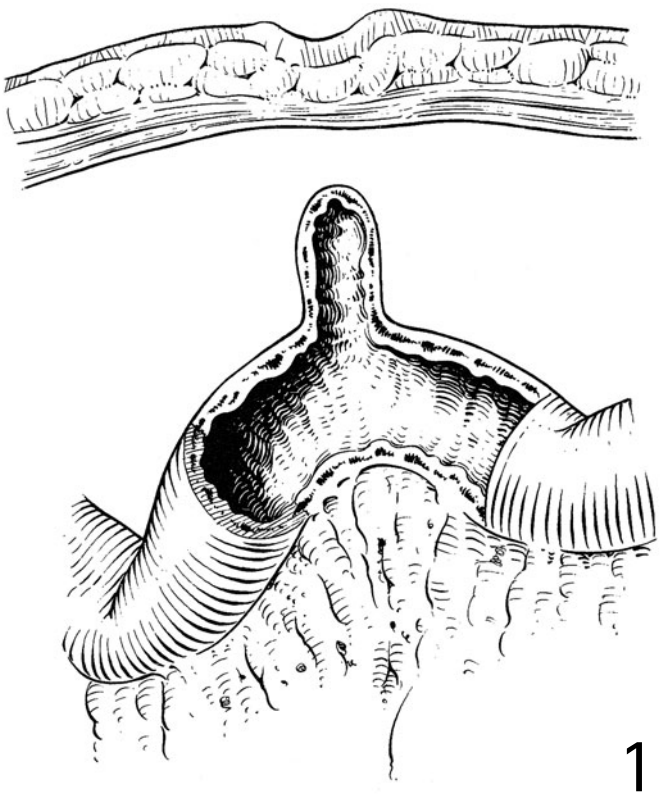
The vitellointestinal (omphalomesenteric) duct is an embryonic communication between the yolk sac and the midgut. This communication normally disappears at about the sixth week of fetal life. Persistence of the duct between the intestinal tract and the umbilicus, or persistence of its embryonic blood supply, results in a variety of lesions that usually present in early infancy, but occasionally appear later in life.

TYPES OF ANOMALIES

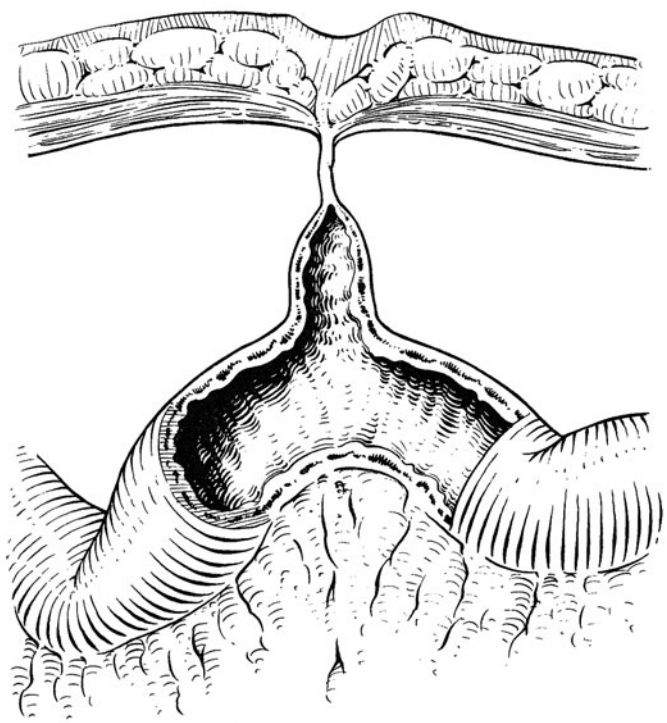
A Meckel's diverticulum represents persistency and patency of the inner intestinal component of the vitellointestinal tract (see Illustration 1). In a small proportion of patients there will be a fibrous band extending from the apex of the Meckel's diverticulum to the undersurface of the umbilicus (see Illustration 2), but more often there is a band representing the remnants of the vitelline vessels joining the Meckel's diverticulum to the mesentery of the small bowel (see Illustration 3). Meckel's diverticulum has diverse clinical presentations (Table 45.1), but frequently remains quiescent throughout life. The chances of an asymptomatic Meckel's diverticulum causing symptoms later in life are such that en passant removal of the structure when it is observed during operation for some other reason is arguably justified in children, but is probably not justified in adults in whom the chances of it becoming symptomatic are much less likely.

Table 45.1 *Presentation of Meckel's diverticulum*

Melena and anemia	Ectopic gastric mucosa in the Meckel's diverticulum releases hydrochloric acid, which causes ulceration of adjacent ileum, producing major gastrointestinal bleeding
Abdominal pain (Meckel's diverticulitis)	Inflammation of a Meckel's diverticulum, particularly if long and with a narrow lumen, causes clinical features similar to those of acute suppurative appendicitis
	Perforation of an adjacent ileal ulcer may also cause an inflammatory mass, pneumoperitoneum and peritonitis
Intussusception	A Meckel's diverticulum may invert and act as a lead point for an intussusception
	This is responsible for about 2% of intussusceptions
Meckel's band obstruction	A band extending from a Meckel's diverticulum to the root of the small bowel mesentery or to the umbilicus may cause a loop of bowel to become entangled around it, producing a bowel obstruction
Incidental findings	During laparotomy for other conditions, e.g., appendicitis, a Meckel's diverticulum may be found

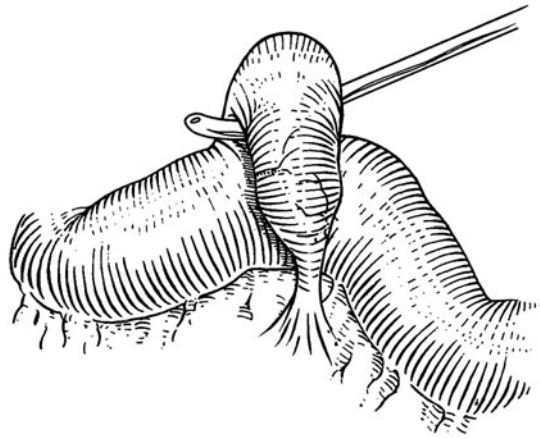


1 The typical appearance of a Meckel's diverticulum is shown in cross-section. Parts of the inner surface may contain ectopic pancreatic or gastric mucosa. Ectopic gastric mucosa produces hydrochloric acid, which can ulcerate adjacent non-gastric mucosa, either in the diverticulum itself or in adjacent ileum, and cause major bleeding. The child may present with 'brick-red' rectal bleeding (often without much abdominal pain), and anemia.



2 The vitellointestinal band (Meckel's band) is the remnant of the duct in which the lumen has been obliterated, but a fibrous cord or band persists. This runs from the deep surface of the umbilicus to the ileum or to a Meckel's diverticulum. There is always a risk that a loop of bowel may become entangled around it, producing intestinal obstruction, often with a closed loop.

3 The Meckel's diverticulum is often bound down by remnants of the vitelline vessels from its apex and is adherent to the mesentery of the ileum. (Division of the vessels at the apex of the peritoneal fold reveals its typical antimesenteric origin.) Loops of bowel can be trapped beneath this band, producing a closed loop obstruction.



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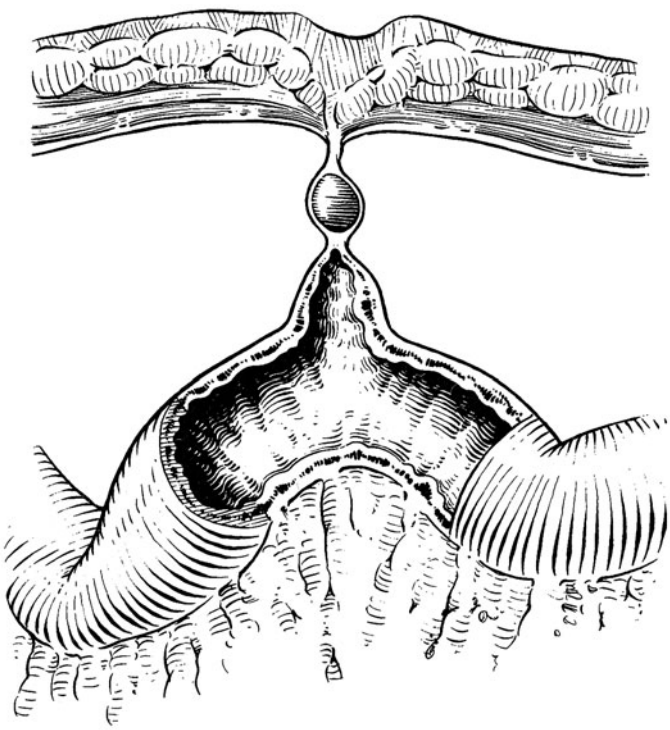


4

4 This form of Meckel's diverticulum has a cord containing a cystic remnant, which may slowly increase in size. The cyst is the result of partial obliteration of the duct. It may become infected and form an abscess; if it has a sinus, pus may discharge from the umbilicus.

BLEEDING MECKEL'S DIVERTICULUM

PRINCIPLES AND JUSTIFICATION

This is the most common clinical presentation of a Meckel's diverticulum, and results from the presence of ectopic gastric mucosa in the lining of the diverticulum. Hydrochloric acid produced by the gastric mucosa causes ulceration of the adjacent small bowel mucosa and, less commonly, of the diverticulum itself. This may result in rapid hemorrhage, which usually presents as relatively painless but profuse 'brick-red' rectal bleeding. The resultant anemia may necessitate blood transfusion, but the bleeding usually stops spontaneously without the need for emergency surgery. The definitive investigation is surgery, but a technetium scan may confirm the presence of ectopic gastric mucosa.

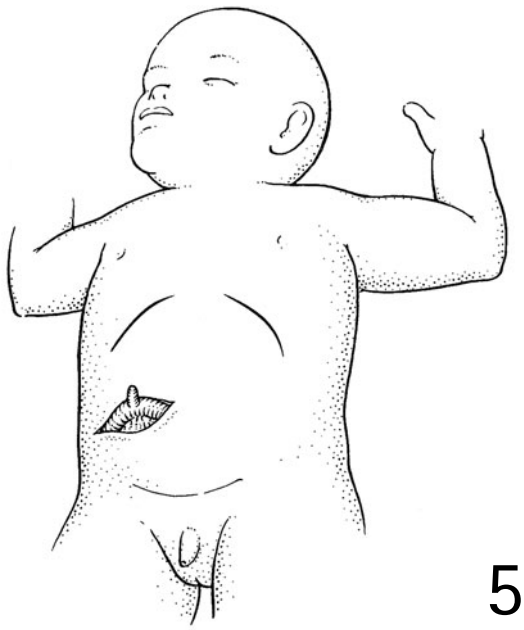
INDICATIONS

Surgery (open or laparoscopic) is indicated where the clinical presentation of major and painless intestinal hemorrhage is consistent with a bleeding Meckel's diverticulum, irrespective of the result of the technetium scan. One advantage of a laparoscopic approach is that the diagnosis can be confirmed using a minimally invasive technique.

PREOPERATIVE

Anesthesia

The procedure is performed under general anesthesia and muscle paralysis, using the same technique as for any acute abdominal procedure. Blood transfusion is required very occasionally in the perioperative period for major blood loss from the ileum, although the operative procedure itself is relatively bloodless. Electrolytes and fluid balance must be monitored. Perioperative antibiotics are administered.



5

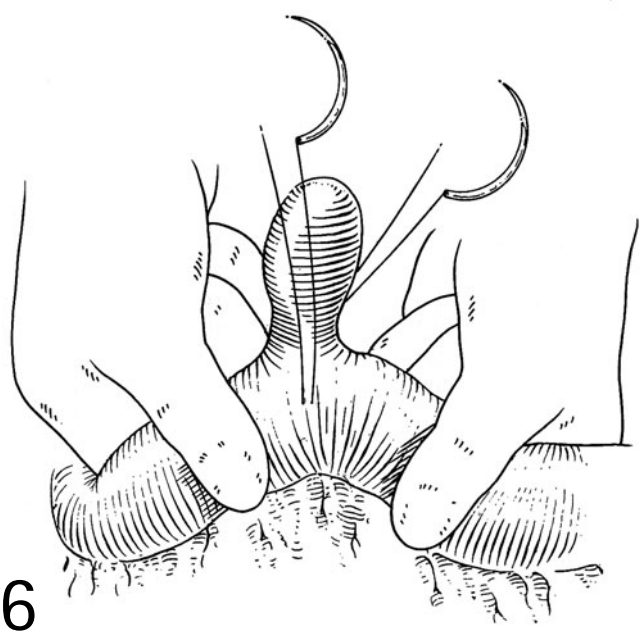
OPERATION

Incision for an open procedure

5 The surgical approach is through a small, right transverse, infraumbilical, muscle-splitting or muscle-cutting incision. This can be extended medially by retracting the rectus toward the midline or even dividing the muscle if necessary. A Meckel's diverticulum can be excised easily through the standard incision used for an appendicectomy. Alternatively, laparoscopic-assisted Meckel's diverticulectomy through an umbilical incision can be employed (see below).

Identification of lesion

At laparotomy for bleeding, the Meckel's diverticulum will not usually be inflamed. A blue discoloration will be seen in the ileum and colon distal to the diverticulum if recent bleeding has occurred. The diverticulum causing bleeding is delivered through the wound.



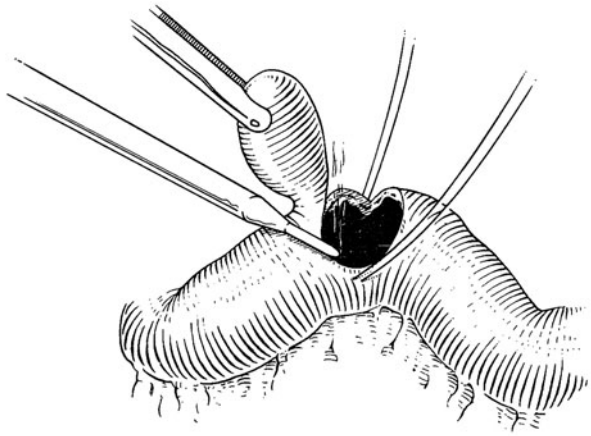
6

Control of ileum and its contents

6 Compression of the ileum with fingers or a non-crushing bowel clamp (fingers are preferred because they are less traumatic to the bowel) reduces the amount of bleeding and soiling that occurs when the ileum is opened and the diverticulum is excised. Packs are placed on either side of the loop of ileum containing the Meckel's diverticulum. Suction is kept nearby to reduce accidental spillage of liquid ileal contents when the ileum is opened. Stay sutures (3/0) are placed on the ileum on either side of the diverticulum.

Excision of diverticulum

7 A longitudinal or oblique elliptical incision is made in the ileum near the base of the diverticulum using scissors or diathermy. It is *essential* that the entire diverticulum is removed, because a remnant of acid-secreting mucosa left at the base of the diverticulum could continue to cause ulceration and bleeding of the adjacent ileum.



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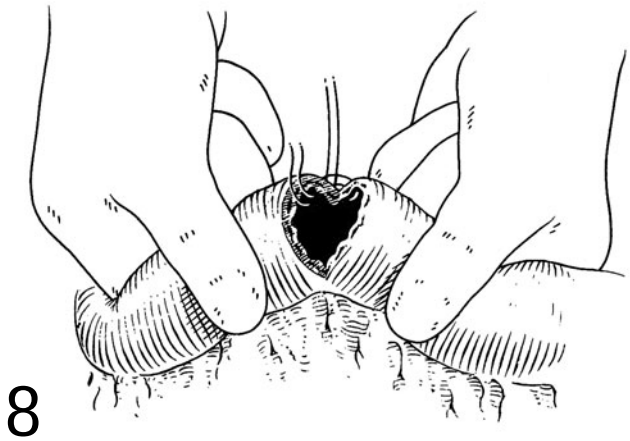


8

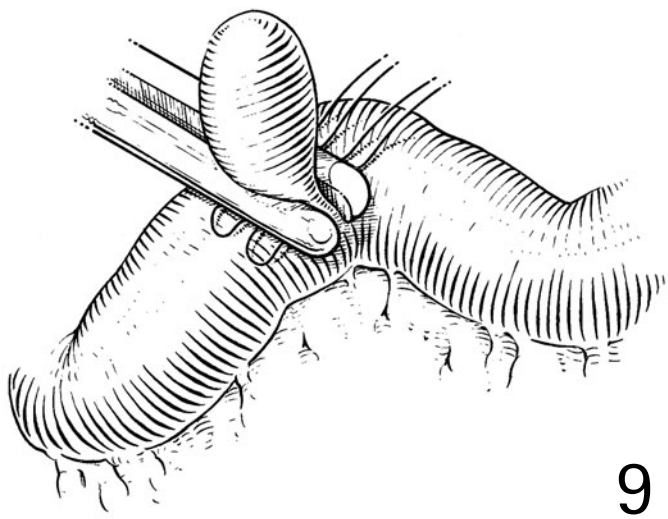
8 The stay sutures are then held apart to transform the longitudinal or oblique elliptical incision into a transverse one. This allows closure of the wound without narrowing the lumen of the ileum.

Closure of ileum

A 4/0 absorbable, continuous or interrupted, all-layers suture is used to close the bowel. A second seromuscular continuous layer is employed by some surgeons.

Alternative method for narrow-necked diverticulum

9 A curved artery forceps or crushing clamp is placed across the base of the diverticulum at 45° or more to the long axis of the ileum. This avoids the narrowing that might be caused by an incision closed longitudinally. Mattress sutures of 3/0–4/0 absorbable material are inserted under the clamp and tied. The diverticulum is cut away at the distal border of the clamp.



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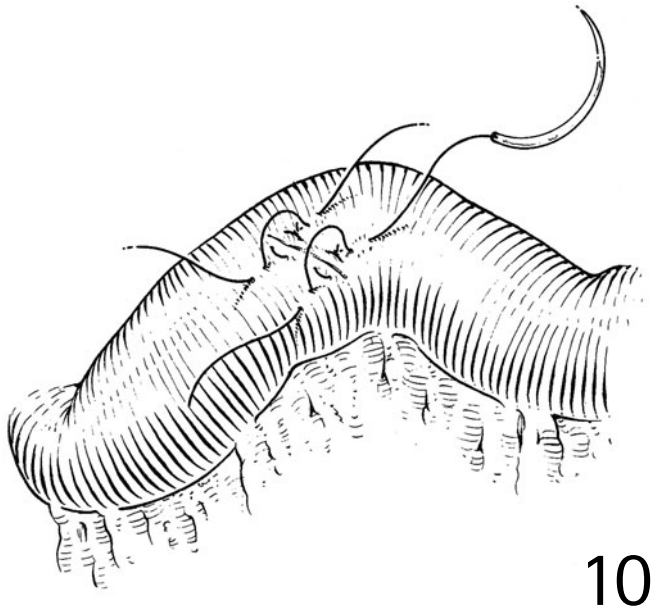
10 The clamp is removed and the line of section is buried by a second layer of sutures.



10

Wound closure

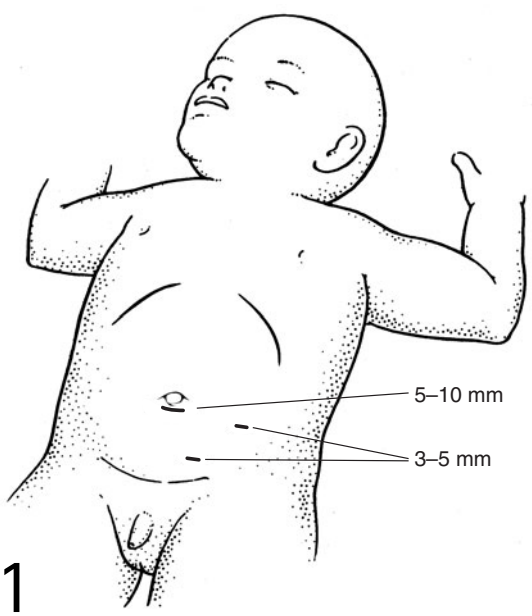
The peritoneal cavity is irrigated with antibiotic saline before closure. The wound is closed in layers with 2/0 or 3/0 absorbable sutures in the standard fashion.

POSTOPERATIVE CARE

Oral fluids can be commenced on about the second day after operation when the abdomen is becoming soft to palpation and there is no nausea, i.e., no clinical evidence of ileus. Most children can be discharged on the second or third day after operation.

Laparoscopic approach

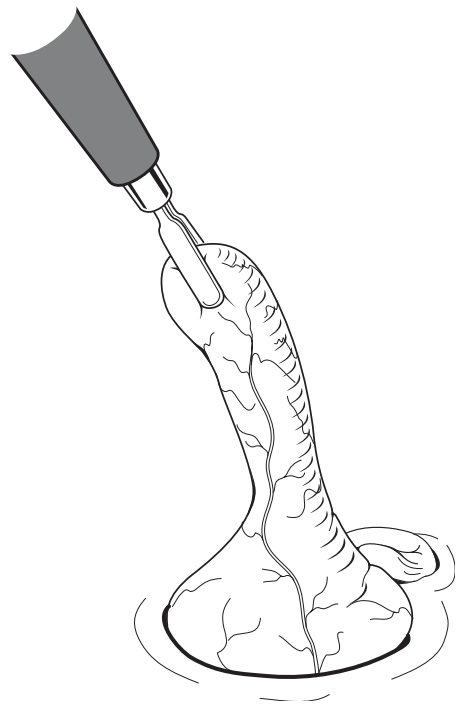
This is now the preferred approach of many surgeons, and offers the advantage of being able to confirm the presence of a Meckel's diverticulum using a minimally invasive technique where there has been diagnostic uncertainty preoperatively.



11

11 Following local anesthetic infiltration with 0.25 percent plain bupivacaine, an infraumbilical incision is made. The linea alba is incised and the peritoneum opened under direct vision. A 5 mm or 10 mm 30° telescope is introduced through a 5 mm or 10 mm umbilical port and secured to the rectus fascia (see Chapter 34). An initial inspection of the peritoneal cavity is performed. One or two 5-mm working ports can be introduced on the left side of the abdomen, the size and location being determined by the age of the child.

12 Once located, the Meckel's diverticulum can be delivered through the umbilical port opening (sometimes requiring enlargement) and the diverticulum is removed outside the abdomen using the same technique as described above.



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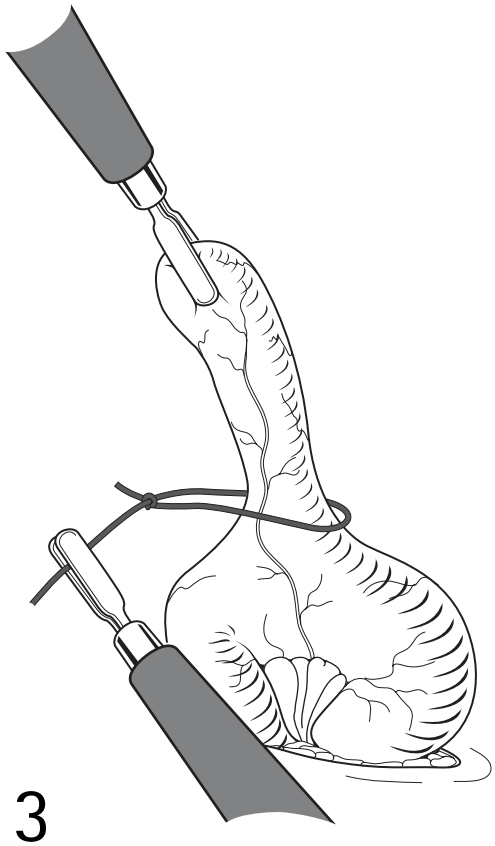


13

13 Alternatively, particularly where the diverticulum is quite narrow at its base, it can be dissected from the ileum laparoscopically: the vitelline vessels are either divided by electrocautery or included in two endoloops placed around the base of the Meckel's diverticulum. Care is taken both to ensure the whole diverticulum is removed (to avoid leaving ectopic gastric mucosa behind) and to ensure that the caliber of the lumen of the ileum is not compromised. The Meckel's diverticulum is divided between the endoloops and removed through the umbilical port. An alternative and equally effective method involves resection using an endo-GIA stapler.

MECKEL'S DIVERTICULITIS

PRINCIPLES AND JUSTIFICATION

Meckel's diverticulitis is an unusual presentation in children. When it does occur, the child is assessed clinically as having acute appendicitis, but at laparotomy the appendix is found to be normal and there is an inflammatory process involving a Meckel's diverticulum. In all operations for suspected appendicitis in which the appendix is found to be normal at laparotomy or on laparoscopy, the distal 100 cm of ileum should be inspected to exclude inflammation of the Meckel's diverticulum as being responsible for the symptoms. The inflamed Meckel's diverticulum is often palpable in the abdomen on opening the peritoneum. An inflammatory mass around a Meckel's diverticulum may result from perforation of an ulcer in the ileum adjacent to the base of the diverticulum. Perforation may also cause pneumoperitoneum and peritonitis.

INDICATIONS

The indication for surgery for Meckel's diverticulum is when the appendix is found to be normal at the time of surgery for suspected appendicitis and an adjacent inflammatory mass (inflamed diverticulum) is identified.

Extensive inflammatory mass

14 If there is severe or long-standing inflammation of the diverticulum causing edema and involving the surrounding ileum, or if the base of the Meckel's diverticulum is very broad, it is appropriate to excise it with a small sleeve of ileum and perform an end-to-end small bowel anastomosis along conventional lines. Otherwise, the inflamed Meckel's diverticulum is excised in the same way as for a bleeding Meckel's diverticulum.

PREOPERATIVE

Anesthesia

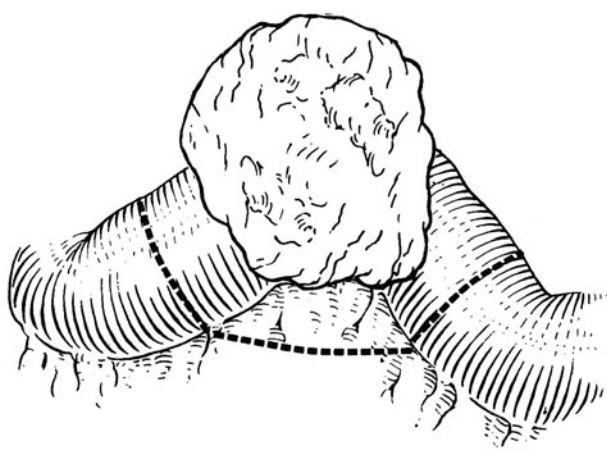
The procedure is performed under general anesthesia and muscle paralysis using the same technique as for acute appendicitis. Perioperative antibiotics are administered.

OPERATION

Indication and exposure

When the diagnosis is known or suspected before the operation, a small, transverse, right infraumbilical incision can be used. If the diagnosis is made during operation at the time of exploration for suspected appendicitis, the inflamed Meckel's diverticulum and adjacent ileum can be delivered easily through the appendicectomy wound and the diverticulum excised outside the abdomen. If a laparoscopic approach to appendicectomy has been performed, the Meckel's diverticulum can be removed:

- laparoscopically,
- laparoscopically assisted, by delivering the diverticulum through the umbilical port, or
- by converting to an open approach, depending on the expertise and experience of the surgeon.



14

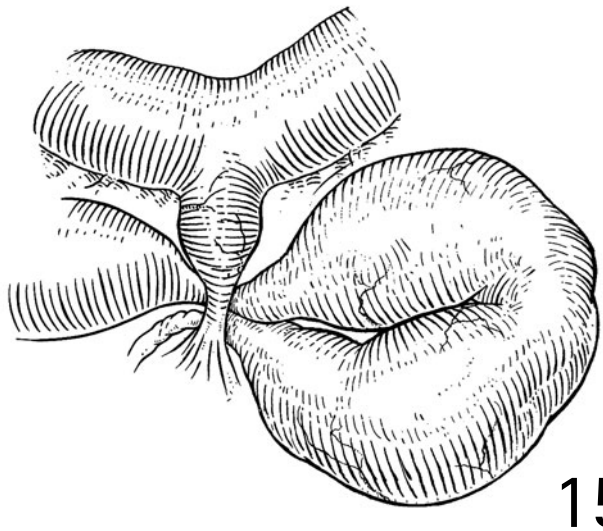
MECKEL'S BAND OBSTRUCTION

INDICATIONS

These children present with a distal small bowel obstruction, the exact cause of which usually cannot be determined clinically. Laparotomy is performed for obstruction. Closed loop obstruction is common. A laparoscopic approach can be used by the experienced laparoscopic surgeon, but the distended loops of small bowel may compromise visualization of the cause of the obstruction.

OPERATION

15 A right supraumbilical or subumbilical incision is standard where the pathology is not certain before operation. The band running from the Meckel's diverticulum is identified and divided, releasing the entrapped loops of bowel. Most commonly, the band runs to the root of the small bowel mesentery (see Fig. 45.2), but may be attached to the undersurface of the umbilicus (see Fig. 45.3).



15

INTUSSUSCEPTION OF MECKEL'S DIVERTICULUM

Meckel's diverticulum is the most common cause of intussusception in which a pathologic lesion at the lead point is identified. It may occur at any age during childhood, but is most common in the first 2 years of life.

INDICATIONS

Peritonitis or other clinical evidence of the presence of ischemic bowel is an absolute indication for surgery in patients with intussusception. Otherwise, the child is treated initially by gas (or barium) enema. Where the intussusception is due to a Meckel's diverticulum, however, enema reduction is unlikely to be successful, and surgery is indicated because the enema has failed to reduce the intussusception.

OPERATION

Incision

A right, transverse, supraumbilical incision is deepened as a muscle-cutting incision through the rectus abdominis muscle.

16

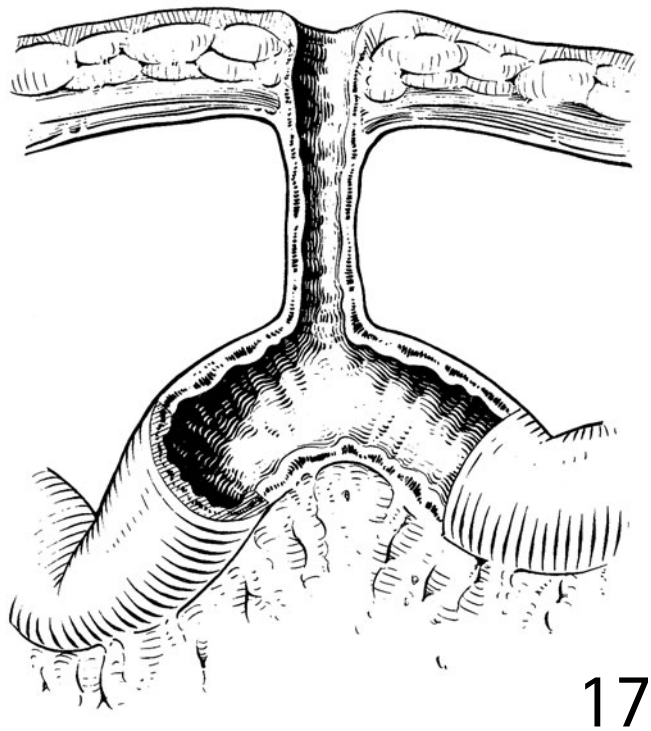
Technique

16 The intussusception is located and an attempt is made to reduce it manually by gentle compression of the colon in a proximal direction at the level of the lead point. Much of the intussusception can be reduced, but when there is a Meckel's diverticulum at the lead point the final portion often cannot be reduced and must be resected in continuity with the diverticulum. On other occasions, the diverticulum will become evident once the intussusception has been fully reduced.

COMPLETE PERSISTENCE OF THE VITELLOINTESTINAL DUCT (FISTULA)

PRINCIPLES AND JUSTIFICATION

17 When the entire vitellointestinal (omphalomesenteric) duct persists and remains patent, there is an open communication between the ileum and the umbilicus. This allows intermittent discharge of the contents of the ileum (gas and ileal fluid) and causes periumbilical excoriation. The tract may be of sufficient caliber to allow prolapse of the ileum as a 'pair of horns'.



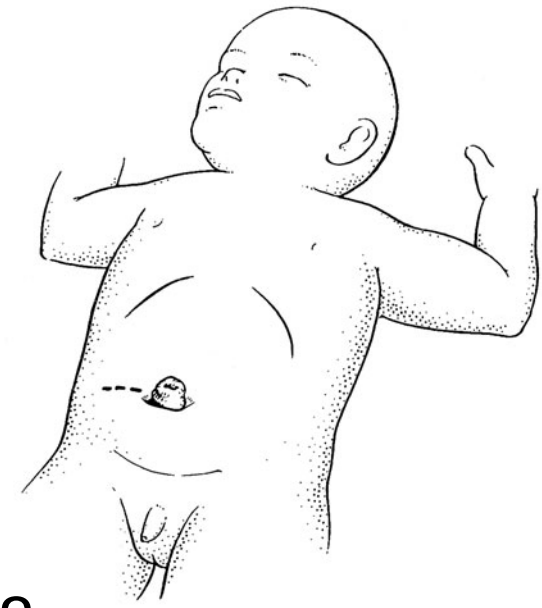
17

DIAGNOSIS

Escape of air and feces through an opening in the umbilicus is pathognomonic of a patent vitellointestinal tract. When there is doubt about the nature of the discharge from the umbilicus (often because it is intermittent), a sinogram will usually demonstrate direct communication with the ileum.

INDICATIONS

Surgery is indicated in all cases where a patent vitellointestinal tract has been demonstrated.



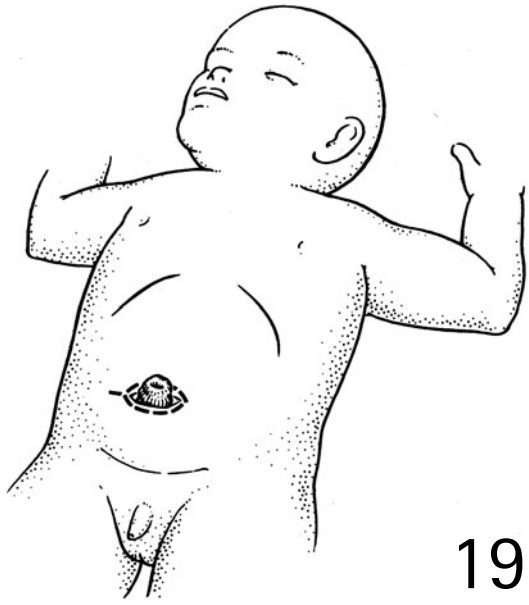
18

OPERATION

Incision

18 A right transverse incision immediately lateral to the umbilicus through the rectus muscle allows adequate exposure of the vitellointestinal tract from within, avoiding the need to circumcise the umbilicus. After dissection of the tract, the defect in the umbilicus can be repaired from its deep surface.

19 Alternatively, an incision in the umbilicus at the mucocutaneous junction, circumcising the external opening of the patent vitellointestinal tract, can be made. The tract is mobilized by separating it from the tough fascia of the linea alba surrounding it, and the peritoneum is opened. To improve exposure, the incision may need to be extended lateral to the umbilicus by dividing the medial 1–2 cm of rectus abdomin abdominis muscle, including the anterior rectus sheath, medial fibers of the rectus abdominis muscle, and the posterior rectus sheath.



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20

Dissection of tract

20 Dissection of the vitellointestinal tract is continued into the peritoneal cavity, where its communication with the ileum is readily seen.

Division at junction with ileum

Two stay sutures are placed on the ileum and the vitellointestinal tract is divided at this point. The communication with the ileum may be narrow, resembling a fibrous cord, or broad, as in many patients with a Meckel's diverticulum. The ileum is closed in one or two layers with 3/0 or 4/0 absorbable sutures.

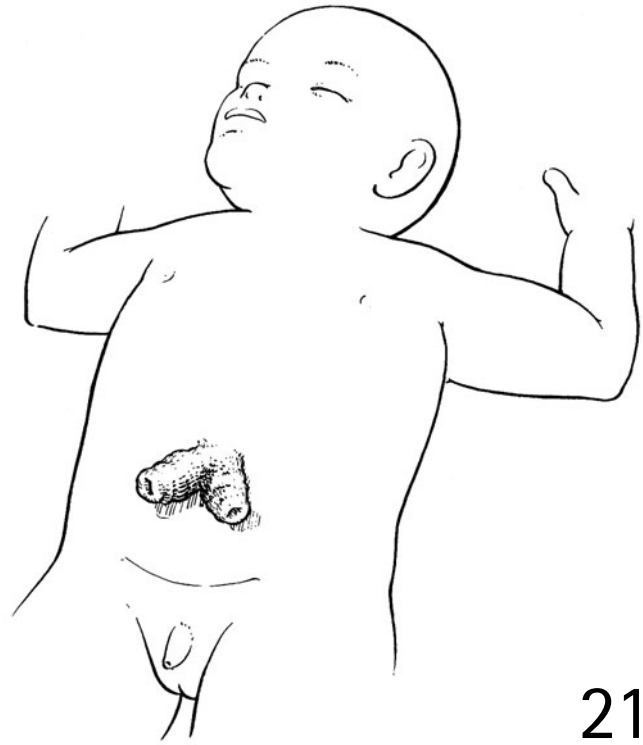
Reconstruction of umbilicus

The defect in the umbilical ring is repaired in the same way as for an umbilical hernia. When the incision has been extended to the right, the posterior and anterior rectus sheath should be closed with 3/0 absorbable sutures.

PROLAPSE OF PATENT VITELLOINTESTINAL TRACT

PRINCIPLES AND JUSTIFICATION

21 When the channel is a short and broad vitellointestinal tract, the ileum may intussuscept through it onto the surface of the umbilicus, producing a double-horned 'Y-shaped' segment of bowel, inside out, with a lumen evident on each horn.



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OPERATION

As long as the bowel is not necrotic (which is rare), it should be reduced manually, after which the vitellointestinal tract can be excised as a semi-elective procedure. In extremely rare situations there is a single-horned prolapse of the vitellointestinal tract: this is seen in the neonatal period and suggests that there is an atresia of one horn, which will necessitate excision of the vitellointestinal tract and end-to-end ileoileostomy to reconstitute gastrointestinal continuity. This is usually an isolated lesion, not associated with other abnormalities.

ECTOPIC MUCOSA

PRINCIPLES AND JUSTIFICATION

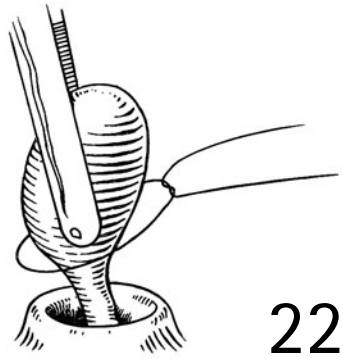
Discharge of mucus from the umbilicus may be caused by a small sequestrated nodule of ectopic alimentary mucosa (representing persistence of the external part of the vitellointestinal tract). It appears as a shiny, spherical, deep-red nodule in the depths of the umbilical cicatrix, but may be pedunculated. Crusts may form on the cicatrix and on the surrounding skin. It should be distinguished clinically from the more common umbilical granuloma, which presents as a small mass of heaped pink or greyish granulation tissue producing a chronic seropurulent discharge; the granuloma can be treated with topical application of silver nitrate.

CONTRAINDICATIONS

If a sinus opening is evident, it is likely that all or part of the vitellointestinal tract is present and patent. Likewise discharge of air or fecal fluid suggests complete patency of the vitellointestinal tract (vitellointestinal fistula). In this situation, surgical excision of the whole tract will be required.

OPERATION

22 Pedunculated ectopic mucosa can be ligated within the umbilical ring; alternatively; a wider based lesion may be excised by diathermy dissection across its base. Suture is required only occasionally.



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46

Duplications of the alimentary tract

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HISTORY

Duplications of the alimentary tract have also been termed giant diverticula, enterogenous cysts, ileum duplex, and inclusion cysts. Calder described the first case in 1733. Ladd introduced the term 'duplications of the alimentary tract' in 1937. He also listed a set of three criteria for diagnosis: (1) the presence of a well-developed coat of smooth muscle; (2) epithelial lining that represents some part of the alimentary tract; and (3) intimate attachment to some part of the alimentary tract.

PRINCIPLES AND JUSTIFICATIONS

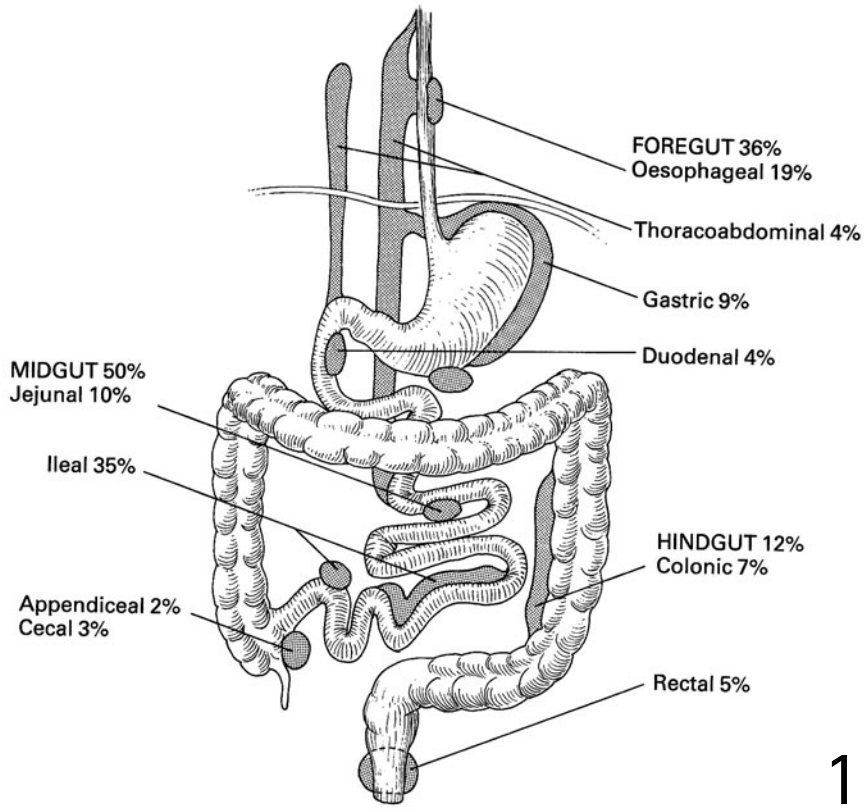
Alimentary tract duplications are congenital malformations that may be found anywhere from mouth to anus. Duplications are found in about 1 in 4500 autopsies. The most common duplication is cystic and located on the mesenteric aspect of the small or large intestine. Multiple duplications are seen in about 10 percent of patients. As many as 30 percent of patients with thoracic or thoracoabdominal duplications have additional duplications below the diaphragm.

Duplications are lined by alimentary tract mucosa and usually share a common smooth muscle wall and blood supply with the adjacent gut. Ectopic acid-producing gastric mucosa may be present in approximately 20 percent of duplications, and can cause peptic ulceration, bleeding, and perforation. Most duplications cause symptoms in infancy or early childhood and most are diagnosed by the age of 2 years, but some present as an incidental finding in patients with other conditions or may be identified on routine prenatal ultrasound. In adults, duplications can sometimes present with adenocarcinoma of the mucosa.

The pathogenesis for the development of duplications is unknown, but cystic duplications are thought to develop secondary to a split notochord mechanism and tubular duplications secondary to partial twinning. Cystic duplications may be associated with spinal cord and vertebral anomalies, and tubular duplications may be associated with urinary tract, spine, and central nervous system anomalies. Foregut duplications probably occur during the normal division of the foregut into respiratory and esophageal structures, and may therefore contain elements of respiratory tract such as cartilage and bronchial epithelium (bronchogenic cyst), elements of esophagus such as smooth or striated muscle and squamous epithelium (esophageal duplication), or elements of both (foregut duplication).

DISTRIBUTION AND VARIETY

1 The distribution of 495 alimentary tract duplications in 455 patients shows that the most common location is in the midgut, and most of these duplications are located in the ileum. Ectopic alimentary tract mucosa is seen in many duplications. In particular, ectopic acid-producing gastric mucosa can be found in any location and can cause bleeding, perforation, and fistulae.



1

Most duplication cysts are diagnosed prior to the age of 2 years, and about 50 percent are diagnosed before 5 months of age. There is an overall equal sex distribution.

Presentation

Patients may present with a mass or with symptoms related to the location and size of the cyst. Intestinal duplications can serve as a lead point for intussusception and can cause a localized volvulus of the small bowel. Ectopic acid-producing gastric mucosa can cause peptic ulceration of the normal intestinal mucosa, with resultant painless gastrointestinal bleeding or perforation. Duplication cysts may also cause intestinal obstruction due to mass effect, particularly those in the small bowel, which are located on the mesenteric surface between the leaves of the mesentery (Fig. 46.1). Foregut duplication cysts may present with obstructive symptoms involving the esophagus, trachea, or both.

Some duplication cysts are diagnosed on routine prenatal ultrasound. The differential diagnosis in these cases includes ovarian cyst, mesenteric or omental cyst, lymphangioma, choledochal cyst, and, less commonly, cysts of solid organs such as the kidney, spleen, liver, or pancreas. The parents should be counseled by a pediatric surgeon prenatally. Postnatally the infant should be evaluated by a pediatric surgeon and should have a repeat ultrasound. If the child is asymptomatic and the cyst is still present and less than 4 cm in diameter, it is reasonable to manage it expectantly with repeated ultrasounds. Ovarian cysts will often resolve over time, whereas duplication cysts will not. Symptomatic cysts should be resected expeditiously, and

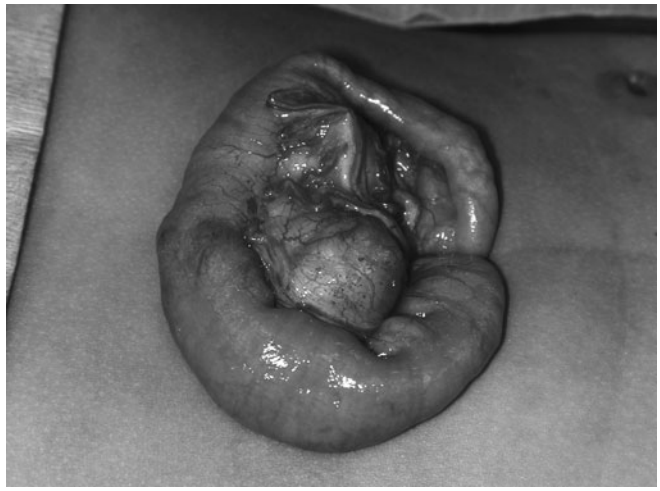


Fig. 46.1 Child with a small bowel cystic duplication in the mesentery undergoing laparoscopic-assisted resection. A small (5 mm) trocar was placed in the left lower quadrant and a large (12 mm) trocar was placed in the umbilicus. The involved small bowel segment was pulled out through the umbilicus and excision with anastomosis was performed.

asymptomatic cysts can either be resected electively or carefully followed.

TREATMENT

Because alimentary tract duplications may be complicated by infection, bleeding, intestinal obstruction or volvulus, and the development of malignancy, many surgeons believe that they should be treated by early complete excision when possible. Sometimes complete excision is not achievable, e.g., in cases of long segmental tubular duplications or complex duodenal or retroperitoneal duplications. Development of adenocarcinoma of the mucosal lining is rare, with only a few cases having been described in the literature. It is therefore acceptable to manage asymptomatic duplications that are not amenable to resection conservatively, as long as the patient is followed on a regular basis. Unresectable symptomatic duplications should be managed by partial resection in order to deal with the particular symptom.

PREOPERATIVE ASSESSMENT AND PREPARATION

Associated anomalies

The associated anomalies that may be present can be predicted based on the location of the duplication.

Foregut duplications may be associated with vertebral anomalies. Midgut duplications may have associated malrotation or atresia of the bowel. In patients with hindgut lesions, genitourinary duplications, anorectal malformations, and bladder extrophy are sometimes seen. Associated anomalies are often picked up clinically or when imaging is being used in the work-up of the patient. When multiple anomalies are found in the same patient, it is important that the management of the patient be planned carefully, since a multidisciplinary approach involving various specialists may be necessary.

Imaging diagnostics

X-RAY

X-ray of the abdomen may reveal a mass effect in the case of large abdominal cysts. Chest X-ray will often show a mass, usually in the middle or posterior mediastinum, and associated vertebral anomalies can often be identified.

ULTRASOUND

Ultrasound will determine whether a mass felt on physical examination is cystic or solid and its relationship to the intestine. Often a characteristic outer hypoechoic rim and an inner echogenic rim can be seen, a sign that is sometimes referred to as a 'bowel signature'.

Computed tomography (CT) or magnetic resonance imaging (MRI) may be helpful in cases where ultrasound is unclear or cannot be done for technical reasons. These modalities are particularly useful for the investigation and delineation of foregut duplications, duodenal duplications, and rectal lesions. Chest CT with contrast is very useful for the visualization of foregut duplications, and provides important information for the preoperative planning of the procedure (Fig. 46.2).

TECHNETIUM SCANNING

Technetium scanning is used to identify ectopic gastric mucosa in the cyst, which is present in about 20 percent of cases. This technique is most useful in the investigation of the child with painless rectal bleeding, for whom the differential diagnosis includes both Meckel's diverticulum and intestinal duplication. It is also useful in the investigation of the child with an asymptomatic prenatally diagnosed duplication, in whom demonstration of ectopic gastric mucosa may push the surgeon to recommend excision rather than observation.

ESOPHAGOGASTRODUODENOSCOPY

Esophagogastroduodenoscopy may be helpful to identify ulcers or strictures, and better to define the anatomy prior to operative repair of duplication cysts in the upper gastrointestinal tract.

CONTRAST RADIOGRAPHY

Upper and lower intestinal contrast radiography may also be helpful for elucidating the anatomy. Barium swallow will demonstrate a typical anterior indentation of the esophagus in cases of foregut duplication cysts.

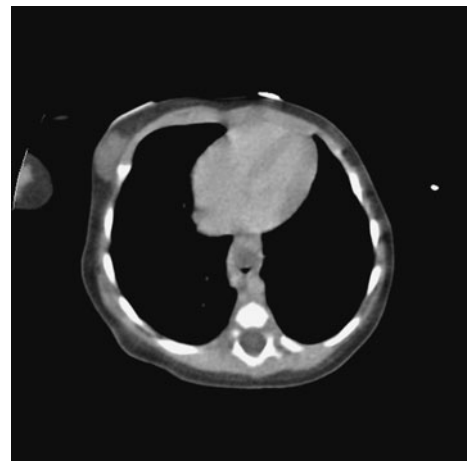
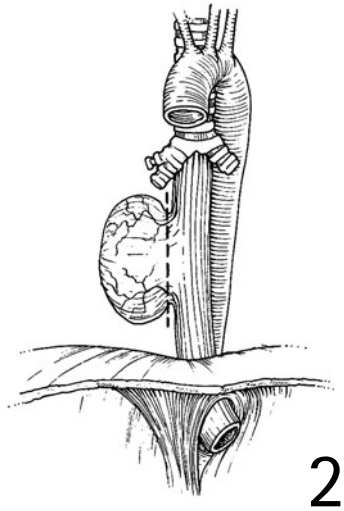


Fig. 46.2 Computed tomography of chest with contrast of an infant showing a foregut duplication in the lower part of the chest, anterior to the esophagus.

OPERATION

Esophageal duplications

2a–c Esophageal duplication cysts and foregut duplication cysts are usually intramural, noncommunicating, cystic lesions. They are most commonly located to the right side of the esophagus (Illustration 2a), although those in the proximal esophagus are often situated between the esophagus and the trachea. A posterolateral thoracotomy provides excellent exposure for most esophageal duplications, although many surgeons are now using a thoracoscopic approach, which provides excellent exposure to the esophagus. Thoracoscopy minimizes postoperative pain, decreases hospital stay, and provides a better cosmetic result. Proximal foregut duplications may be removed more easily through a supraclavicular cervical incision. The phrenic and vagus nerves and the thoracic duct should be carefully identified to avoid injury. Esophageal duplication cysts are excised by dissecting the duplication off the esophageal wall at the base; it is important to remove residual mucosa from the wall of the esophagus (Illustrations 2b and 2c). If the esophagus is inadvertently entered, it can be repaired primarily and covered with a pleural flap. If an esophageal stricture or ulcer was noted on preoperative esophagoscopy or swallow study, it may be necessary to perform a segmental resection of the esophagus.



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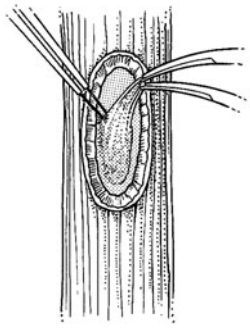

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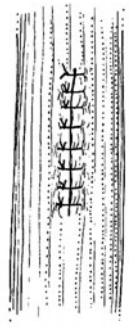
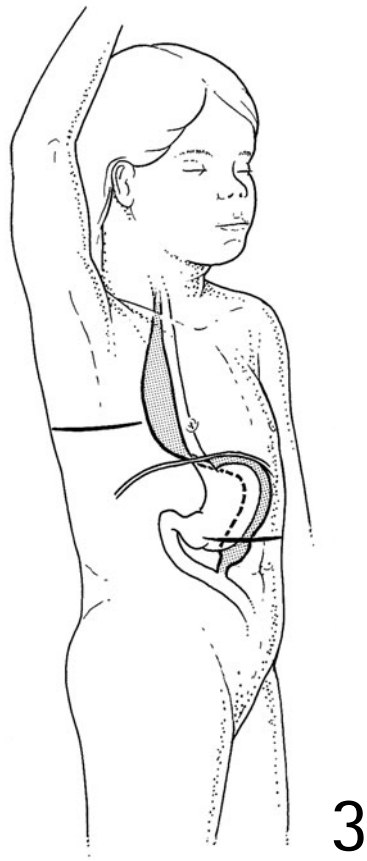


2c

Thoracoabdominal duplications

3 Thoracoabdominal duplications are commonly located to the right of the esophagus in the posterior mediastinum. They communicate with the stomach or small intestine through the diaphragm, and may also communicate with the spinal canal (neurenteric cysts). These lesions are best approached through a combination of thoracotomy and laparotomy, or through a thoracoabdominal incision.

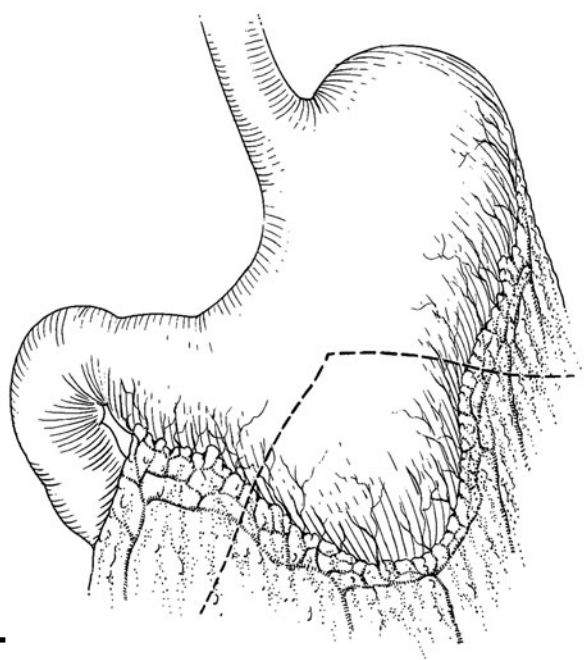


3

It is recommended to start with a posterolateral thoracotomy or thoracoscopy and excise the intrathoracic component first. This involves dissecting the duplication off any attachments in the chest. Sometimes it is attached to the vertebrae and must be carefully dissected off the bone. If an intraspinal component is present and a laminectomy is necessary for complete removal, a neurosurgeon should be involved.

After the duplication is free of any attachments in the chest, it is traced distally to the diaphragm, where it is divided between ligatures. Next, a complete resection is completed through a laparotomy or laparoscopy. The abdominal part of the duplication is often a tubular lesion communicating with the small intestine, although it sometimes ends blindly along the greater curve of the stomach.

Occasionally, thoracoabdominal duplications present with hemoptysis due to erosion into the lung secondary to peptic ulceration. Treatment in these cases may also require a segmental lung resection.

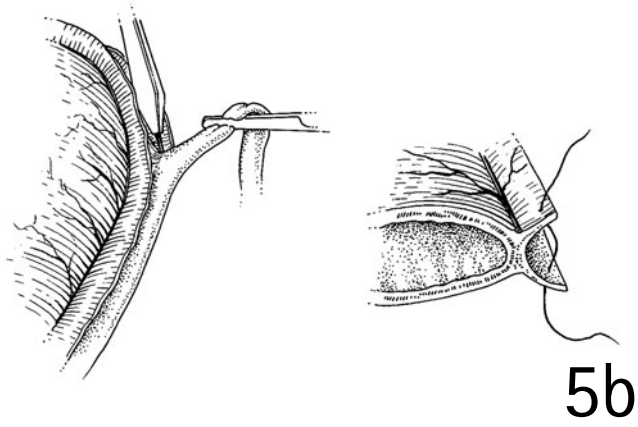


4

Gastric duplications

4 Duplications of the greater curvature can be treated by complete resection. As with esophageal duplications, most gastric duplications are contained within the stomach wall and can be resected without entering the gastric lumen. Laparoscopic resection has also been described.

5a–c If the duplication cyst involves an extensive part of the stomach, partial resection and stripping of the residual mucosal lining is an alternative.



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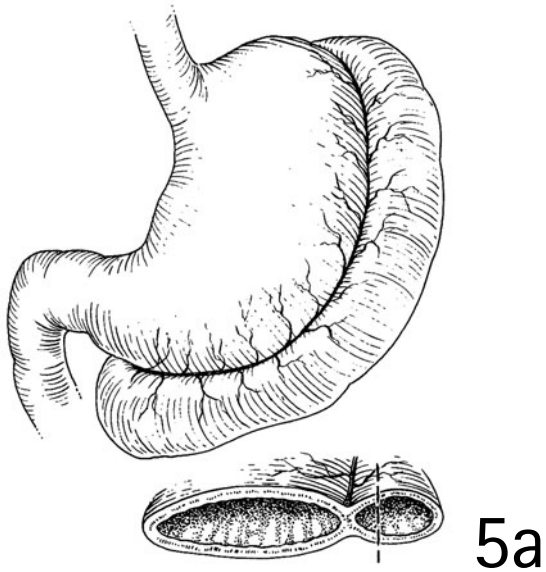

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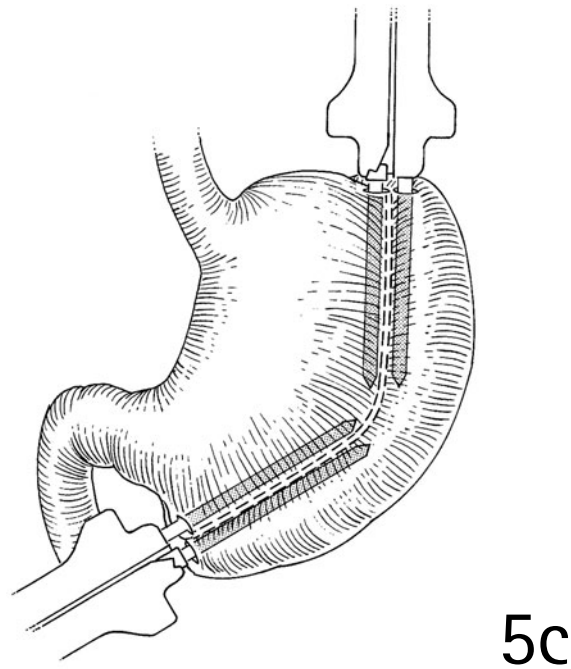


5c

Duodenal duplications

Duplications in the duodenum often present with vomiting and abdominal pain, but due to the close proximity to the biliary tree and pancreas, symptoms and findings related to these organs can be part of the initial presentation. Duodenal duplication cysts of the first, third, and fourth portion can usually be excised using a technique similar to that described for esophageal and gastric duplications.

Duodenal duplications attached to the second portion of the duodenum should undergo preoperative or intraoperative radiographic visualization of the bile ducts and pancreatic ducts (Fig. 46.3). If the biliary or pancreatic ducts are involved, complete excision will be difficult. Partial excision and mucosal stripping are an acceptable alternative in these cases. In some cases, a small bowel Roux-en-Y loop can be brought up and a cyst-enterostomy created, similar to what has been described for the treatment of pancreatic pseudocysts.

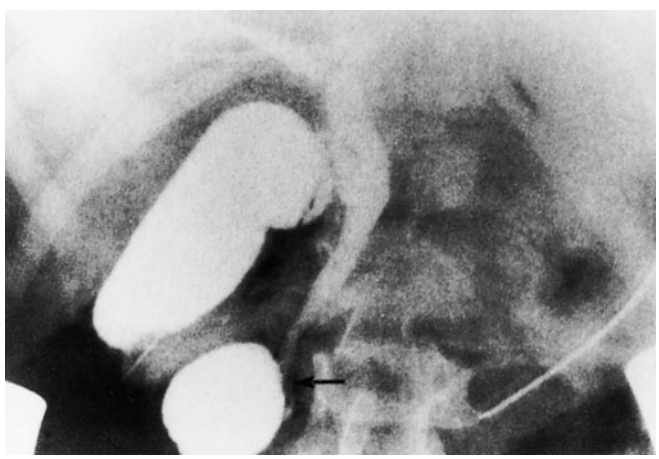
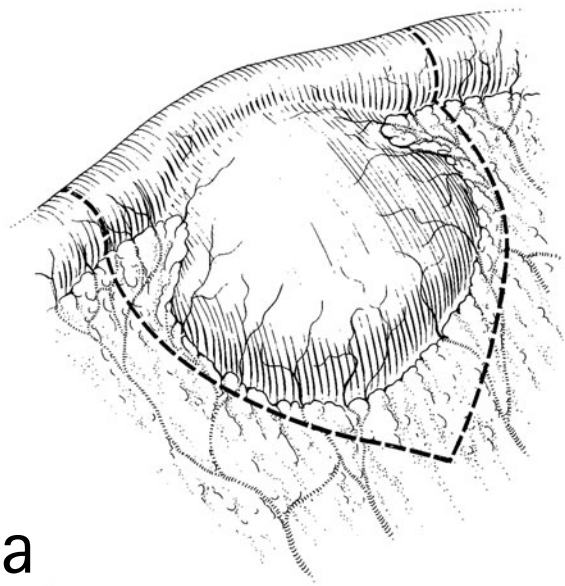


Fig. 46.3 Intraoperative cholangiogram showing a duplication cyst communicating with the distal common bile duct.

Small intestinal duplications

6a–c Cystic lesions of the jejunum or ileum are the most common duplications and are usually excised without difficulty. Because of the common muscular wall and blood supply the cyst shares with the normal bowel, cyst and normal bowel are usually resected as one specimen. This procedure can often be performed using laparoscopic techniques. A more detailed description of the laparoscopic approach is given under the colon section below.



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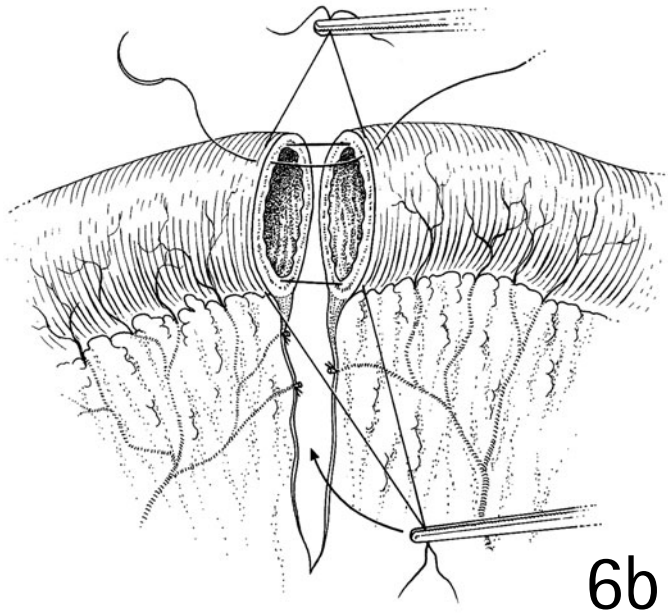

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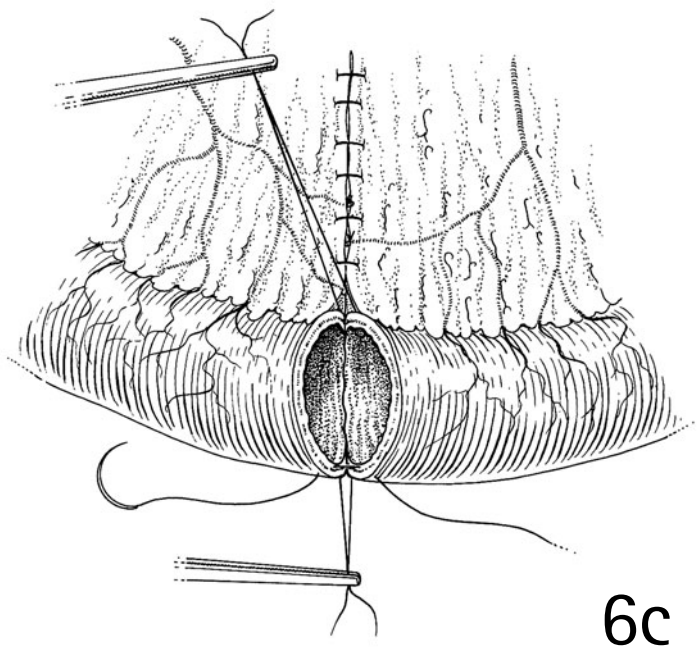

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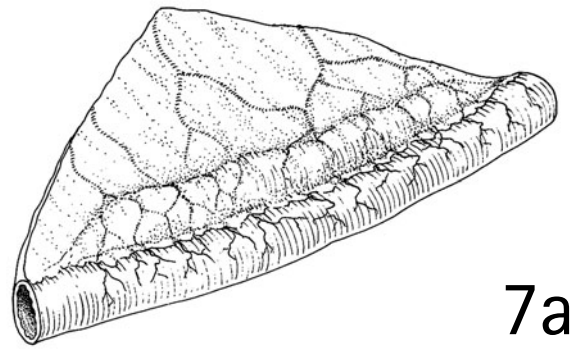

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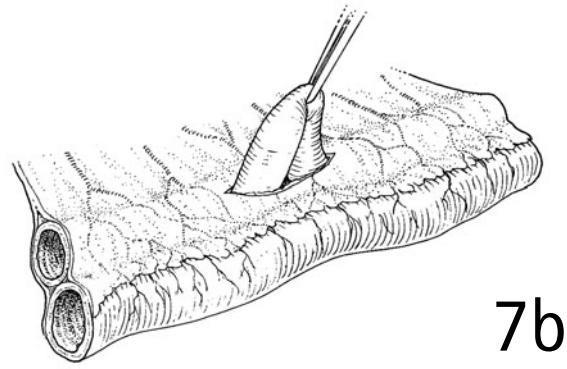

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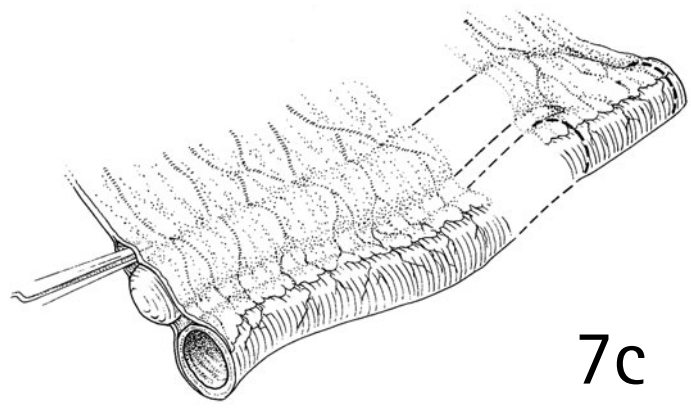


7c

7a–c Duplications that are separate from the intestine and located within the mesentery can be removed by careful separation of the two leaves of mesentery and division of the vessels on one side, and enucleation of the duplicated bowel.

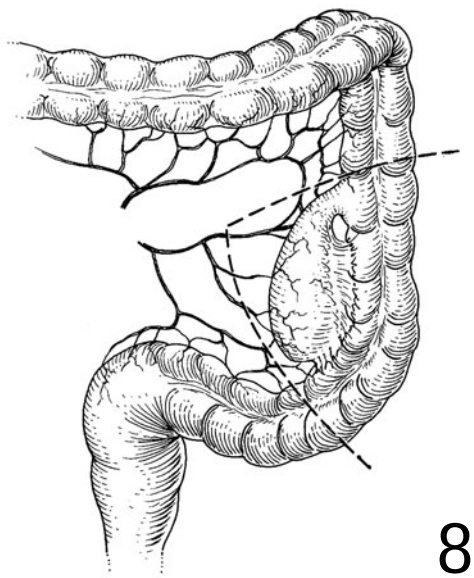
Short tubular duplications can be excised in continuity with adjacent intestine, but care should be taken to obtain complete excision at the proximal and distal ends of the lesion, where the distinction between normal and duplicated bowel is difficult to detect.

Very long tubular duplications, where remaining intestinal length may be insufficient if completely excised, pose a greater surgical challenge. Treatment possibilities include the following:

- Submucosal resection, where the mucosal lining is stripped using multiple longitudinal seromuscular incisions.
- If a proximal communication exists, an alternative to resection is to establish a distal communication by fenestration of the common wall to prevent a blind loop. In many cases this will be the best option, as the shared blood supply makes resection of long segments of tubular duplications very difficult.
- No surgical treatment and close follow-up: this approach may be appropriate if the patient is asymptomatic and no communication exists between the cyst and the bowel.

Colonic and appendiceal duplications

8 Cystic duplications and most short tubular duplications may be excised directly. The colon is then repaired with an end-to-end anastomosis.



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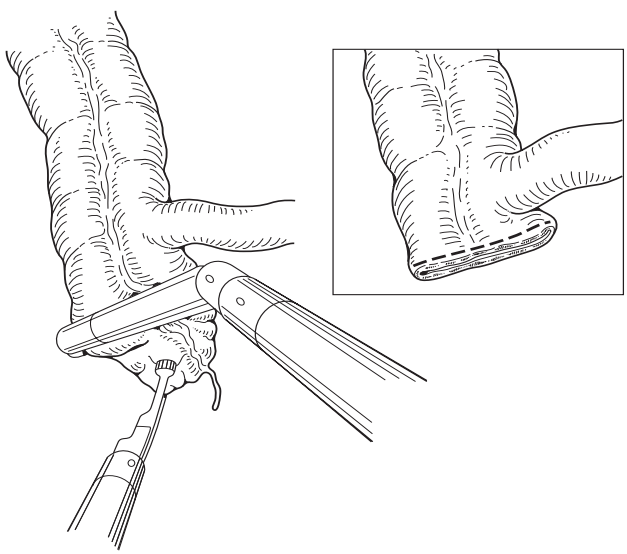
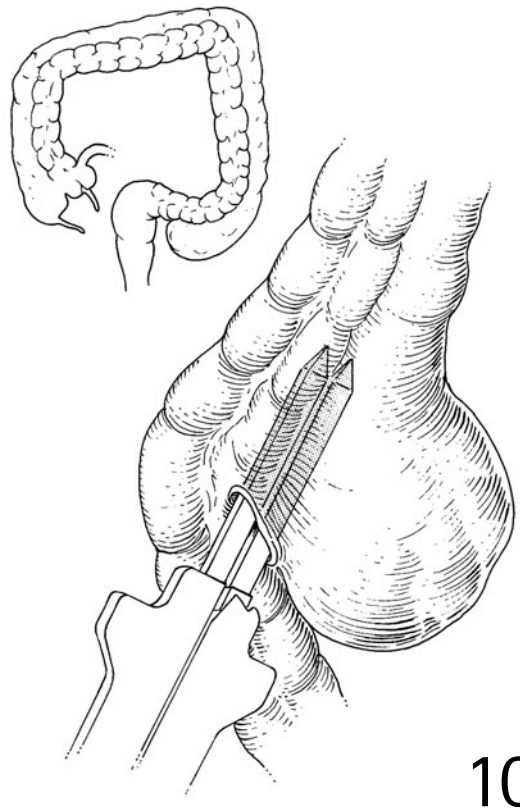


9

9 The operation can also be performed using a laparoscopic or laparoscopic-assisted technique. Initially, an umbilical port is placed for exploration and to assess whether the operation can be completed laparoscopically. This is followed by the placement of two additional working ports. A 12 mm port is necessary for the endoscopic linear stapler, and this port can also be used for removal of the specimen. If it is anticipated that a laparoscopic-assisted approach is necessary, one port site is placed over the location of the duplication so that it can be used as part of the open incision, if one is required.

10 Total colonic duplications are long tubular duplications involving the entire colon. They are located on the antimesenteric border or medial to the normal colon and not in the mesentery. The duplication usually communicates proximally with the normal bowel. If the duplication also communicates distally and involves most of the colon and rectum, no treatment is necessary. If the tubular duplication does not communicate distally, a communication must be established, and if a small communication is present, this opening may need to be enlarged. Fenestration can be established by excising a piece of the common wall.



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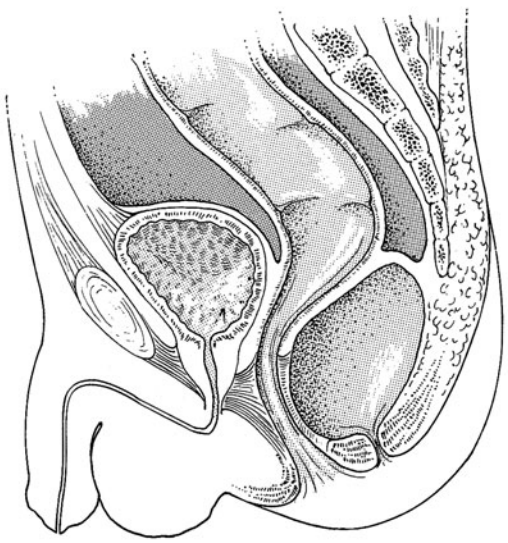

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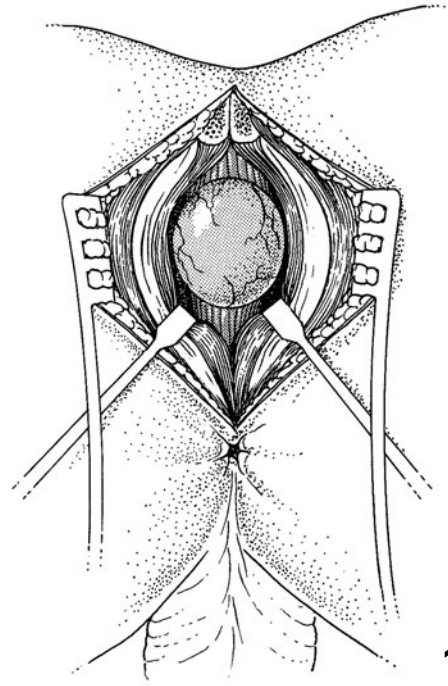
Rectal duplications

11 Rectal duplications are cystic and located in the retrorectal space. There are often fistulas to the anal canal or perineal region. The presentation may include constipation, rectal abscess, rectal bleeding, prolapse of the rectum, urinary tract infection, and hemorrhoids. If the cyst is infected, it should initially be drained and then later excised when the inflammation has resolved. Spinal abnormalities are common and should be looked for.

12 Treatment options include transanal or posterior sagittal exposure and excision, or transanal fenestration of the common wall if the cyst is large. Fenestration is a good alternative in cases where the cyst is infected or a fistula between the cyst and the rectum is already present. All patients should receive full bowel preparation prior to the operation.



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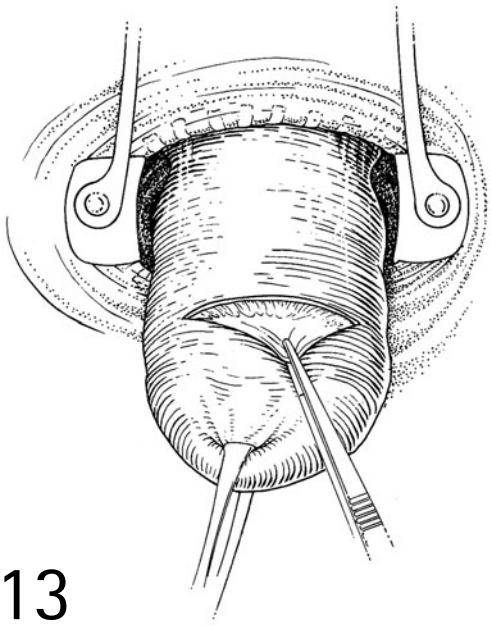

13

13 Small submucosal rectal lesions can be excised through a small incision in the mucosa over the cyst. To make the dissection easier, the cyst should preferably not be aspirated until the end. After excision, the mucosa is closed with absorbable sutures.

In most cases, rectal duplications can be safely excised without a defunctioning colostomy.

Retroperitoneal duplications and spinal involvement

Retroperitoneal duplications can be very large and difficult to remove. It is important to identify adjacent and involved structures to prevent inadvertent injury during the operation. The precise procedure and exposure are dictated by the location and symptoms of the duplication.

Intradural and extradural spinal duplications should always be treated in collaboration with a neurosurgeon.

POSTOPERATIVE CARE

The patient should be monitored and followed up according to the extent of the procedure and the findings during the operation.

Patients who are managed by incomplete excision or fenestration and have cyst tissue left behind should be followed closely, with repeat ultrasound examinations on a regular basis.

OUTCOME

Generally, patients with the most commonly found cystic duplications of the small and large intestine have an excellent outcome, with several series reporting no mortality or significant morbidity after surgical management. The same applies to short tubular gastric duplications and isolated rectal duplications. Patients who present with thoracoabdominal duplications, complete obstruction of the gastrointestinal tract, bleeding, perforation, and involvement of the mesenteric vessels often have a more difficult clinical course. Associated anomalies, when present, may have a significant impact on long-term outcome.

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47

Intussusception

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INTRODUCTION

An intussusception is an invagination of one part of the intestine (the intussusceptum) into another part of the intestine (the intussuscipiens). It most commonly occurs in a proximal to distal direction. The commonest area to be affected is the ileum, intussuscepting into the cecum and ascending colon. The incidence is highest in infants between the ages of 4 and 10 months, but it has also been reported in neonates and adults.

ETIOLOGY

The most common etiological factor in the so-called 'idiopathic group' is a preceding viral illness, either of the upper respiratory tract or a gastroenteritis. The resulting viremia stimulates the gut-associated lymphoid tissue with enlargement and edema of Peyer's patches on the luminal surface of the distal small bowel. At the time of laparotomy, these enlarged Peyer's patches can frequently be found at the lead point of an intussusception along with marked lymphadenopathy in the mesentery. There has been recent debate as to whether the vaccine produced against rotavirus leads to an increase in intussusception. Between 2 and 12 percent of patients will have a pathologic lead point. This most commonly is a Meckel's diverticulum, but polyps, duplication cysts, and solid tumors have also been described. A pathologic lead point should always be suspected in a child who presents outside of the normal range or who has multiple episodes. The condition is seen to occur more commonly in boys unless there is a pathologic lead point, in which case the incidence is equally distributed.

CLINICAL PRESENTATION

Colicky abdominal pain with the infant characteristically 'drawing up the legs' is one of the first clinical signs of intussusception. The child will initially be well between the spasms, but will later become pale and lethargic. As intestinal obstruction develops, the vomiting will become bilious. Bright-red rectal bleeding mixed with mucus, the so-called 'redcurrant jelly stool', will be seen in a quarter of cases and more frequently if formal rectal examination is performed. A palpable mass is felt in the right upper quadrant or epigastrium, along with a distinguishable feeling of emptiness in the right iliac fossa (Dance's sign). Tenderness with evidence of peritonism indicates ischemia or perforation of the intestine. Marked hemodynamic instability with depletion of the intravascular volume can be encountered in children with intussusception. Tachycardia and decreased capillary return should prompt vigorous resuscitation. The diagnosis of intussusception should always be borne in mind when dealing with a child with shock of unknown cause.

At the time of presentation, the findings on the plain abdominal radiograph are often variable and non-specific, and may only be contributory in 50 percent of cases. The X-ray may have a normal appearance if the presentation is early, or may show a relative paucity of gas, possibly in the right iliac fossa (Fig. 47.1). As time progresses, the features of small bowel obstruction with dilated, gas-filled loops and the soft tissue mass of the intussusceptum may be seen. It is important to note that the possibility of intussusception is usually raised clinically but the diagnosis is usually established radiologically, by ultrasound. Therefore if the diagnosis of intussusception is suspected, ultrasound is now the first-line investigation.

ULTRASOUND IN THE DIAGNOSIS OF INTUSSUSCEPTION

Ultrasound is the technique of choice for establishing the presence of an intussusception and should be performed in all cases of suspected intussusception and especially those in which the clinical findings are equivocal. Ultrasound is highly

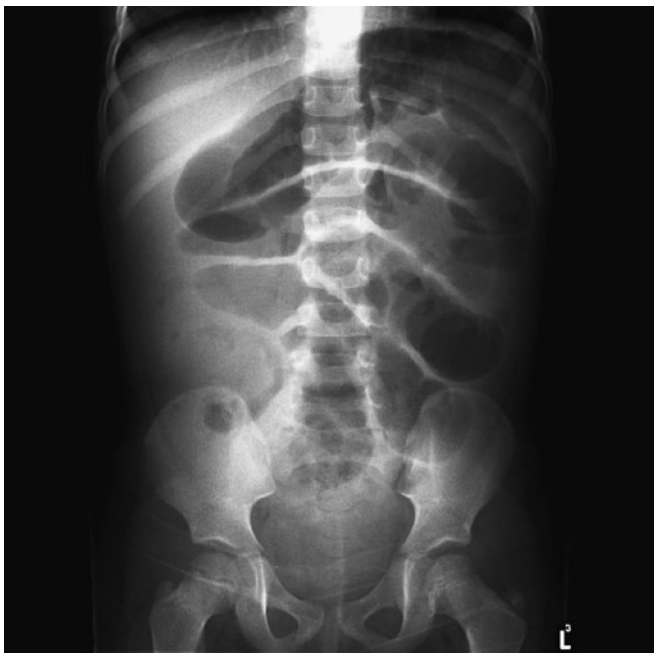


Fig. 47.1 Plain abdominal X-ray of the abdomen of a child with intussusception showing small bowel obstruction with paucity of gas in the right iliac fossa.

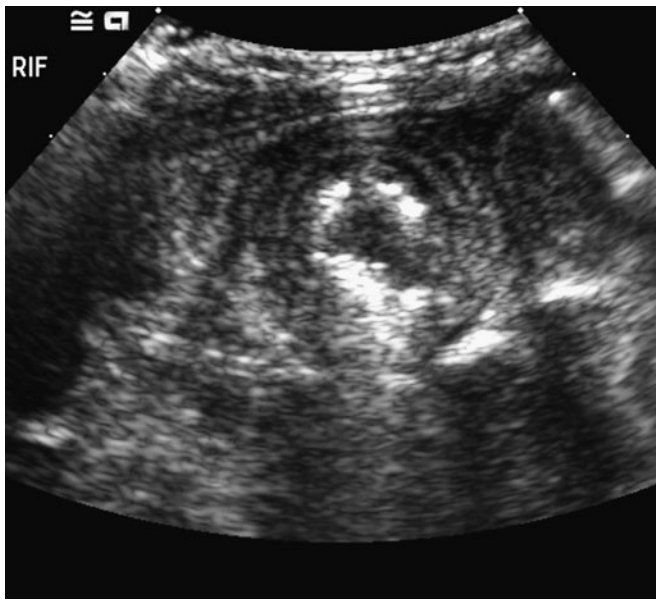


Fig. 47.2 Ultrasound scan (transverse section) showing the typical 'target' sign of intussusception.

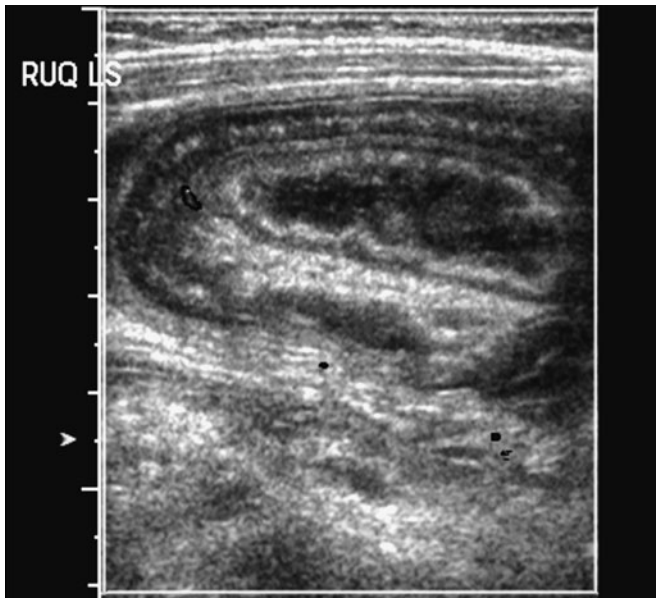


Fig. 47.3 Ultrasound scan (longitudinal section) showing the oval-shaped intussusception ('pseudokidney') with echogenic mucosal walls.

accurate in the diagnosis of intussusception, with a sensitivity of 98–100 percent and a specificity of 88–100 percent. The technique is universally available and does not involve ionizing radiation. False-negative studies are rare, even for less experienced operators, and therefore the absence of intussusception on ultrasound should generally exclude the diagnosis. Ultrasound may also be able to give information on alternative diagnoses.

Ultrasound is performed with the child supine. The characteristic appearance of an intussusception is a series of concentric rings on the transverse view and an oval multilayered mass on the longitudinal view, the layers representing the invaginated layers of the bowel wall and edematous mucosa (Figs 47.2 and 47.3). The most common site for an intussusception is in the right hypochondrium, but the intussusception may extend as far down as the rectum. Ultrasound may not always be able to demonstrate the full extent of the intussusception, as gas-filled loops of small bowel may exclude visualization in the mid-abdomen and the pelvis.

Color Doppler can be used to assess the vascularity of the intussusception. If no color flow is seen, the intussusception may be of longer duration and may be harder to reduce non-surgically, but the absence of color flow does not necessarily imply that the intussusception is avascular or necrotic and, all other factors being satisfactory, radiological reduction should be attempted. Lymph nodes are a common finding on ultrasound and are usually a reflection of the underlying inflammatory process, but may represent the lead point, especially if associated with mucosal thickening.

Ultrasound is also useful for assessing the presence and extent of free intraperitoneal fluid. Small amounts of fluid are commonly seen on ultrasound, but larger amounts of fluid may indicate bowel perforation and if this is a clinical concern, a plain X-ray may be helpful.

NON-OPERATIVE MANAGEMENT

Once the diagnosis of intussusception has been confirmed, and after liaison between the surgeon and the radiologist,

the patient would normally proceed to radiologic reduction.

Radiological reduction of intussusception

Once the diagnosis of intussusception has been confirmed on ultrasound, radiological reduction will be attempted provided the patient is adequately resuscitated and there are no contraindications, such as free intraperitoneal air or signs of peritonism. A history longer than 24 hours or absent color flow on Doppler is not considered a contraindication but may indicate that reduction may be more difficult and should be undertaken with caution.

Radiological reduction should only be attempted in centers also offering pediatric surgery in case of complication or unsuccessful reduction. Published standards of care are available.

Radiological reduction is performed using air under fluoroscopic guidance. It is also possible to use water or air under ultrasound guidance. Fluoroscopy allows visualization of the whole abdomen, and therefore early detection of perforation, but uses ionizing radiation. Ultrasound uses no radiation but can only follow the head of the intussusceptum. Hydrostatic reduction using barium or water-soluble contrast media is generally no longer used, as air reduction has been shown to be safer, has a higher reduction rate, and gives a lower patient radiation dose.

Before any radiological reduction, the patient must have good intravenous access and be fully resuscitated. A nasogastric tube should be passed and prophylactic antibiotics administered. Analgesia can be given at the surgeon's discretion.

When using air reduction, a catheter or feeding tube is placed in the patient's rectum; the anus is occluded either by strapping the buttocks tightly together with tape or by the radiologist gripping the patient's buttocks together between the fingers. Some devices are becoming available to occlude the anus, but the value of these is not yet proven. The catheter is connected to a device or system that can deliver air at consistent pressure, which can be set by the operator. The system must include a safety device to prevent the pressure unexpectedly exceeding that set by the operator. The child may be supine or prone, but it is easier to observe the child if he or she is supine. A control image is obtained and then air is slowly introduced into the large bowel until the first set pressure is reached, typically 80 mmHg equivalent. This slow start allows visualization of the head of the intussusceptum and prevents it being missed if reduction is very rapid (Fig. 47.4).

Multiple attempts will then be made to reduce the intussusception, usually three times for 3 minutes each at three increasing pressures: 80, 100, and 120 mmHg. This will be monitored very closely under fluoroscopic guidance so that if perforation does occur, it is immediately detected and the procedure can be terminated immediately, before a pneumoperitoneum causes splinting of the diaphragm and respiratory arrest, or increased vasovagal stimulation causes a cardiac arrest (Fig. 47.5).

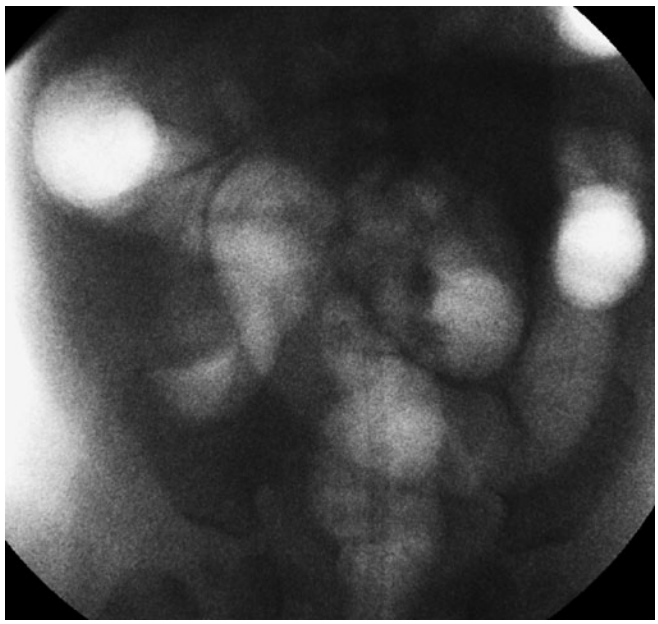


Fig. 47.4 Appearance of colon during pneumatic reduction. The intussuscipiens can be seen as a filling defect on the left side of the image. Air fills the more distal colon.

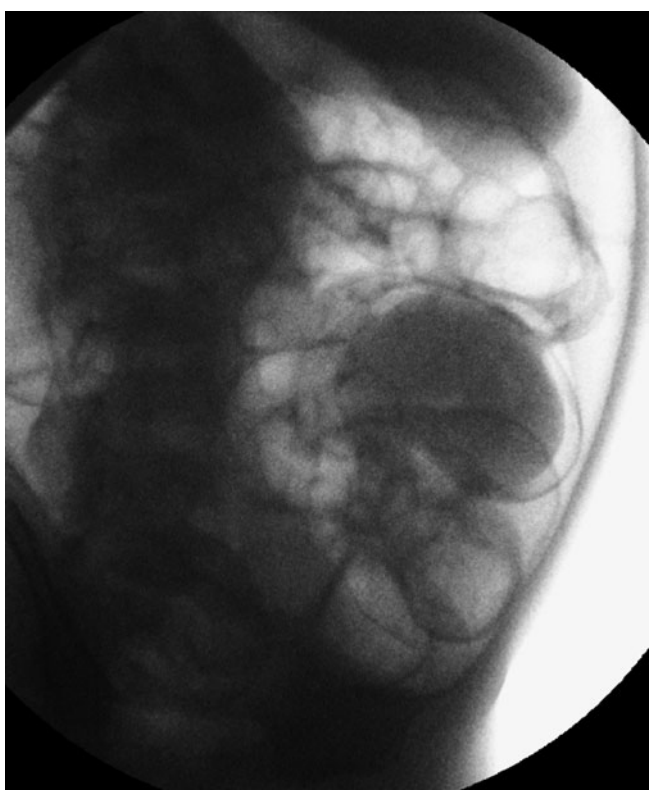


Fig. 47.5 Pneumoperitoneum occurring during attempted reduction. Free air is seen over the right side of the image.

The procedure is also terminated if air is seen to track along the sides of the intussusceptum – the 'air dissection' sign – as this implies that attempted reduction will be unsuccessful (Fig. 47.6).

Reduction is achieved when air flows freely into the small bowel (Fig. 47.7).

The procedure is often distressing for the child, and for the parents if they are present in the room. However, only a few centers in the UK give sedation, as there is some evidence that when the child cries and performs a Valsalva maneuver, the brief rise in intra-abdominal pressure assists with the reduction. The Valsalva maneuver may in addition have a protective effect against perforation because external abdominal pressure decreases the transmural gradient. More importantly, if there is a sudden deterioration in the child's clinical condition, this is more likely to be detected in an unsedated child.

If the intussusception is reduced as far as the ileocecal valve but not through it, it is now considered worthwhile to allow the child to settle for a few hours, as edema at the ileocecal valve may resolve and a further radiological attempt at reduction can be made to avoid the need for surgery (Fig. 47.8). A second attempt is usually made at 4–6 hours if the patient is stable and the surgeon and radiologist concur.

Success rates vary amongst institutions, but current guidelines suggest that a success rate of at least 70 percent should be achieved. Perforation rates are typically 1–2 percent, and the consenting parents should be warned that this would necessi-

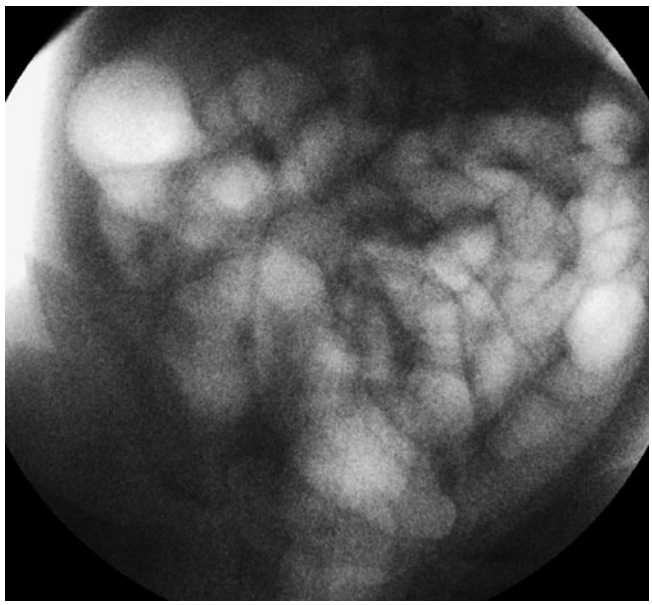


Fig. 47.7 Filling of small bowel with air, indicating successful reduction.

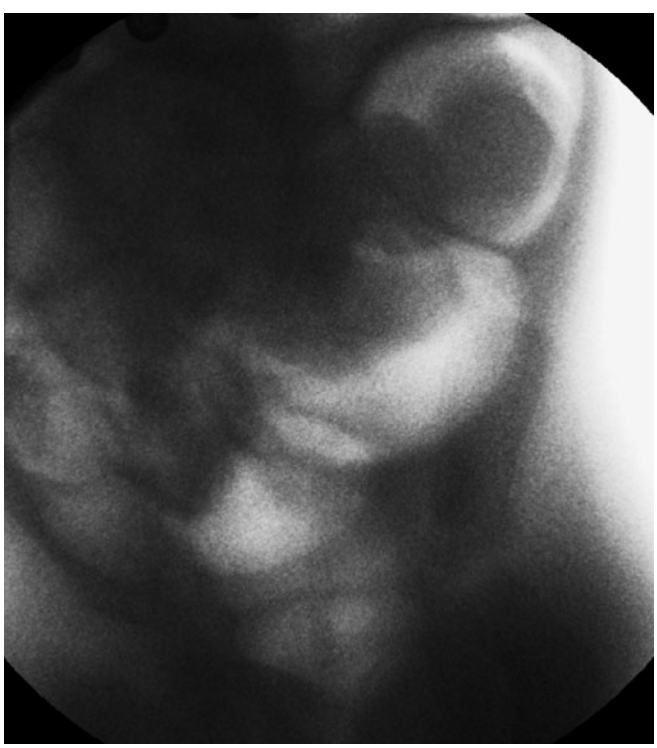


Fig. 47.6 Air dissection along the side of the insussusception.

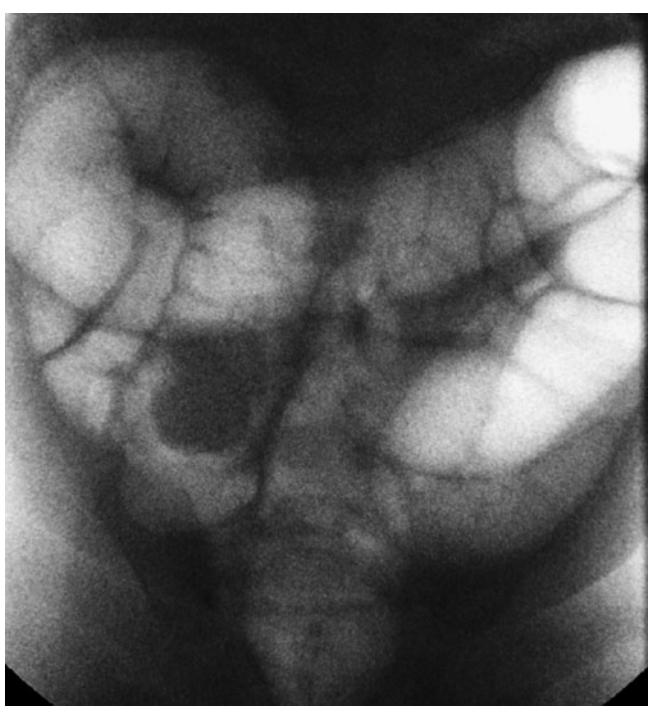


Fig. 47.8 Persistence of intussusception at the ileocecal valve.

tate immediate surgical intervention. A significant pneumoperitoneum requires immediate decompression by the radiologist, usually by the placing of an 18 G needle into the abdomen. Perforation with air reduction does not result in widespread contamination of the peritoneal cavity, as the large bowel is usually empty of stool as a result of the preceding diarrhea and also because air escapes and rises to the top of the abdomen rather than washing contaminated contents around within the abdomen, as would occur with fluids. Perforation is most likely to be secondary to bowel necrosis and not excessive pressures.

The principles outlined above also pertain to ultrasound-guided reduction. A tube is passed per rectum and a good seal is obtained at the anus. Water is introduced via the tube connected to a reservoir, which is then raised above the patient. To generate a pressure equivalent to 120 mmHg, the reservoir must be 150 cm above the patient. The intussusceptum already identified is then followed on ultrasound as it passes back along the colon. Reduction is complete when fluid is seen to pass retrogradely into the small bowel. Ultrasound reduction can be technically difficult for the operator if there is marked small bowel obstruction, with distended gas-filled loops, as air does not allow the passage of ultrasound and the image may be difficult to obtain. Nevertheless, avoiding the use of ionizing radiation is a considerable advantage. Recent studies have demonstrated the possibility of using air reduction under ultrasound guidance with some success.

INDICATIONS FOR OPERATIVE REDUCTION

Indications for operative reduction include:

- initial evidence of peritonism or perforation;
- perforation during radiological reduction;
- failure of radiological reduction;
- third-time presentation (presentation well beyond the usual age range).

PREOPERATIVE PREPARATION

To minimize delay, arrangements should be in place for emergency surgery for all infants undergoing attempted radiological reduction.

Fluid resuscitation should continue and the infant's hemodynamic status should be regularly assessed. Broad-spectrum antibiotics should be administered if not previously given prior to attempts at radiological reduction, and a nasogastric tube is passed.

Informed consent is obtained and should include the possibility of bowel resection, especially of the terminal ileum.

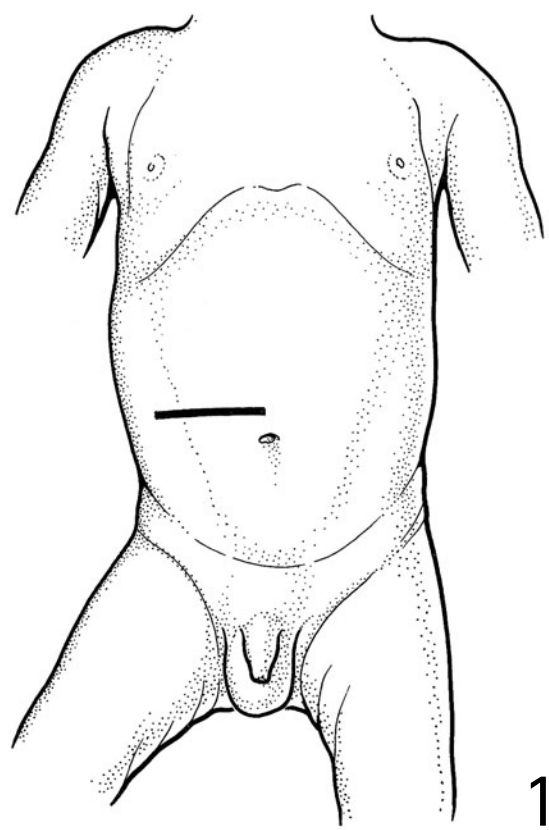
Anesthesia

General anesthesia with endotracheal intubation and muscle paralysis is used.

OPERATION

Incision

1 A right transverse skin incision is made either above or below the level of the umbilicus depending on the presence of a mass or radiological indication of the site of the intussusception. The lateral abdominal muscles, rectus sheath, and rectus muscle are divided to provide adequate and safe exposure.



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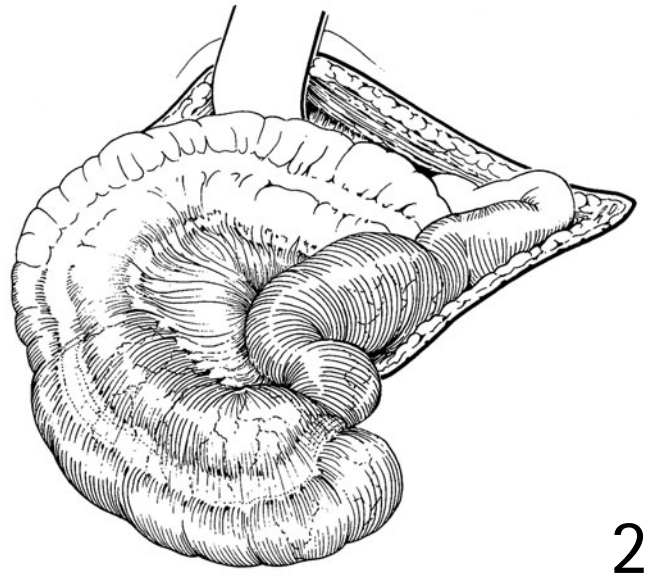
Reduction of intussusception

2 The affected bowel is delivered from the abdominal cavity to facilitate reduction. This often involves division of the peritoneal attachment of the right colon and cecum using sharp dissection.



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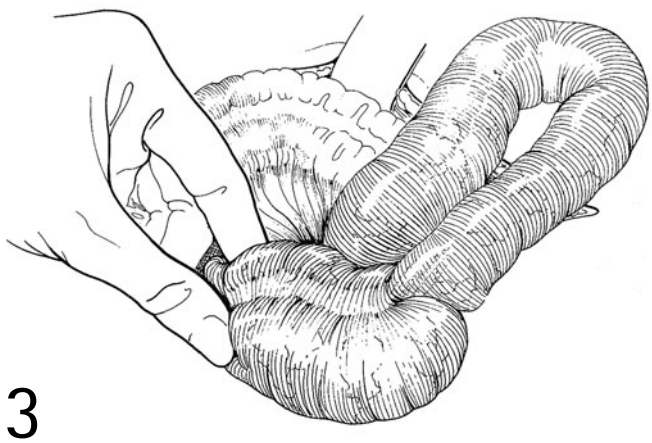
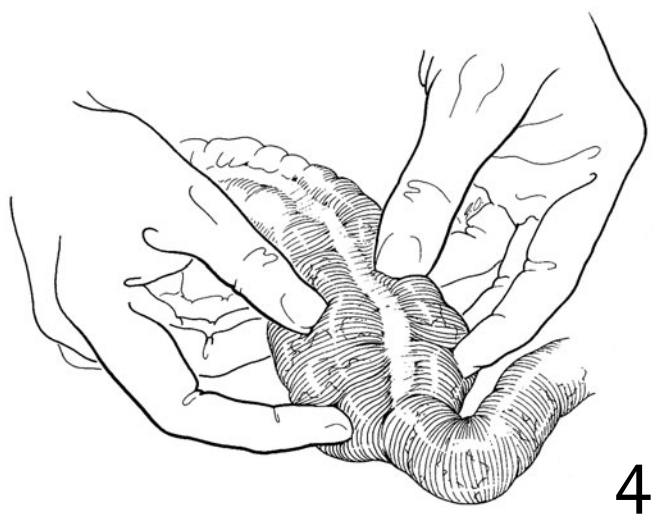


3

3 Once the affected segment has been delivered, all other parts of the intestine are returned to the abdomen. Reduction is achieved by gently applying pressure on the apex of the distal part of the intussusception. Grip on the intestine can be facilitated by the use of a gauze swab. Traction on the proximal intestine should be avoided, but a gentle pull may establish the direction in which to apply the reducing squeeze.

4 Reduction of ileum through the ileocecal valve requires patience. Forefingers and thumbs are used to apply gentle squeeze to the apex of the intussusception while pulling back the cecal wall. The gut should be palpated to rule out a pathological lead point, bearing in mind that an edematous ileocecal valve or Peyer's patch can mimic an intraluminal mass. Careful palpation and knowledge of the likely etiology, particularly in the young infant, should avoid unnecessary resection.



4

Resection

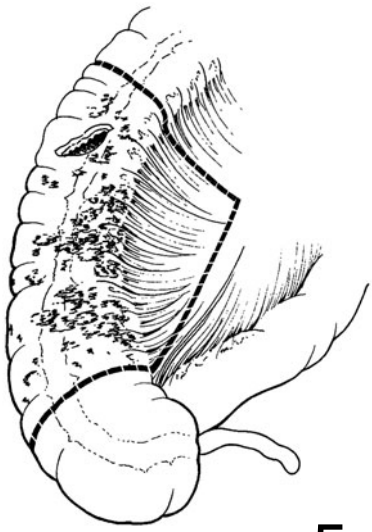
5a–c Resection will be necessary if:

- the intussusception cannot be reduced;
- there is necrotic or compromised bowel after reduction;
- there is a pathological lead point.

The intussusception should be reduced as far as possible to minimize the extent of any resection. Resection is usually segmental and rarely extends to formal right hemicolectomy.

If the intussusception is extensive, i.e., beyond the splenic flexure, consideration should be given to examining the duodenojejunal flexure for coexistent malrotation. If present, a formal Ladd's procedure should be performed.

In the presence of a transverse incision below the level of the umbilicus, it is acceptable to perform an incidental appendicectomy provided the adjacent cecal wall is normal.



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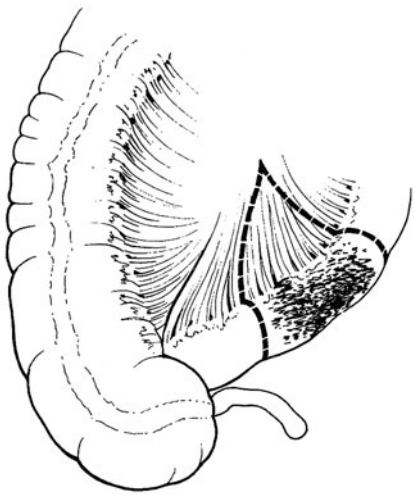

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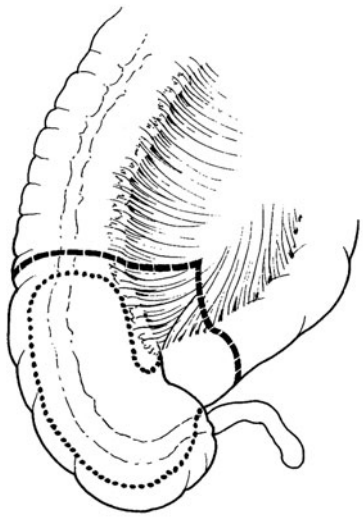

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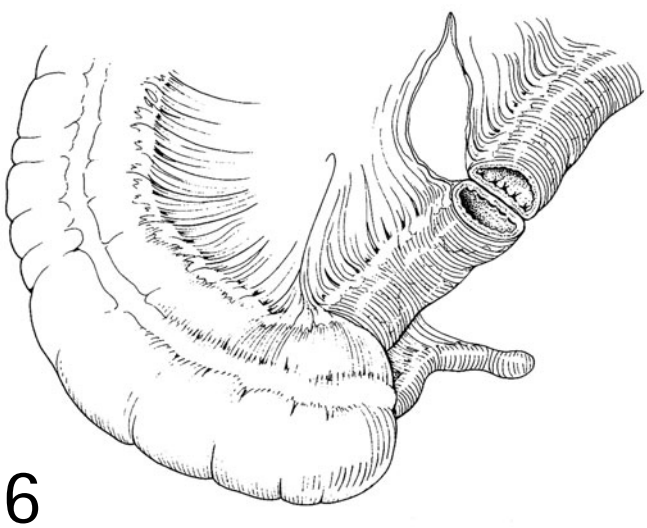


6

Anastomosis

6 Suction of the proximal and distal bowel avoids the need for bowel clamps. Primary anastomosis is almost always possible and is performed with a single layer of interrupted, extramucosal sutures. Knots are placed on the serosal surface of the bowel. The choice of suture material will depend on the surgeon's preference. Any mesenteric defect is closed in similar fashion. The need for expeditious surgery in an unstable infant or doubt about the viability of the resection margins may require the formation of adjacent stomas. Closure of the stomas is performed when the infant's condition has improved.

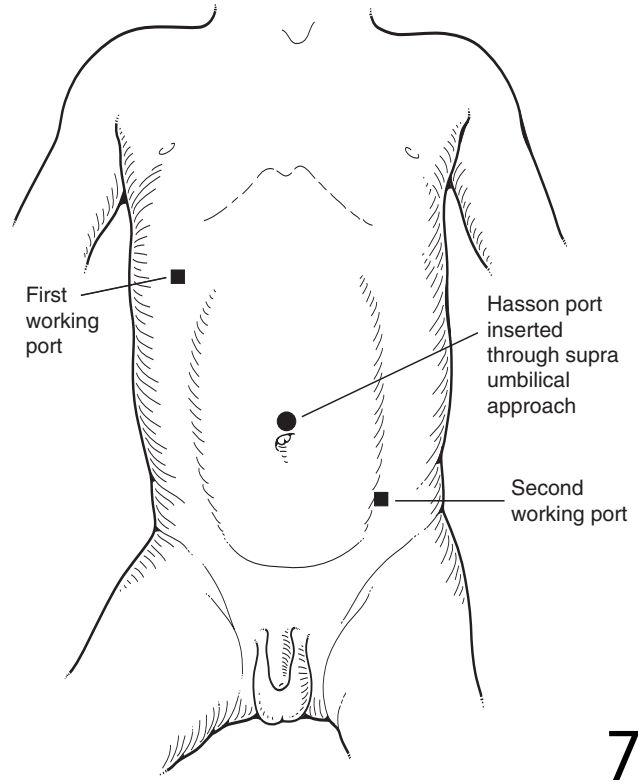
It may be possible to over-sew a perforation using interrupted, extramucosal sutures provided the edges are clean and viable.

Laparoscopic reduction

The techniques of minimally invasive surgery are currently being applied to infants with intussusception. Potential advantages are the ability to diagnose full reduction when this is unclear after pneumatic reduction, thus avoiding laparotomy or cases of recurrence. Reduction with minimal invasive surgery is more problematic, but if achievable, avoids the trauma of open access and significantly reduces the risk of intraperitoneal adhesions.

Pneumoperitoneum is raised according to surgical preference (see Chapter 34). Access ports are inserted in the right upper quadrant and left lower quadrant, at right-angles to the small bowel mesentery. Reduction is effected by a combination of taxis and traction using atraumatic graspers.

Conversion to an open procedure, if necessary, can be performed through a small and accurately placed incision.



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POSTOPERATIVE CARE

Appropriate intravenous fluids are continued postoperatively. Nasogastric decompression is continued until the volume decreases, indicating the return of intestinal function. Oral fluids are commenced within 24 hours, but in the presence of resection may be delayed a further 24 hours. Continued antibiotic therapy is dictated by surgeon's preference and the presence of peritoneal contamination at the time of laparotomy.

A high temperature is common in the first 24–48 hours postoperatively and usually subsides without specific treatment.

OUTCOME

Recurrences following pneumatic reduction are reported in up to 8 percent of cases. Further radiological reduction should be attempted, but a second recurrence should raise the possibility of a pathological lead point.

Deaths from intussusception have been reported and relate to failure to establish the diagnosis and inadequate resuscitation.

Excision of the ileocecal valve may predispose to anemia secondary to depletion of vitamin B12 or the development of gall stones from loss of bile salts, but both are uncommon in clinical practice.

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48

Appendectomy: open and laparoscopic approaches

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HISTORY

In 1554, a 7-year-old French girl developed abdominal pain and diarrhea that ominously progressed to signs of right lower quadrant peritonitis. In an era lacking antibiotic and fluid support, she quickly succumbed to overwhelming peritoneal sepsis. Post-mortem examination confirmed the clinical diagnosis of 'iliac passion' caused by free perforation in the region of the cecum. Her physicians, perhaps spurred on by recent descriptions of the appendix by DaVinci (1492) and Vesalius (1545), published a careful description of her appendix and its involvement in the disease. Thus began a debate on the role of the appendix and its surgical therapy that lasted for over three centuries. Progress was made in the early nineteenth century with the cautious recommendation to drain abscesses clearly localized to the right lower quadrant. Encouraged by early success, the indications expanded to include operation for the diagnosis of spreading peritonitis. In 1886, Reginald Fitz published convincing evidence that the appendix was the cause of this most common malady and, importantly, that operation should occur within 48 hours of symptom onset. News of Fitz's work traveled across the Atlantic to London, where Treves instantly popularized appendectomy by curing King Edward VII in 1902. In Canada, a country doctor named Abraham Groves is said to have diagnosed and successfully removed an inflamed appendix of a 12-year-old boy while on rounds of his farming area. This event apparently took place in 1883, three years before the Fitz landmark presentation. However, credit for the first published report goes to Claudius Amyand from England who, in 1736, removed an inflamed perforated and fistulizing appendix from the scrotum of an 11-year-old boy.

The almost pedantic modern state of appendectomy belies a long history of bold innovation and honest communication with colleagues. Today the debate remains fresh, with advances in preoperative care, diagnostic imaging, antibiotics, and laparoscopic removal. The search for new ways to improve care that started with a 7-year-old girl in France has now relieved the suffering of millions from this ancient, most common abdominal surgical emergency.

PRINCIPLES AND JUSTIFICATION

Indications

Appendectomy has been recommended for appendicitis and other conditions of the appendix for over 100 years. The recent availability of a reliable laparoscopic approach should not change the basic indications. Candidates for laparoscopic appendectomy undergo the same careful preoperative diagnostic evaluation as those children who will have an open procedure. Prompt operation by either approach is the most effective way to prevent the inexorable progression of acute appendicitis to full-thickness necrosis and perforation. Most surgeons believe these sequelae will occur within 48 hours of the onset of symptoms; earlier perforation may be expected in young children or when there is complete obstruction of the lumen such as by a fecalith. Once perforated, operative drainage and appendectomy are not problematic, but are vital for survival in most circumstances. Occasionally a child with a well-contained perforation will benefit from intravenous antibiotics, percutaneous drainage, and delayed appendectomy. In these otherwise stable patients, appendectomy performed acutely has not been as successful. The delayed or interval appendectomy, if performed at all, may be approached by laparoscopic or open methods after approximately 6 weeks, or later.

Appendectomy is also recommended for small (< 2 cm) carcinoid tumors at the tip of the appendix, other benign conditions of the appendix, and as part of Ladd's procedure for midgut malrotation. Incidental removal of a normal appendix is often a part of other gastrointestinal operations,

such as pull-through procedures for Hirschsprung's disease in small children. Whether a normal appendix should be removed in an older child not undergoing a gastrointestinal procedure is open to debate.

Contraindications

Most children with perforated appendicitis can and should undergo immediate operation for control of intra-abdominal sepsis and prevention of recurrent disease. However, stable patients with a well-contained perforation will benefit from the non-operative approach detailed above. A laparoscopic appendectomy cannot be justified for patients with advanced perforation and sepsis or a large appendiceal mass. In addition to the complications of laparoscopic dissection under these circumstances, the benefits of laparoscopy such as early hospital dismissal are negated by the need for inpatient treatment that includes intravenous fluids, antibiotics, and pain control for resolving peritonitis. It is often too difficult to assemble these therapies on an outpatient basis. For these reasons, it is essential to continue the time-honored practice of abdominal examination under anesthesia before appendectomy. If a large immobile mass has been palpated, a laparoscopic approach should probably be abandoned in favor of an open procedure or conservative therapy.

Incidental appendectomy is relatively contraindicated as part of an abdominal procedure when the intestine will not otherwise be opened. A normal appendix should not be removed if vascular or other prosthetic materials are used.

PREOPERATIVE

Timely operation despite a paucity of corroborating preoperative evidence is still the best approach to children with persistent right lower quadrant pain. Mortality from perforation has been prevented by antibiotics and fluid support, but long-term complications are still seen. For this reason, a 10–15 percent negative laparotomy or laparoscopy rate is the accepted norm.

The approach to the diagnosis of acute appendicitis in this era of sophisticated preoperative imaging still relies on the history of a visceral prodrome that progresses to localized somatic symptoms in the right lower quadrant. The delight of treating children with appendicitis is the challenge of a history that is not always classical, but the physical examination will reveal the diagnosis. In most instances, no further testing is required. When the diagnosis is questionable, computed tomographic scanning has become popular; fast, high-resolution, lower radiation scans have taken much of the guess work out of the diagnosis, but whether or not eventual outcome is improved is still debated. Over-utilization and radiation dose remain a concern as the search for other preoperative investigations continues. Ultrasonography has been very helpful, not only to visualize an inflamed appendix, but also, more importantly, to exclude other abdominal and pelvic conditions, especially in teenage girls. Radiographic examination is non-specific, except to identify a fecalith, and barium studies are usually contraindicated.

Children with appendicitis are evaluated for degree of sepsis and dehydration. Most will have vomited or not eaten for more than 24 hours. For this reason alone, intravenous isotonic fluid resuscitation before operation is mandatory for all patients with appendicitis. Superimposed sepsis will indicate close attention to volume support before operation. Infectious complications are best prevented and treated in all patients by using perioperative antibiotics. The typical regime is designed to control aerobes and anaerobes of the lower gastrointestinal tract. In the author's experience, the approach of universal antibiotic coverage for all patients with appendicitis has resulted in wound infection and intra-abdominal abscess rates of 2.6 percent and 4.4 percent respectively for gangrenous or perforated appendicitis. The classic 'triple antibiotic' therapy is effective, and several newer medications achieve similar results.

The most efficient way to control preoperative pain is immediate appendectomy, but if delayed, intravenous narcotics are acceptable.

Patients undergoing a laparoscopic procedure benefit from nasogastric and bladder decompression prior to operation. Once again, the abdomen should be palpated before insufflation to rule out the presence of a large fixed mass.

Psychological preparation of the child and family is essential for a successful procedure. For many patients, this is their first introduction to the hospital and surgery. The family is informed of what to expect during the hospital stay as well as of the resources available to them and their child. Most children usually just want to know when they will be able to go home, but find these explanations reassuring.

Informed consent includes the location of the incision, the expected postoperative course, the negative appendectomy rate, the possibility of drains, infectious, and hemorrhagic complications, and the remote chance of a stoma.

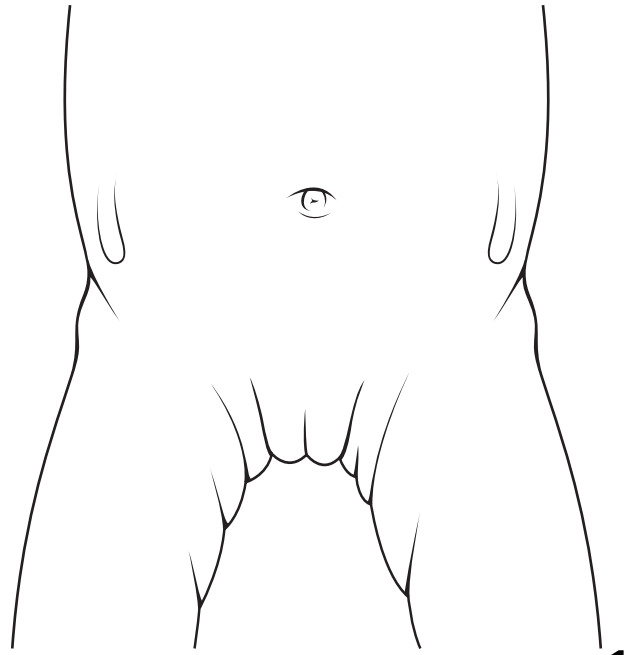
Anesthesia

Apart from pain control, premedication is generally unnecessary. A general anesthetic with muscle relaxation provides the best environment for surgical exposure. Local or regional anesthetic blocks are not helpful in infected or septic cases.

OPERATION

Open appendectomy

1 The principles of skin incision for appendicitis include a transverse right lower quadrant skin crease approach, allowance for extension medially or laterally, and avoidance of the large periumbilical fat pad of the plump child. With the child supine on the operating table, and positioned closer to the surgeon's side when using an adult table, the abdomen is palpated for the presence of a mass. If detected, the incision is adjusted to be in close proximity to the mass, allowing for local topography and possible extension as needed. The slender child with point tenderness could tolerate appendectomy through a very low, short skin crease incision that can be extended medially if required. This approach is risky and best avoided by placing the incision just above and slightly medial to the anterior superior iliac spine. The incision then courses medially through or below McBurney's point. Lateral extension is also possible to the flank, while medial enlargement crosses the rectus sheath. As a general rule, a higher and longer incision will provide more room for exploration, a satisfactory and faster dissection, but a more prolonged and painful hospital stay.



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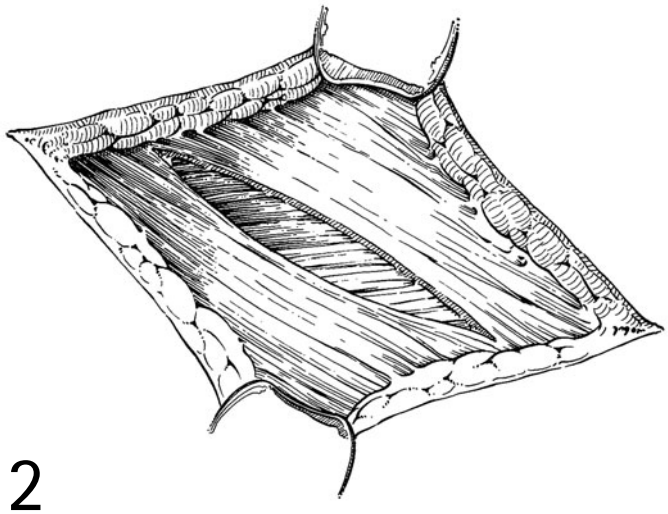
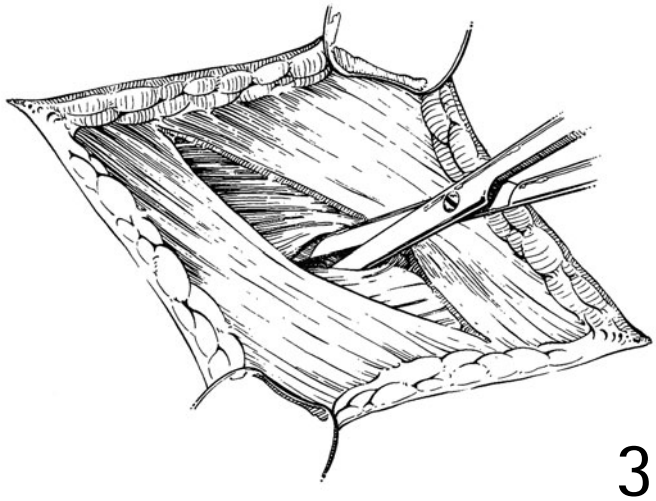


2

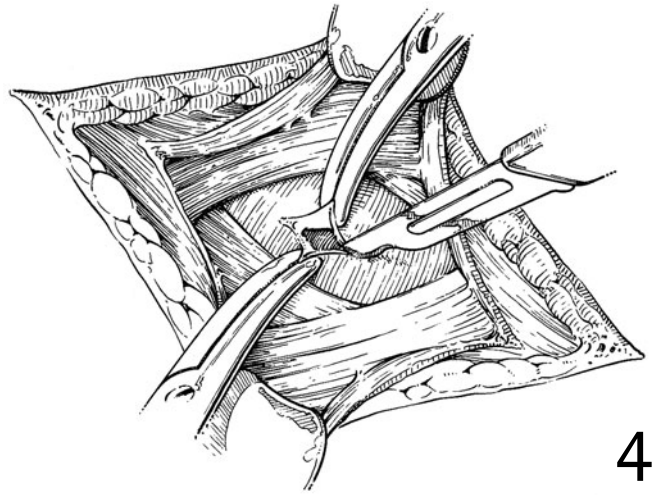
2 Following sharp dissection, the external oblique muscle is split along the direction of the fibers, out to and including the muscular belly. A liberal split of the external oblique muscle will provide adequate operative exposure.

3 The internal oblique and transverse muscles are split in the direction of their fibers using blunt-tipped scissors. Generally, both muscle bellies are split in tandem, as the transverse muscle is nearly parallel with the internal oblique muscle. The aponeurosis of the transverse muscle occurs more laterally, and the fibers may run more obliquely from lower right to upper left than those of the internal oblique. Once split over a short distance, small retractors are then placed to continue the muscle division in the direction of the fibers.



3

4 The peritoneum is grasped after blunt dissection of the transversalis fascia and lateral preperitoneal fat. Edema from appendiceal inflammation may obscure the peritoneum laterally; it is best identified medially. On opening the peritoneum with a scalpel, free fluid is suctioned and may be sent for culture. The peritoneal opening is enlarged with scissors.



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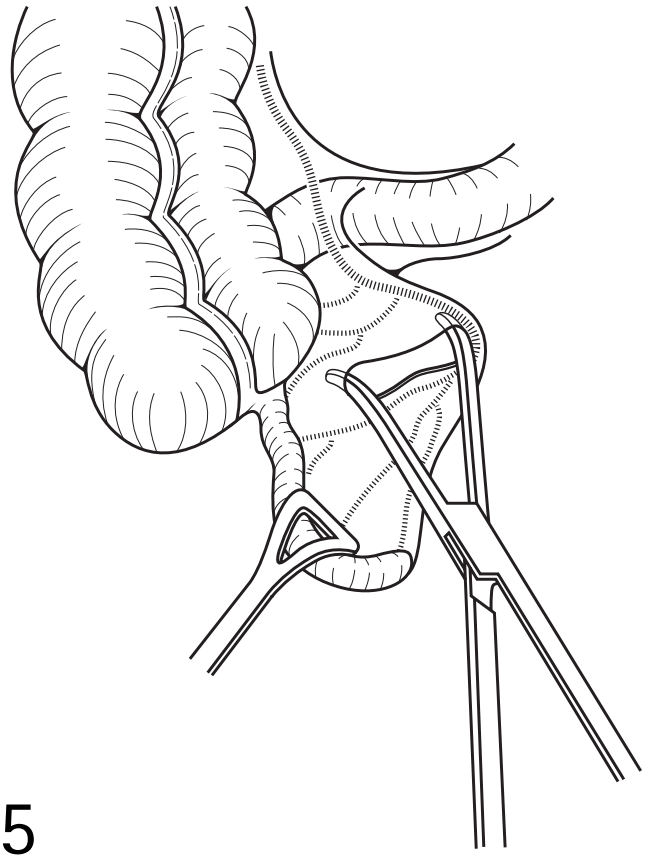
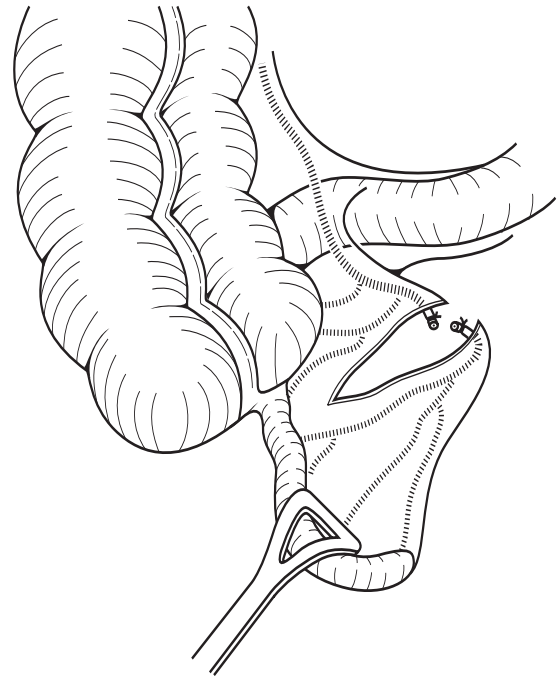


5

5 The key to successful exposure of the appendix is delivery of the cecum into the wound. The anterior tenia coli is grasped and the cecum delivered using an up-and-down rocking motion, first pulling down then up. If the cecum cannot be delivered easily, lateral peritoneal attachments may require dissection under direct vision using cautery. This division should never be performed blindly or medially to the appendix. An appendiceal mass is almost always bound down laterally and inferiorly. Medial attachments are usually to the terminal ileum and its mesentery. These should be divided once the mass has been safely delivered through the wound.

A difficult dissection becomes easier with wound extension. The muscle-splitting incision can be extended medially across the anterior and posterior rectus sheaths using cautery. The rectus muscle is retracted and the inferior epigastric vessels divided between hemostatic ties. Lateral incision enlargement is also helpful if the wound is too medial or further dissection is required in the flank.

6 The inflamed appendix and any overlying omentum should not be allowed to touch the wound once delivered. Attached omentum is divided between hemostatic ties, and the mesoappendix is gently grasped with large Babcock forceps encircling, but not touching, the appendix. With traction on the Babcock forceps, the appendix is easily controlled and any further lateral attachments divided with cautery. The mesoappendix is then serially divided between hemostats and tied in sequence. The inflamed mesentery does not hold ties well. Excessive upper traction during tying may lead to delayed knot slippage and hematoma. The mesenteric division should continue to Treves' fold and end with careful evaluation of a small group of veins on the inferior cecal wall. These veins usually do not require division, but excessive traction on the appendix may tear them.



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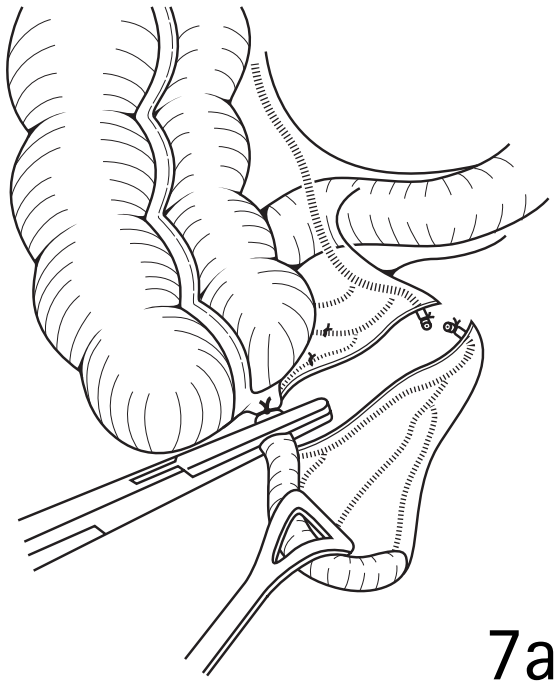
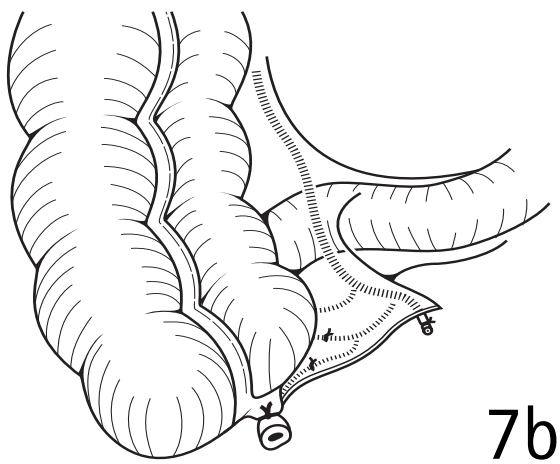


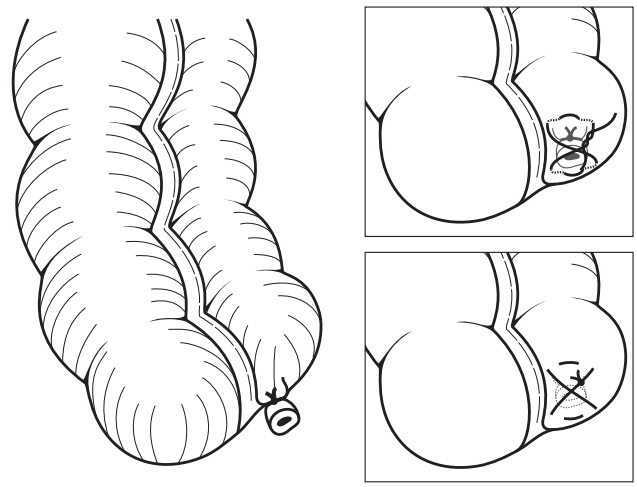
7a



7b

7a,b The base of the appendix is crushed 5 mm above its origin and the clamp is drawn distally a few more millimeters. The area crushed is then tied with an absorbable suture and the appendix removed by sharp division just proximal to the clamp. The mucosa of the remaining stump is cleansed with antiseptic solution and may be cauterized. Stump inversion is not necessary but, if performed, a Z-shaped, purse-string, absorbable suture is placed through the seromuscular base of the cecum, taking care to avoid the inferior cecal veins. If the stump is inverted with the help of a hemostat, it is then removed from the operating field. Rarely, the appendix is pedicled on a very broad and inflamed base that cannot be safely ligated with a free tie. In this case, a double row of inverted Lembert sutures will satisfactorily avoid a blown stump.

8 Before replacing the cecum, the operative field and mesoappendix ties are inspected for hemostasis. The distended cecum may prove difficult to return to the abdomen. Gentle alternative finger compression avoids tearing by forceps or stump blow-out from excessive pressure.



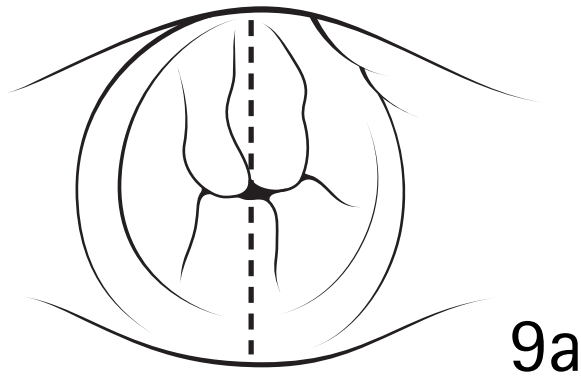
8

The pelvis and right paracolic spaces are suctioned and irrigated with saline if pus is found. Large, well-developed abscesses may be drained using closed wound suction through a separate stab wound. This is rare in children, since most fibrinopurulent debris can be removed atraumatically and the areas irrigated clear. When there has been free perforation, the surrounding bowel loops and omentum should be checked for abscesses. Intra-abdominal antibiotics are not helpful when perioperative antibiotics have been used.

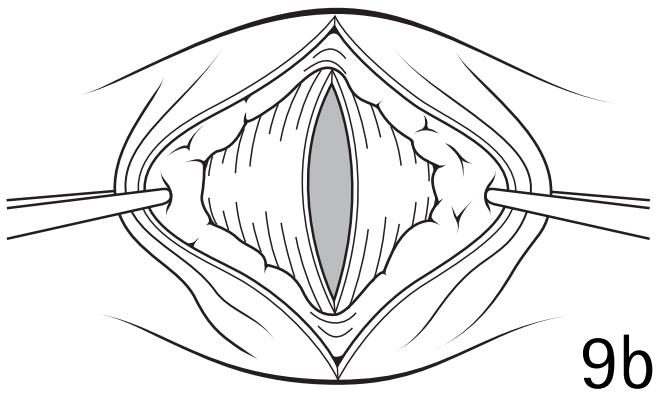
The wound is closed in layers with absorbable sutures after irrigation of each consecutive layer. The peritoneum is run, and the internal oblique and transverse muscles are closed at the same time by apposition of the internal oblique epimysium using interrupted, absorbable sutures. The external oblique aponeurosis is closed with a running suture, followed by Scarpa's fascia and skin. Even when there has been perforation and gross contamination, the skin is usually closed with an absorbable, running, subcuticular suture.

Laparoscopic appendectomy

9a–c A 10–12 mm vertical intraumbilical incision provides the most cosmetic approach for the laparoscopic port and is easily extended as needed. In smaller children, a curvilinear incision is necessary. It may also be wise to open the umbilicus in the small child rather than attempt percutaneous placement of these ports. If an open technique is used, a horizontal mattress suture around the port provides satisfactory air seal without the necessity of using a more complex cannula. The abdomen is insufflated with carbon dioxide in a standard fashion and the laparoscope is inserted for diagnosis and assessment of operative feasibility.



9a



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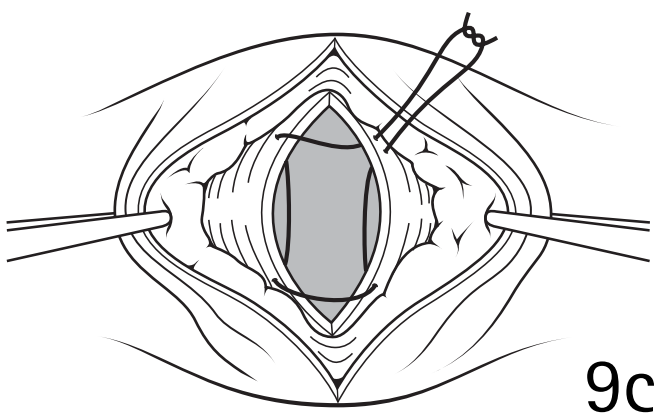

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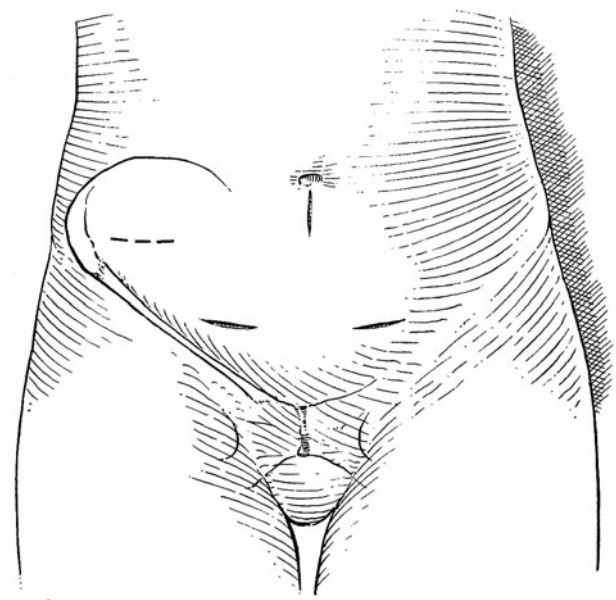
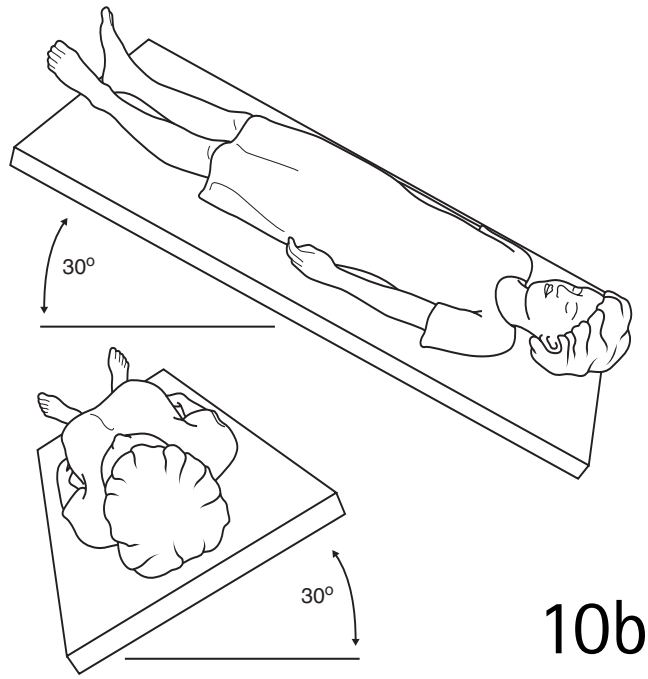


10a

10a Two separate 3–5-mm ports are placed in the lower abdomen under laparoscopic control to avoid the inferior epigastric vessels. An optional third port may be necessary for more complex dissection. Exact port positioning varies with the size of the child and location of the appendix. Generally, a right lower quadrant and suprapubic port will suffice. There are many modifications of port placement and dissection, including a single umbilical port for laparoscopic identification and transumbilical removal.

10b Visualization of the appendix is enhanced by Trendelenburg positioning and rotation of the table to the patient's left.



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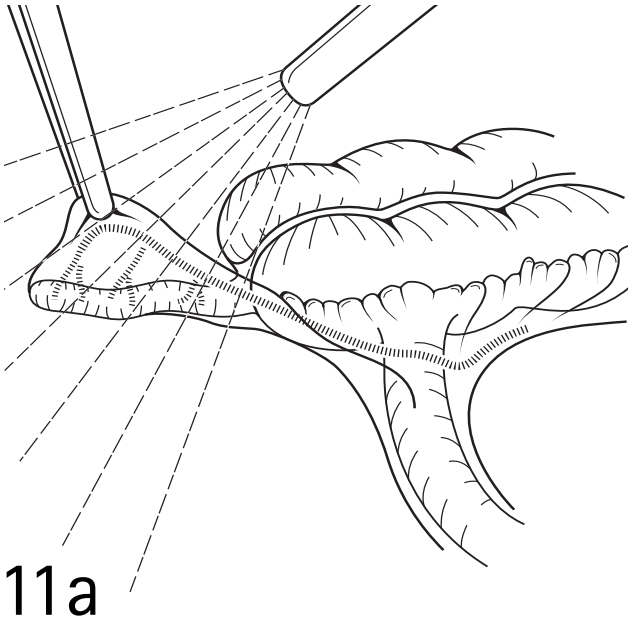
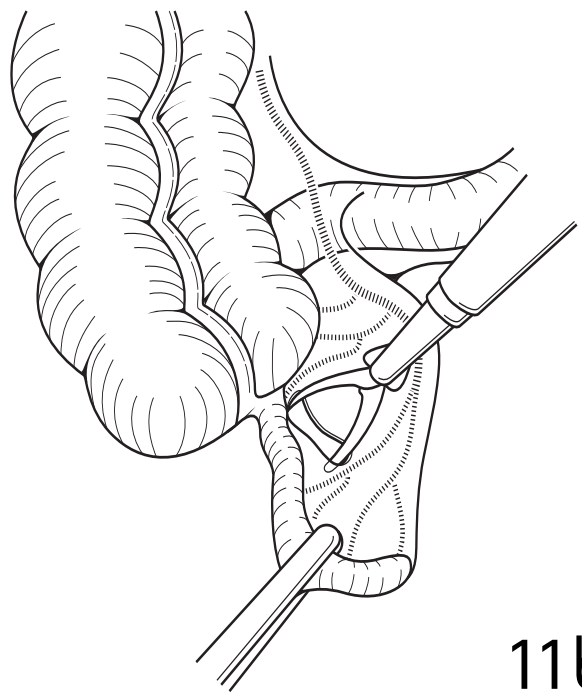


11a

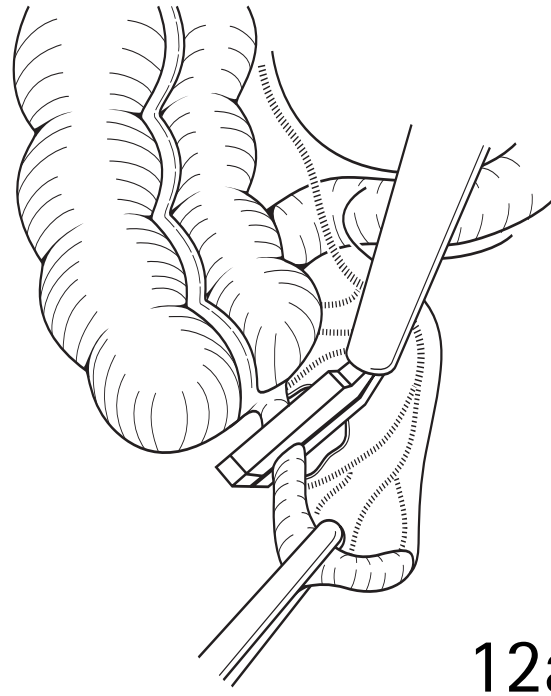
11a The appendix is grasped by the free edge of the mesentery or with laparoscopic Babcock forceps through the right lower quadrant port. With traction of the appendix, any lateral peritoneal attachments are divided to free the appendix and its mesentery. The dissection does not require extension onto the cecum, as it will not be elevated through the wound. Lateral mobilization of the cecum is only desirable if the appendix is to be pulled through the umbilical port for extracorporeal appendectomy. Even then, it is usually not necessary.

11b A mesenteric window is created near the base of the appendix. This is accomplished through one of the lateral ports using blunt dissection and cautery. Care is taken to visualize the cecal wall throughout the maneuver.

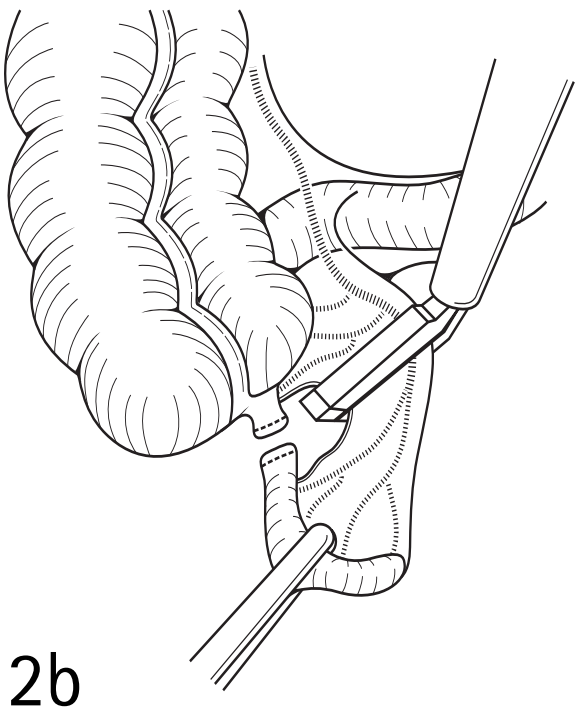


11b

12a The order of appendiceal and mesenteric division is a preference of technique. Once pedicled on the cecum, the appendiceal stump is ligated with suture or the gastrointestinal stapler. Care must be taken with the stapler to ensure that only the base is ligated, avoiding the cecal wall. This step often precedes mesenteric division when staplers are used.



12a



12b

12b The mesoappendix is divided using cautery, clips, or a stapler. Many surgeons find cautery is sufficient. In certain cases, visualization of the mesoappendix is improved by prior stapled division of the appendix through the mesenteric window. The larger stapling instruments may be inserted through the umbilical port under direct vision with a 3 mm or 5 mm camera placed through one of the other ports.

The appendix is then removed through the sheath of the largest trocar so that the infected appendix does not touch the intra-abdominal wall. If the appendix is large, it is placed in a sterile bag and removed through the largest wound following trocar removal. After a final check for hemostasis, the abdomen is suctioned and irrigated as necessary. The laparoscopes and trocars are removed, the abdomen desufflated, and the umbilicus closed with fascial sutures. The 3 mm incisions require subcutaneous closure; however, 5 mm incisions require fascial closure.

POSTOPERATIVE CARE

If the appendix was not perforated, a nasogastric tube is not required and the patient begins oral diet within 12–24 hours. Antibiotics are discontinued after 2 postoperative doses. Intravenous narcotics are given on demand or by patient-controlled analgesia pump for older children during the first day. Pain control transitions to oral medication as needed on the second postoperative day. Most children are up and active the day after surgery and are discharged home that day or early the next.

The amount of peritoneal contamination and visceral inflammation will determine the need for nasogastric tube decompression and nil-by-mouth status in patients with perforated appendices. Intravenous fluid resuscitation is vital for the first 24 hours after operation to compensate for ongoing third space losses from peritoneal inflammation. Antibiotics are continued until the temperature is normal (37 °C) and the white blood cell count < 10^9/L with absence of left shift (< 80 percent neutrophils). Occasionally bladder catheter decompression is necessary. Patients are encouraged to ambulate, deep breathe, and cough. Intravenous narcotics may be continued for 2–3 days.

Complications

INFECTIONS

The harbinger of infection is persistence of fever, white blood cell count elevation, and pain. It is rare that patients with entirely normal vital signs and white blood cell count will return with an abscess, but recrudescence of these symptoms will signal the complication. Wound infections may be drained locally, but intra-abdominal abscesses will require antibiotics and probable drainage. Intra-abdominal abscesses usually occur in the right pelvis, but may be found anywhere, including between bowel loops and under the diaphragm. Percutaneous drainage, under radiologic control, and antibiotics have been very successful. Large phlegmonous infections are not amenable to drainage and may require several weeks to subside on intravenous antibiotics. These patients often do not eat well and may require parenteral nutrition.

HEMORRHAGE

Significant postoperative hemorrhage requires re-operation to control bleeding and prevent infection.

STUMP LEAK

A blown appendiceal stump can present with diffuse feculent peritonitis and sepsis and requires immediate re-operation. Occasionally stump leaks have been discovered only after continued leakage of feculent material from the wound or percutaneous drain in otherwise stable outpatients. Such situations may be managed non-operatively with the hope that the tract will eventually seal. If not, re-operation to excise and close the tract will effect a cure. Persistent drainage may also indicate retained foreign material such as a fecalith that escaped detection at initial operation.

OUTCOME

In an update of a standard protocol described in this chapter, 648 children were treated for appendicitis with perioperative antibiotics, primary wound closure, and no drainage. Discharge criteria for complicated appendicitis followed the protocol described above. There were no wound infections in patients with simple appendicitis, and the deep infection rate was 0.56 percent. Children with complicated appendicitis had infection rates of 2.6 percent for wound and 4.4 percent for intra-abdominal abscesses. Hospital stay was 1.39 ± 0.89 for simple appendicitis, 2.97 ± 1.25 for gangrenous, and 6.31 ± 3.51 for ruptured appendicitis. These results indicate the efficacy of perioperative intravenous antibiotics and careful peritoneal and wound cleansing to decrease infection rates in children.

FURTHER READING

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49

Necrotizing enterocolitis

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HISTORY

Necrotizing enterocolitis (NEC) is a devastating disease of infancy and the most common serious acquired gastrointestinal disease in the newborn infant. The term necrotizing enterocolitis was first coined in the 1950s when used to describe infants who died with necrotic lesions of the gastrointestinal tract, but it did not become recognized as a distinct clinical entity until the 1960s, when a number of authors began reporting their experience with this disease. With improvements in neonatal intensive care over the past four decades, the incidence of NEC is increasing as more babies born at the extreme limits of prematurity now survive. Recent estimates place the incidence at approximately 0.5 percent of all live births and at between 3 and 5 percent of low-birthweight infants born prematurely. Concurrent with this increase in incidence has been an extensive amount of time and energy devoted to exploring the etiology and pathogenesis of this disease by a number of individuals and groups worldwide. Despite these efforts, our understanding of the processes contributing to the development of NEC remains limited.

PRINCIPLES AND JUSTIFICATION

Necrotizing enterocolitis represents a disease state comprising a wide spectrum of intestinal pathology and resulting systemic manifestations. At one end of the spectrum lie infants with minimal systemic upset with a distended abdomen and in whom the diagnosis of NEC is made on the pathognomonic radiographic finding of pneumatosis intestinalis. In the absence of evidence of intestinal perforation or clinical deterioration, such infants are often managed conservatively by resting and decompressing the intestinal tract and administering broad-spectrum antibiotics. Following 7–10 days of such conservative management, feeds may be slowly reintroduced. Whilst some of these infants may develop complications of NEC such as a post-inflammatory intestinal stricture requiring surgery, the majority recover without the need for operative intervention.

At the other end of the spectrum are infants with extensive intestinal involvement and gross systemic upset, often with failure of one or more organ systems. These infants almost invariably require surgical intervention and often require resection of one or more lengths of gangrenous intestine. Unfortunately, in these severely ill babies the intestinal involvement is only part of the overall disease process, and the development of multi-system organ failure requires prolonged periods of intensive care. Mortality in this severely affected group is as high as 20–40 percent.

There is a group of infants who present with similar, if not identical, abdominal and systemic symptoms and signs to those found in infants with NEC but who are found to have a focal intestinal perforation, often in the terminal ileum, with preservation of the remainder of the gastrointestinal tract. This is often associated with indomethacin therapy used to encourage closure of a patent ductus arteriosus. Whether focal intestinal perforation is one end of the spectrum of NEC is frequently debated. The presentation and principles of management are identical to those of infants with NEC and they are often grouped together.

Symptoms and signs

The onset of symptoms of NEC appears to be inversely related to the gestational age at birth and birth weight, such that full-term infants who develop NEC (primarily but not exclusively those with congenital cardiac abnormalities) do so in the first few days of life, whereas preterm infants of low birth weight develop symptoms most commonly between 15 and 20 days of age. The etiological factors implicated in the

development of NEC are an immature intestine, mesenteric hypoxia or ischemia, bacterial colonization of the intestine, and the presence of feed within the intestinal lumen. The precise pathogenesis remains poorly understood.

The physical condition of infants with NEC varies. Initial symptoms may be subtle, such as increased gastric aspirates, mild abdominal distension, and flecks of blood in the stool. However, some infants present with cardiovascular collapse, bilious vomiting, gross abdominal distension with tenderness, and the passage of frank blood per rectum. Additional presenting features include abdominal erythema or discoloration, the presence of an abdominal mass, and non-specific signs of sepsis, including fever and hypovolemia. In addition, abnormal laboratory tests are often present, including thrombocytopenia, raised C-reactive protein concentration, and high or low white blood cell count.

Radiographic findings

The radiographic findings in infants with NEC vary from one case to another, but the presence of pneumatosis intestinalis on abdominal radiograph is pathognomonic of NEC. Other radiographic findings include portal venous gas, dilated loops of intestine (one or more of which may appear to be 'fixed' on serial radiographs), ascites, and, in the presence of intestinal perforation, pneumoperitoneum (free gas within the peritoneal cavity). Radiographs of infants with focal perforation may have none of these findings other than pneumoperitoneum. The presence of pneumoperitoneum may be difficult to detect, and radiographs of infants with NEC or suspected NEC should be carefully examined for the presence of free gas.

Indications for surgery

The initial management of all infants with NEC consists of appropriate resuscitation and support of all failing organ systems. The majority of infants require mechanical ventilation and in many the cardiovascular, renal, and hematological systems also require support. Conservative management therefore comprises 7–10 days of intestinal rest and broad-spectrum antibiotic treatment, with frequent clinical review to identify those infants in need of surgical intervention.

Indications for surgery (Box 49.1) in the setting of acute NEC are contested among surgeons and remain based largely on clinical judgment and previous experience. Absolute indications for surgical intervention reported in the literature are the presence of free gas within the peritoneal cavity, the continued deterioration of an infant despite maximal medical therapy, and the presence of an abdominal mass with ongoing intestinal obstruction or sepsis. Among these, the most widely accepted indication for surgery is pneumoperitoneum. The other 'relative' indications for surgical intervention are controversial.

In addition to surgery during the acute stage of NEC, a number of infants require surgery at a later stage to treat complications of NEC regardless of whether they have had surgery during the acute stage of their illness. The most common indication is the presence of an intestinal stricture.

Box 49.1 *Indications for surgery in infants with acute necrotizing enterocolitis*

Absolute indications
Pneumoperitoneum
Clinical deterioration despite maximal medical treatment
Abdominal mass with persistent intestinal obstruction or sepsis

Relative indications
Increased abdominal tenderness, distension and/or discoloration
Fixed intestinal loop
Portal vein gas
Positive paracentesis
Thrombocytopenia

Surgical approach

There is no general consensus among surgeons concerning the ideal surgical management of NEC. Current surgical options include primary peritoneal drainage (PPD) or laparotomy. Primary peritoneal drainage was first reported in 1975 by Marsh and Ein as a means of stabilizing and improving the systemic status of premature infants with intestinal perforation secondary to NEC. Since then, PPD has been reported as a definitive therapy rather than an intermediary for laparotomy. The majority of surgeons currently consider that PPD is best reserved for small infants (less than 1000 g in weight) with evidence of pneumoperitoneum. However, whether these infants are best served by PPD or laparotomy is unknown, and the results of trials comparing these two treatment modalities are eagerly awaited.

Minimal access surgery has become commonplace in recent years within the field of pediatric surgery, but has not been reported extensively in the setting of NEC. Certainly performing any of the operations outlined below in a neonate using a laparoscopic approach would be technically demanding and potentially unsafe. However, diagnostic laparoscopy may assist surgical decision making in infants with NEC. In infants who are critically unwell yet who lack a specific indication for surgery and for whom laparotomy may have disastrous consequences, laparoscopy allows visualization of the intestine and a more informed decision to be made concerning the need for laparotomy based on the condition of the intestine.

PREOPERATIVE ASSESSMENT AND PREPARATION

Infants with NEC are nursed on a neonatal intensive care unit with continuous monitoring. Prior to surgery, infants should

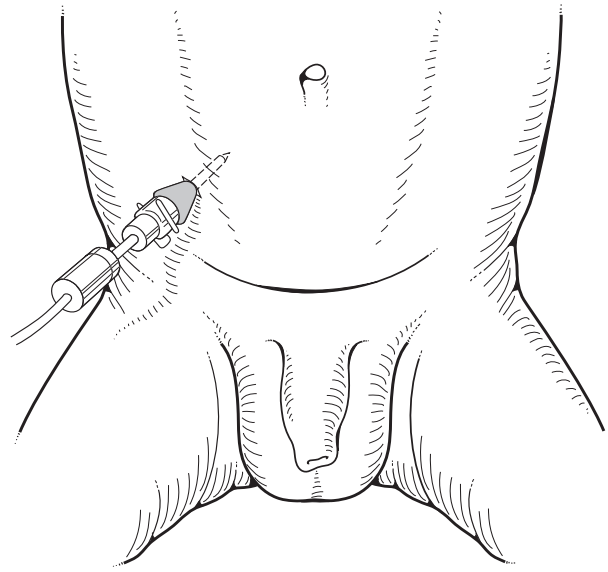
be fully resuscitated and cardiovascularly stable with adequate blood pressure. Of particular importance in the setting of NEC is correction of coagulopathy and thrombocytopenia prior to surgery, ensuring that blood products are available for immediate use during and after the operative period if required. A proportion of infants with NEC develop T-antigen activation in which red cells become sensitive to hemolysis during blood transfusion. Hemolysis can be avoided by the administration of compatible blood products (low titer anti-T fresh frozen plasma, washed platelets, and red blood cells). It is our practice routinely to test all infants with NEC for T-antigen activation prior to the administration of blood products.

OPERATIONS

Diagnostic laparoscopy

Laparoscopy may be performed on the neonatal intensive care unit under fentanyl anesthesia with muscle paralysis. High-frequency oscillatory ventilation and inhaled nitric oxide therapy do not preclude laparoscopy.

1 Following preparation of the abdomen, a 3 mm Hasson cannula is inserted in the right or left iliac fossa under direct vision and carbon dioxide pneumoperitoneum is established with a maximum pressure of 15 mmHg and maximal flow rate of 2 L/min. A 30° laparoscope is inserted and the intestine and intraperitoneal contents are inspected.



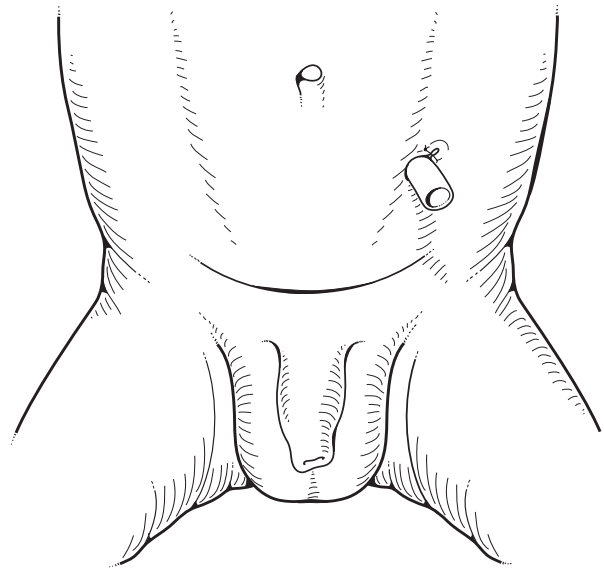
1

In cases in which there is perforation with peritoneal contamination, an additional incision may be made in the abdominal wall for the introduction of a peritoneal suction/irrigation device. If a drain is considered beneficial, a soft Penrose drain may be positioned in one of the port sites. Identification of frankly gangrenous intestine during laparoscopy may be considered an indication for laparotomy.

Primary peritoneal drainage

Primary peritoneal drainage can be performed at the cotside with the infant on the neonatal intensive care unit. The site of drain insertion is usually the left iliac fossa, although the right iliac fossa may also be considered if the lower border of the liver can clearly be palpated above the insertion site. The upper quadrants of the abdomen should be avoided to limit visceral damage.

2 With the infant sedated, local anesthetic is infiltrated into the skin and subcutaneous tissues. A small (0.5–1 cm) incision is made in the skin and blunt dissection used to expose first the fascia and then the peritoneum, which is carefully opened to avoid damage to the underlying bowel. Gas or meconium-stained fluid may be released from the peritoneal cavity and a microbiological swab should be taken to direct future antibiotic therapy. A soft drain such as a Penrose drain is inserted into the peritoneum and sutured in place to the skin to allow continued drainage of the peritoneal cavity. Stiff drains, including intravenous cannulae, are best avoided as they may perforate the intestine.

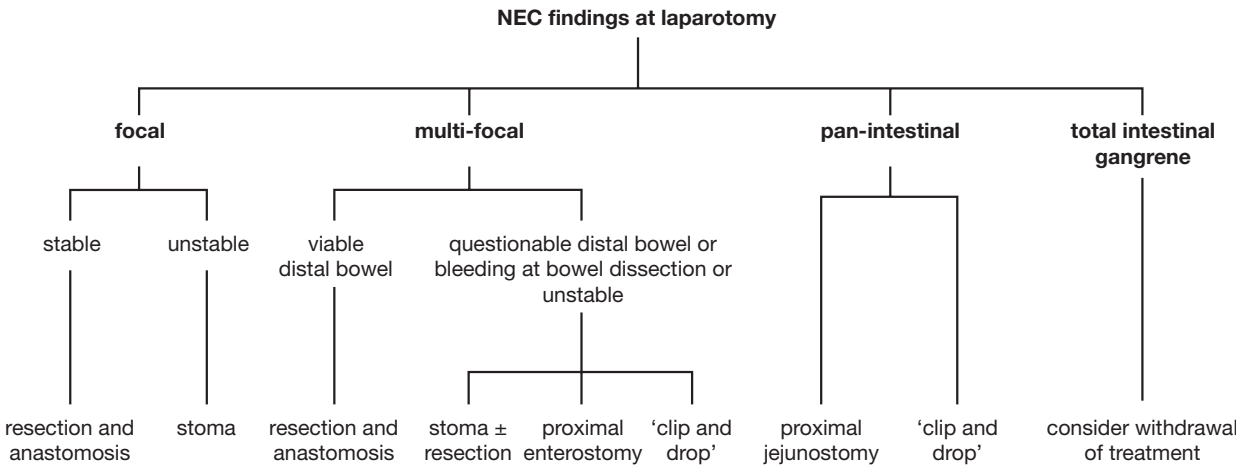


2

Laparotomy

The principal surgical objectives of laparotomy in acute NEC are to control sepsis, remove gangrenous bowel, and preserve as much bowel length as possible. Within these objectives, a number of options exist. The patient's weight and clinical status as well as the extent of the disease influence the choice of surgical procedure. At laparotomy, the extent of the disease can be classified as focal when it is limited to a single intestinal segment, multifocal if it includes two or more intestinal segments with more than 50 percent of the small intestine viable, and pan-intestinal when the majority of small and large bowel is involved with less than 25 percent viable bowel remaining. Laparotomy is performed under general anesthesia and may be performed on the neonatal intensive care unit or in the operating theater, depending on the stability of the infant and local practice and policy.

3 The authors' preferred approach to the operative management at laparotomy of infants with NEC is illustrated.



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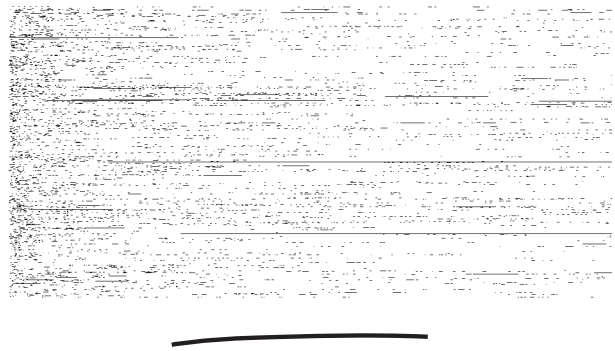


INCISION

4 A standard transverse supraumbilical incision is used for laparotomy. This may be slightly to the right side initially, allowing for further extension of the incision to the left side if necessary.

Care should be taken to limit damage to the thin anterior abdominal wall in extremely premature infants. The skin is incised and the underlying subcutaneous tissues and muscle can be safely divided with point diathermy, which has the added advantage of providing a degree of hemostasis. When entering the peritoneal cavity, particular attention should be given to the liver, which is often very large and extremely fragile in infants with NEC. Capsular liver damage should be avoided at all costs, as hemorrhage from any injury to the liver may have disastrous consequences and result in death.

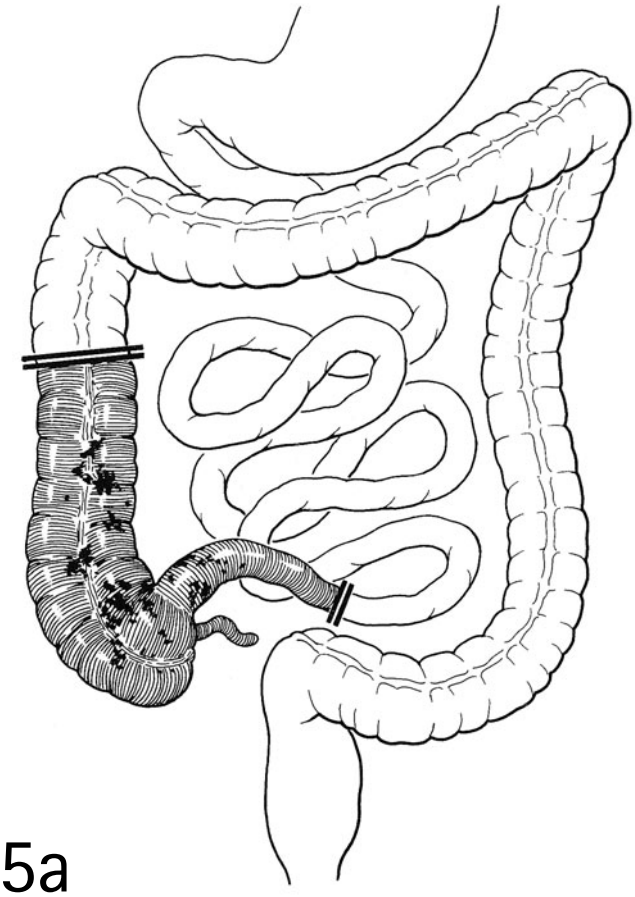
Operations for focal necrotizing enterocolitis

For infants with focal NEC, whether perforated or not, and for infants with a focal intestinal perforation with no evidence of NEC, there are two surgical options at laparotomy, namely resection followed by primary anastomosis and resection followed by stoma formation. Traditionally, the recommended surgical approach has been to perform resection followed by stoma formation. However, resection followed by primary anastomosis has been gaining popularity in recent years. It has the advantage of restoring intestinal continuity in one operation and avoids the potential complications associated with stoma formation. However, there is a risk of anastomotic leakage or stenosis with primary anastomosis. Infants selected for primary anastomosis should be stable during the perioperative period, and the resection margins and remaining intestine should be healthy with good perfusion. Neither the weight of the child nor the presence of peritoneal contamination secondary to intestinal perforation affects the decision of whether to perform primary anastomosis or to fashion a stoma.

RESECTION WITH PRIMARY ANASTOMOSIS

The standard laparotomy incision in used. The affected segment of intestine is delivered through the wound and placed on povidone-iodine-soaked gauze to minimize the risk of further peritoneal contamination.

5 The affected segment along with its associated mesentery is resected, ensuring hemostasis of the mesenteric vessels, which can be satisfactorily achieved using bipolar diathermy. A single-layer extramucosal, seromuscular anastomosis is performed using an appropriately sized (4/0–6/0 in the premature neonate) interrupted monofilament suture. The first two stitches are placed on opposite sides of the intestine to give stability. The anastomosis is then completed, laying the knots on the outside of the lumen. Finally, the mesenteric defect is closed. In instances in which there is a discrepancy in the circumference of the two ends to be anastomosed, it may be necessary to cut one of the ends at an angle to increase the circumference for anastomosis.



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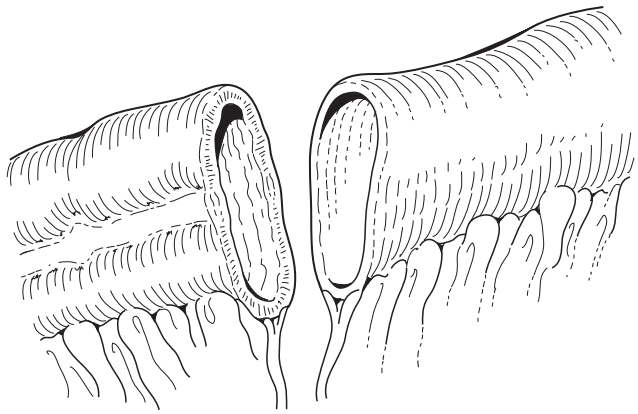

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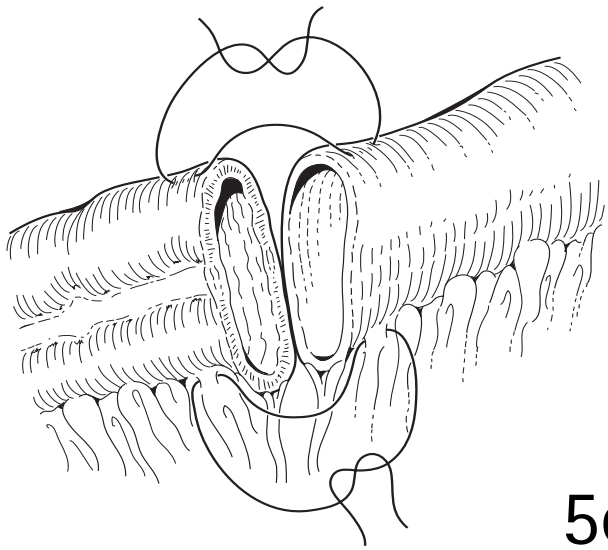

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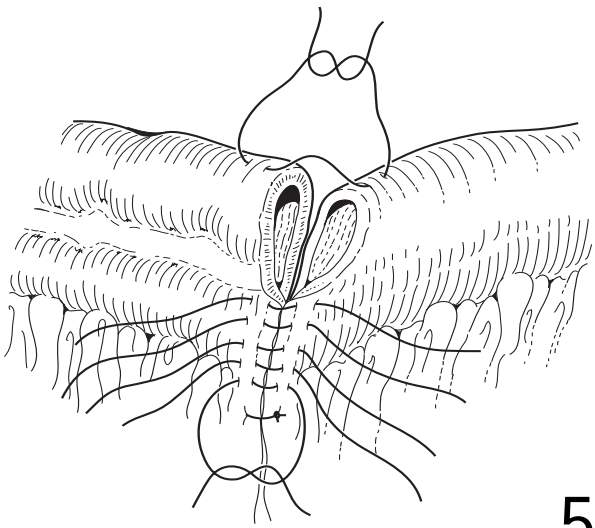
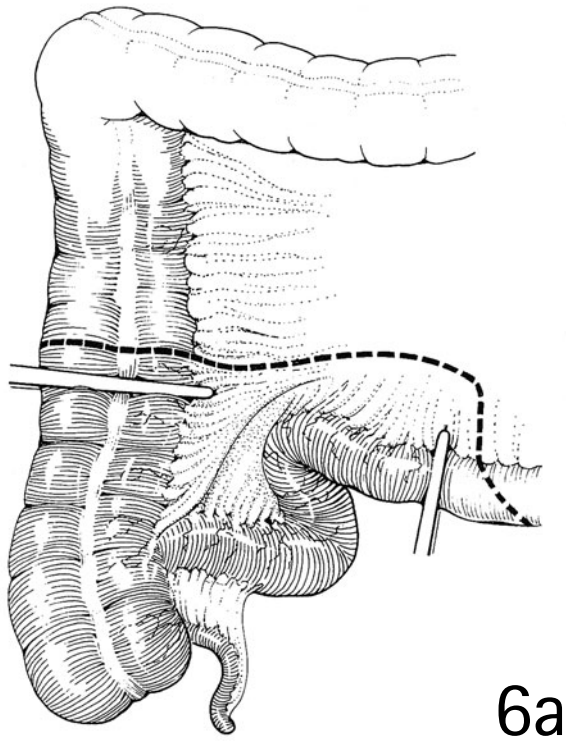


5d

Providing an adequate length of intestine remains, primary anastomosis may be used following resection of any part of the intestinal tract from the jejunum to the sigmoid colon, including cases of total colonic NEC requiring total colectomy and ileorectal anastomosis.



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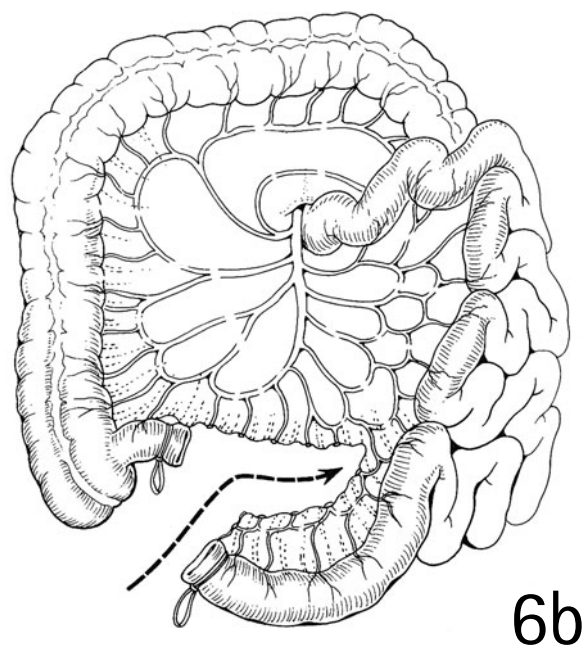
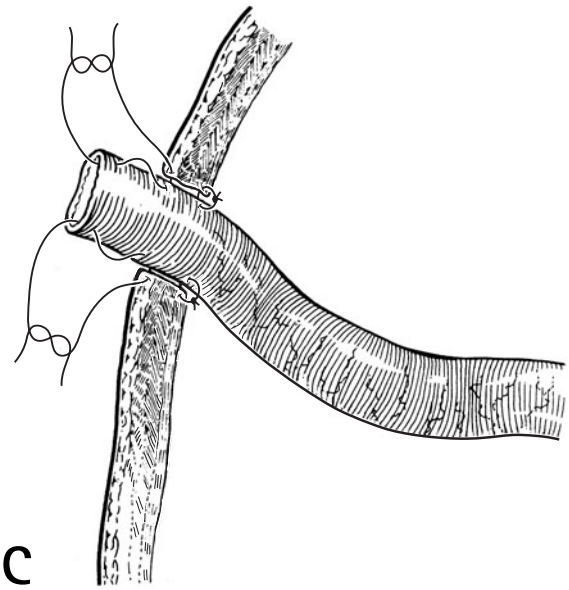


6b

RESECTION AND STOMA FORMATION

6a–e In infants with focal NEC who are too unstable to undergo the procedure of primary anastomosis, or when the resection margins or remaining intestine are of doubtful viability, or when it is not possible to ascertain the condition of the intestine distal to the resected region, a stoma is formed following resection. The stoma is fashioned into one end of the abdominal incision unless the mobility of the mesentery is inadequate, in which case the stoma may be positioned anywhere on the anterior abdominal wall. Whenever possible, a mucous fistula is also fashioned next to the proximal stoma. Wide separation of the proximal stoma and mucous fistula is to be avoided, as this would require a full laparotomy rather than a more limited procedure when restoring intestinal continuity. When it is not possible to bring the distal resection margin to the wound, it may be clipped or over-sewn.



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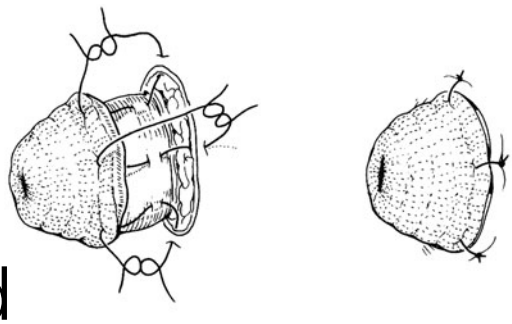

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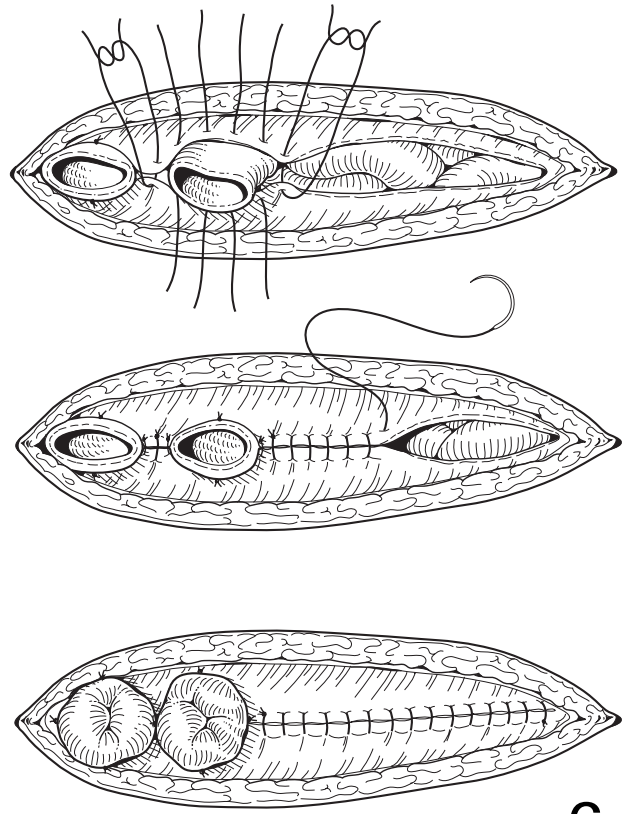


6e

When fashioning the stoma, a limited number of sutures is used to secure the intestine to the anterior abdominal wall to avoid compromising the blood supply. It is not mandatory to secure the serosal to the fascial layer of the abdominal wall, although some surgeons believe that this reduces the risk of stomal prolapse. Similarly, the choice of whether to evert the mucosa as a mature stoma is left to the individual. The mucous fistula is attached to the skin in a similar fashion, but may be left flush with the wound. When the stoma is fashioned away from the main incision and through a separate 'stab wound' type of opening, care must be taken when delivering the bowel through the anterior abdominal wall to ensure that the tract is neither too small (which may lead to stenosis) nor too large (which may predispose to prolapse) and that the bowel passes through en masse without stripping of the serosa.

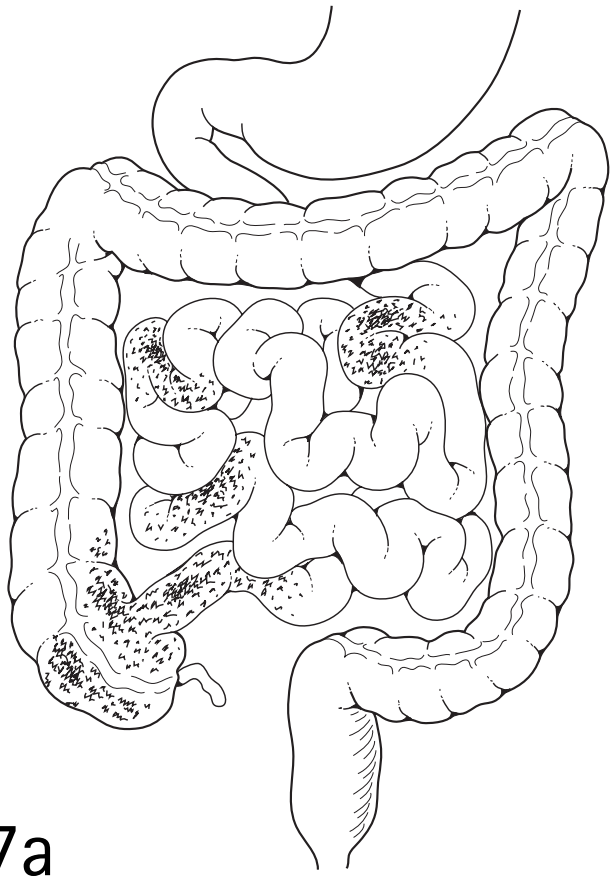
Operations for multifocal necrotizing enterocolitis

When there are multiple separate segments of diseased intestine separated by lengths of healthy bowel, there are a number of surgical options at laparotomy, depending on the overall condition of the infant, the viability of any potential resection margins, and the mobility of the remaining intestine and mesentery. Of prime concern in such cases is the excision of all gangrenous intestine whilst preserving as much bowel length as possible. Measuring the remaining healthy intestine may be used as a guide to expected outcome. In a stable child with minimal peritoneal soiling and healthy resection margins, it is possible to perform multiple resections and multiple primary anastomoses. However, we advise against performing more than two anastomoses due to the increased risk of complications.

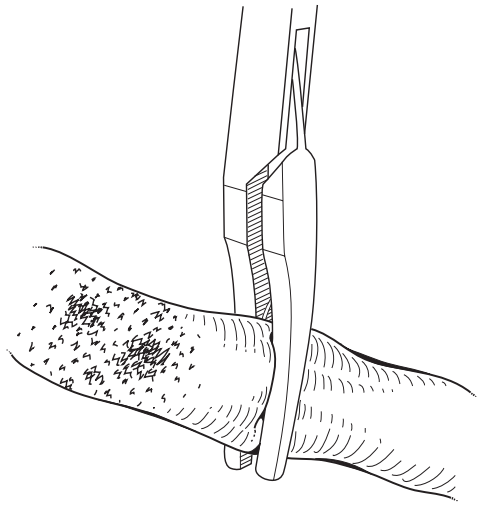
Stoma formation may also be used in such cases of multifocal disease. If this approach is selected, the most proximal resection margin should be brought out as a stoma. In some cases of multifocal disease, a combination of stoma formation and primary anastomosis may be most appropriate. Resection margins suitable for primary anastomosis may be joined and areas of more doubtful viability may be exteriorized as a stoma. Similarly, if an anastomosis has been performed following resection of one segment of diseased intestine but it is not possible to determine the viability of the

colon, a stoma may be formed below a primary anastomosis. This approach is preferable to the formation of multiple stomas and requires the closure of the stoma rather than a full laparotomy to restore intestinal continuity at a later stage.

In addition, there are two other operations described specifically for use in the presence of multifocal disease, both of which have the aim of preserving as much bowel length as possible.



7a



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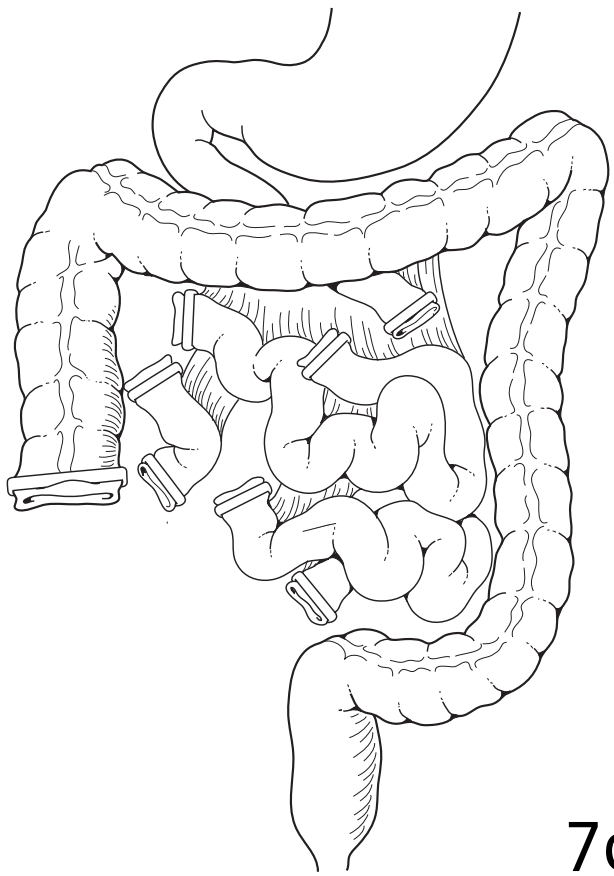


7c

'CLIP AND DROP' OPERATION

7a–c This technique allows for removal of gangrenous intestine and also avoids stoma formation. All segments of grossly non-viable or perforated bowel are resected and the peritoneal cavity irrigated. The ends of remaining bowel are clipped using Ligaclips and returned to the abdomen. This is followed by a second-look laparotomy, ideally with anastomosis, not longer than 72 hours later. This procedure may be combined with stoma formation as appropriate.

PATCH, DRAIN, AND WAIT

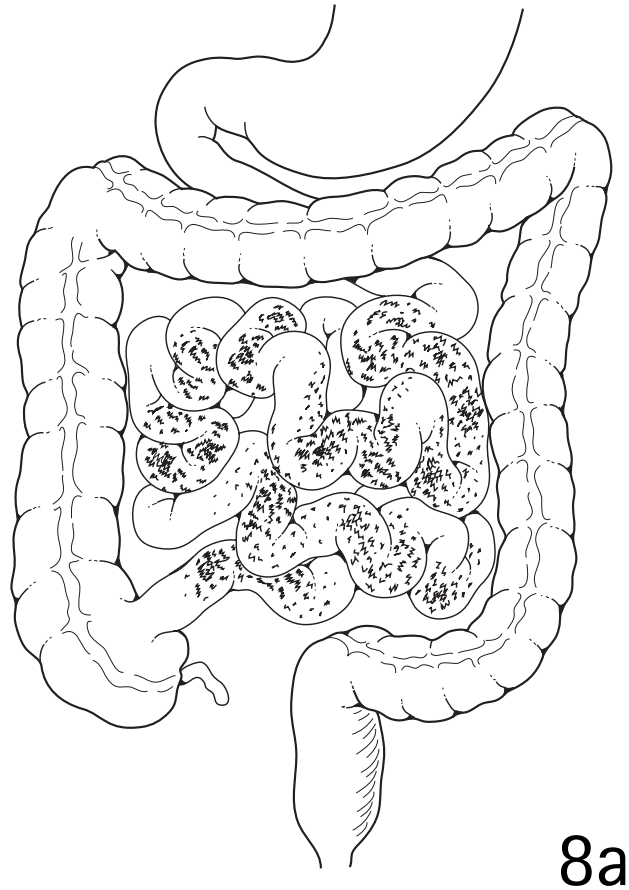
The basic principle of this technique, reported by Moore, is to resect no bowel and do no enterostomies. At laparotomy, areas of intestinal perforation are patched closed and bilateral Penrose drains are inserted running from the undersurface of the diaphragm into the pelvis, with exit sites in both lower quadrants. After at least 14 days, a second laparotomy is performed to examine the outcome of this procedure and the need for further surgery. The authors have no experience with this technique, and other centers have not reported its use in the literature.

Operations for pan-intestinal necrotizing enterocolitis

The techniques described thus far are of particular use for the infant with one or more short segments of NEC. Infants with NEC affecting a large proportion of the gastrointestinal tract pose a particularly difficult problem, and treatment of this group remains controversial. The surgical principles in these children are difficult if not impossible to fulfill. Due to the length of bowel involved, it is often not possible fully to remove all gangrenous intestine whilst salvaging adequate length for sustainable life. It is for these reasons that in the infant with pan-intestinal NEC who is unstable and critically ill, some surgeons would perform an 'open and close' laparotomy, ascertaining the futility of further treatment with subsequent withdrawal of therapy. However, when there is doubt, a number of techniques may be utilized with the aim of allowing time for stabilization of the infant's general condition and the chance of some healing of the gastrointestinal tract to occur. Due to the severity of the disease, the mortality in these cases remains high regardless of the surgical approach. The procedures available are the 'clip and drop' and 'patch, drain, and wait' approaches, as described above, and the formation of a proximal defunctioning jejunostomy.

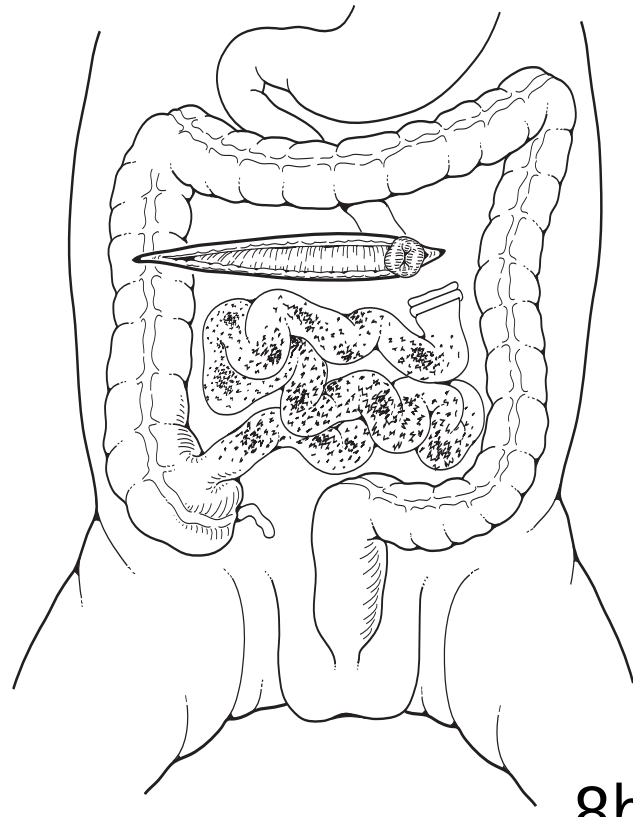
PROXIMAL JEJUNOSTOMY

Surgical creation of a proximal jejunostomy in the presence of pan-intestinal disease allows decompression and defunctioning of the diseased intestine but does not remove gangrenous segments and may permit continued bacterial translocation. This procedure is useful in neonates with NEC affecting the majority of the intestine, but the high morbidity and mortality rates should be carefully considered.



8a

8a,b The jejunum is divided proximally to the most diseased section of intestine and the distal resection margin over-sewn. The proximal margin is exteriorized as a stoma as previously described, either through the end of the wound or at a separate site.



8b

Once the infant is more stable, the intestine has had a chance to recover from the initial insult, and the infant has had the opportunity to grow, a second laparotomy is performed. A definitive procedure is undertaken and the stoma closed. A specific complication of forming a high jejunostomy is the risk of massive stomal output resulting in fluid and electrolyte imbalance. This may expedite the need for second laparotomy and stomal closure.

POSTOPERATIVE CARE

The postoperative care of infants with NEC is invariably on the neonatal intensive care unit with continuous monitoring. Usually respiratory and sometimes cardiovascular support is necessary. A systemic inflammatory response often develops in response to either NEC or surgery (or a combination of both), and failure of one or more organ systems may ensue, requiring appropriate intervention. Sepsis is a common problem during the postoperative period despite the routine use of broad-spectrum antibiotics for 10 days. Feeds are reintroduced 10 days following surgery, using a cautious regime and increasing as tolerated.

Intestinal stricture

Infants who do not tolerate feeds should be investigated for the presence of an intestinal stricture. Following anastomosis, a stricture at the anastomotic site should also be considered. In suspected cases, a gastrointestinal contrast study may prove the diagnosis. Treatment is by laparotomy, with resection of the strictured intestine and primary anastomosis.

Closure of stoma

Once the infant is thriving and has achieved satisfactory weight gain, any stoma may be closed and intestinal continuity restored. Typically this is after a period of approximately 2–4 months. Prior to closing any stoma, a contrast study should be undertaken of the distal intestine to ensure the absence of intestinal stricture requiring resection. The stoma is closed by performing an anastomosis between the two ends of bowel in an identical fashion to that described for primary anastomosis in the setting of acute NEC.

OUTCOME

Necrotizing enterocolitis appears to be associated with a significant long-term morbidity, although the long-term outcome for infants with NEC is poorly reported. The most serious gastrointestinal complication of NEC is short bowel syndrome, with an incidence of up to 23 percent in NEC survivors. Neurodevelopmental implications of NEC have not been fully investigated. Only approximately 50 percent of the neonates with NEC are neurodevelopmentally normal. However, the neurological sequelae in these children appear to be related to underlying prematurity and other complications thereof rather than to NEC itself.

The mortality rate of neonates with NEC depends on the severity of the disease, associated anomalies, weight, and gestational age. Overall mortality in a study of 83 neonates from our institution requiring laparotomy for NEC was 30 percent. The acute mortality rate was higher (67 percent) in patients with pan-intestinal involvement of the disease compared to patients with multifocal NEC (30 percent) or focal disease (12 percent). Causes of death included multisystem organ failure ($n = 10$), sepsis ($n = 14$), and congenital cardiac abnormality ($n = 1$). In a recent study of 51 infants with NEC weighing less than 1000 g, including 44 who underwent surgery, outcome did not appear to be related to surgical procedure performed. However, the mortality in this group of extremely small infants was 27 percent during the acute illness and 49 percent at a median follow-up of 24 months. While the surgical outcome of treatment for NEC may be successful in the majority of cases, the long-term morbidity and mortality associated with such premature infants should not be overlooked.

FURTHER READING

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50

Anorectal malformations

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HISTORY

Anorectal malformations have been described for centuries. Previously, most children with these malformations received an operation consisting of the creation of an orifice on the perineum. With this simple procedure, many children survived, probably because the rectum was located very close to the skin. However, many died, probably because the rectum was located high in the pelvis. In 1835, Amussat reported, for the first time, suturing of the rectal wall to the skin edges, which could be considered the first anoplasty.

For many years, surgeons performed a perineal operation, without a colostomy, for the so-called low malformations. High imperforate anus, on the other hand, was usually treated with a colostomy performed during the neonatal period, followed by an abdominoperineal pull-through sometime later in life. The specific recommendation was often to pull the intestine as close to the sacrum as possible to avoid trauma to the genitourinary tract. Stephens performed the first objective anatomic studies of human specimens with these defects, and in 1953 proposed an initial sacral approach to separate the rectum from the urinary tract and preserve the puborectalis sling (considered a key factor in maintaining fecal continence). He also suggested opening the abdomen, if necessary, after the sacral approach. Following Stephens' recommendations, several different surgical techniques were proposed. The common denominator in all these techniques was the protection and utilization of the puborectalis sling. In 1980, a new approach, the posterior sagittal anorectoplasty, allowed direct exposure of this important anatomic area. With this approach it became possible to correlate the external appearance of the perineum with the operative findings, and subsequently the clinical results. The approach has implications for understanding the anatomy of these defects, terminology, classification, and most importantly treatment.

PRINCIPLES AND JUSTIFICATION

Incidence

Anorectal malformations occur in 1 in 4000 neonates, slightly more commonly in boys than in girls. The most common defect in girls is a rectovestibular fistula followed by a rectoperineal fistula. Contrary to what is claimed in most of the published literature, girls with rectovaginal fistulas are rare. Most of the 'rectovaginal fistulas' reported in the literature are probably cases of misdiagnosed cloacas or rectovestibular fistulas. Therefore, the third most common defect in girls is persistent cloaca. The most common defect in boys is a rectourethral fistula, followed by a rectoperineal fistula. Rectobladderneck fistulas in boys represent 10 percent of the entire group of defects. Imperforate anus without fistula in both boys and girls is unusual and represents only 5 percent of the entire group of defects. The estimated risk of having a second child with an anorectal malformation is approximately 1 percent and four times that if the first child is a girl with rectoperineal fistula.

Classification

The classification shown in Box 50.1 is proposed because it is therapeutically oriented.

Box 50.1 *Classification of anorectal malformations*

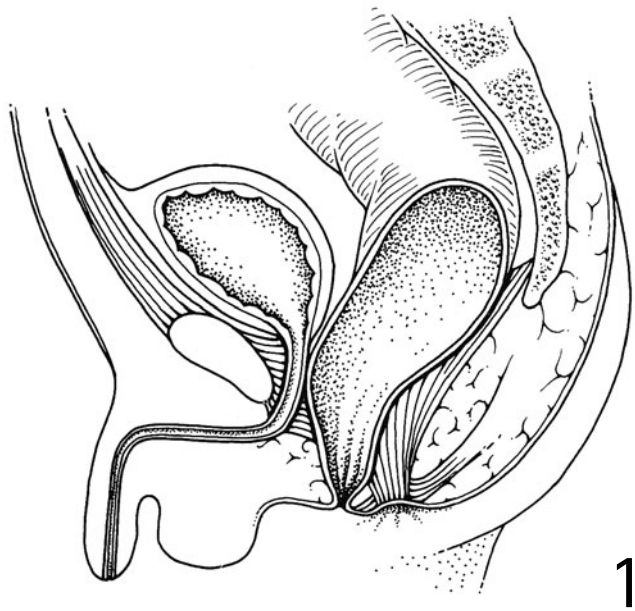
Boys
Perineal (cutaneous) fistula
Rectourethral fistula
  Bulbar
  Prostatic
Rectobladderneck fistula
Imperforate anus without fistula
Rectal atresia

Girls
Perineal (cutaneous) fistula
Vestibular fistula
Persistent cloaca
Imperforate anus without fistula
Rectal atresia

BOYS

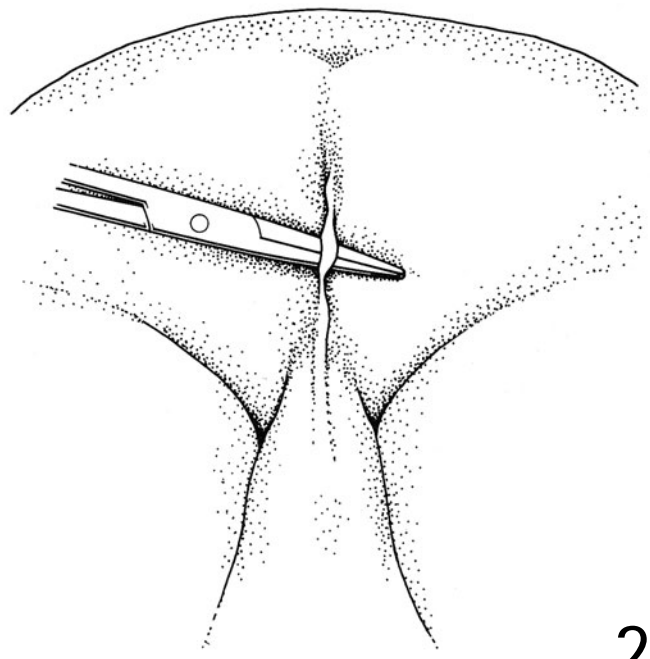
PERINEAL (CUTANEOUS) FISTULA

This type of defect is also known as a low imperforate anus. The rectum is located within most of the sphincter mechanism. Only the lowest part of the rectum is anteriorly displaced.



1

2,3 Sometimes the fistula follows a subepithelial midline tract opening along the midline perineal raphe, scrotum, or penis. The perineal findings in this kind of defect include a prominent skin tag, below which an instrument can be passed, known as a 'bucket-handle' malformation (Illustration 2), a black or white, ribbon-like, midline structure that represents a subepithelial fistula filled with meconium, or a very well-formed anal dimple suggesting the presence of a very low defect (Illustration 3). The diagnosis is established by perineal inspection. No further investigations are required.



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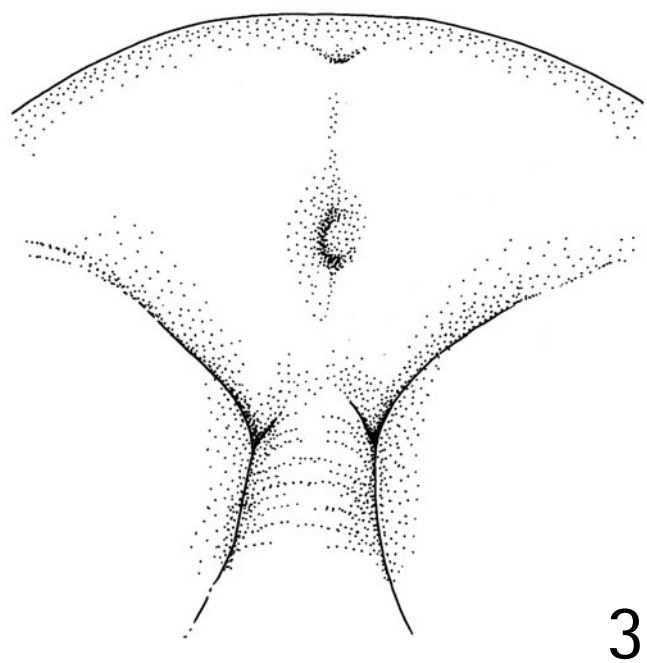
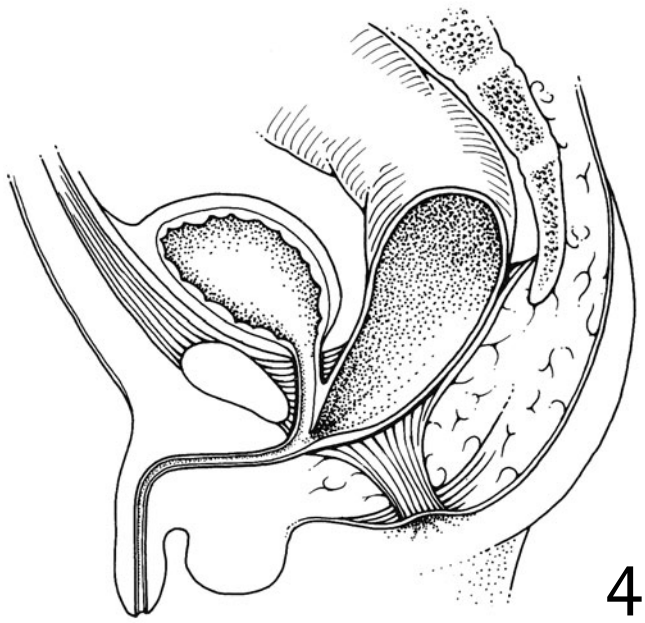


3

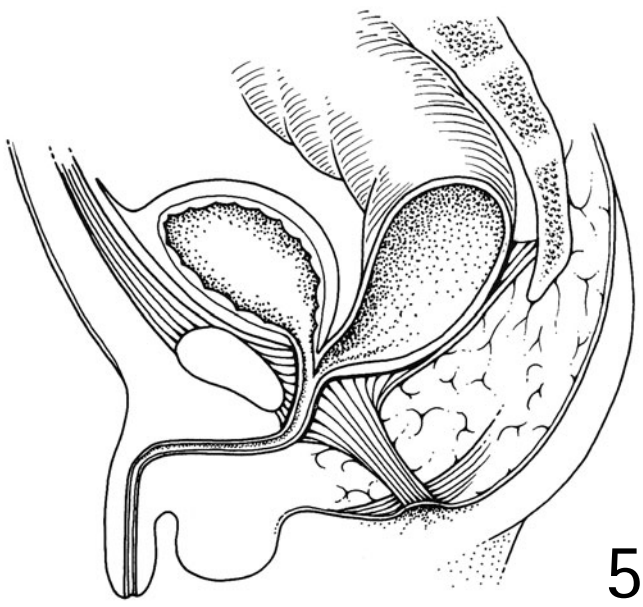
RECTOURETHRAL FISTULA

This is the most common defect in boys.



4

4,5 The rectum may communicate with the lower part of the urethra (bulbar urethra) or with the upper urethra (prostatic urethra). Immediately above the fistula site, the rectum and urethra share a common wall with no plane of dissection.

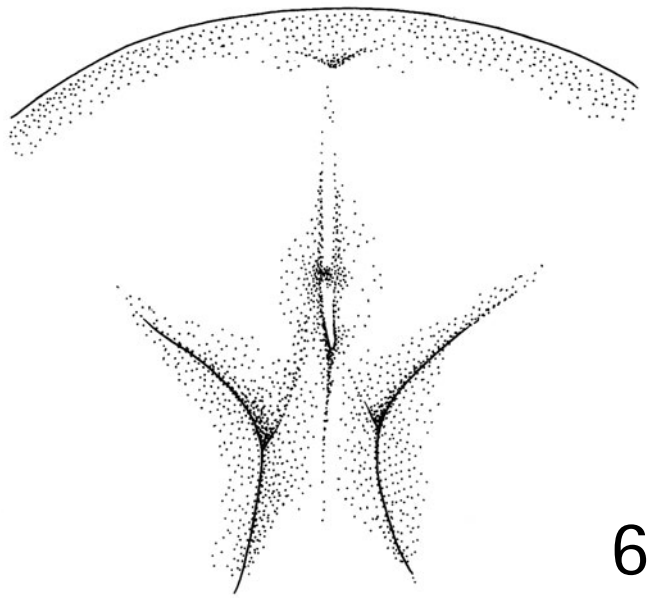


5

This anatomic fact has important technical and surgical implications.

The rectum is surrounded laterally and posteriorly by the levator muscle mechanism. Between the end of the rectum and the perineal skin, there is a portion of striated voluntary muscle called the 'muscle complex'. The contraction of the levator muscle pushes the rectum forward. The contraction of the muscle complex elevates the skin of the anal dimple. At the level of the skin, and located on both sides of the midline, there is a group of voluntary muscle fibers called parasagittal fibers.

6 Patients with rectourethral bulbar fistulas usually have a normal sacrum and a 'good-looking perineum' consisting of a prominent midline groove.



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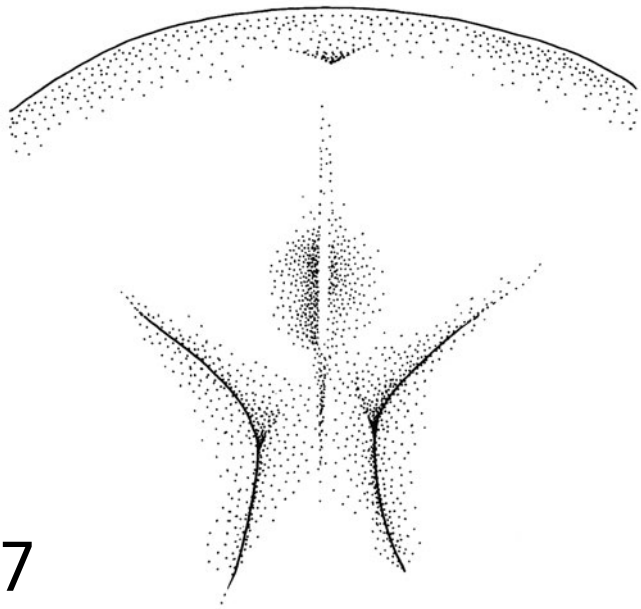
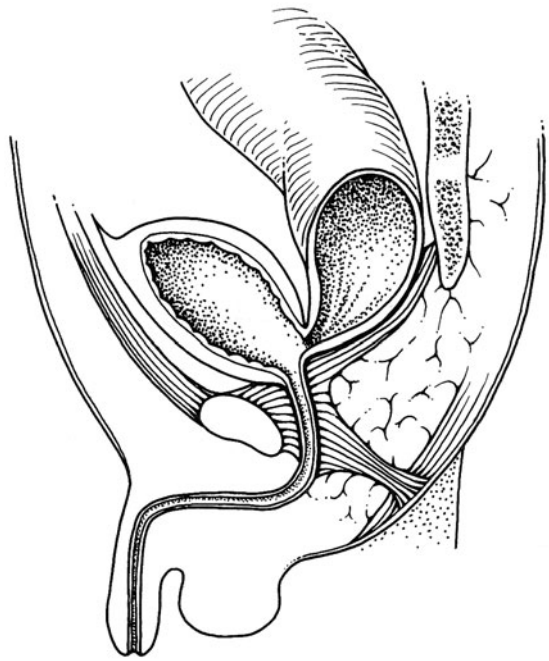


7

7 Patients with rectoprostatic fistulas tend to have a higher incidence of an abnormal sacrum, underdeveloped sphincter mechanism, and flat perineum. The external sphincter (anal dimple) is often located very close to the scrotum. Exceptions exist, however. Neonates with rectourethral fistulas may pass meconium through the urethra, usually after 20 hours of life, which is an unequivocal sign of rectourethral fistula.

RECTOBLADDERNECK FISTULA

8 In these malformations, the rectum communicates with the urinary tract at the bladderneck. Levator muscle, muscle complex, and parasagittal fibers are often poorly developed. The sacrum is often deformed or absent. The entire pelvis seems to be underdeveloped, and its anteroposterior diameter seems to be foreshortened. The perineum is usually flat (see Illustration 7). For all these reasons, the prognosis for bowel function is poor.



8

IMPERFORATE ANUS WITHOUT FISTULA

In these cases the rectum is completely blind and is almost always found at the same level as in cases with rectourethral bulbar fistula. The sacrum and sphincteric mechanism are usually normal, and therefore these patients have a good prognosis. This is a common malformation in patients with Down's syndrome.

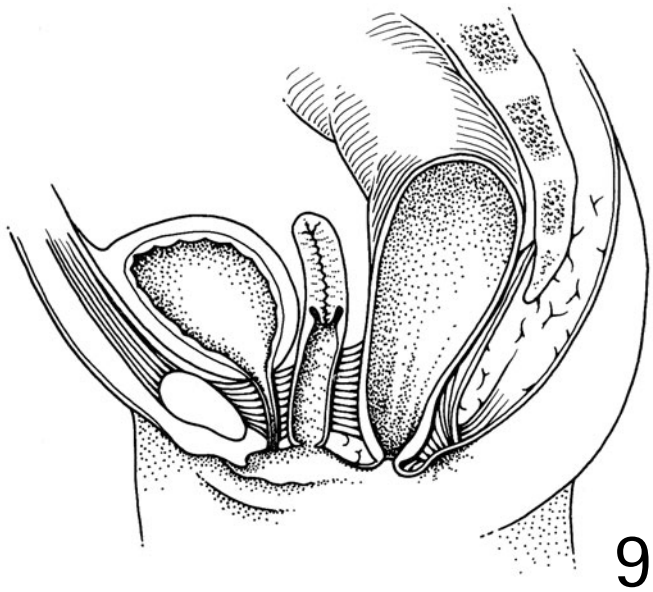
RECTAL ATRESIA

This is a very unusual defect; occurring in only 1 percent of cases. These are the only patients with imperforate anus who are born with a normal anal canal. Externally the anus looks normal, and the malformation is often discovered during an attempt to take a rectal temperature or after the onset of symptoms and signs of low intestinal obstruction. About 2 cm from the anal verge there is an atretic or stenotic area. The upper blind rectum is usually located very close to the anal canal. The sacrum is normal, the sphincteric mechanism is excellent, and therefore the prognosis is good.

GIRLS

PERINEAL FISTULA

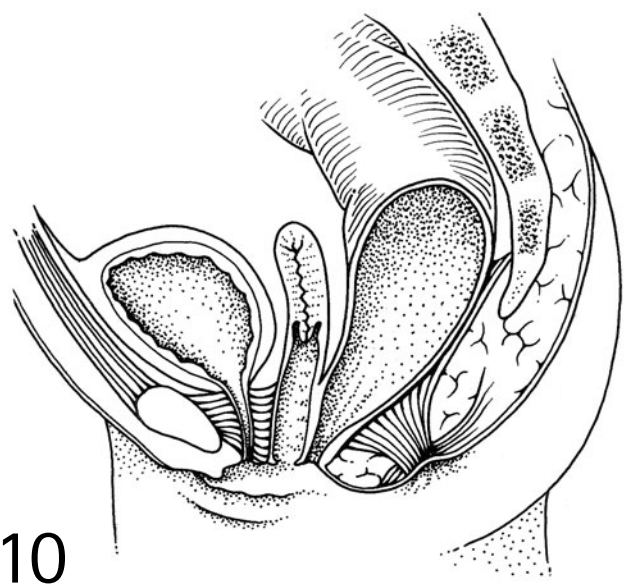
9 This defect is equivalent to the perineal fistula described for boys. The rectum and vagina are well separated. The sphincteric mechanism is very good, and therefore the prognosis is also good.



9

VESTIBULAR FISTULA

This is the most common defect seen in girls. It has an excellent functional prognosis. Unfortunately, this is the most common type to suffer a failed repair.



10

10 The intestine opens in the vestibule of the female genitalia immediately posterior to the hymen. The most pertinent anatomic characteristic of this defect is that immediately above the fistula site the rectum and vagina share a very thin common wall. These patients usually have good muscles and a normal sacrum. The diagnosis is established by perineal inspection. These patients are commonly mislabeled as having rectovaginal fistula, which only reflects an imprecise inspection of the newborn genitalia.

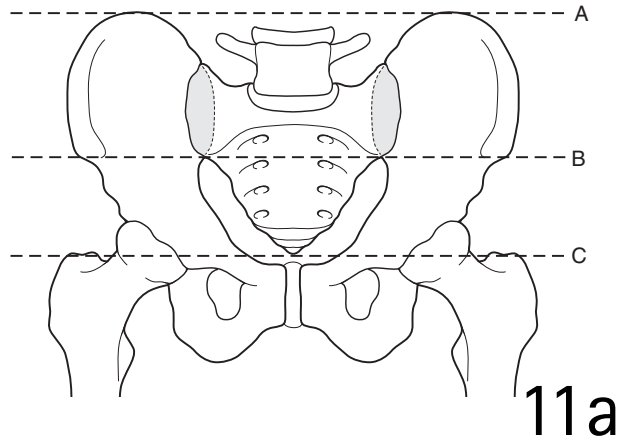
IMPERFORATE ANUS WITHOUT FISTULA AND RECTAL ATRESIA

These defects in girls have the same anatomic characteristics as those described in boys, and therefore have similar prognostic implications.

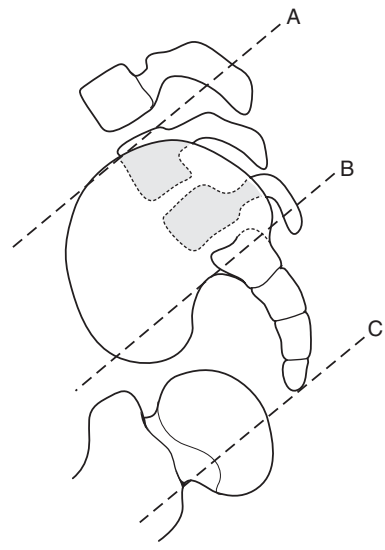
ASSOCIATED DEFECTS

SACRUM AND SPINE

The sacrum is often abnormal in these types of malformations. There appears to be a very good correlation between the degree of sacral development and the final functional prognosis. Traditionally, the number of vertebrae has been the most useful criterion for evaluation of the sacrum. One missing vertebra does not have important diagnostic implications. More than two absent vertebrae represent a poor prognostic sign.

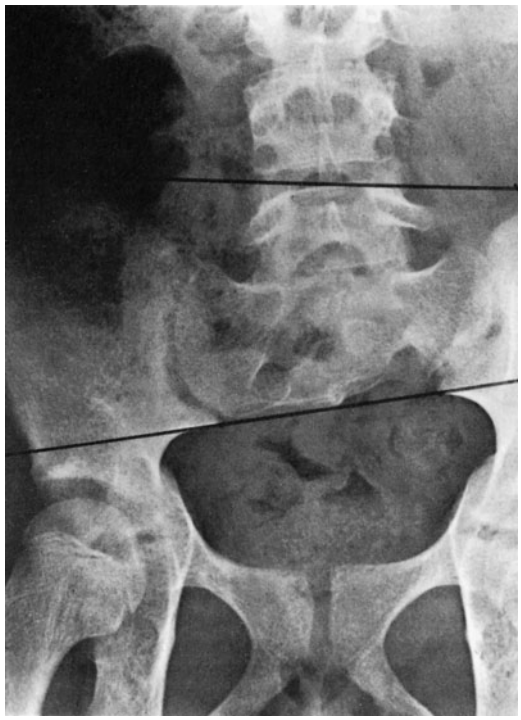


11a



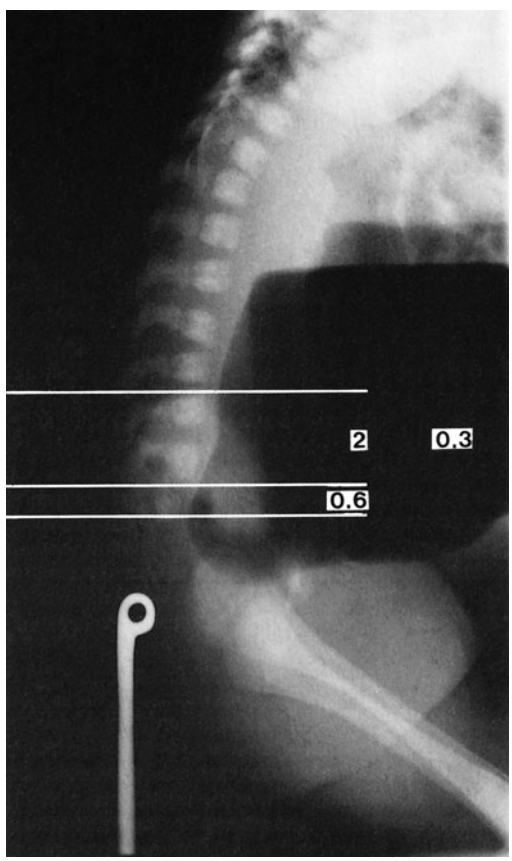
11b

11a,b In order to improve the prognostic accuracy based on sacral abnormalities, a sacral ratio was created that expresses the degree of sacral development. For this measurement, three lines are drawn: line A extends across the uppermost portion of the iliac crests; line B joins both inferior and posterior iliac spines; and line C runs parallel to lines A and B and passes through the lowest radiologically visible sacral point. In 100 normal children the ratio of the distances BC:AB was between 0.7 and 0.8 in both anteroposterior and lateral projections. Children with anorectal malformations suffer from different degrees of sacral hypodevelopment, with the ratio varying between 0 and 0.8. A ratio of less than 0.4 usually signifies a poor functional prognosis.



12a

12a,b Two different examples of sacral abnormalities and poor ratios.



12b

Higher spinal abnormalities include hemivertebrae located in the lumbar or thoracic spine. The prognostic implications of these types of defects in terms of bowel and urinary control are not known.These patients often need treatment for scoliosis.

UROGENITAL DEFECTS

The frequency of associated urogenital defects varies from 25 percent to 50 percent. The reported variation may reflect the accuracy and thoroughness of the urologic investigations in different institutions. In the authors' series, 48 percent of the patients (55 percent of girls; 45 percent of boys) had associated urologic anomalies. Patients with persistent cloaca or rectovesical fistulas have a 90 percent chance of having significant associated urologic abnormality. Children with minor defects (perineal fistula) have less than a 10 percent chance of suffering from an associated urologic defect. The most common urologic malformation associated with imperforate anus is absent kidney, followed by vesicoureteric reflux. Hydronephrosis, urosepsis, and metabolic acidosis from poor renal function represent the main sources of mortality in neonates with anorectal malformations. Patients with anorectal malformations should have an ultrasonographic study of the abdomen during the first 24 hours after birth, and if this study shows some abnormalities, a thorough urologic evaluation is indicated.

OTHER DEFECTS

Other congenital malformations are commonly associated with anorectal malformations, including esophageal atresia, duodenal atresia, and cardiovascular defects.

Management of anorectal malformations during the neonatal period

Two important questions must be answered during the first 24 hours of life: what are the associated anomalies, and what operation is required, a newborn pull-through or a colostomy?

BOYS

The decision-making algorithm used by the author for the management of newborn males with anorectal malformations is shown in Figure 50.1. Associated malformations must be investigated. In more than 80 percent of boys, perineal inspection and urinalysis provide enough clinical evidence to make a clinical diagnosis. If a perineal fistula exists, the patient may be treated with a minimal posterior sagittal anorectoplasty in the newborn period. The presence of a flat bottom and the demonstration of meconium in the urine are an indication for a diverting colostomy. The colostomy decompresses the intestine in the neonatal period, provides access for a contrast study to define the anorectal anatomy and will subsequently provide protection against infection during the healing process after the main repair.

It is important to wait 20–24 hours before making a decision, for these patients do not show abdominal distension during the first few hours of life. Even if a perineal fistula is present, meconium is not usually seen on the perineum until 20–24 hours after birth. In fact, a significant amount of intra-abdominal pressure is required for the meconium to force its way through a perineal or urinary fistula. A significant amount of intraluminal rectal pressure is required to reach a level high enough to overcome the voluntary muscle tone that keeps the most distal part of the rectum compressed. It must be remembered that, in most cases of anorectal malformation, the most distal part of the rectum is surrounded by a striated muscle mechanism that keeps the rectum collapsed (see Illustrations 1, 4, and 5). To distend that most distal part of the rectum, it is necessary to exert significant intraluminal pressure. Radiologic evaluations performed during the first hours of life are, therefore, unreliable.

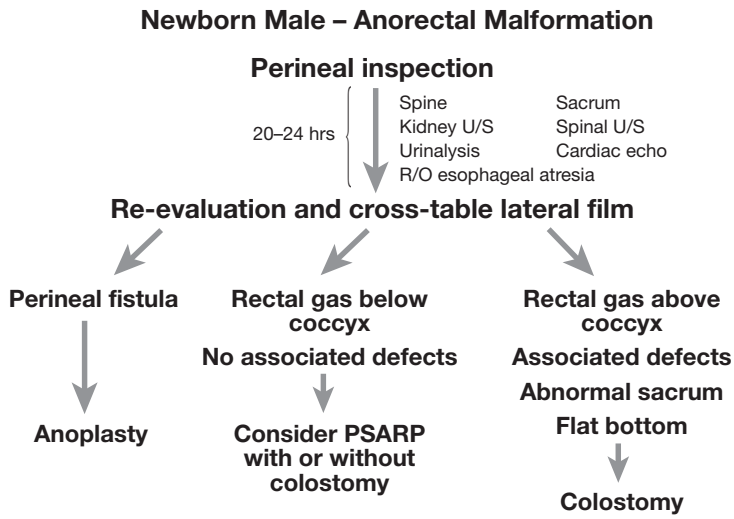


Fig. 50.1 Decision-making algorithm for the management of newborn boys with anorectal malformations. PSARP; (Pull-through) ileostomy ureteroileostomy.

Shortly after birth, intravenous fluids must be administered. A nasogastric tube is inserted to keep the stomach decompressed and thus avoid the risk of vomiting and aspiration. Antibiotics are administered and an ultrasonographic study of the abdomen is performed to rule out the presence of other anomalies (mainly urologic). A piece of gauze is placed on the tip of the penis, and the nurses are then instructed to check for particles of meconium filtered through this gauze.

If after 20–24 hours of observation there is no clinical evidence indicating the need for a colostomy or a perineal operation, the patient must have a radiologic evaluation. A cross-table lateral film with the patient in the prone position helps to determine the position of the rectal pouch. The anal dimple is marked with radio-opaque material. If the rectum is visible below the coccyx, the patient can undergo a primary newborn repair, provided the surgeon is experienced with this technique. If the image is questionable, it is preferable to construct a diverting colostomy.

After recovering from the colostomy, the patient is discharged from the hospital. If the patient is growing well and has no other associated defects (cardiovascular or gastrointestinal) that require treatment, he is re-admitted at 4–8 weeks of age for a posterior sagittal anorectoplasty. This early repair can be performed safely only if the surgeon has experience in dealing with the delicate anatomy of the infant.

Performing the definitive repair at 1 month of age has important advantages for the patient, including less time with an abdominal stoma, less size discrepancy between proximal and distal intestine at the time of colostomy closure, simpler anal dilatation, and no recognizable psychologic sequelae from painful perineal maneuvers. In addition, at least theoretically, placing the rectum in the right location early in life may represent an advantage in terms of acquired local sensation.

Some surgeons have proposed a primary repair of all anorectal malformations during the neonatal period without a protective colostomy. There is no question that this can be done and that it has the potential of avoiding the morbidity related to the formation and closure of a colostomy. However, the disadvantages that the anatomy of the neonate is not as well defined as in older patients. Also, the diagnostic tests used to determine the level of the defect are not accurate enough, and the surgeon is actually subjecting the patient to a blind exploration of the perineum. If the rectum is located high in the abdomen, the surgeon may damage other structures during the search for the rectum. Such structures include the posterior urethra, seminal vesicles, vas deferens, and ectopic ureters. In addition, there is a risk of dehiscence and infection because the stool is not diverted.

FEMALES

A decision-making algorithm for the initial management of females is shown in Figure 50.2. Perineal inspection usually provides more information in girls than in boys. The principle of waiting 20–24 hours before making a decision is again valuable.

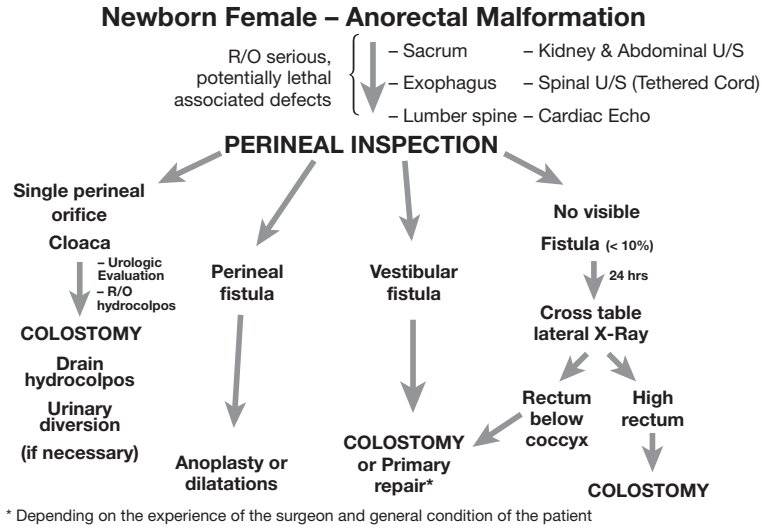
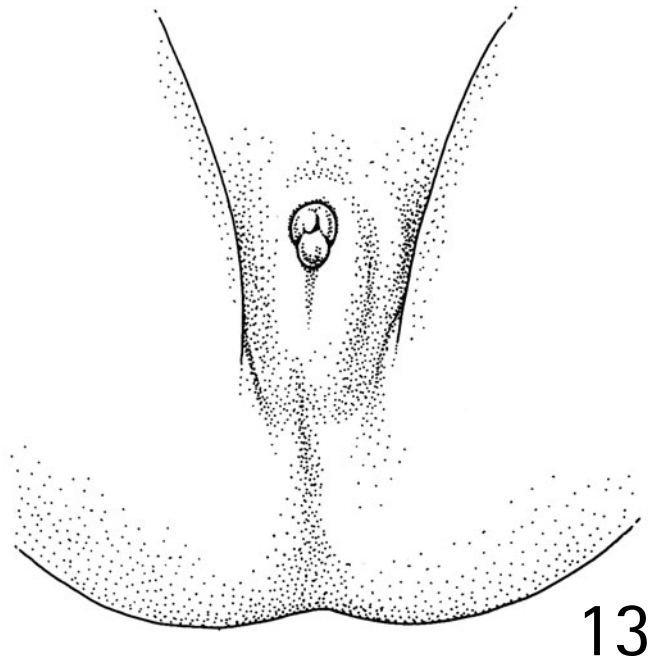


Fig. 50.2 Decision-making algorithm for the management of newborn girls with anorectal malformations.

13 The presence of a single perineal orifice is pathognomonic for a cloaca. Because of their complexity, these defects are dealt with separately in Chapter 51.



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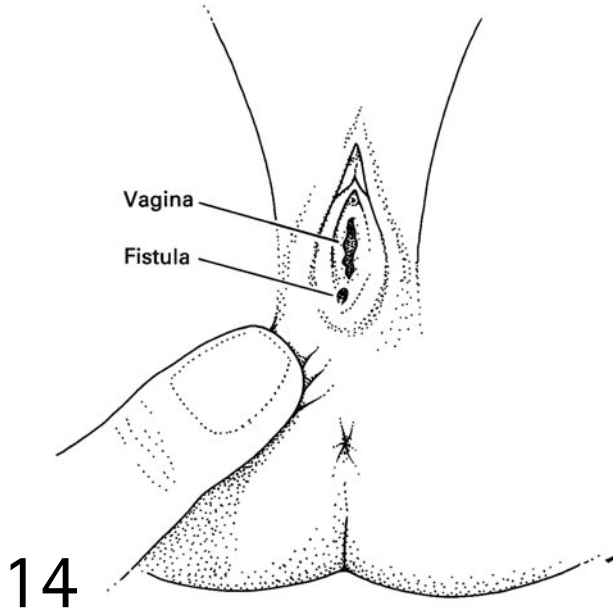


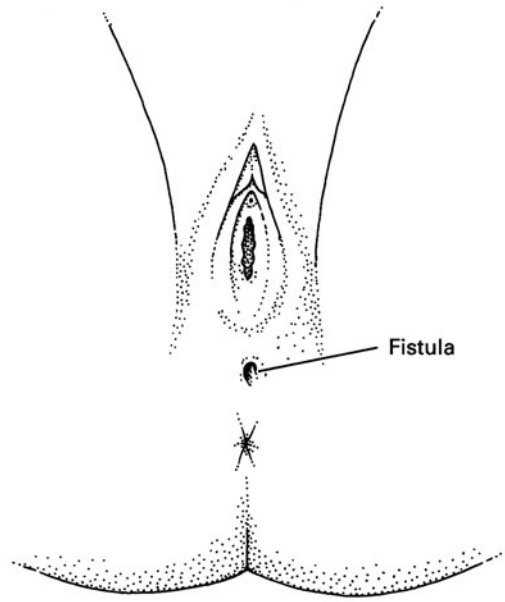
14

14 Perineal inspection may reveal the presence of a vestibular fistula, which is the most common condition in girls. In cases of imperforate anus with rectovestibular fistula, the rectal orifice is located within the vestibule and outside the hymen. A true rectovaginal fistula is an extremely rare anomaly.

These patients can undergo a primary repair via a posterior sagittal approach, either in the newborn period or following a period of dilatations provided the surgeon has adequate experience and a meticulous technique is utilized. The author's preference is the newborn period. A colostomy followed by the definitive repair is also an acceptable and safe approach. These fistulas are usually large enough to decompress the gastrointestinal tract. Occasionally, the fistula is too narrow and the patient will suffer from abdominal distension. In these patients the fistula may first be dilated in order to facilitate emptying of the rectum. The defect is then repaired with a limited posterior sagittal operation.

Patients with vestibular fistula are the ones who most often suffered from a failed attempt at primary repair without a colostomy. In addition, patients with this particular defect are usually continent after a successful operation. An infection and/or dehiscence must be considered an unacceptable complication, not only because it is undesirable, but also because it may damage the continence mechanism and change the final functional prognosis.

15 The presence of a rectoperineal fistula is the simplest defect in the spectrum of female malformations. These patients can be treated with a minimal posterior sagittal anoplasty, without a colostomy, during the neonatal period.



15

Most girls with imperforate anus have a fistula (95 percent). Sometimes, after 20–24 hours of observation, the neonate's abdomen may become distended and yet there is no evidence of meconium passing through the genitalia. In such a case, the baby probably suffers from imperforate anus without fistula. The neonate is a candidate for radiologic evaluation, using the same principles discussed for male neonates above (see Illustrations 1 and 2).

OPERATIONS

Colostomy

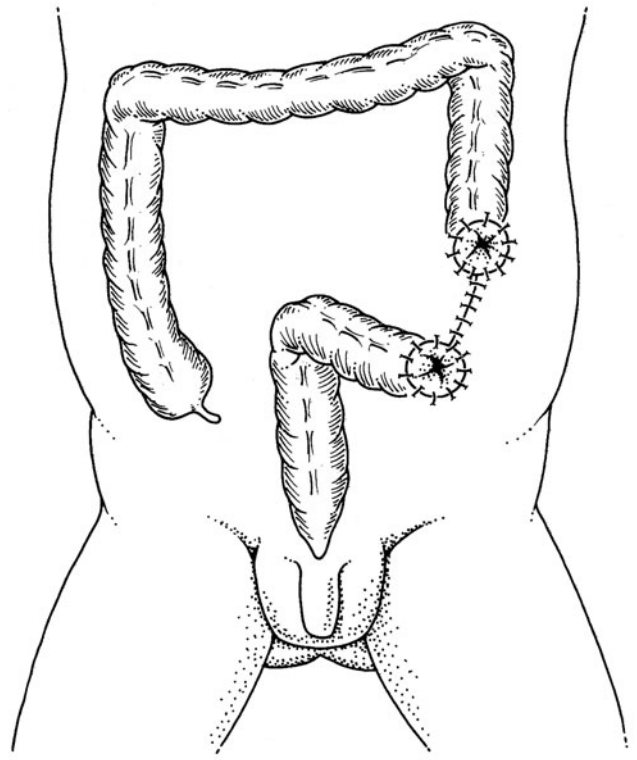
A descending colostomy with separated stomas is preferable for the management of anorectal malformations. Transverse colostomies have several disadvantages: the mechanical preparation of the distal colon before the definitive repair is much more difficult and, in the case of a large rectourethral fistula or rectobladder fistula, the patient often passes urine into the colon, where it remains and is absorbed, leading to metabolic acidosis. Also, during distal colostography it is more difficult to distend the distal rectum and define the anatomy. Patients with transverse colostomies are more likely to develop a megarectosigmoid. A more distal colostomy does not allow significant absorption of urine. Loop colostomies often permit the passage of stool from the proximal stoma into the distal intestine, which can cause urinary tract infections and impaction of stool in the distal rectal pouch. Prolonged dilatation of the rectal pouch may provoke irreversible intestinal damage, which translates into severe constipation later in life.

Colostomy prolapse is more common with loop colostomies and those created in a mobile portion of the colon.

A colostomy created too distally in the area of the rectosigmoid colon may interfere with mobilization of the rectum during the pull-through procedure. The incidence of prolapse in descending colostomies is almost zero, owing to the fact that the proximal stoma is opened immediately distal to the fixed descending colon.

During the opening of the colostomy, the distal intestine must be irrigated to remove all the meconium, preventing the formation of a megasigmoid.

16 The colostomy is constructed through a left lower quadrant oblique or transverse incision. The proximal stoma is exteriorized through the upper and lateral part of the wound and the mucous fistula is placed in the medial or lower part of the wound. The mucous fistula is made very small to prevent prolapse. The stomas should be separated enough to allow the use of a stoma bag, which covers only the functional stoma.



16

High-pressure distal colostography

Before the definitive repair, distal colostography is performed. It is the most valuable and accurate diagnostic study for anorectal malformation. Water-soluble contrast medium is instilled into the distal stoma, which fills the distal intestine and enables demonstration of the location of the blind rectum and the precise site of a rectourinary fistula. The rectum is surrounded by striated muscle, which keeps it collapsed and prevents filling of the most distal part. This may give the erroneous impression of a very high defect and may prevent demonstration of a rectourinary fistula, which is always located at the most distal part of the rectum. To avoid this problem, the contrast medium must be injected with considerable hydrostatic pressure under fluoroscopic control. The use of a Foley catheter is recommended; it is passed through the distal stoma, the balloon is inflated (2–5 mL), and it is pulled back as far as possible to occlude the stoma during the injection of the contrast medium. This maneuver permits exertion of enough hydrostatic pressure (syringe manual injection) to overcome the muscle tone of the striated muscle mechanism, fill the rectum, and demonstrate the urinary fistula when present.

In cases of rectourethral fistula (prostatic and bulbar), the surgeon knows precisely where to find the rectum. In cases of rectobladderneck fistulas, the surgeon does not expect to find the rectum through the posterior sagittal approach and so avoids a blind perineal dissection. In this latter case, the surgeon can prepare the patient for an additional laparoscopy or laparotomy to mobilize a very high rectum.

Definitive repair

INCISION

All anorectal malformations can be corrected by the posterior sagittal approach. The size of the incision depends on the specific defect. The patient is placed in the prone position with the pelvis elevated. An electric stimulator is used to elicit muscle contraction during the operation as a guide to remain exactly in the midline. An incision that starts in the lower portion of the sacrum and extends anteriorly to the anal sphincter is necessary for rectoprostatic fistulas. Smaller incisions (limited posterior sagittal anorectoplasty) are adequate for defects such as vestibular fistula. Perineal fistulas require a very small posterior sagittal incision (minimal posterior sagittal anoplasty).

The anatomic relationship of the rectum to genitourinary structures is complex. The separation of the rectum from these structures represents the most risky part of the procedure.

About 90 percent of male defects can be repaired via the posterior sagittal approach without entering the abdomen.

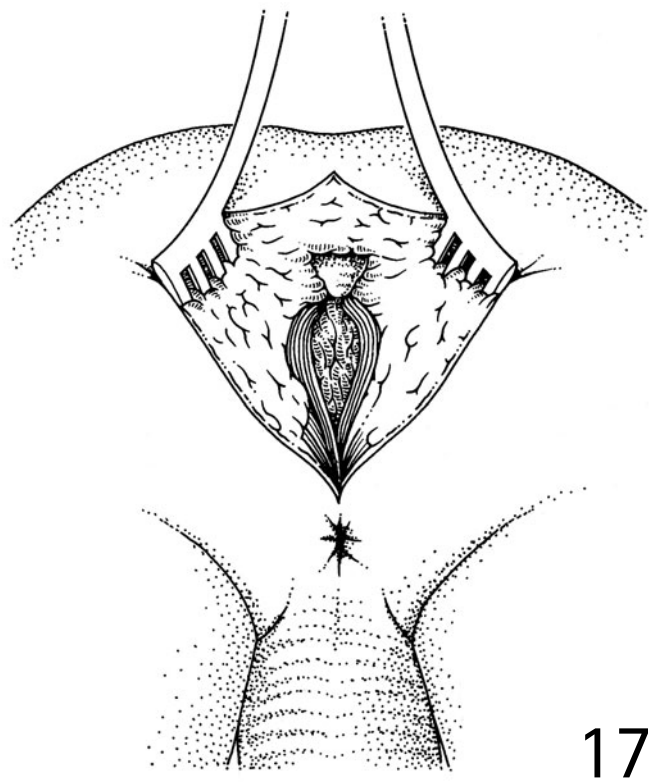
PERINEAL FISTULAS

The repair of these defects consists of a small anoplasty with minimal mobilization of the rectum, sufficient for it to be transposed and placed within the limits of the external sphincter. This is a meticulous operation and can be done during the neonatal period without a colostomy. The most common complication during the repair of this defect is a urethral injury, which can be avoided by placing a urethral catheter and taking particular care during the dissection of the anterior rectal wall. These patients, both boys and girls, have an excellent prognosis, even without an operation, provided the anal orifice is not strictured. These patients have problems with bowel control under only three circumstances: first, if they are subjected to an inadequate surgical technique, second, if they are not treated adequately to prevent constipation, fecal impaction, and overflow pseudoincontinence, and third if they have significant, associated spinal problems.

RECTOURETHRAL FISTULA

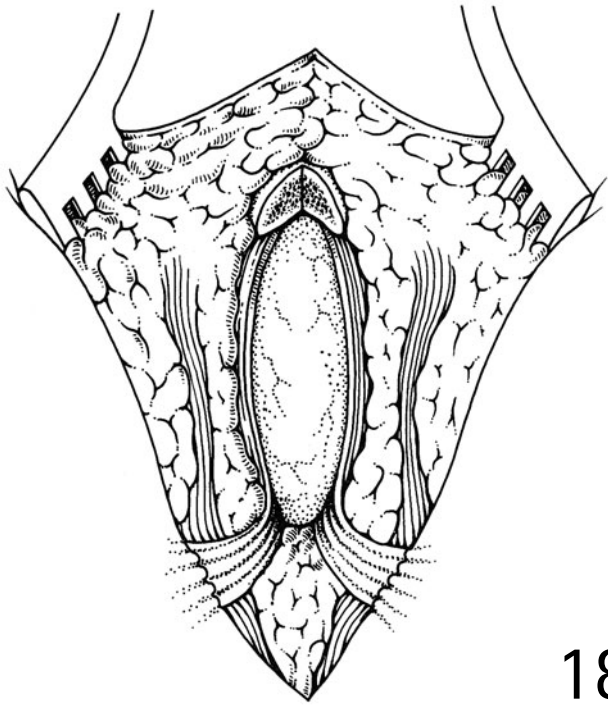
A Foley catheter is inserted through the urethra. In about 15 percent of cases, this catheter goes into the rectum rather than into the bladder. To avoid this, the catheter must be intentionally directed anteriorly by the use of a lacrimal probe inserted in the distal tip of the catheter or with a Coude catheter to find its correct path. Occasionally the catheter must be positioned intraoperatively under direct vision once the fistula is visualized.

17 The skin is opened through a midsagittal incision, and the parasagittal fibers and muscle complex are divided exactly in the midline by use of fine-needle cautery. The fibers of the muscle complex run perpendicular and medial to the parasagittal fibers. The crossing of the muscle complex fibers with the parasagittal fibers represents the anterior and posterior limits of the new anus. These limits can be seen most clearly with the use of an electrical stimulator. The levator muscle, which lies deep in the incision, is then divided in the midline. The higher the malformation, the deeper the levator muscle is found. The levator muscle fibers run parallel to the skin incision. Levator muscle and muscle complex are in continuum.



17

18 When all muscle structures have been divided, the rectum can be seen. In cases of rectourethral bulbar fistulas, the intestine is prominent and it almost bulges into the wound. In cases of rectoprostatic fistulas, the rectum is located much higher, just under the coccyx, and is not as prominent. In cases of rectobladderneck fistulas, the rectum is not visible through this approach, and searching for it risks injuring other structures.



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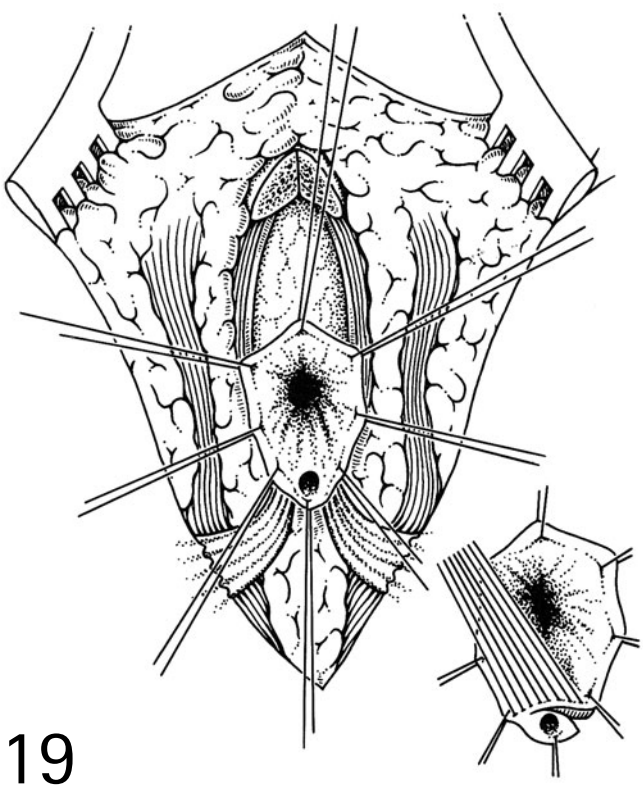


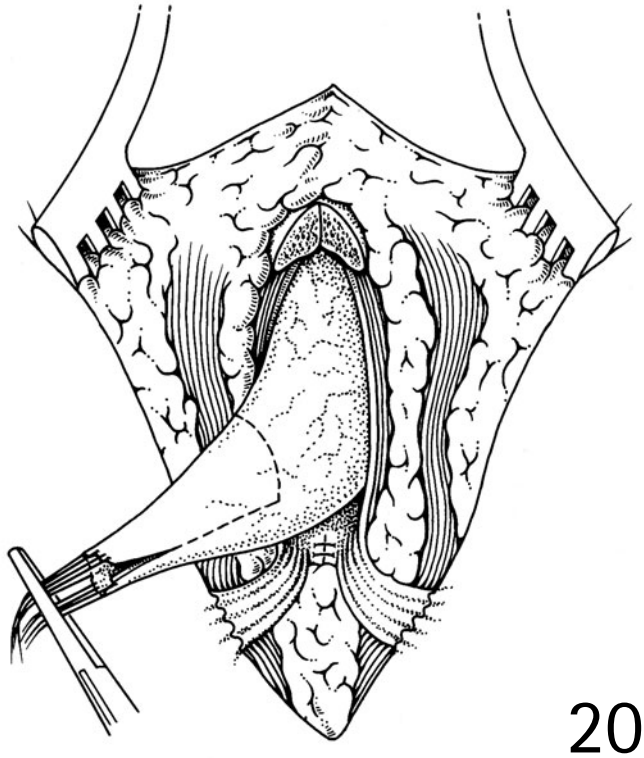
19

19 Two silk sutures are placed in the posterior rectal wall on both sides of the midline. The rectum is opened between the sutures and the incision is continued distally, exactly in the midline, down to the fistula site. Temporary silk sutures are placed on the edges of the open posterior rectal wall for traction.

The anterior rectal wall immediately above the fistula is a thin structure. There is no plane of separation between rectum and urethra in that area. A plane of separation must be created in the common wall. Multiple 6/0 silk sutures are placed through the rectal mucosa immediately above the fistula in semi-circumferential fashion. The rectum is then separated from the urethra, creating a submucosal plane for approximately 5–10 mm above the fistula site. During this delicate dissection, it is very helpful to dissect the rectum laterally, very close to the rectal wall until both dissections (lateral and medial) meet, separating the rectum completely from the urinary tract. Once the rectum is fully separated, a circumferential perirectal dissection is performed to gain enough rectal length to reach the perineum. The rectum is surrounded by a conspicuous whitish fascia. The dissection must be performed between this fascia and the rectal wall to avoid damage to the innervation of the bladder and genitalia.

In cases of a fistula opening into the bulbar urethra, the dissection necessary to pull the rectum down to the perineum is minimal, whereas in cases of prostatic fistula the perirectal dissection is considerable. In both cases, enough rectal length must be gained in order to perform a comfortable, tension-free anastomosis between the rectum and the skin. As traction is exerted on the mobilized rectum, some grooves can be seen in the rectal wall, which demonstrate the tension lines that hold the rectum. These indentations are nerves and vessels that must be divided. Patients with lower defects treated with this approach suffer more constipation than patients with higher defects.

20 Once the rectum has been fully mobilized, a decision must be made concerning the need for tailoring of the rectum. The size of the rectum can be evaluated and compared with the available space so that its size matches the limits of the sphincter. If necessary, the rectum can be tapered by removing part of the posterior wall. The rectal wall is reconstructed with two layers of interrupted, long-lasting, absorbable sutures. The anterior rectal wall is often damaged to some degree as a consequence of the separation between rectum and urethra. To reinforce this wall, both smooth muscle layers can be sutured together with interrupted 5/0 long-lasting, absorbable sutures. The urethral fistula is sutured with long-term, absorbable sutures.



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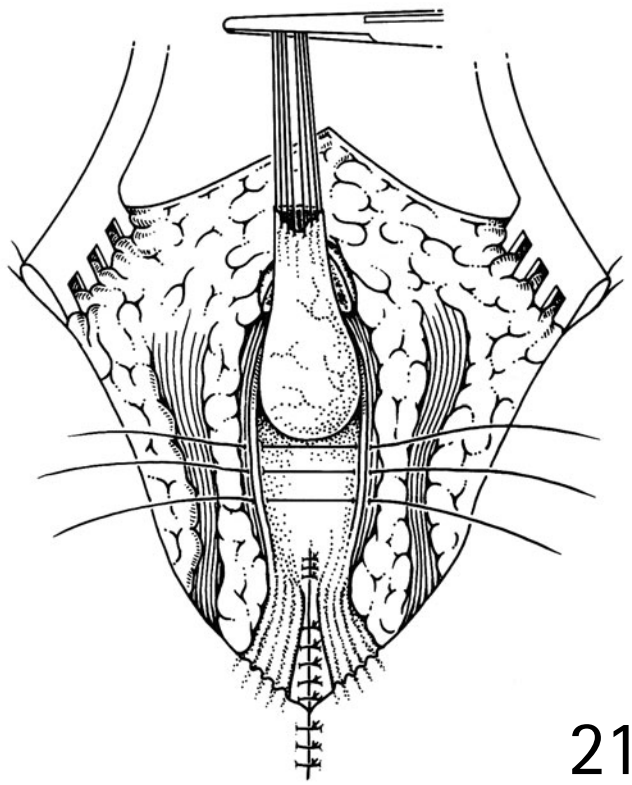
The tapering of the rectum must always be done on the posterior rectal wall. The part of the intestine that will be adjacent to the closed urethral fistula must be normal rectal wall to avoid a recurrent rectourethral fistula.

21 The rectum is placed in front of the levator muscle and within the limits of the muscle complex and external sphincter. The electrical stimulator is helpful in identifying the limits of the muscle structures. Anterior and posterior limits of the external sphincter are temporarily marked with silk sutures. In cases where the incision is extended anteriorly beyond the limits of the sphincter, it is necessary to repair the anterior perineum with interrupted, long-term, absorbable sutures to bring together both anterior limits of the external sphincter. Long-lasting absorbable sutures are placed on the posterior edge of the levator muscle. The posterior limit of the muscle complex must also be re-approximated behind the rectum. These sutures should include part of the rectal wall in order to anchor it and help to avoid rectal prolapse.



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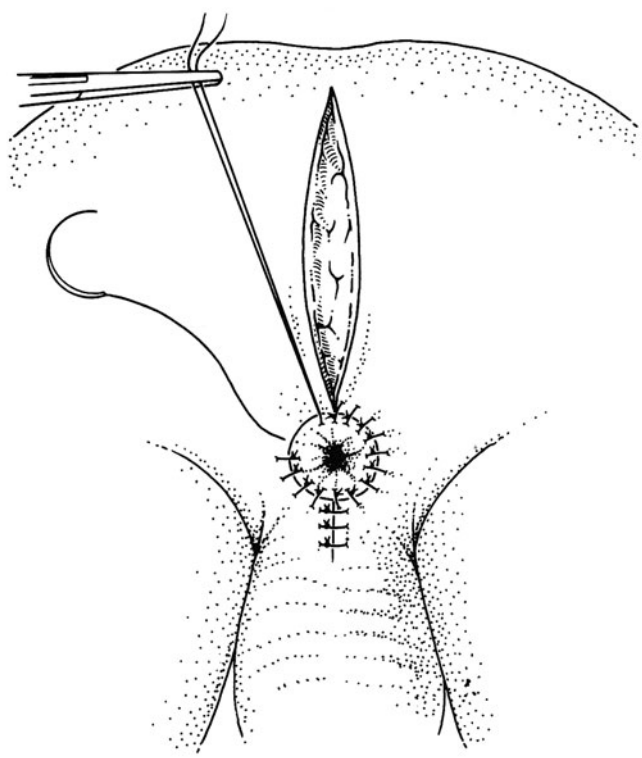
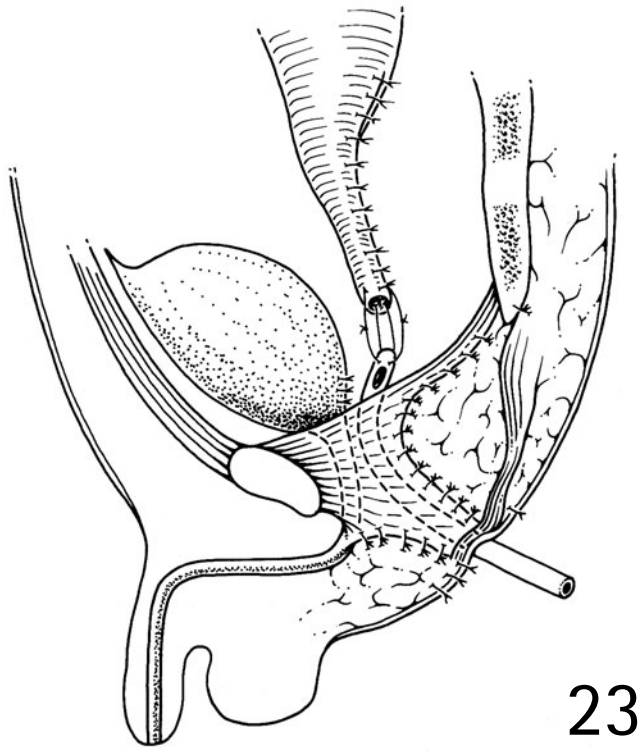


22 The anoplasty is performed with 16 interrupted, long-lasting, absorbable sutures. Anoplasty sutures are placed under slight tension, so that once cut, the anus retracts slightly. The wound is then closed, bringing together corresponding sphincteric structures in the midline.

The Foley catheter is left in place for 7 days. The patient receives broad-spectrum antibiotics for 2 days.

RECTOBLADDERNECK FISTULA

23 For the repair of rectobladderneck fistulas, the entire lower part of the patient's body from chest down is included in the sterile field, so that the surgeon can work simultaneously in the abdomen and the perineum. The initial approach is posterior sagittal. All the muscle structures are divided in the midline. The presacral space in front of the levator muscle and within the limits of the muscle complex and external sphincter is defined. This is where the rectum will subsequently be placed. This incision can also be done supine with legs lifted to the baby's chest.



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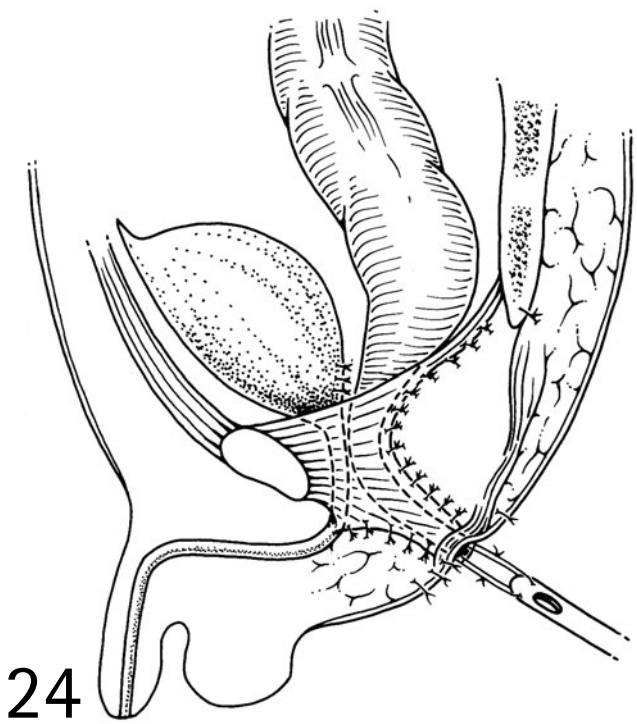
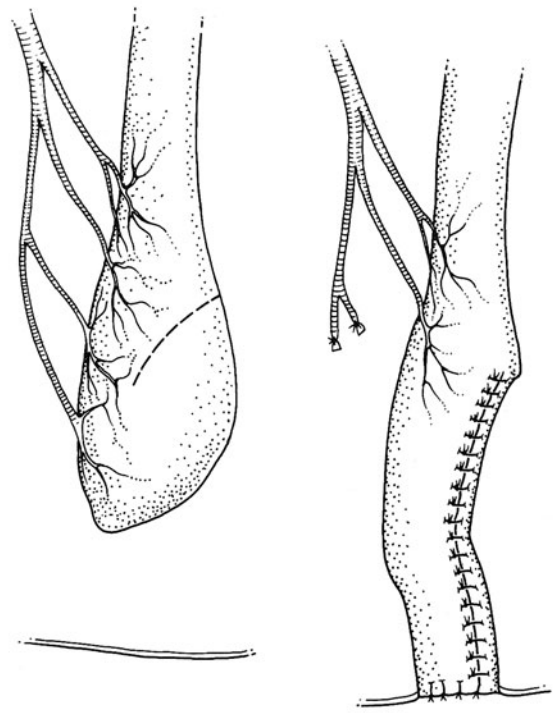
The abdomen is entered either via laparotomy or laparoscopically, and the rectosigmoid colon is mobilized. In this very high defect, the rectobladderneck fistula is located approximately 2 cm below the peritoneal reflection, and the rectum communicates with the urinary tract in a T fashion, which means that there is no common wall between the distal part of the rectum and the urinary tract. This facilitates the dissection, which is minimal. The surgeon must be careful to avoid damage to the vas deferens, which run very close to the bowel. With a laparoscopic technique, separation of the rectum from the bladderneck is straightforward. Gaining adequate length, particularly with a high rectum, is challenging, and must be meticulous so as to avoid devascularizing the distal rectum.



24

24 The rectum is separated from the bladderneck, and the bladder end of the fistula is closed with interrupted absorbable sutures.

25 As this is a very high defect, mobilization of the rectum to reach the perineum requires special maneuvers. Ligation of the inferior mesenteric vessels as high as possible, very close to their origin near the aorta, would mobilize the rectum, but would probably compromise the blood supply of the rectum because the arcades that connect the middle colic vessels with the inferior mesenteric ones may have been interrupted at the time of the colostomy creation. An alternative is to ligate the most distal branches of the inferior mesenteric vessels close to the rectum. If this is done, the more proximal branches of the inferior mesenteric vessels must be left intact to guarantee a good blood supply to the rectum. This last maneuver is possible only because the rectum has an excellent intramural blood supply.



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If it is necessary to gain extra length, a plasty of the distal dilated portion of the rectum can also be performed. A combination of these maneuvers usually allows the intestine to reach the perineum, provided the colostomy does not interfere with the pull-through of the rectum. This can be anticipated by the use of colostography, which demonstrates the precise length of intestine available from the colostomy to the end of the rectum. If the colostomy was placed too distal in the sigmoid, the surgeon may have to take down the mucous fistula and leave it on a Hartman's pouch to allow for the pull-through. The rectum should be preserved and never discarded as it performs a vital reservoir function. The rectum is tapered, if necessary, and then pulled down through the pelvis. The anoplasty is performed as previously described.

IMPERFORATE ANUS WITHOUT FISTULA

About 5 percent of patients have imperforate anus without a fistula. This is the likely defect in patients with Down's syndrome. In both boys and girls, the rectum lies about 2 cm from the perineal skin. Most of these patients have a very good sacrum and good muscles. The fact that these patients have no fistula does not necessarily mean that the repair is simpler. The rectum must be carefully separated from the urethra, because the two structures have a common wall. The rest of the repair must be performed as described for the rectourethral fistula type of defect.

RECTAL ATRESIA AND STENOSIS

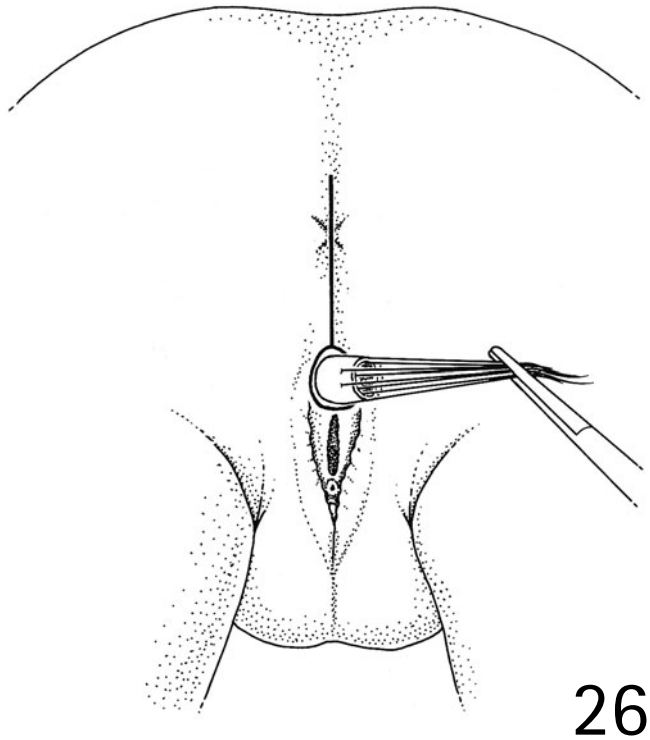
These defects are repaired through a posterior sagittal approach. The entire sphincteric mechanism is divided in the midline. The upper rectal pouch is opened, as well as the small distal anal canal. An end-to-end anastomosis is performed under direct vision, followed by a meticulous reconstruction of the muscle mechanism posterior to the rectum. The wound is closed following the principles already described.

Repair in girls

PERINEAL FISTULA

The treatment of perineal fistula in girls is the same as that discussed for boys.

Vestibular fistula



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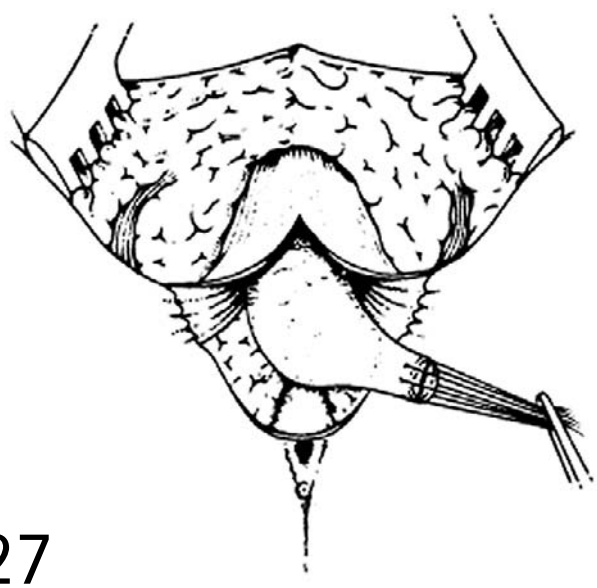
26 Most surgeons underestimate the complexity of this defect. Multiple 6/0 silk sutures are placed at the edge of the fistula in order to exert uniform traction on the rectum to facilitate its dissection. The incision used to repair this defect is shorter than that used to repair rectourethral fistulas in boys. The incision continues around the fistula into the vestibule in a racket-like fashion. All the sphincteric mechanism is divided in the midline until the rectal wall is located. A characteristic whitish fascia covers the rectum posteriorly and must be divided. This helps to locate the plane of dissection during mobilization of the rectum. Once the rectal wall has been identified, a lateral dissection is performed from the posterior midline, while placing traction on the fistula to make the plane of dissection more obvious. It is vital to be adjacent to the rectal wall, and clean away the thin white fascia that envelops it. The surgeon must be in this key plane in order to be able to mobilize the rectum.

Hemorrhoidal vessels are usually found on the lateral aspects of the rectum. The most delicate part of this dissection is the anterior rectal wall. The rectum and the vagina share a common wall, which is often very thin. This thin wall has no plane of separation and the surgeon has to make two walls out of one. This dissection is performed using a very fine needle cautery and suction, which allows each one of the little vessels that are encountered during the procedure to be meticulously cauterized. It is continued up to the point where rectum and vagina separate and have full-thickness walls. A characteristic areolar tissue between the two full thickness walls identifies this point in dissection. The most common error in performing this operation is incomplete separation of the vagina and rectum. This may create a tense anastomosis between the rectum and the skin, which may provoke dehiscence and recurrence of the fistula.



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27 Once the dissection has been completed, the electrical stimulator is used to determine the limits of the sphincteric mechanism. The anterior limit of the external sphincter and the anterior edge of the muscle complex are reapproximated as previously described creating perineal body. The levator muscle is usually not exposed and therefore does not have to be reconstructed. The muscle complex must, however, be reconstructed posterior to the rectum. The anoplasty is performed as previously described.

VAGINAL FISTULA

Imperforate anus with a true rectovaginal fistula is extremely rare. The term is often misused, and patients with rectovestibular fistula or cloaca are commonly incorrectly described as having a rectovaginal fistula. For a patient with cloaca that is misdiagnosed in this way, the surgeon might repair the rectum but leave the urogenital sinus intact, which would thereafter require a complete re-operation.

A true rectovaginal fistula requires a full posterior sagittal incision. The operation is essentially the same as that described for a vestibular fistula, except that it is necessary to dissect much more of the rectum to gain enough length to pull it down to the perineum.

POSTOPERATIVE CARE

Patients generally have a smooth postoperative course. Pain is not a prominent symptom, except in those patients who have undergone a laparotomy.

In cases of rectourethral fistula in boys, the urethral catheter is left in place for 7 days. If the urethral catheter is accidentally dislodged, the patient must be observed for spontaneous voiding, which usually occurs. Attempts to reintroduce a urethral catheter can be dangerous and must be avoided. Occasionally patients suffer from bladder spasms due to the presence of the balloon in the bladder. This is an indication to remove the catheter earlier than planned.

Intravenous antibiotics are administered for 48 hours. An antibiotic ointment is applied locally for 7 days. The patient is discharged after 2 days in cases of a posterior approach without a laparotomy, and after 3–5 days in cases of an abdominal approach.

Two weeks after the operations, anal dilatations are started. On the first occasion, a dilator that fits snugly into the anus is used to instruct the parents, who must carry out dilatation twice daily. Every week, the size of the dilator is increased until the rectum reaches the desired size, which depends on the patient's age (Table 50.1). Once the desired size is reached, the colostomy can be closed. The frequency of dilatations may be reduced once the dilator of desired size passes easily. This reduction should occur according to the following schedule: at least once a day for 1 month; every third day for 1 month; twice a week for 1 month; once a week for 1 month; and every 2 weeks for 3 months.

Table 50.1 *Size of dilator required in different age groups*

Age group	Hegar dilator size
1–4 months	12
4–8 months	13
8–12 months	14
1–3 years	15
3–12 years	16
Over 12 years	17

After the colostomy is closed, patients often suffer from severe diaper rash as a consequence of multiple bowel movements. The number of bowel movements eventually decreases, and patients develop their own bowel movement pattern. This pattern has a very significant prognostic value by 6 months after the closure of the colostomy. A baby who has one to three bowel movements each day, remains clean between bowel movements, and pushes during each bowel movement – indicating that there is some feeling during the defecation process – has, in general, a good functional prognosis, and therefore is likely to respond to toilet training. On the other hand, an infant who passes stools constantly, without any evidence of feeling or pushing, usually has a poor functional prognosis and will need bowel management. In addition, on the basis of the results obtained in the authors' series, it may be possible to predict the final functional result from the precise anatomic diagnosis and the status of the sacrum.

FUNCTIONAL DISORDERS AFTER REPAIR OF ANORECTAL MALFORMATIONS

Most patients who have undergone repair of anorectal malformations suffer from some degree of functional disorder due to congenital deficiencies that are not correctable.

Deficiencies in sensation

Except for patients with rectal atresia, most patients are born without an anal canal. This means that they do not have the exquisite sensation that normally resides in this anatomic area. Most patients, however, still preserve a vague sensation called proprioception, generated from distension of the rectum, and therefore stretching of the voluntary muscles around it. Liquid stools, which do not distend the rectum, are not felt by most of these patients.

Sphincteric mechanism

Anorectal malformations are represented by a spectrum of defects. Most of these patients have a sphincteric mechanism represented by parasagittal fibers of the external sphincter, muscle complex, and levator muscle, with different degrees of development, which varies from almost non-existent muscles to almost normal sphincteric mechanism. Therefore, most of these patients have a limited capacity to hold stools inside the rectum.

Coordinated rectosigmoid motility

Most patients with anorectal malformations suffer from abnormal rectosigmoid motility. Patients who have undergone a surgical procedure in which the rectosigmoid colon

was removed, as in older endorectal procedures, do not have a normal fecal reservoir, but have a segment of sigmoid or descending colon pulled down to the perineum. They have a tendency to pass stool constantly, similar to patients with a perineal colostomy.

On the other hand, patients who have undergone repair in which the rectosigmoid colon was preserved (e.g., posterior sagittal anorectoplasty, sacroperineal pull-through, or simple anoplasty) behave as if they had too large and floppy a fecal reservoir. Clinically, this translates into varying degrees of constipation. Mild cases of constipation can be treated very efficiently with laxatives, and the children usually live a normal life. Severe cases of constipation, particularly if they are not treated properly, may lead to fecal impaction, constant soiling, and, therefore, overflow pseudoincontinence. This constipation seems to be more severe in patients with lower defects. Patients with vestibular fistula in particular are more prone to these problems. An ectatic distended colon (sometimes associated with a loop or transverse colostomy) leads to megarectosigmoid and eventually provokes severe constipation.

COMPLICATIONS

In the author's series of 1750 patients, the following complications were encountered.

Wound infection

Twenty-three patients suffered wound infections and mild dehiscences of the posterior sagittal incision. The infection affected only the skin and subcutaneous tissue and had no repercussions on the function of the sphincteric mechanism.

Anal strictures

Six patients suffered from an intractable anal stricture, which required a secondary operation; a very clear correlation was found in these cases with intraoperative devascularization of the distal rectum.

The protocol of dilatations was not followed by several patients, who were lost to follow-up for several weeks. When they returned, they were suffering from an anal stricture. This stricture was only a ring-like fibrous band at the mucocutaneous junction, which was easily treated. This was different from a long, narrow stricture secondary to ischemia.

It is important to emphasize that before the advent of the posterior sagittal approach, most surgeons would try to create a very large neo-anus to avoid strictures and cumbersome anal dilatations. At that time, surgeons did not recognize the existence of a sphincteric mechanism. With new concepts based on objective knowledge of the sphincteric mechanism obtained by direct visualization, the surgeon is obliged to create an anus no bigger than the size of the external sphincter, and usually one that is smaller than a normal anus for that age. In addition, the new anus is surrounded by voluntary muscle that keeps it closed. If it is not dilated, the rectum will heal narrow or closed. Therefore, anal dilatations are necessary. Tapering of the rectum was not responsible for stricture formation in any of the patients.

Constipation

This was the most common functional disorder observed in this series.

Transient femoral nerve pressure

This occurred in three adolescent patients owing to excessive pressure in the groin during a posterior sagittal operation. This problem can be avoided by adequate cushioning of the patient's groin area.

Ureteric injury

Ectopic ureters were injured in two cases. In one case the ureter was injured during the search for the rectum in a high malformation. This patient required a laparotomy for the ureteric re-implantation during the same procedure. This complication did not occur again, since the importance of a good distal colostogram done preoperatively was recognized. Defining the anatomy prevents the surgeon from looking for a very high rectum via a posterior sagittal approach. A good distal colostogram in boys shows exactly where the rectum opens into the urinary tract and will allow the surgeon to identify those patients who have a rectobladderneck fistula in whom the rectum is too high to be mobilized from below. The second patient was a girl with a persistent cloaca and the ureteral injury occurred during the separation of the bladder from the vagina. This patient underwent a ureteric re-implantation via a posterior sagittal approach.

Neurogenic bladder

Difficulty voiding after a posterior sagittal approach has occurred only in patients with a very abnormal or absent sacrum or with a myelomeningocele, in whom the presence of a preoperative neurogenic bladder can be predicted.

Neurogenic bladder following a posterior sagittal approach in patients with favorable anatomy can occur due to nerve damage during a defective technique, where the surgeon does not follow the principles of the posterior sagittal approach and veers off the midline. In addition, placing Weitlander's retractors deeper than is necessary may compress the nerves that come from the sacral area, causing a neurogenic bladder.

Difficulty voiding after the operation in patients with a normal sacrum has been observed only in cases of complex

cloacas with hydrocolpos. These patients are born with a very large bladder that seems unable to contract well.

Medical management for fecal incontinence

As shown in Table 50.2, there remains a significant number of patients who suffer from fecal incontinence, despite optimal anatomic surgical reconstruction used for the repair of their malformations (approximately 25 percent). For these patients, a program of bowel management is useful. This consists of training the parents and children to clear out the colon once a day with the use of enemas or colonic irrigations and to avoid bowel movements between irrigations by adherence to a specific diet and sometimes medication.

In addition to the 1750 patients followed by the authors, more than 500 patients have been referred for management of fecal incontinence secondary to an operation for imperforate anus performed in other institutions. All these patients are evaluated clinically and undergo a contrast enema study, voiding cystourethrography, radiologic evaluation of their sacrum, and magnetic resonance imaging. These evaluations allow the patients to be classified into the following groups.

NOT TRAINABLE

These are patients who have a poor sacrum, poor muscles, a very high defect, and a poor bowel movement pattern. The best treatment for these patients is a bowel management program. Because of the nature of their original defect and on the basis of statistics, these patients will always suffer from fecal incontinence. Therefore, time should not be wasted on biofeedback programs, behavior modifications, or re-operations, and, perhaps more importantly, false expectations should not be created for the family. The bowel management program allows the patient to remain clean all day in order to be socially accepted.

This group is usually divided into two categories.

CONSTIPATED

These patients have undergone a procedure in which the rectum was preserved, as in anoplasties, a sacroperineal approach or posterior sagittal approach. This group of patients tends to suffer from constipation. Management consists of the use of enemas or colonic irrigation, with volumes of fluids large enough to clean a large rectosigmoid colon. It is not usually necessary to use any kind of diet or medication, because the constipation contributes to the patients remaining completely clean between enemas.

PATIENTS WITH A TENDENCY TO HAVE DIARRHEA

This group of patients has undergone a type of procedure in which their original rectosigmoid colon was resected, as in the Kiesewetter, Soave, or Rehbein types of operation. They have a natural tendency to suffer from the constant passing of liquid stools. A contrast enema shows that the colon runs straight from the splenic flexure down to the anus, and colonic haustrations are apparent all the way down to the perineum. They never suffer from constipation. This is the group that is most difficult to keep clean. Management consists of colonic irrigation followed by a very strict constipating diet and agents that slow colonic motility, such as loperamide hydrochloride. Patients in whom colonic motility fails to slow down are the rare candidates for a permanent colostomy.

TRAINABLE

These patients were born with a favorable type of defect (vestibular fistula, perineal fistula, rectourethrobulbar fistula), a good sacrum, and a good sphincteric mechanism, and underwent an operation that placed the rectum in the correct position. In addition, these patients have a good bowel movement pattern. They undergo a behavior modification program to train them to have voluntary bowel movements, but they may need additional help with laxatives to treat their constipation.

CANDIDATES FOR A RE-OPERATION

These patients were born with a favorable type of defect, good sacrum, and good sphincteric mechanism, and yet they underwent an operation that placed the rectum in the wrong place, as demonstrated clinically and confirmed by magnetic resonance imaging. Repositioning the rectum within the limits of the sphincteric mechanism may improve the functional result.

Table 50.2 *Clinical results in the most common defects*

Type of fistula	Perineal	Vestibular	Bulbar	Prostatic	Bladderneck
Total cases evaluated	39	97	83	71	29
Voluntary bowel movement (%)	100	92	82	73	28
Soiling (%)	21	36	54	77	90
Totally continent (%)[a]	90	71	50	31	13
Urinary incontinence (%)	None	4[b]	2[b]	8[b]	18[b]

[a]Voluntary bowel movement and no soiling.

[b]Patients with absent sacrum, meningocele, or severe associated urologic abnormalities.

CANDIDATES FOR A SIGMOID RESECTION

There is a subgroup of patients who were born with a defect that had a good prognosis and who underwent a technically good operation but suffer from severe constipation and severe megasigmoid colon. Often such patients' constipation was not aggressively managed postoperatively. They are incapable of emptying the rectosigmoid colon and suffer from chronic soiling and overflow pseudoincontinence. Prior to considering a sigmoid resection, a laxative test is performed. The test is carried out over a period of several days. First, the colon is disimpacted with enemas. This process is radiologically monitored. Then the laxative requirement for each individual patient is determined by trial and error. Once the right dose is reached, as demonstrated by an X-ray film that shows a clean colon, the patient's ability to have bowel control can be determined. If the patient is continent, but in order to remain clean requires an enormous dose of laxatives, a sigmoid resection, preserving the rectum and creating an anastomosis between the descending colon and the rectum above the peritoneal reflection, can reduce that laxative requirement dramatically. If the patient is incontinent, a bowel management regimen is implemented, and a sigmoid resection is contraindicated. Resecting colon in such a patient would change an incontinent, constipated child into an incontinent patient with loose stools, which is a much more difficult situation to manage.

OUTCOME

Each type of defect has a different prognosis. Traditionally, defects have been classified as high, intermediate, or low. This type of classification represents an over-simplification that should be avoided. The so-called high defects often include individual malformations with very different prognoses for bowel function.

Medical management, as described above, allows most patients to remain clean for 24 hours and allows them to live a normal social life. This does not mean, however, that they have normal bowel function. Without medical help, many of these patients will suffer from fecal incontinence.

When medical management is given, all patients remain clean. Table 50.2 shows the clinical results obtained in patients with the most common types of defects. When the sacrum is very abnormal, most patients remain fecally incontinent for life. All patients with low defects are continent.

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51

Cloaca

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HISTORY

A persistent cloaca is a malformation in which the rectum, vagina, and urethra are fused into a single common channel. This defect is considered one of the most formidable challenges in pediatric surgery and represents the extreme in the spectrum of complexity of female anorectal and urogenital malformations. Treatments traditionally involved repair of the rectal component of the malformation, leaving the urogenital sinus alone, planning its repair in a second stage, or performing a combined abdominoperineal approach with vaginal and rectal pull-through. Some treatments were adequate for certain malformations, but not for others. The perineal approach to the urogenital sinus was useful for low defects but not for higher defects. Similarly, the abdominal approach was required for some anomalies but not for others. These approaches were often limited in their exposure and thus could not clearly define the complex anatomy of the defect, and as with surgery for anorectal malformations, the urinary sphincter and anorectal sphincter were matters of speculation because the precise anatomy was not visualized.

Hendren compiled the most comprehensive reports on the secondary reconstruction of these cases, and emphasized a global approach to the simultaneous repair of the entire anomaly, with particular focus on the urologic reconstruction. The posterior sagittal approach for the repair of imperforate anus was used to repair a cloaca for the first time in 1982, and led to the operation described as the posterior sagittal anorectovaginourethroplasty (PSARVUP). This approach allowed for direct exposure of the complex anatomy and the voluntary muscles of urinary and fecal continence and provided an approach of the entire spectrum of defects.

PRINCIPLES AND JUSTIFICATION

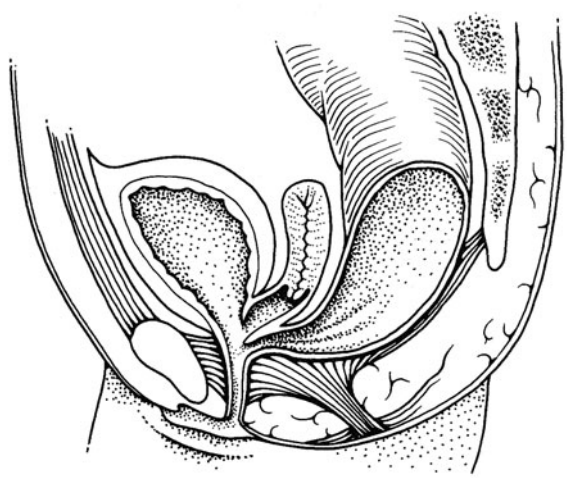
Incidence

Cloacal anomalies probably occur in 1 in 20 000 live births. They occur exclusively in girls. Persistent cloaca was, in the past, considered an unusual defect, and a high incidence of rectovaginal fistulas was reported in the literature. In retrospect, it seems that cloaca is a much more common defect than reported, as imperforate anus with rectovaginal fistula is an almost non-existent defect, occurring in less than 1 percent of all cases. Most patients with persistent cloaca were probably erroneously thought to have a rectovaginal fistula. Many of those patients underwent surgery, had the rectal component of the malformation repaired, but were left with a persistent urogenital sinus.

The goals of treatment of cloaca include the achievement of bowel control, urinary control, and sexual function.

Classification

1 In cloacal malformations, the length of the common channel varies from 1 cm to 10 cm, which has important technical and prognostic implications. When the common channel is shorter than 3 cm, patients usually have a well-developed sacrum and good sphincters. When the common channel is longer than 3 cm, this usually suggests a more complex defect and the patient often has a poor sphincter mechanism and poor sacrum.



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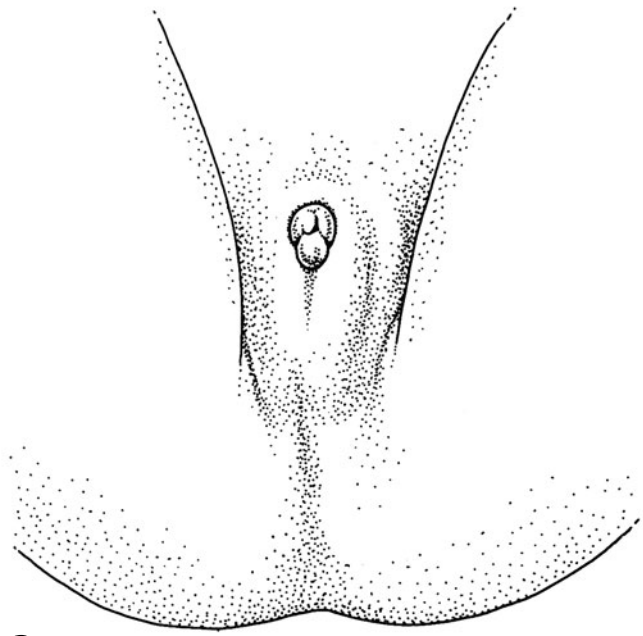

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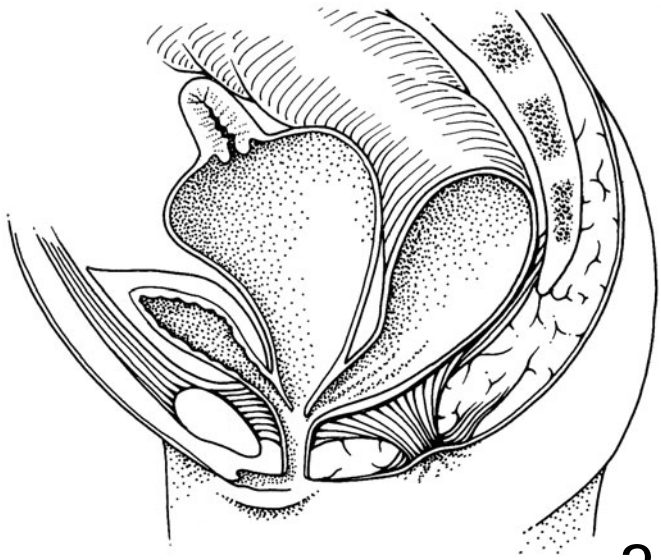
2 The diagnosis of persistent cloaca is a clinical one. Careful separation of the labia discloses a single perineal orifice, which is pathognomonic of a cloaca. These patients often have small external genitalia. Sometimes patients with cloacas have a palpable lower abdominal mass that represents a distended vagina (hydrocolpos). Failure to recognize the presence of a cloaca in a neonate may be dangerous, as more than 90 percent of these patients have important associated urologic problems.

Management of cloacal malformations during the neonatal period

Once the clinical diagnosis of a cloaca has been established, the next step is to perform an urgent urologic evaluation. Abdominal and pelvic ultrasonography is the most important screening test to rule out the presence of hydronephrosis, hydroureter, and/or hydrocolpos.

3 In more than 30 percent of these cases, the vagina is abnormally distended and full of mucous (hydrocolpos). The distended vagina may compress the trigone, interfere with the drainage of the ureters, and produce megaureters. The most common error at this stage is to perform only a colostomy in a patient with severe obstructive uropathy, as this can lead to acidosis and urinary sepsis. The dilated vagina can also become infected, which is called 'pyocolpos' and may lead to vaginal perforation and peritonitis. Such a large vagina may ultimately represent a technical advantage at the time of the main repair, because having more vaginal tissue will facilitate its reconstruction.



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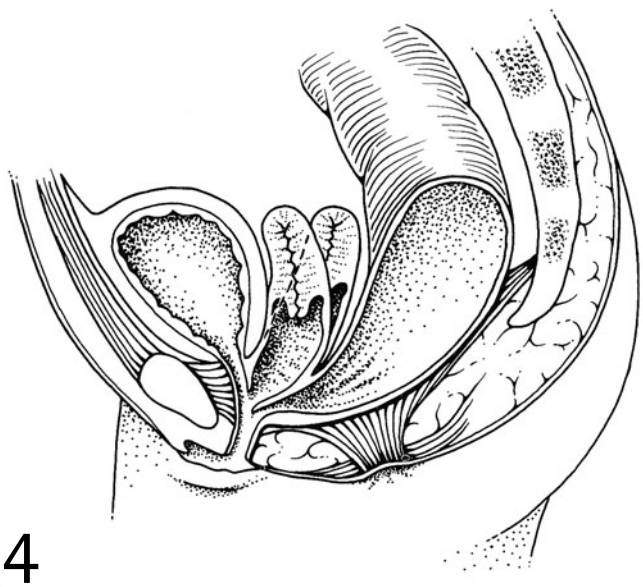
If a hydrocolpos is correctly identified and drained, obstruction of the urinary tract is usually relieved, making urinary diversions such as a vesicostomy, ureterostomy, or nephrostomy unnecessary. Rarely, a near-atresia of the urethra exists and a vesicostomy may be required.

Attempts to drain the urinary tract through the single perineal orifice (common channel) by way of intermittent catheterization or dilatations is not recommended, as it is unpredictable whether the catheter will enter the bladder or the vagina. This particularly applies in cases of long common channels. Blind dilatations of the single external orifice may also provoke local damage that can interfere with the future repair.



4

4 The vagina and uterus commonly show varying degrees of septation. The rectum usually opens in between the two hemivaginas.

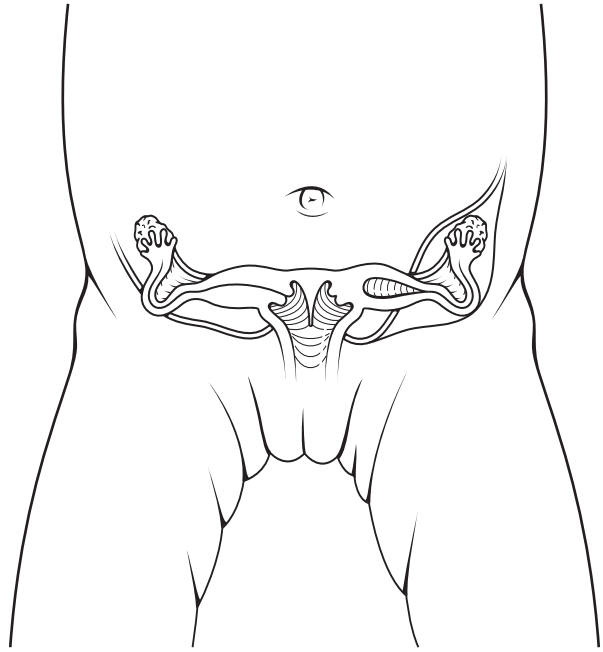
ASSOCIATED DEFECTS

Genitourinary defects

Patients with persistent cloaca have a 90 percent chance of having an associated genitourinary abnormality. Hydronephrosis, urosepsis, and metabolic acidosis represent the main source of morbidity and mortality in newborns with anorectal malformations. Thus, a thorough urologic investigation is mandatory.

Müllerian anomalies

Some of these patients may also suffer from cervical or vaginal atresias or stenoses. When undetected, these may interfere with the drainage of menstrual blood during puberty. These patients can develop hematometra, hematocolpos, or intra-abdominal pseudocysts from retrograde menstruation. The gynecologic anatomy can be ascertained during the main repair (if the abdomen is entered) or at the time of the colostomy closure.



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Associated spinal, sacral, cardiac, and gastrointestinal anomalies occur, as described for patients with anorectal malformations (see Chapter 50).

OPERATIONS

Endoscopy

An endoscopy is recommended for babies with cloaca to try to determine the anatomy. With the right equipment, this can be done in the newborn period at the time of the colostomy. The specific purpose of this procedure is to determine the length of the common channel, the status of the bladderneck and the presence or absence of one or two cervices.

Two well-characterized groups of patients with cloaca exist. These two groups represent different technical challenges and must be preoperatively recognized. The first is comprised of patients who are born with a common channel shorter than 3 cm. Fortunately, these patients represent over 60 percent of the entire group of cloacas. The cloacas in the majority of these patients can be repaired with a posterior sagittal approach only, without a laparotomy. The second group is comprised of patients with longer common channels. This group of patients usually needs a laparotomy, followed by a decision-making process that requires considerable experience, and special training in urology. Therefore, these patients should be referred to centers dedicated to the repair of these defects.

Table 51.1 *Short versus long cloaca*

	Group A	Group B
Common channel	Short, < 3 cm	Long, > 3 cm
Type of operation	Only posterior sagittal	Posterior sagittal and laparotomy
Length of procedure	3 hours	6–12 hours
Hospitalization	48 hours	Several days
Associated urological defects (%)	59	91
Incidence in our series (%)	62	38
Voluntary bowel movements (%)	68	44
Urinary continence (%)	72	28
Average number of operations[a]	9	18
Intra-operative decision making	Relatively easy, reproducible operation	Complex, delicate and technically demanding[b]

[a] Including orthopedic, urologic, cardiac, and general.
[b] Including bladder and vaginal separation with or without the following procedures: Bladder/vagina separation. Ureteral catheter, Ureteral re-implantation. Vesicostomy, Cystostomy, Bladder neck reconstruction or closure, Vaginal switch, Vaginal replacement, (Rectum, Colon, Small bowel.)

Colostomy

All babies with a cloaca need a colostomy. It is important to perform the colostomy proximally enough to avoid it interfering with the repair of the malformation. In other words, the surgeon must leave enough redundant distal rectosigmoid to allow a pull-through, and for potential use of colon for vaginal replacement. (See the colostomy section in Chapter 50.)

DRAINAGE OF THE HYDROCOLPOS

During the opening of the colostomy, it is mandatory to drain the hydrocolpos when present. If the hydrocolpos is not large enough to reach the abdominal wall above the bladder, it can be drained with a tube. Because a significant number of these patients have two hemivaginas, the surgeon must be certain that the tube inserted into the hydrocolpos is really draining both of them. Occasionally, the surgeon has to open a window in the vaginal septum in order to drain both with a single tube. Sometimes the hydrocolpos is so large that it may even produce respiratory distress; such giant vaginas may be drained directly, connecting the vaginal wall to the abdominal wall as a tubeless vaginostomy.

On rare occasion, patients with cloaca are unable to empty their bladders because they suffer from a near atresia of the common channel. In such circumstances, the baby may require a vesicostomy, or a suprapubic cystostomy.

DISTAL COLOSTOGRAPHY AND CLOACAGRAM

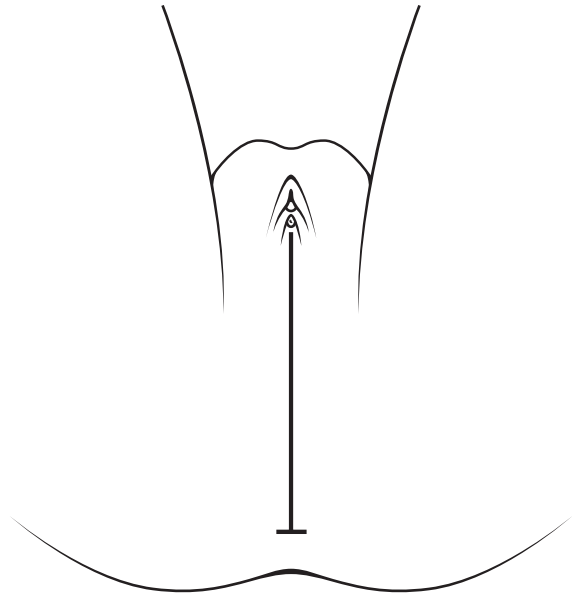
After the patient has recovered from the colostomy, a high-pressure distal colostogram injection of contrast through the single perineal orifice will help define the cloacal anatomy. This study can demonstrate the location of the rectum, and demonstrate the vaginas or hemivaginas, and often assess for vesicoureteral reflux. It is a vital study to help plan the definitive repair.

Definitive repair

The goal of the operation is to separate the rectum from the vagina and place it within the sphincteric mechanism. The urethra and vagina need to be mobilized so that each is a separate orifice at the perineum.

CLOACAS WITH COMMON CHANNEL SHORTER THAN 3 CM

6 With the patient in the prone position, a long midsagittal incision is performed that extends from the middle portion of the sacrum through the sphincter mechanism and down into the single perineal opening. All of the muscle structures are divided in the midline.



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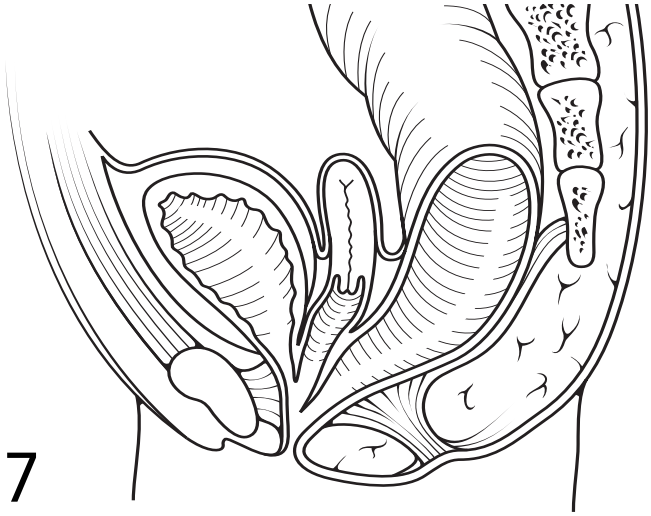

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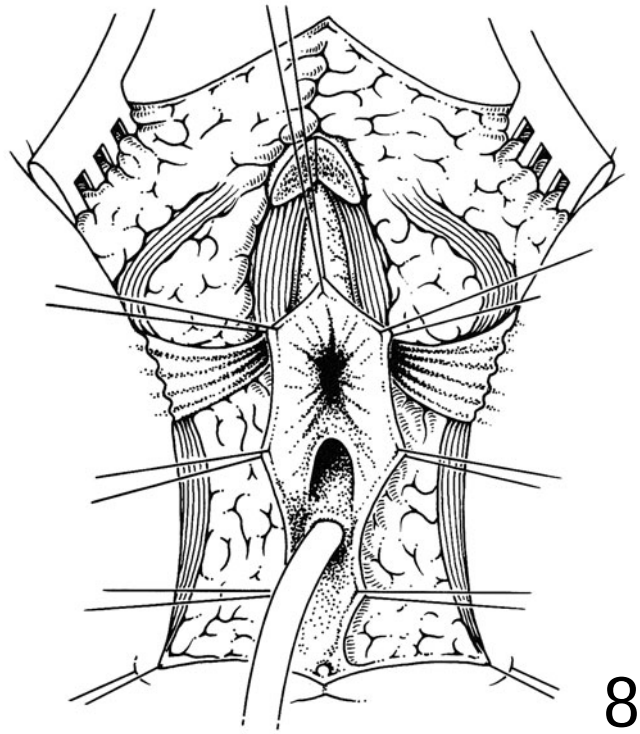
7 Low cloacal malformations (less than 3 cm) are usually associated with a well-developed sacrum, a normal-appearing perineum, and adequate muscles and nerves. Therefore a good functional prognosis is expected.

8 The incision is continued all the way down to the single perineal orifice, exposing the entire malformation. The entire sphincter mechanism is divided in the midline.



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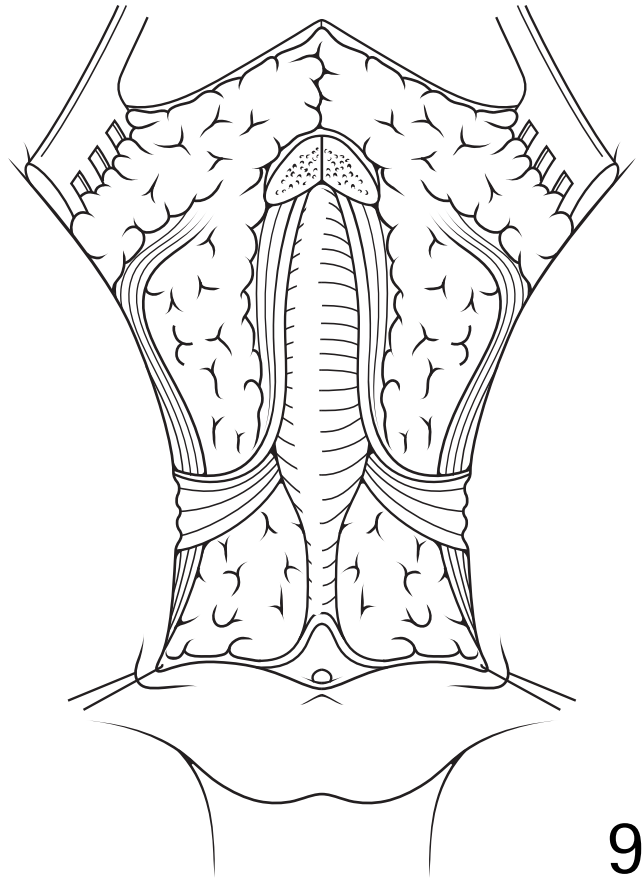
9 The first visceral structure to be found is usually the rectum. The surgeon must be prepared to find bizarre anatomic arrangements of rectum, vagina, and urethra.

At this stage, the surgeon has an objective idea of the complexity of the defect and can directly measure the length of the common channel. If the common channel is shorter than 3 cm, it will usually be possible to mobilize the entire urogenital sinus (vagina and urethra together), as well as the rectum, without opening the abdomen.



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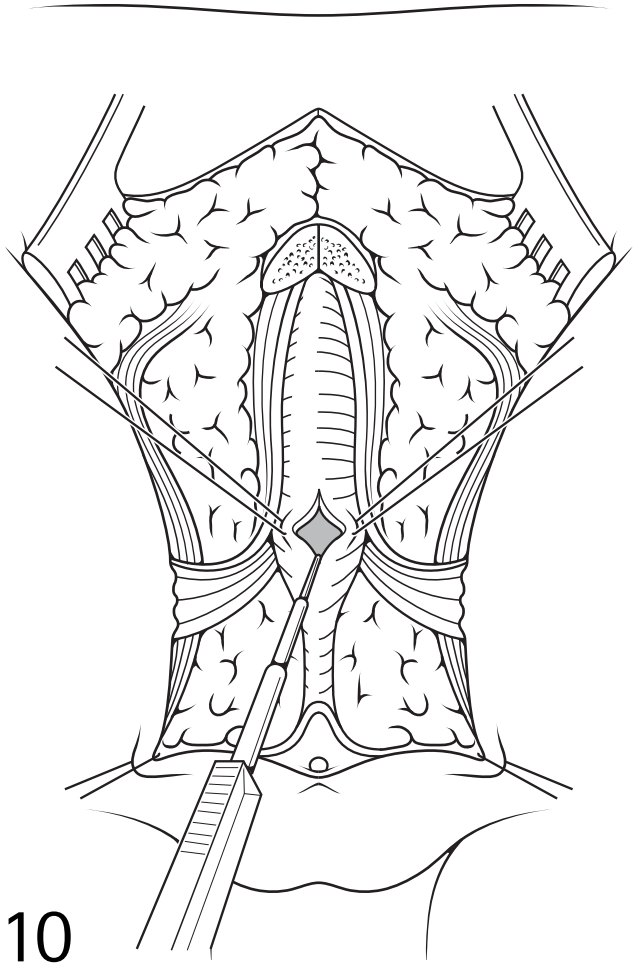

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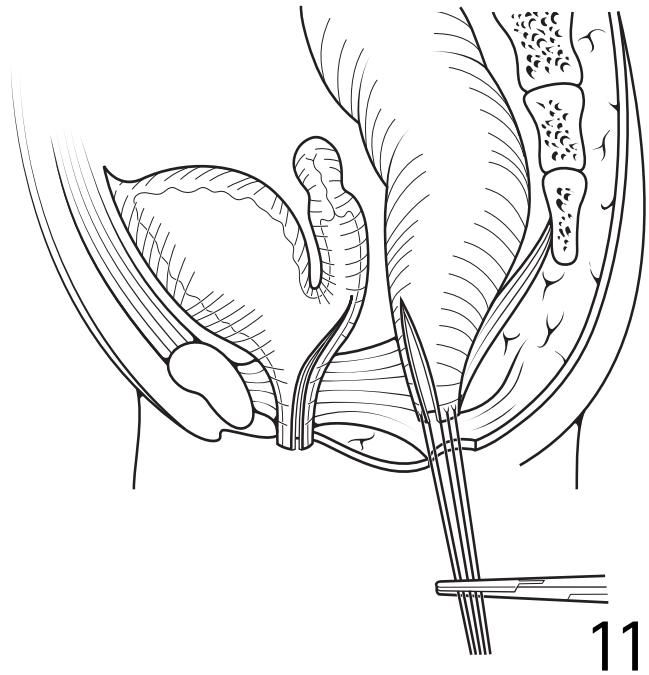
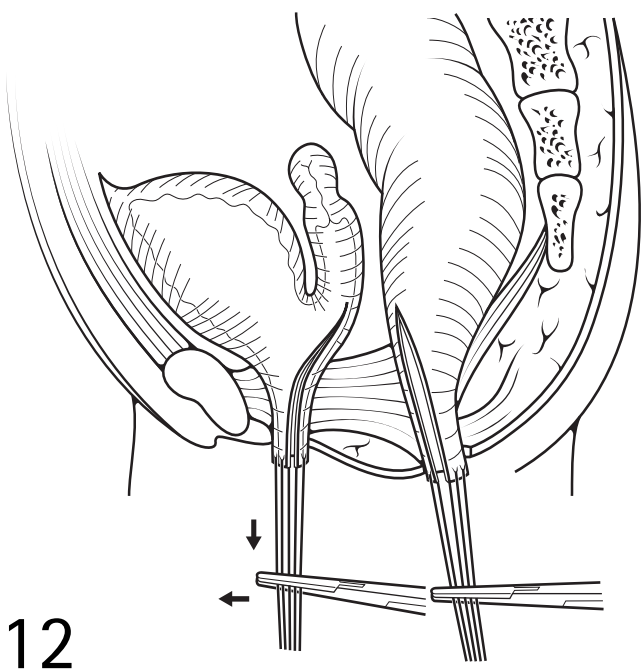
10 The rectum is opened in the midline and silk stitches are placed along the edges of the posterior rectal wall. The incision is extended distally through the posterior wall of the common channel. The entire common channel is exposed, which allows the length of the common channel to be measured and confirmed under direct vision.

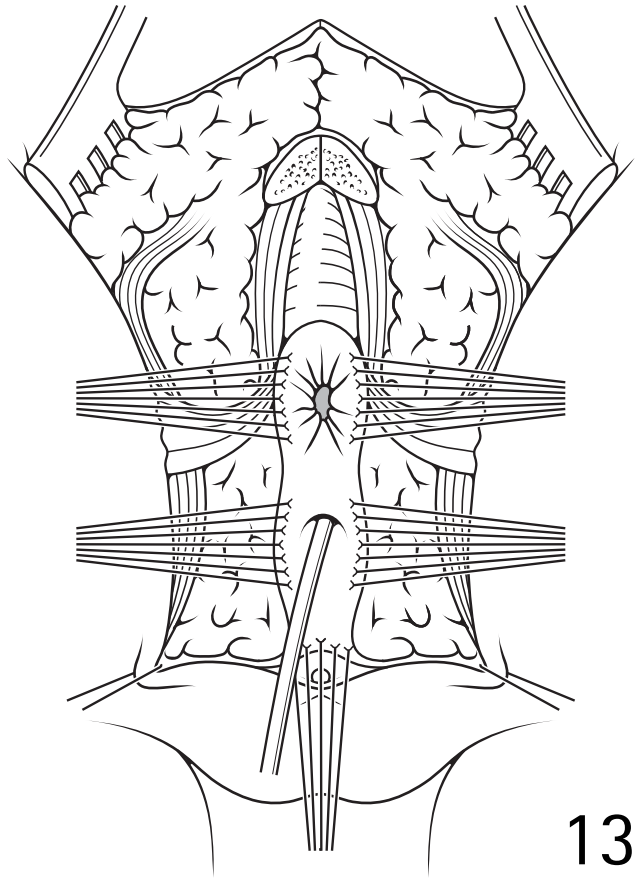
11 The next step consists of separating the rectum from the vagina. This is performed in the same way as described for the repair of rectovestibular fistula (see Chapter 56). Rectum and vagina share a common wall.





12 In the past, we used to separate the vagina from the urinary tract, which was a technically challenging, and not very reproducible maneuver with significant morbidity. Now, once the rectum has been completely separated from the vagina, we perform what we call a total urogenital mobilization. Total urogenital mobilization consists of the mobilization of both the vagina and urethra as a unit without separating one from the other. After the rectum has been separated, multiple silk stitches are placed, incorporating the edges of the vagina and the common channel, in order to apply uniform traction on the urogenital sinus to be mobilized.

13 Another series of fine stitches is placed across the urogenital sinus approximately 5 mm proximal to the clitoris. The urogenital sinus is transected full thickness between the last row of silk stitches and the clitoris, taking advantage of the fact that there is a natural plane between it and the pubis. Working in a bloodless field, one can very rapidly reach the upper edge of the pubis, where an avascular structure – the suspensory ligaments of the urethra and bladder – can be identified that gives support to the vagina and bladder. While applying traction to the multiple stitches, these ligaments are divided, which immediately provides significant mobilization of the urogenital sinus. With this maneuver, one can gain between 2 cm and 3 cm of length.



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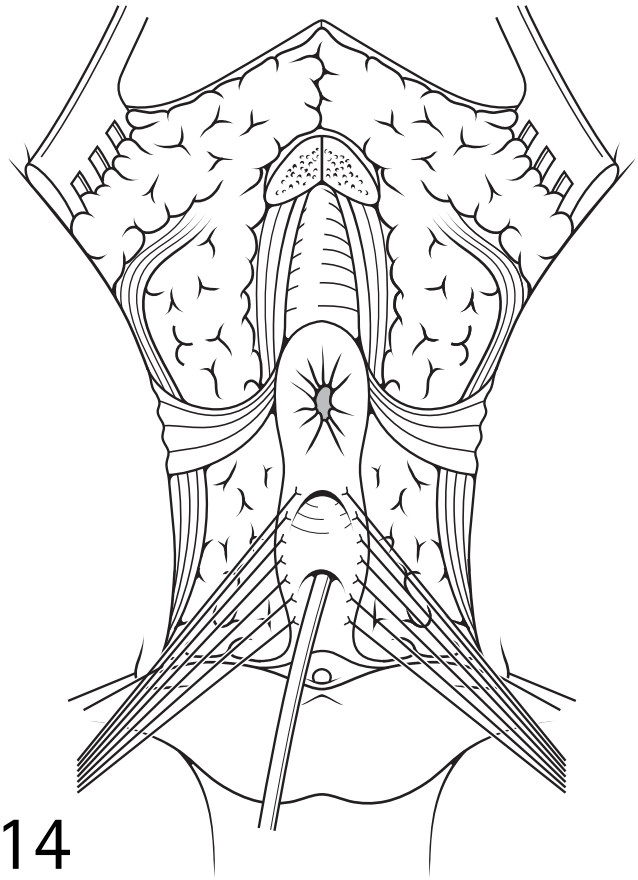
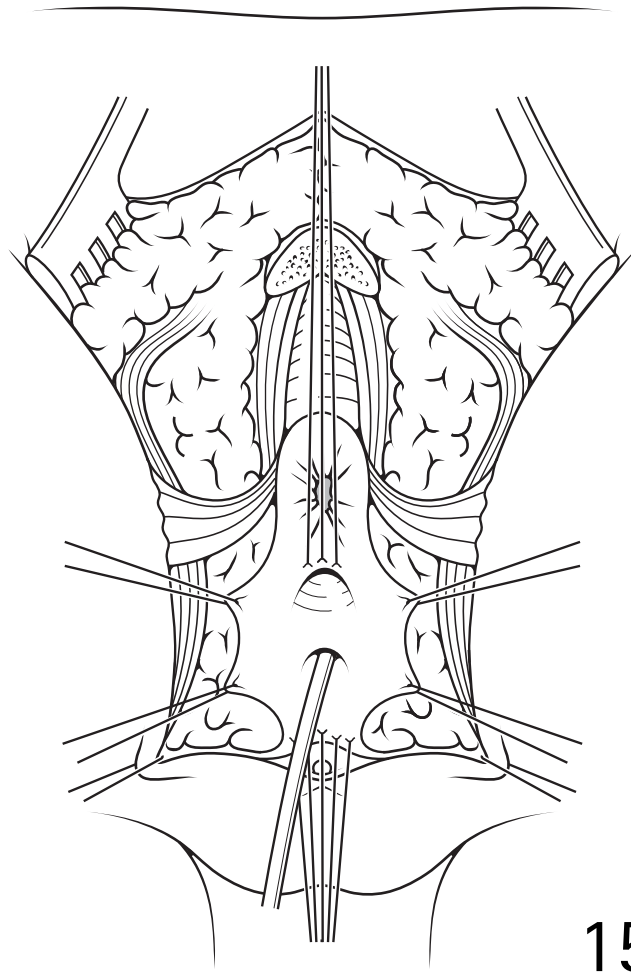


14

14 Additionally, one can then dissect the lateral and dorsal walls of the vagina. This dissection is enough to repair about 60 percent of all cloacas.

15 The urogenital mobilization has the additional advantages of preserving an excellent blood supply to both the urethra and vagina and of placing the urethral opening in a visible location to facilitate intermittent catheterization when necessary. It also provides a smooth urethra that can be catheterized easily. What used to be the common channel is divided in the midline, creating two lateral flaps that are sutured to the skin of the patient's new labia. The vaginal edges are mobilized to reach the skin and to create a good-looking introitus. The limits of the sphincter are electrically determined. The perineal body is reconstructed, bringing together the anterior limit of the sphincter. The rectum is then placed within the limits of the sphincter.



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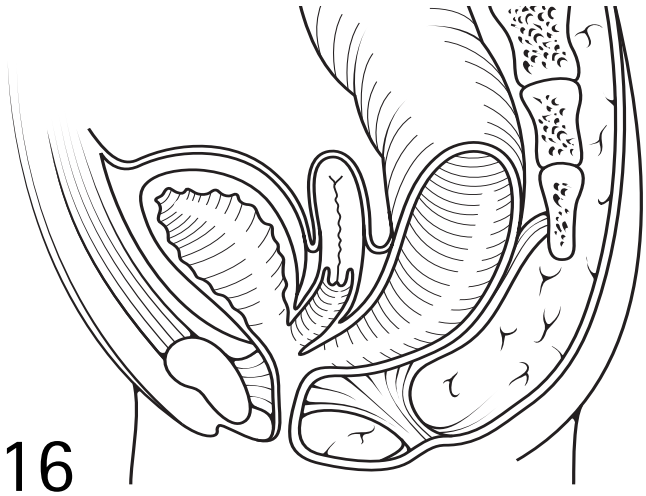


16

CLOACAS WITH A COMMON CHANNEL LONGER THAN 3 CM

16 When the endoscopy shows that the patient has a long common channel, the surgeon must be prepared to face a significant technical challenge.

A surgeon who has minimal experience in performing this repair, or lacks special urological training, should ask for the assistance of an appropriately experienced specialist.

The patient is prepared with a total body preparation so that the surgeon can switch between the prone position and an abdominal approach, as described for rectobladderneck fistula.

The rectum is separated from the vagina and urethra. A very long common channel (more than 5 cm) cannot be repaired by total urogenital mobilization alone, and therefore the channel should be left in place so that it can be used later for intermittent catheterization. In this situation, an attempt should be made to separate the vagina from the urinary tract either from below or from the abdomen by placing multiple 6/0 silk stitches that take in the vaginal wall to try to create a plane of dissection between the vagina and the urinary tract. This is a very delicate, meticulous, and tedious maneuver. With this dissection from the perineum one can gain a separation of the vagina from the urinary tract for approximately 2 cm. The rest of the separation must be completed through the abdomen.

A midline laparotomy is recommended; the bladder is opened in the midline and feeding tubes are placed into the ureters to protect them. A very large common wall between the vagina and the bladder exists, and both the ureters run through this common wall. The ureters sometimes have to be skeletonized during this separation process, and therefore require protection.

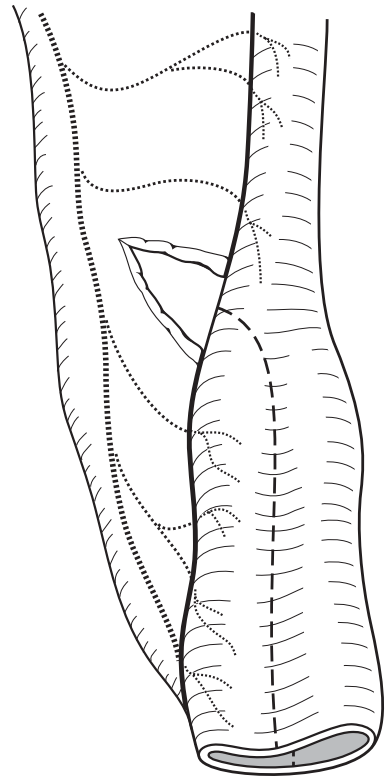
The surgeon must be familiar with the different techniques of ureteral re-implantation because these patients may require this operation during the procedure. Once in the abdomen, the patency of the Mullerian structures can be confirmed by passing a No. 3 feeding tube through the fimbriae of the fallopian tubes and injecting saline solution. If one of the systems is not patent, we recommend its excision, with very careful attention being paid to avoiding damage to the blood supply of the ovary.

When both Mullerian structures are atretic, we recommend leaving them in place, and following the patient closely so that a decision can be made when she reaches puberty.

The procedure continues with the placement of traction sutures are placed in the single uterus or in both hemiuteri. Traction sutures are also placed in the dome of the bladder. With the use of traction on both structures, dissection is initiated between the urinary tract and the vagina. This dissection is continued all the way down to meet the previous dissection initiated from below. Once the vagina(s) has been separated from the bladder and urethra, the surgeon has to a make decisions based on the specific anatomic findings. A single, mid-sized vagina must be separated from the urinary tract, with care being taken to preserve its blood supply from the uterine vessels. When the vagina is found to be too short, some form of vaginal replacement is required.

Vaginal replacement

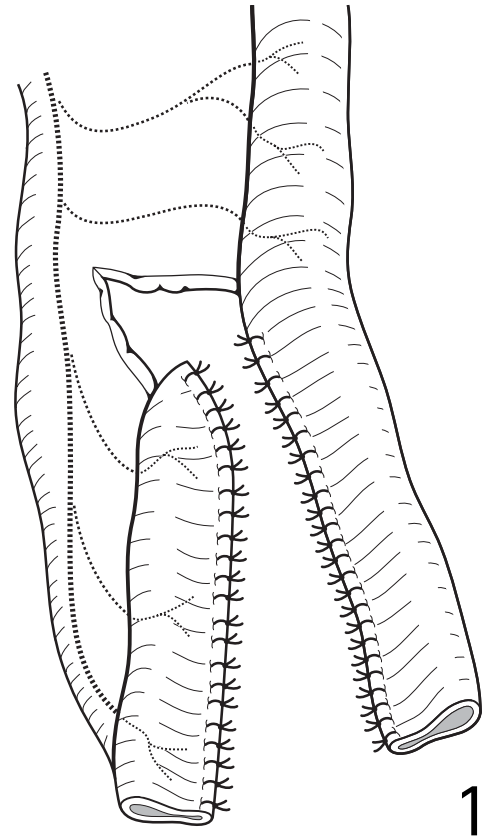
The vagina can be augmented or totally replaced with bowel when it is very small and is located very high or in cases of absent vagina. The choices are rectum, colon, or small bowel.



17a

RECTUM

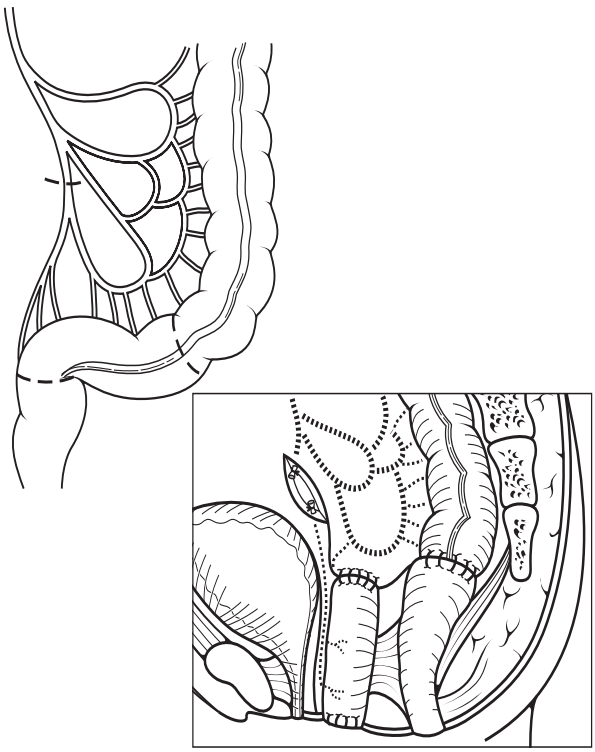
17a,b Replacing the vagina with rectum is only feasible in patients who have a megarectum that is large enough or long enough to be divided into a portion with its own blood supply, which will form the new vagina, and a portion with enough circumference to reconstruct an adequate-sized rectum. The blood supply of the rectum will be provided transmurally from branches of the inferior mesenteric vessels.



17b

COLON

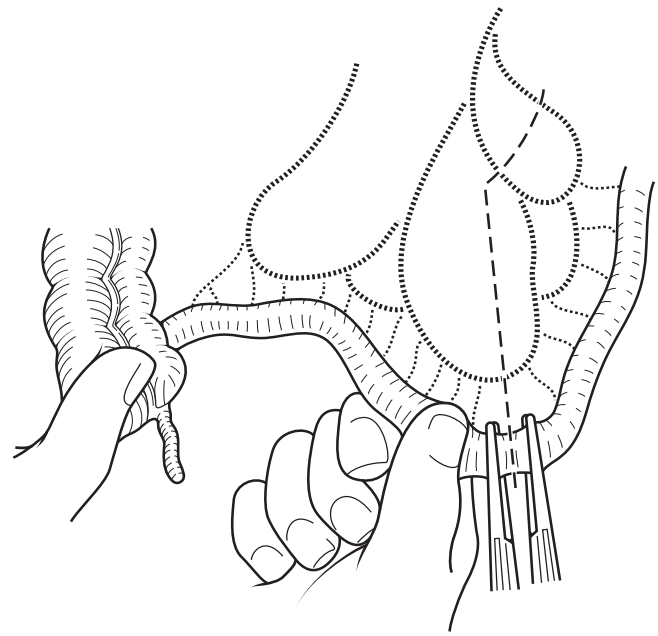
18 Although the colon appears to be an ideal substitute to replace the vagina, this type of reconstruction is sometimes inhibited by the location of the colostomy. When available, the sigmoid colon is preferable. The most mobile portion of the colon must be used in order to have a piece that has a long mesentery. When the patient has internal genitalia or a little cuff of vagina or cervix, the upper part of the bowel used for replacement should be sutured to the upper vagina. When the patient has no internal genitalia (no vagina and no uterus), the vagina is created and left with its upper portion blind; the vagina can then be used for sexual purposes but not for reproduction.



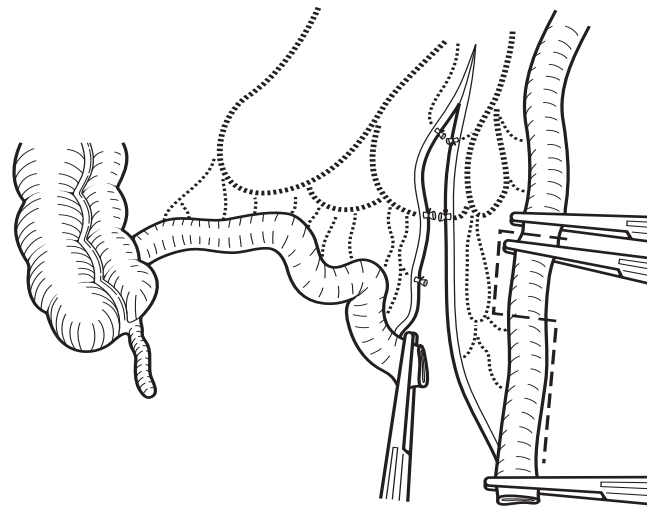
18

SMALL BOWEL

19a,b When small bowel is chosen for reconstruction, the most mobile portion is utilized. The mesentery of the small bowel is longer in an area approximately 15 cm proximal to the ileocecal valve. A segment of this portion of the small intestine is selected, preserving its mesentery.

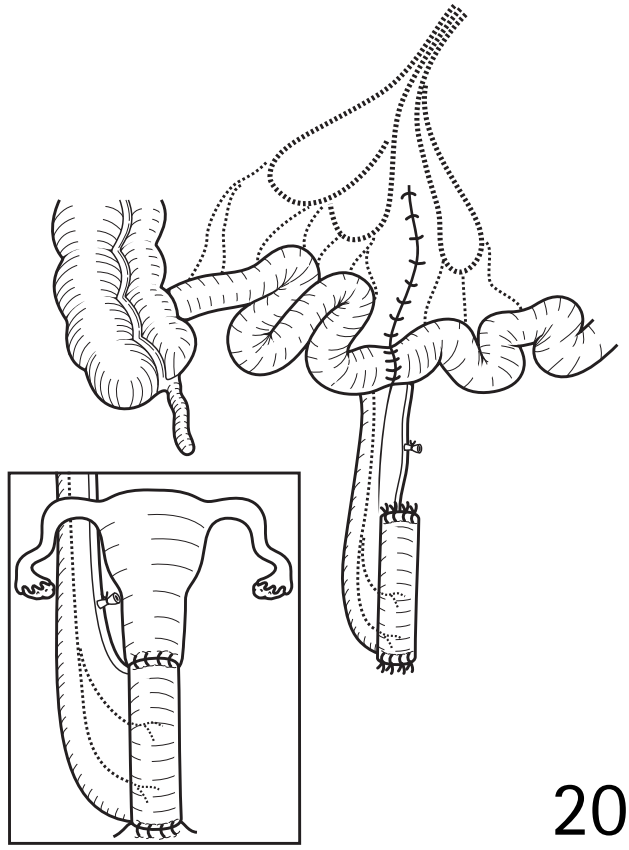


19a



19b

20 The continuity of the small intestine is re-established with an end-to-end anastomosis. Two more anastomoses are necessary: the upper one between the segment of small intestine and the upper vagina, and the lower one between the lower part of the intestine and the perineal skin (new labia).



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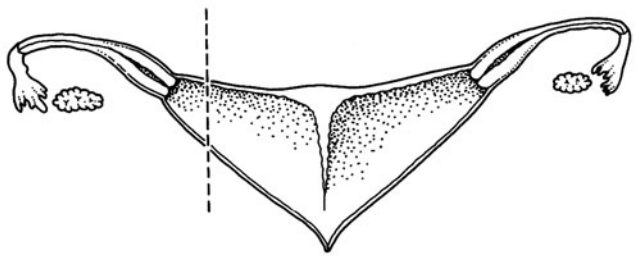
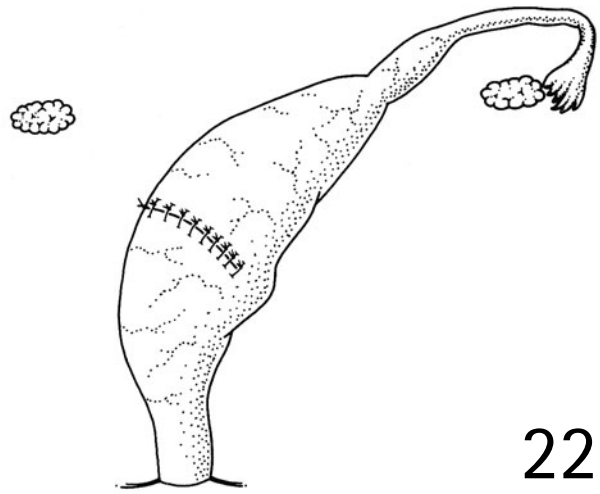


21

Vaginal switch maneuver

21 This maneuver is only applicable in cases with a specific anatomic variant consisting of a long common channel and two hemivaginas with bilateral hydrocolpos. After separation of the vagina from the urinary tract has been completed, the vagina may be too short to fill the gap between vagina and perineum, and it is therefore impossible to move the vagina down. The transverse diameter of both hemivaginas together may be long enough to reach the perineum, provided one of the hemiuteri is sacrificed. During separation of the vagina from the urinary tract, the blood supply to both hemiuteri and hemivaginas reaches the hemiuterus laterally and must be preserved until a decision is reached concerning the type of vaginal mobilization to be performed. One of the hemiuteri and the ipsilateral fallopian tube are resected, with particular care given to preserving the blood supply of the ovary. The blood supply of the hemivagina of that side is sacrificed. The blood supply of the contralateral hemivagina is preserved and is sufficient for both hemivaginas. The vaginal septum is resected, and both hemivaginas are tubularized into a single vagina, taking advantage of their combined long lateral dimension.

22 Then, what used to be the dome of the hemivagina where the hemiuterus was resected is turned down to the perineum. This is an excellent maneuver, but it can only be performed when the anatomic characteristics fulfill the requirements described.



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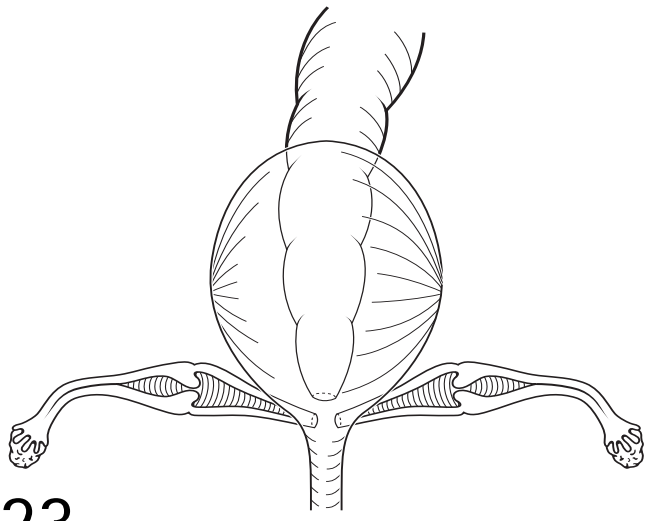

23

23 In the highest type of cloaca, one may find two little hemivaginas attached to the bladderneck or even to the trigone of the bladder. In these cases, the rectum also opens in the trigone. The separation of these structures is done abdominally. Unfortunately, when that separation is completed, patients are often left with no bladderneck or with a very severely damaged bladderneck. At that point, the surgeon must have enough experience to make a decision about whether to reconstruct the bladderneck or to close it permanently. In the first situation, most patients will need intermittent catheterization to empty the bladder, and there is no guarantee that the bladderneck reconstruction will work. In the second situation (permanent closure of the bladderneck), a vesicostomy is created, and the patient will require a continent diversion-type of procedure at the age of urinary continence (3–4 years old). In this particular type of malformation, the patient also needs a vaginal replacement, which should be done in the way previously described.

COMPLICATIONS

Urethrovaginal fistula

Urethrovaginal fistula used to be the most common and feared complication in cases of persistent cloaca, but with the advent of the total urogenital mobilization maneuver, this complication has essentially been eliminated.

Acquired vaginal atresia

Ischemic vaginal fibrosis can occur secondary to an excessive dissection in an unsuccessful attempt to mobilize a very high vagina. To avoid this complication, one of the described vaginal replacement maneuvers should be selected.

POSTOPERATIVE MANAGEMENT AND COLOSTOMY CLOSURE

Postoperatively, patients generally have a smooth course. Pain is not a problem, except for those who have undergone a laparotomy.

The Foley catheter is usually retained for 2–3 weeks. In our series, about 20 percent of the cloaca patients with a common channel shorter than 3 cm require intermittent catheterization to empty the bladder. Patients with common channels longer than 3 cm will require intermittent catheterization 70–80 percent of the time. Therefore, we leave the Foley catheter in place as long as the patient shows signs of edema in the genitalia, and if the urethral meatus is not perfectly visible, we prefer to keep the Foley catheter in place. Once we are able to see the urethral orifice, we remove the Foley

catheter in the clinic and then observe the baby to see if she is capable of emptying the bladder. If the baby cannot pass urine, we teach the caregiver to pass the catheter intermittently. In cases of very long common channels, we prefer to leave a suprapubic tube in place.

Intravenous antibiotics are administered for 48 hours. Prophylactic antibiotics are administered orally to avoid urinary tract infections during the period of catheterization. Antibiotic ointment is applied locally for 8–10 days. Most patients go home after 2 days, or after 3–4 days for those who required a laparotomy.

Two weeks after the repair, anal dilatations are started, following the protocol described for all patients with anorectal malformations.

For patients who had a suprapubic tube, a cystostogram is performed 1 month following surgery to verify the patency of the urethra and rule out the possibility of urethrovaginal fistulas or a urethral stricture. Intermittent clamping of the tube is commenced, and the residual urine is measured as an indicator of the efficiency of bladder function. The suprapubic tube remains in place until we have evidence of a good bladder function or the caregiver learns to catheterize the bladder when required.

At the time of colostomy closure, an endoscopy should be performed to ensure that the repair is intact. If the cloacal repair did not require an abdominal approach, inspection of the Mullerian structures (as already described) should be performed at the time of colostomy closure.

OUTCOME

A patient who has one to three bowel movements per day, remains clean between bowel movements, and shows evidence of feeling or pushing during bowel movements, has a good bowel movement pattern and usually a good prognosis. This type of patient is trainable. A patient with multiple bowel movements or one who passes stools constantly without showing any signs of sensation or pushing usually has a poor functional prognosis.

Cloacas represent a spectrum of defects that can be subclassified on the basis of potential for bowel and urinary control, for which the length of the common channel seems to be the most important prognostic factor.

Many patients suffer from a deficient emptying mechanism of the bladder. However, they do not have the typical 'Christmas tree' type of neurogenic bladder of spina bifida patients, but rather have a flaccid, smooth, large bladder that does not empty completely. Fortunately, most patients with cloacas have a very good bladderneck. The combination of a good bladderneck and a floppy, flaccid bladder makes these patients ideal candidates for intermittent catheterization, which keeps them completely dry. There are two exceptions. One is patients who have a very long common channel, in which the hemivaginas are attached to the bladderneck, and, after these structures have been separated, the patients are left with no bladderneck or a very damaged bladderneck. The second exception is a small number of patients who are born with separated pubic bones. These patients have no bladderneck congenitally and they eventually require a continent diversion type of operation. These patients could be described as having covered exstrophies.

Table 51.2 shows the clinical results obtained in our series of cases with cloacal malformations.

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Table 51.2 *Clinical results in cloacas*

Common channel	Short < 3 cm	Long > 3 cm
Voluntary bowel movement	50/70 (71%)	18/44 (41%)
Soiling	50/79 (63%)	34/39 (87%)
Totally continent[a]	25/50 (50%)	5/18 (28%)
Constipated	34/85 (40%)	17/45 (38%)
Urinary function		
Normal	59/82 (72%)	11/54 (20%)
Dry with intermittent catheterization	23/82 (28%)	43/54 (80%)
Through the urethra	16	20
Through Mitrofanoff or similar	7	23

[a] Voluntary bowel movement and no soiling.

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52

Rectal biopsies

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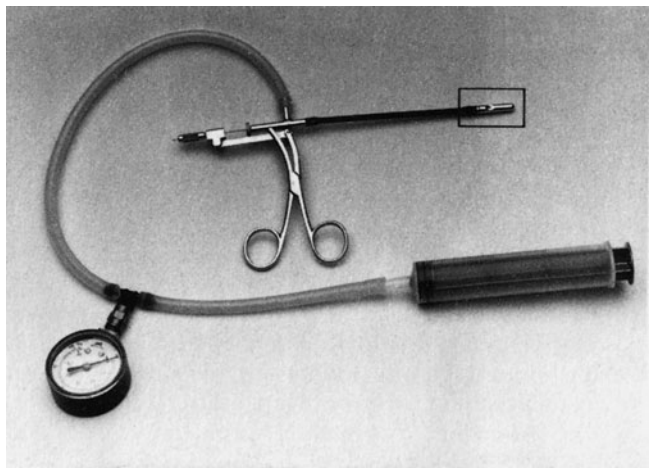
Hirschsprung's disease should be considered in neonates presenting with abdominal distension, delay in passing meconium, and bile vomiting, and in the older child with intractable constipation. Although the diagnosis may be suggested by a contrast enema showing the 'transitional zone' or by anorectal manometry, it can only be established with certainty by histologic examination of the affected, aganglionic bowel wall. This can be achieved most easily by obtaining a biopsy of the mucosa and, most importantly, the submucosa of the rectum.

PRINCIPLES AND JUSTIFICATION

Biopsy of the rectum using a suction biopsy tube is a common procedure in pediatric surgery and has generally superseded the former techniques of open rectal biopsy and punch biopsy with a sigmoidoscope or speculum. The technique needs to be carried out with meticulous attention to detail in order to obtain a suitable diagnostic specimen of rectal mucosa with sufficient submucosa attached on each occasion. The procedure may be performed in the ward or clinic without anesthesia, and is painless provided the biopsy is taken at least 2.5 cm above the anal verge in the neonate and 3.5 cm in the older child (i.e., above the sensitive zone of the anal canal). Prophylactic antibiotics are given.

INSTRUMENTATION

1,2 The original suction biopsy instrument was devised by Noblett and consists of a blunt-ended tube with a 3 mm side hole 1 cm from the tip attached to suction tubing. There are marks on the body of the instrument to indicate the level of biopsy. It has an in-line manometer to measure the suction pressure. When inserted into the rectum and suction applied, a portion of the superficial rectal wall is drawn into the side hole. Triggering the concealed circular knife then completes the biopsy. Other similar biopsy instruments are available, all using the principle of the Noblett biopsy forceps.



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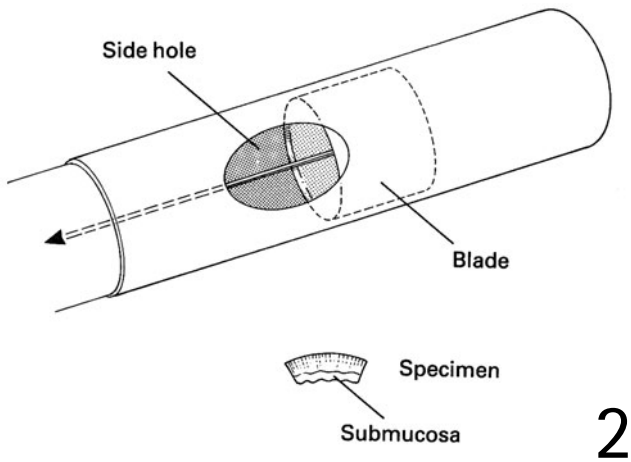

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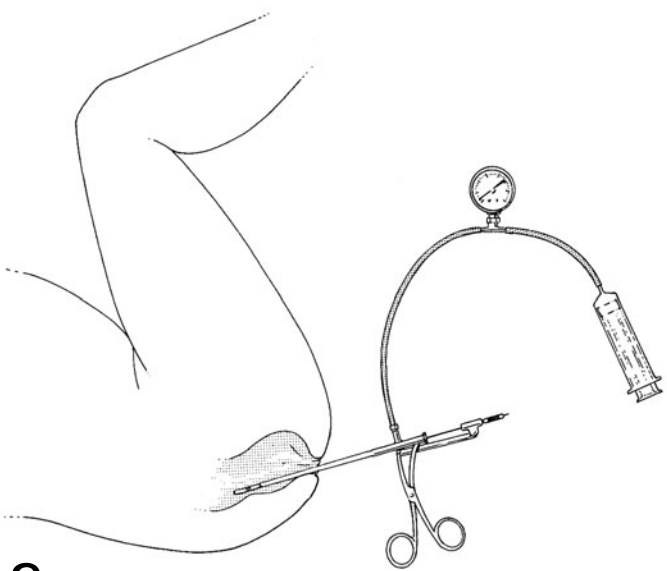


3

OPERATIVE TECHNIQUE

3 The neonate is usually held in the lithotomy position, although the left lateral knees-bent position is more comfortable for the older child. It is essential to confirm that vitamin K has been given to neonates. If the rectum is full, a finger may be passed alongside the instrument, holding it firmly against the rectal wall. The lubricated instrument is inserted into the anus and the side hole positioned initially about 2 cm above the dentate line. This is the minimum distance and avoids the normal hypoganglionic zone and diagnostic confusion. The biopsy specimen should always be taken from the posterior or lateral rectal wall because of the increased risk of perforation into the rectovesical or rectovaginal pouch of the peritoneal cavity if the biopsy is full thickness and anterior.

Suction is then applied to a maximum pressure of not more than 300 mmHg (20–30 cmH_2O) by drawing on a 20 mL syringe attached to the suction tubing. After 2–3 seconds, the knife is triggered and the instrument withdrawn. The end of the suction biopsy instrument is cautiously unscrewed and the biopsy specimen removed with a needle. The specimen is usually about 3×1 mm, and the critical submucosa can be recognized as a definite whitish layer (see Illustration 2). The procedure may then be repeated at 3 cm and 4 cm above the anal verge.

A rectal examination should be carried out after completing the biopsy to exclude active bleeding, and the patient should be observed carefully for at least a further hour.

The method of processing suction biopsies must be ascertained before the procedure, as dictated by specific laboratory requirements. However, fresh specimens are usually requested, and the biopsy material should be placed on a piece of moistened filter paper marked with the level. It is essential to avoid drying out during transport. Although definitive diagnosis usually requires a combination of paraffin section histology and histochemistry (e.g., acetylcholinesterase activity), a diagnosis based on frozen section of the biopsy is possible.

POSTOPERATIVE CARE

Complications

The possible complications are as follows:

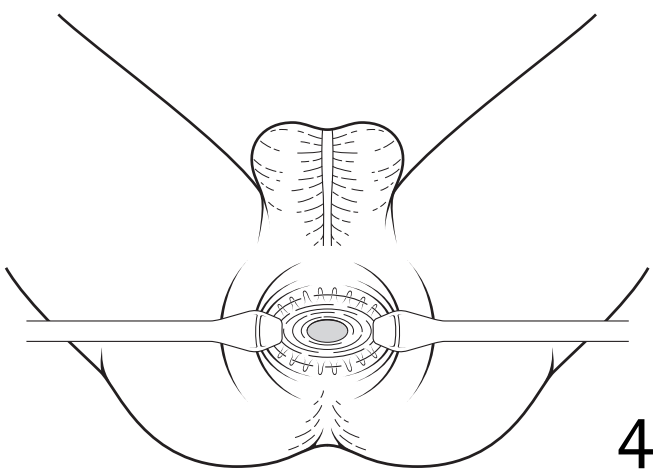
- Inadequate specimen retrieval. This can be avoided by meticulous attention to detail, ensuring that the biopsy instrument is always cleaned carefully after use and that the blade is sharpened at regular intervals.
- Perforation. In one study, full-thickness biopsies were identified histologically in 1 percent of 406 patients undergoing 1340 consecutive biopsies. Although these perforations can generally be treated conservatively with antibiotics, nasogastric suction, and intravenous fluids, a laparotomy may be needed.
- Bleeding.
- Pelvic sepsis. This occurs as a result of perforation into the perirectal tissues.

OPEN RECTAL BIOPSY

Open rectal biopsy is needed when the specimen obtained with the suction biopsy instrument is inadequate or where the instrument is unavailable.

Position

The neonate or infant is held in the lithotomy position; an older child will need to be placed in stirrups in the lithotomy position. Prophylactic broad-spectrum antibiotics are given.



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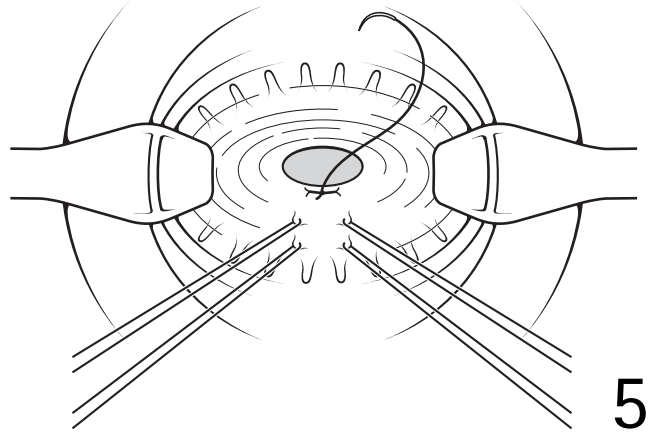
Procedure

4 The anal orifice is digitally dilated. It is held open either with a Park's retractor (or self-retaining retractor) or by an assistant holding two small Langenbeck's retractors.

5 Commencing 1 cm above the dentate line, two parallel sutures are placed in the posterior rectal wall 0.5 cm apart. With the assistant applying traction on these two stay sutures, a second set of sutures is placed a further centimeter proximally on the posterior rectal wall. Traction is now exerted on all four stay sutures and a final suture is placed in the midline 1 cm above the second set of sutures. This suture is tied and the needle is left intact, as it will be used to repair the defect once the specimen has been taken.



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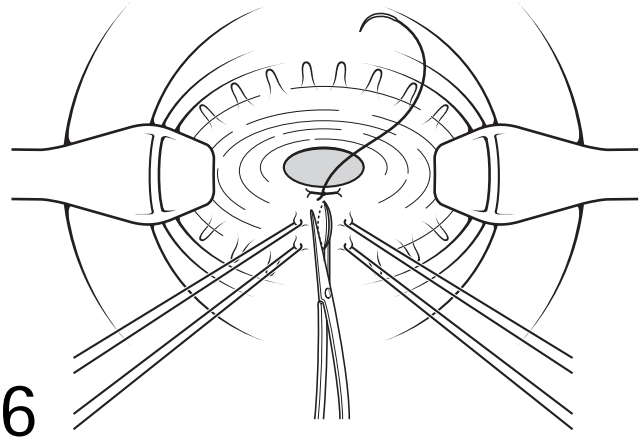

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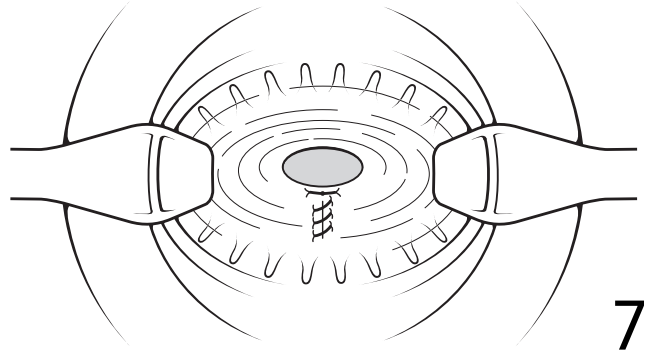
6 Using sharp-pointed scissors, a specimen comprising mucosa/submucosa or full thickness of the rectal wall is taken between the stay sutures commencing 2 cm *above* the dentate line and continuing as a strip to reach the apical suture. The specimen is submitted for histopathologic examination as in the suction technique.

7 Hemostasis may be achieved with bipolar diathermy or, more usually, by suturing the defect with a running locking suture from above.



7

Complications

- Hemorrhage.
- Infection.

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53

Laparoscopic pull-through in the neonate

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HISTORY

Harold Hirschprung described the disease that bears his name at a pediatric conference in 1886 after observing two children die from repeated bouts of enterocolitis. Although recognized as a disease entity in the early 1900s, the pathophysiology of Hirschsprung's disease (HD) was not clearly elucidated until 1948, when the invariable absence of ganglion cells in the distal bowel was identified in patients afflicted with this disease.

Understanding the importance of distal aganglionosis led Swenson to propose the principles that have subsequently formed the basis for the successful surgical management of this complex disease. He proposed that the aganglionic bowel be resected, with anastomosis of normal ganglionated bowel to the anus. Surgical correction of HD was historically accomplished using two or three stages, with initial fecal diversion, subsequent proctectomy and pull-through procedure, and finally stomal takedown for re-establishing bowel continuity.

In 1980, So and colleagues described neonatal pull-through in a single stage without fecal diversion. More recently, minimally invasive approaches have further improved the surgical treatment of HD.

PRINICIPLES AND JUSTIFICATION

Hirschsprung's disease is a disorder marked by abnormal innervation and motility of the rectum and distal bowel. This dysmotility leads to marked constipation or obstruction, often symptomatic in the newborn period. The severity of the symptoms and time of presentation of the disease can vary, but in its most severe form enterocolitis can present as a life-threatening medical emergency.

Patients with HD are found histologically to have absence of ganglion cells in the affected segment, increased acetylcholinesterase activity in the mucosa and submucosa, and hypertrophied nerve trunks. These findings are associated with the failure of coordinated relaxation distal to the peristaltic wave in the involved segment of bowel. The most distal rectum is always affected by the disease, but the proximal extent varies and can even extend into the small intestine in rare cases. Ninety percent of cases involve only the rectum and/or the sigmoid colon.

Up to 10 percent of patients with HD have a familial form of the disease. Total colonic HD is usually transmitted in a familial fashion. Genetic causes are still unknown, although the RET proto-oncogene on chromosome 10, the endothelin receptor B (*EDNRB*) gene on chromosome 13, and the endothelin 3 (*EDN 3*) gene on chromosome 20 have been most often associated with HD.

Definitive treatment of HD is surgical. Various operations have been reported, but all involve resection of the aganglionic bowel with subsequent pull-through of ganglionated bowel to the anus with anastomosis. Historically, almost all patients with HD were initially diverted. More recently, primary pull-through without enterostomy has been successfully employed in many cases.

The authors prefer primary pull-through for most patients with HD. Contraindications to primary repair include associated life-threatening anomalies, deteriorating general condition or malnutrition, severe unremitting enterocolitis, and massive extended dilatation of the proximal ganglionated bowel. The authors also prefer primary diversion for infants with total colonic aganglionosis, followed by a laparoscopic-assisted Duhamel procedure and simultaneous ileostomy takedown. The Duhamel procedure may offer the advantage of creating a larger rectal reservoir in selected patients with near-total or total colonic HD.

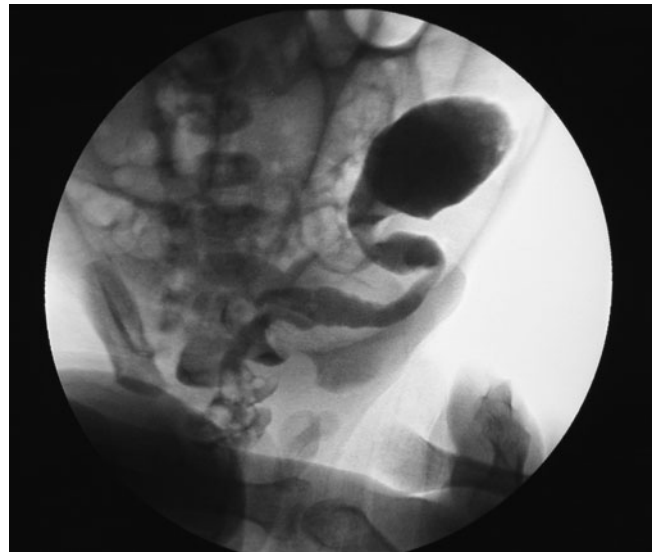
For the usual left-sided Hirschsprung's patient, laparoscopic-assisted transanal pull-through confers several advantages. Laparoscopic biopsies provide a minimally invasive way to confirm the level of aganglionosis prior to division of the colon mesentery or ablation of the rectum. This may be particularly important in neonatal or very young patients with unsuspected long-segment HD (proximal to the splenic flexure), as it allows for a delay in definitive operation until permanent section can confirm the level of aganglionosis, avoiding unnecessary resection of long segments of colon due to a technical error in rapid frozen section analysis. Furthermore, laparoscopy provides a way to develop a tension-free mesocolic pedicle without the morbidity of laparotomy. The pull-through segment can be inspected to ensure the absence of twisting or rotational abnormalities. Overall, the laparoscopic-assisted transanal pull-through allows for versatility in the surgical treatment of all forms of HD and avoids the morbidity of laparotomy.

PREOPERATIVE ASSESSMENT AND PREPARATION

Diagnosis

The diagnosis of HD should be considered in any child with chronic constipation dating back to the newborn period. Failure to pass meconium in the first 24 hours of life is considered abnormal and warrants a work-up for HD. Presentation may include bilious emesis, with plain radiographs consistent with distal bowel obstruction. Failure to thrive in older children or poor feeding in neonates may be associated with the disease. In addition, patients with HD may present with enterocolitis, fever, abdominal distension, and sepsis.

On physical examination, abdominal distension is often a prominent feature. Digital rectal examination may lead to explosive diarrhea following removal of the examiner's finger. Plain radiographs are often suggestive of a distal bowel obstruction, and often show a very dilated colon immediately proximal to the aganglionic segment.



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1 Contrast enema is useful to help identify a potential transition zone. Classically, the colon will have a tapered narrowing at the level of aganglionic bowel, with immediate proximal dilatation. Mucosal cobblestoning can be present and suggests chronic or recurrent enterocolitis. Contrast enema is contraindicated in a patient presenting with acute enterocolitis. In neonates, contrast enema can fail to show a distinct transition zone. Failure to clear the barium on post-evacuation films 12–24 hours later can strongly implicate HD as a possible diagnosis.

Although used less often than rectal biopsy, anorectal manometry with expert clinical oversight can be a sensitive method to diagnose HD. This test is often used in older children to help distinguish HD from encopresis. Findings include failure of relaxation with a distending balloon stimulus, and high baseline resting pressures.

The gold standard for the diagnosis of HD remains a rectal biopsy. In neonates and small infants, suction rectal biopsy allows for detection of the absence of ganglion cells in the submucosal region. In addition, positive findings for HD include hypertrophied nerve trunks and increased submucosal acetylcholinesterase activity. In older children, a full-thickness rectal biopsy may be necessary in order adequately to identify the absence of ganglion cells. In some countries, the elevated submucosal acetylcholinesterase activity will confirm the diagnosis on suction rectal biopsy alone. The disadvantages of full-thickness rectal biopsies include scarring of the posterior rectum, making eventual pull-through more difficult; the need for a general anesthetic; and an increased risk of bleeding, localized infection, and leak. The mucosa just proximal to the dentate line is often hypoganglionated in normal patients, so specimens should be obtained from at least 2 cm proximal to the dentate line.

Preparation

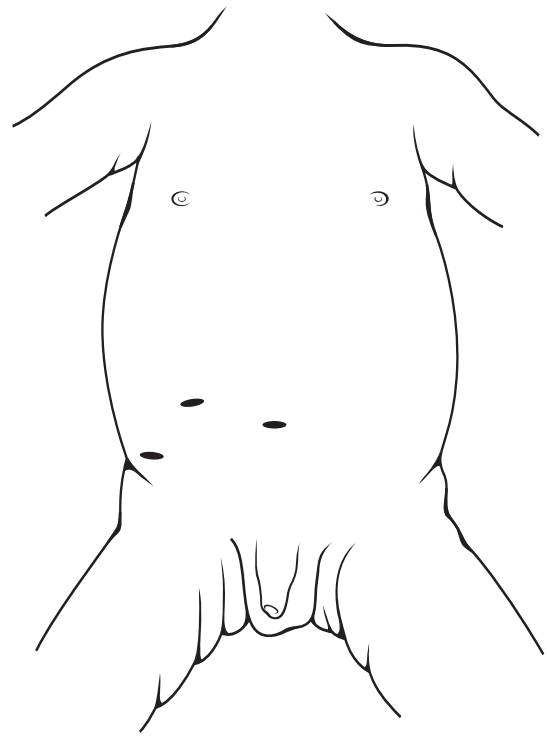
For primary endorectal pull-through in the neonate, rectal irrigations and anorectal stimulation are all that is required for bowel preparation. Older children require the combination of rectal irrigation, enemas, and enteric lavage. All patients receive preoperative intravenous broad-spectrum antibiotics, and older children are also given non-absorbable enteric antibiotics as part of their bowel preparation.

General endotracheal anesthesia is administered and the patient is positioned in the dorsal lithotomy position. Infants can be positioned transversely on the operating table and prepared in a sterile manner circumferentially from nipples to toes. Older children are positioned with stirrups in the dorsal lithotomy position at the foot of the table. Nasogastric suction is used to decompress the stomach, and paralytic agents are used for both the laparoscopic and transanal portions of the procedure.

OPERATION

Port placement

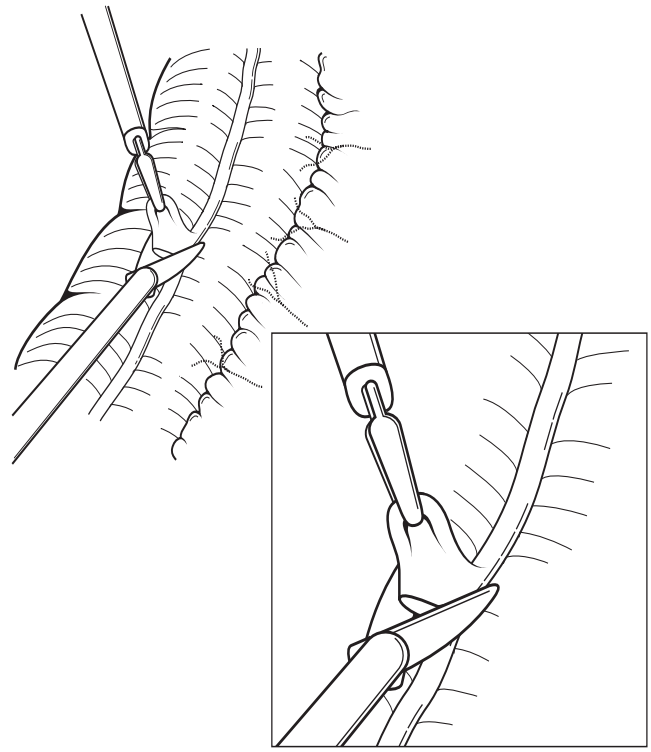
2 The first step in the operation is to determine the level of the transition zone. Usually, three trocars are used. The first is placed through the umbilicus, and the remaining two in the right abdomen. The authors prefer a radially dilating 5 mm trocar for the umbilicus, and 4 mm ports for the other access points.



2

Biopsy

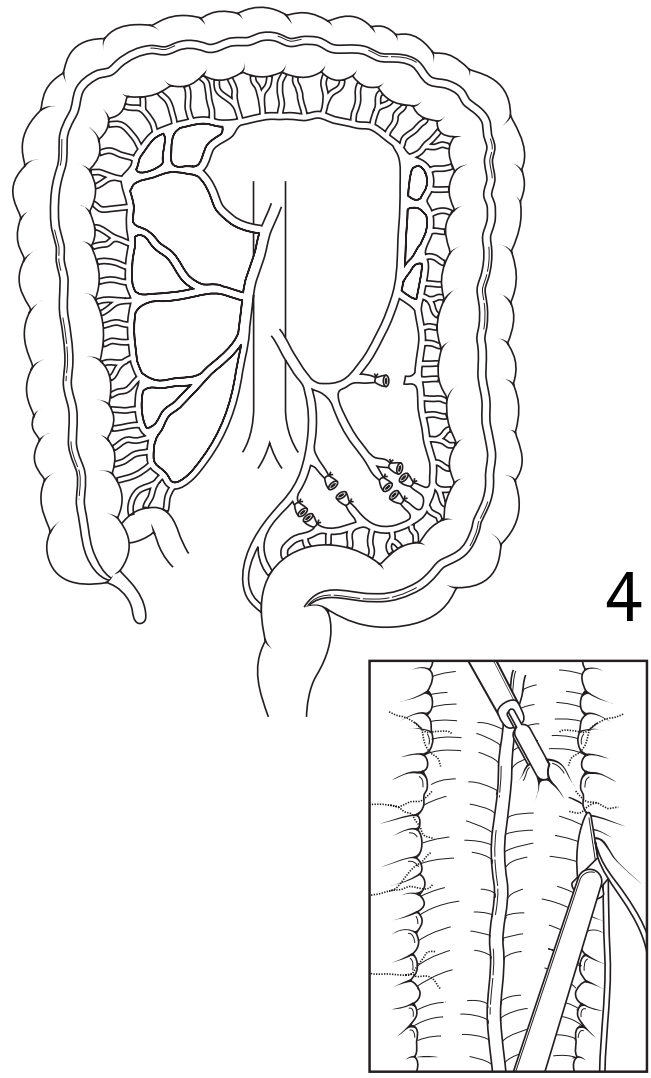
3 Seromuscular biopsies are obtained by grasping the serosa with a fine-tipped grasper and using endoscopic scissors to incise the seromuscular layer down to the mucosa. A small flap is dissected and the seromuscular biopsy is completed with scissors. Perforation or bleeding is closed with 3/0 sutures laparoscopically. These sutures also mark the biopsy sites. Usually three or four biopsies are obtained and sent for rapid frozen section analysis. If all biopsies are abnormal, specimens are taken from the more proximal colon until normal ganglionated bowel is identified. If the transition zone is proximal to the splenic flexure, definitive operation is delayed for permanent section analysis.



3

Laparoscopic division of mesocolon

4 The surgeon should delay division of the mesocolon and the endorectal proctectomy until normal bowel is identified. The mesocolon is divided using hook electrocautery for infants or an ultrasonic scalpel for older children. The mesocolon should be divided close to the aganglionic bowel. In long-segment HD, if the normal ganglionated pull-through segment needs to be pedicled to provide adequate length to bring it down to the pelvis, the mesocolon is divided proximal to the marginal artery in the ganglionated segment to allow for a tension-free anastomosis.



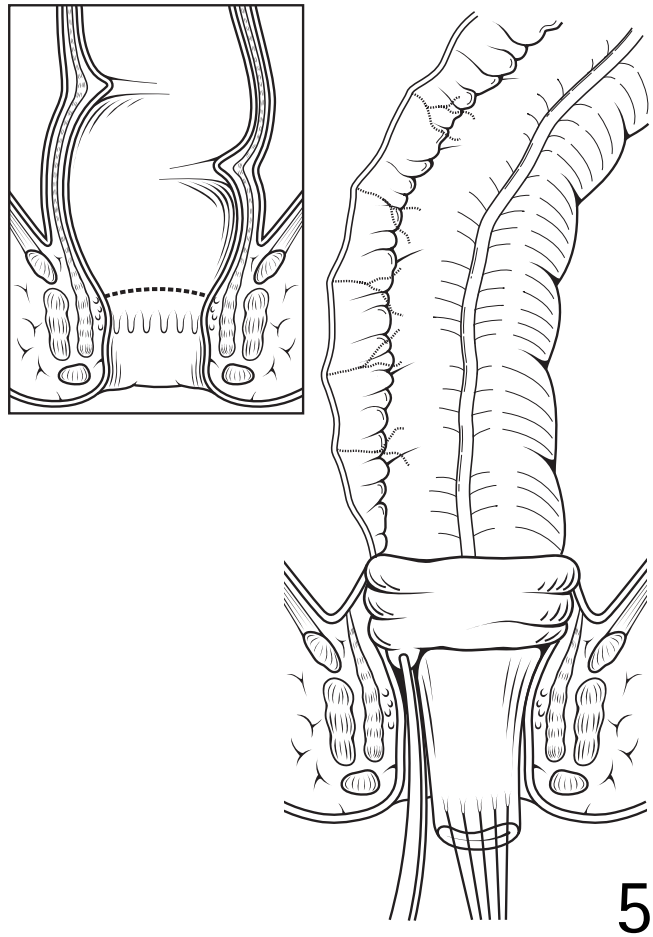
4

Transanal dissection: exposure

Retraction of the anus is accomplished using eight interrupted, circumferential sutures around the anus through the dentate line and the perianal skin 3 cm radially from the anus. When these sutures are tied, the anus is everted, providing excellent exposure and simplifying the submucosal dissection.

Submucosal dissection

5 A circumferential incision is made 2 mm above the dentate line and 3/0 silk sutures are placed through the proximal rectal mucosa to be used as stay sutures for retraction. The endorectal dissection is continued in the submucosal plane between the mucosa and the muscular layers of the rectum. This dissection proceeds until the muscular cuff of the rectal wall freely intussuscepts. Cessation of the bleeding that accompanies the endorectal dissection is another indication that dissection is proximal enough, i.e., the peritoneal cavity and devascularized rectosigmoid colon have been reached.



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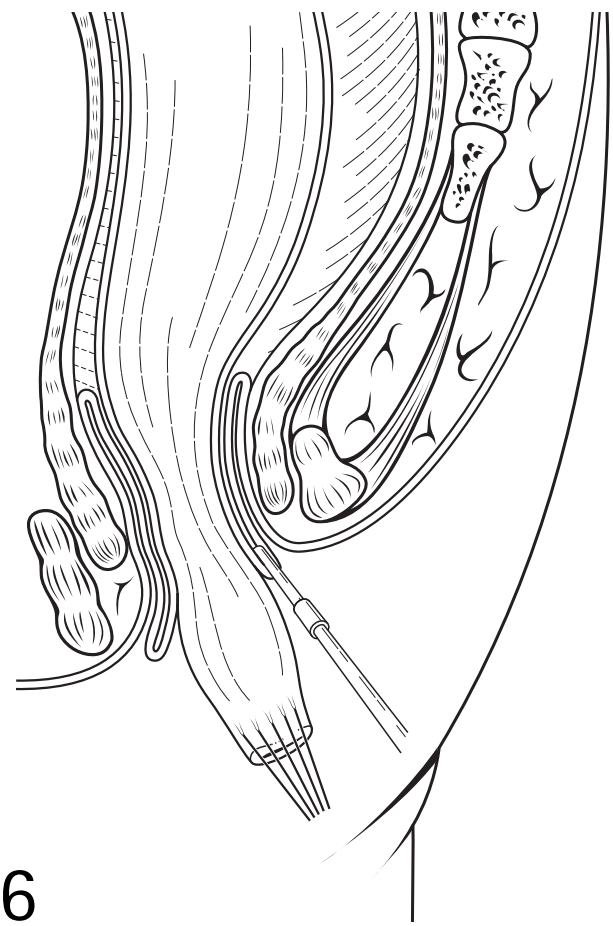


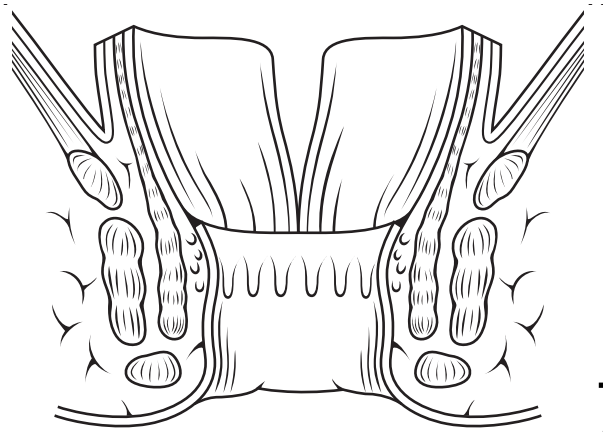
6

Converting the endorectal dissection to full-thickness dissection

6 An incision is made in the posterior wall of the muscular cuff. If a free intraperitoneal plane is then achieved, the muscular cuff is divided circumferentially, converting the submucosal dissection into a full-thickness dissection.

Division of the muscular cuff posteriorly

7 The pull-through colon is then retracted up into the abdomen to allow better visualization of the remaining muscular cuff, which should measure no longer than 5 cm. This muscular cuff is divided posteriorly all the way down to the level of the intended anastomosis. Dividing this cuff is important, as the endorectal dissection does not stretch it as much as transperitoneal dissection. This undivided contracted muscular cuff is prone to constrict the neorectum and prevent the development of an adequate neorectal reservoir.



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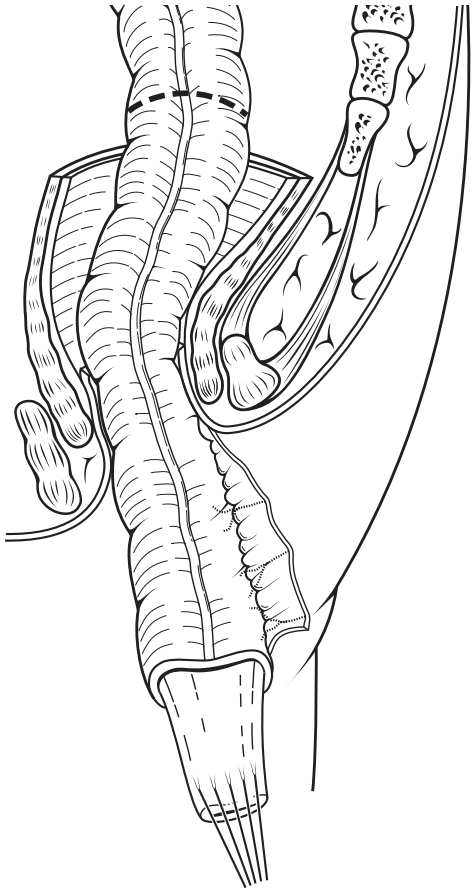


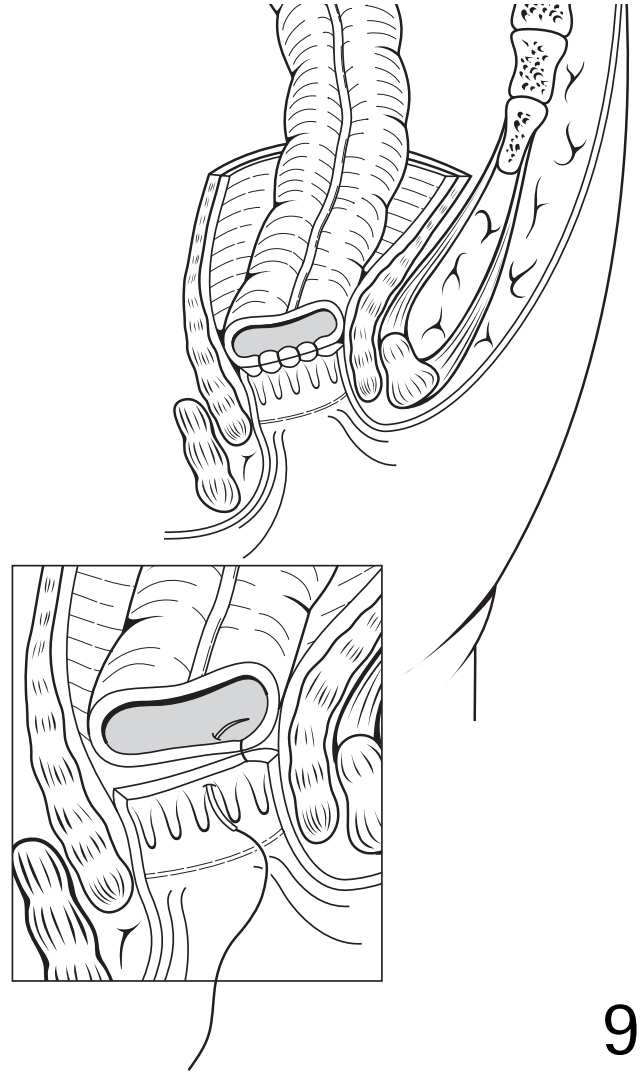
8

Pull-through

8 The aganglionic colon is pulled through the divided muscular sleeve out onto the anus. Previously placed biopsy sutures identify the level of the transition zone. The authors prefer to continue the colon resection 10–20 cm above the biopsy-documented transition zone to avoid problems with potentially dysmotile bowel.

Anastomosis

9 Interrupted, fine, absorbable sutures are placed circumferentially between the pull-through colonic segment and the anus at the dentate line. A watertight anastomosis should be created unless the anastomosis is protected by a diverting stoma.



9

Laparoscopic inspection and closure of the mesenteric defect

The pneumoperitoneum is re-established and the pull-through segment is inspected to ensure that the neorectum is not twisted as it courses into the pelvis. If the pedicle allows a potential space for herniation of small bowel underneath mesentery of the pull-through segment, laparoscopic sutures are placed between the mesentery and the divided sigmoid mesentery. The umbilical port site is closed with a single 2/0 or 3/0 interrupted monofilament suture and the remaining sites are closed with strips.

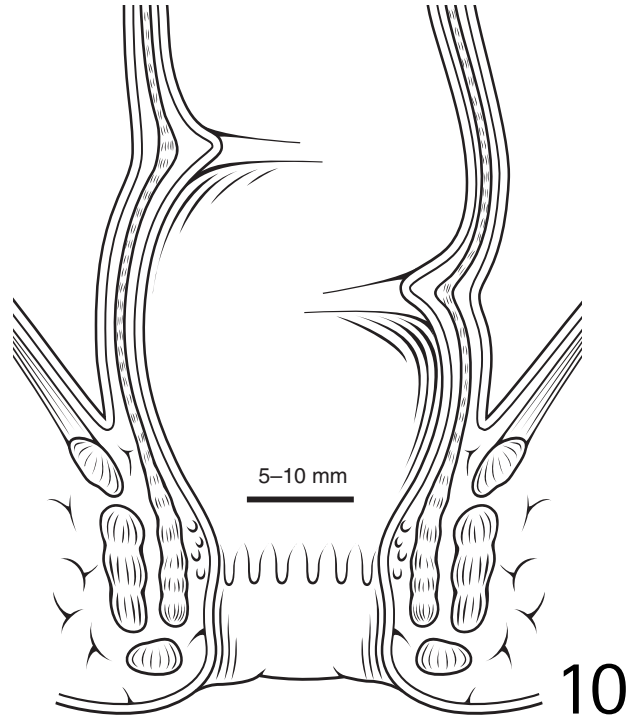
Laparoscopic Duhamel procedure

DIVISION OF THE COLON

For patients with total colon aganglionosis, the authors prefer a laparoscopic Duhamel approach. Initial port placement and biopsies are performed as previously described. The mesocolon is divided close to the colon throughout its length laparoscopically. The right lower quadrant port is upsized to a 12 mm radially dilating trocar and the distal rectum at the level of the peritoneal reflection is divided using a laparoscopic gastrointestinal stapling device. Blunt laparoscopic dissection is used to enlarge the space posterior to the rectum all the way down to 1 cm above the dentate line.

PLACEMENT OF TRANSANAL TROCAR

10 Traction sutures are placed in the mucosa as for endorectal dissection, but an incision is made posteriorly about 1 cm above the dentate line in a transverse direction. A 12 mm radially dilating trocar is then placed through the incision into the dissected space posterior to the rectum inside the peritoneal cavity.



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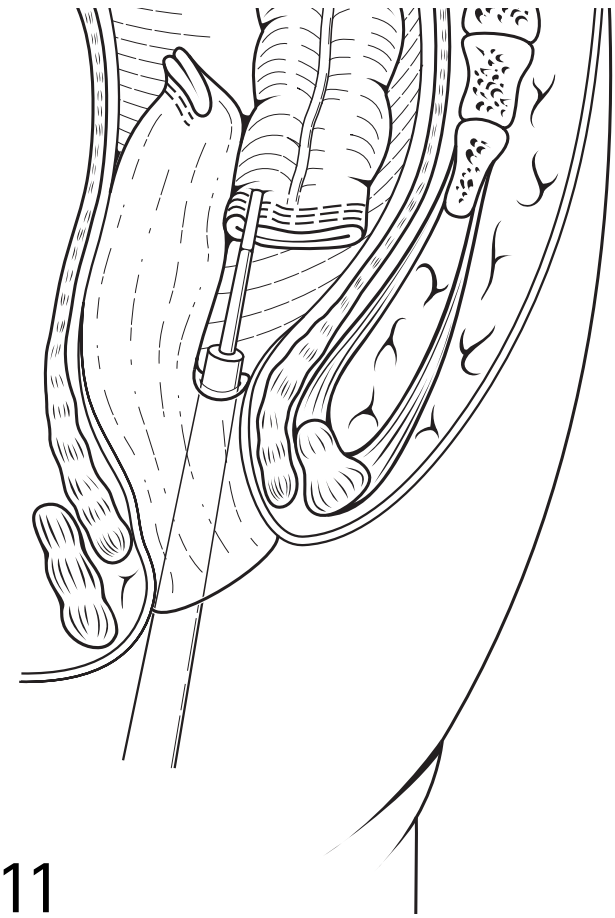


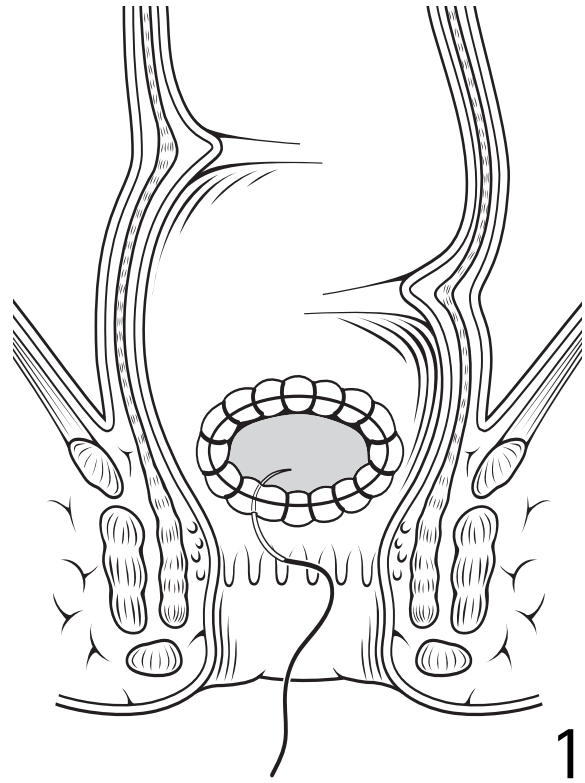
11

PULL-THROUGH

11 The stapled distal end of the colon for resection is grasped with a laparoscopic grasper and is pulled out through the posterior rectal wall trailing behind the trocar. The ganglionated right colon or ileum is brought down through the posterior rectal incision as well, until the level of anastomosis is established. Laparoscopy from above is used to confirm that the pull-through segment is not twisted and that the mesentery is situated posteriorly.

ANASTOMOSIS

12 An anastomosis is then created by suturing the ganglionated pull-through segment to the posterior rectal defect using interrupted absorbable sutures.



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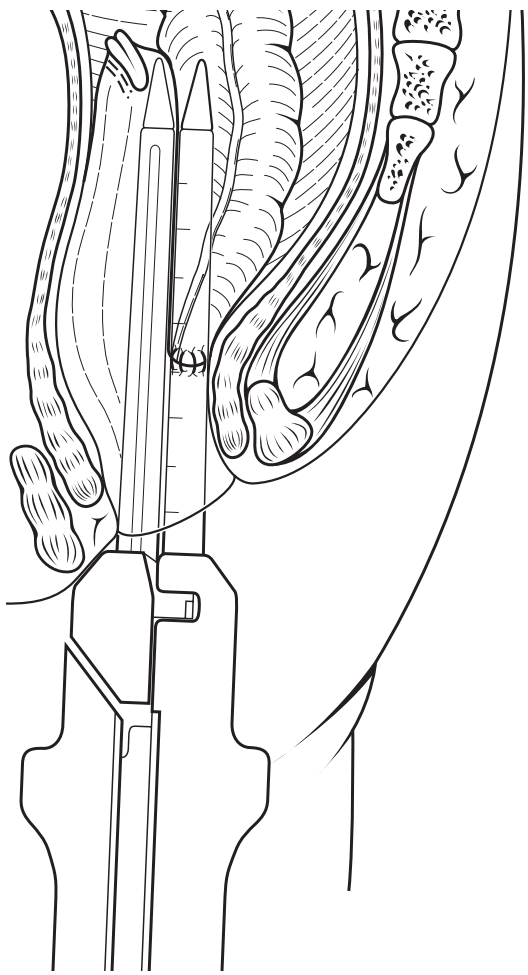
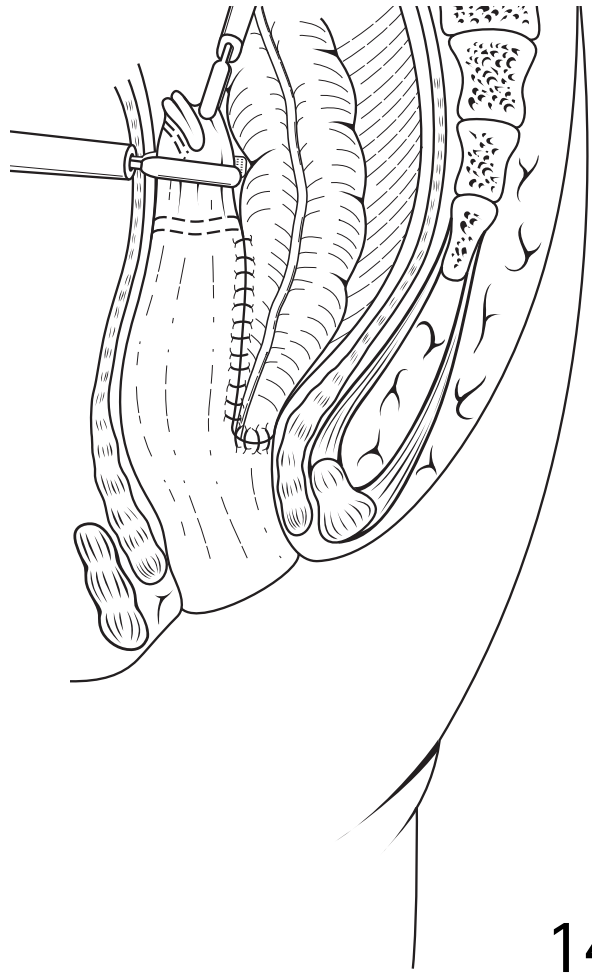


13

CREATING THE RECTAL RESERVOIR

13 One limb of a 6 cm endoscopic gastrointestinal stapler is passed into the patient's remaining aganglionic rectum and one limb is passed into the pull-through segment. Laparoscopy is used once again to verify that the mesentery of the pull-through segment is situated posteriorly to avoid injury during stapling. Stapling of the spur between these structures completes the anorectal anastomosis.



RESECTING THE RECTAL POUCH

14 Laparoscopy is used to determine whether any aganglionic rectal pouch remains after creation of the rectal reservoir. If it exists, it should be excised using an endoscopic stapling device through the 12 mm right lower quadrant port in order proactively to avoid the possibility of a backwash fecaloma.

14

POSTOPERATIVE CARE

Nasogastric decompression is not usually required postoperatively. Patients are offered oral fluids on the day after surgery. Meticulous perineal care with protective creams is required to avoid diaper rash in the neonate. Patients generally have a bowel movement within 24 hours of surgery. They are ready for discharge from the hospital between postoperative days two and four.

OUTCOME

Laparoscopic-assisted transanal endorectal pull-through is a relatively new technique. The largest reported series to date involved 80 patients in 1999. Seventy of these 80 patients were less than 6 months old. Average operating time was 147 minutes, and average blood loss was less than 10 mL. The majority of patients had a bowel movement within the first 24 hours, and average time to discharge was 3.7 days. Conversion to open operation occurred in 2.5 percent of cases.

Enterocolitis and chronic diarrhea are each present in 7.5 percent of patients postoperatively. Anastomotic leak occurred in 2.5 percent of patients, bleeding in 1 percent of patients, and early constipation in 1 percent of patients.

As these techniques are new, long-term outcome data have not yet been published. To date, the authors anecdotally have seen no difference in rates of fecal continence or enterocolitis when compared with open surgical techniques.

FURTHER READING

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54

Malone procedure for antegrade continence enemas

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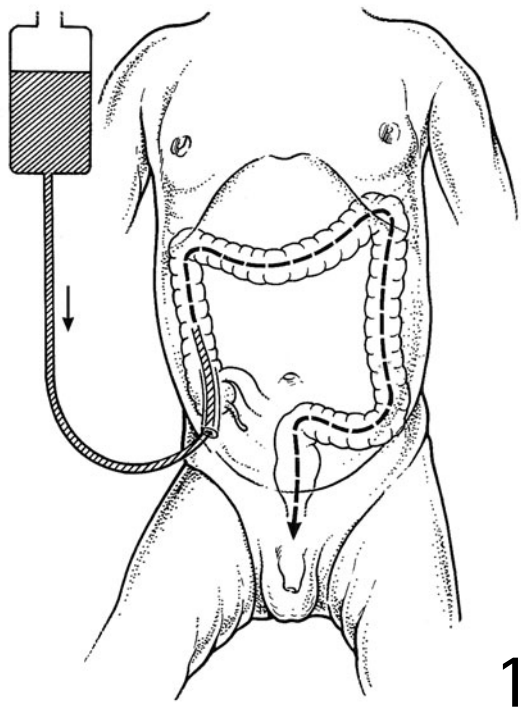
HISTORY

The Malone procedure for antegrade continence enema (MACE) is now accepted as an established treatment for intractable fecal incontinence secondary to conditions such as spinal dysraphism and anorectal malformation. The successful use of the MACE is described in numerous reports in thousands of patients with follow-up of 10 years or more, and it has also been demonstrated that a successful MACE significantly improves quality of life. Technical modifications have been introduced over the years and these are illustrated in this chapter. It is no longer recommended to disconnect the appendix from the cecum (as described in the previous edition of this book), and the in-situ appendix is now the norm. If no other procedure is required, a laparoscopic approach (LACE) is recommended. For patients in whom constipation is a major problem, it may be best to site the conduit in the left colon rather than the cecum. A left-sided MACE can be performed colonoscopically, the percutaneous endoscopic colostomy (PEC), inserting a catheter as one would insert a percutaneous gastrostomy tube. This can then be replaced at a later date by a button or conduit, depending on the patient's wishes. If the appendix is absent or required for a simultaneous Mitrofanoff procedure, the Yang–Monti conduit is now the procedure of choice. The major ongoing complication associated with the MACE is stomal stenosis, which occurs in approximately 30 percent of patients, and this has led to a number of different techniques to construct the stoma.

PRINCIPLES AND JUSTIFICATION

The main indication for the procedure is fecal incontinence secondary to neuropathy and anorectal malformations, which has not responded to conventional therapy. Although the procedure has been used in patients with chronic constipation and complicated Hirschsprung's disease, the author is reluctant to recommend its use under these circumstances. However, if a MACE is used for chronic constipation, it may be best to site it in the left colon colonoscopically, as a clinical trial in the first instance. No patient should have a colostomy without having the opportunity to consider the MACE as an alternative.

1 The MACE combines the principles of the Mitronanoff continent, catheterizable conduit and antegrade colonic washout to produce a continent, catheterizable colonic stoma through which washouts are delivered to produce complete colonic emptying and thus prevent soiling.



1

PREOPERATIVE

Assessment and preparation

Motivation of patients and carers is essential for a successful outcome. Intensive counseling is required and it must be stressed that the MACE is not a 'magic' cure. A rigid, time-consuming regimen is required postoperatively and this is a lifelong commitment. A successful MACE takes approximately 45 minutes every day or on alternate days.

Many patients being considered for the MACE will also have a neuropathic bladder, and it is vital that management of the bladder is assessed simultaneously. In many cases a combined lower urinary tract reconstruction and MACE is appropriate, and double continence rates have been reported in 79 percent of patients with this approach. It is vital, therefore, to have a pediatric urologist involved in the assessment of these patients and in the planning of the operative procedure. Investigations may include ultrasonography, renography, and videourodynamics. In the case of an isolated MACE, usually no special investigations are required, but some authors recommend bowel transit studies to guide them as to where the conduit should be sited, in the cecum for isolated incontinence or in the left colon when performed for chronic constipation.

A preoperative full blood count is recommended, but cross-match is only required when a simultaneous bladder reconstruction is to be performed. For patients with a neuropathic bladder or renal scarring, metabolic renal function should be assessed preoperatively. The author favors a 48-hour bowel preparation program using sodium picosulphate and rectal washouts, together with a 5-day course of antibiotics such as co-amoxiclav. As the patient loses a lot of fluid with the bowel preparation, an intravenous infusion is administered on the night prior to surgery.

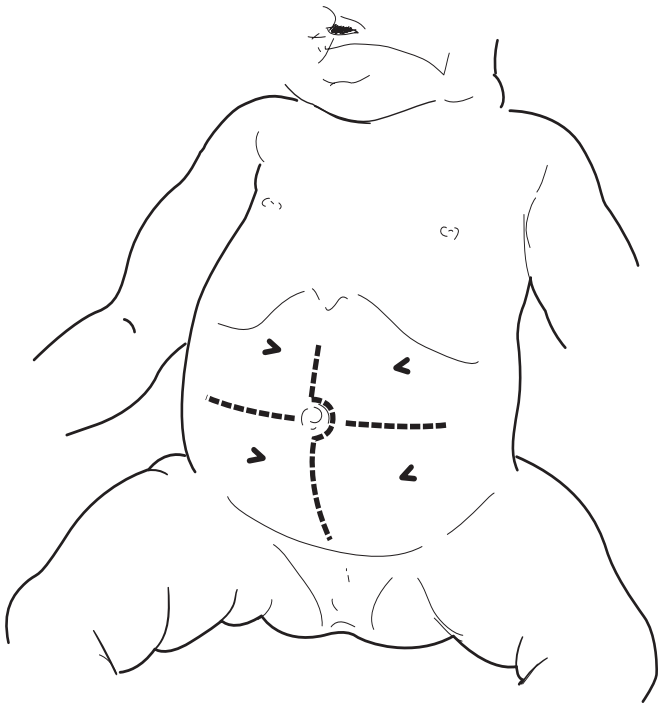
Anesthesia

The operation is performed under general anesthesia, but there are no special requirements.

OPERATION

Incisions

2 When an isolated MACE is performed, a laparoscopic approach is now recommended, but if an antireflux valve is to be created, a right or left lower quadrant muscle-cutting incision is used. A midline incision is better if a simultaneous bladder reconstruction is being carried out. For a cecal MACE, it is usually possible to site the stoma in the umbilicus, but for a left-sided conduit the stoma is usually sited in the left lower quadrant. For patients who are wheelchair-bound it may be necessary to site the stoma on the upper abdomen for ease of access.



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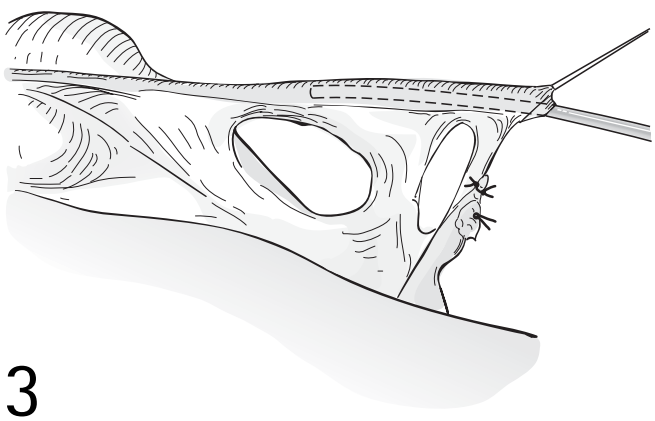


3

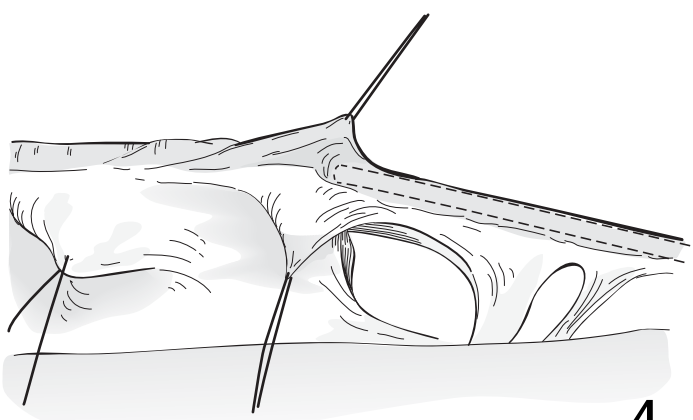
The in situ appendix MACE

PREPARATION OF THE APPENDIX

3 The cecum is mobilized, the tip of the appendix is amputated, and a stay suture is inserted and the appendix stretched to reveal the mesentery. The mesentery is fenestrated between the vessels, as this allows the cecum to be wrapped around the appendix without compromising the blood supply. A 12 Fr catheter is passed through the appendix into the cecum.

CREATION OF CECAL SUBMUCOSAL TUNNEL

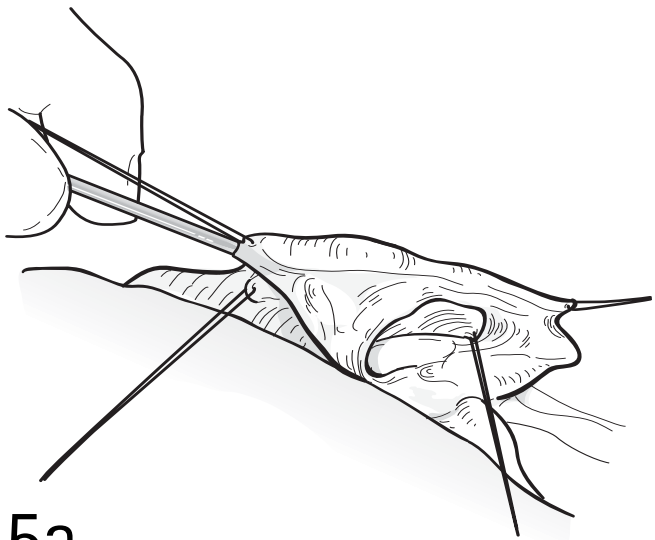
4 A trough down to the submucosa is created along a tenia by a combination of sharp and blunt dissection. As the trough approaches the base of the appendix, a V-shaped incision is created around approximately 60 percent of its circumference; this allows the base of the appendix to be folded into the cecum without kinking. There is no need for a wide trough, as it is not planned to bury the appendix in it, it is simply there to fix the appendix when the cecum is wrapped around it.



4

WRAPPING THE CECUM AROUND THE APPENDIX

5a–c The appendix is folded over onto the exposed submucosa and the cecum is loosely wrapped around the appendix through the fenestrations in the mesentery using a 4/0 polyglycolic acid suture. The suture picks up the seromuscular layer on the cecum on each side and the appendix to anchor it in the tunnel. The wrap is continued until only a short length of appendix sticks out from the tunnel. The stoma is then ready to be created. It is important to anchor the cecum to the back of the anterior abdominal wall where the appendix emerges to prevent twisting and kinking of the conduit.



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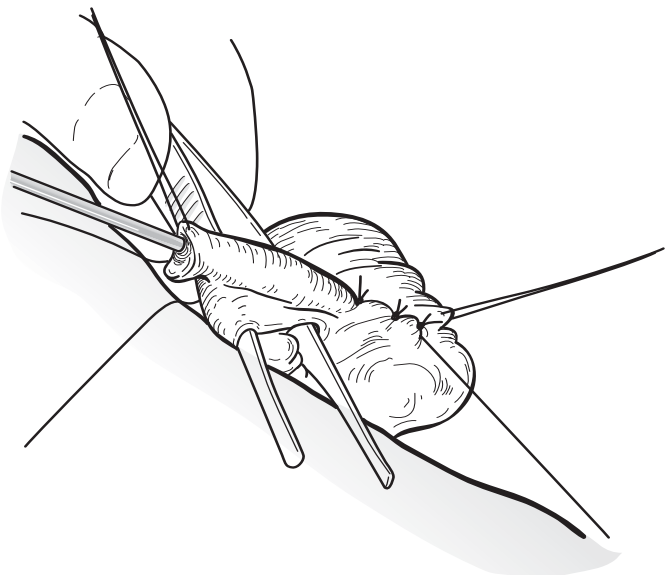

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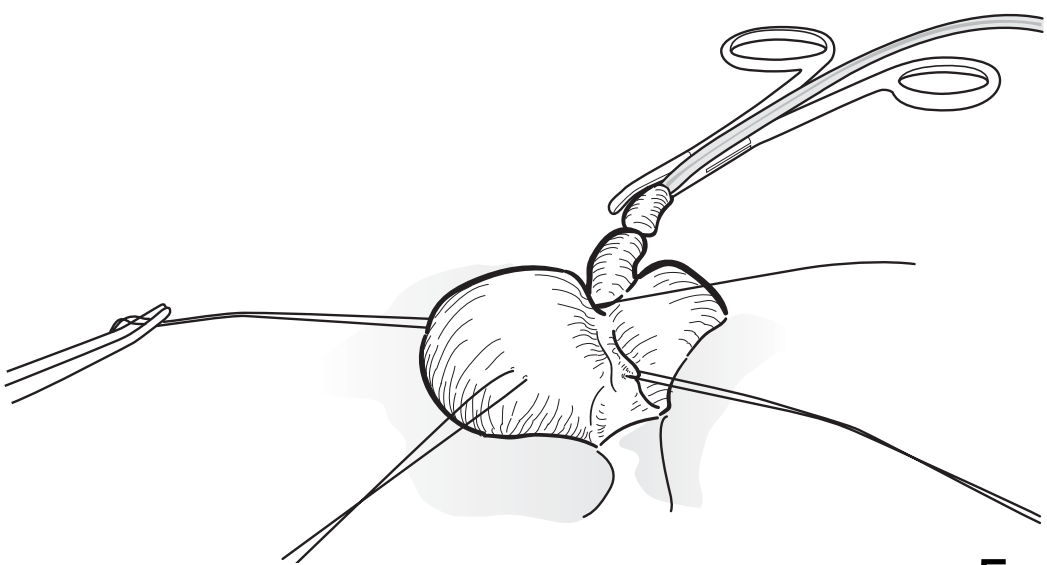


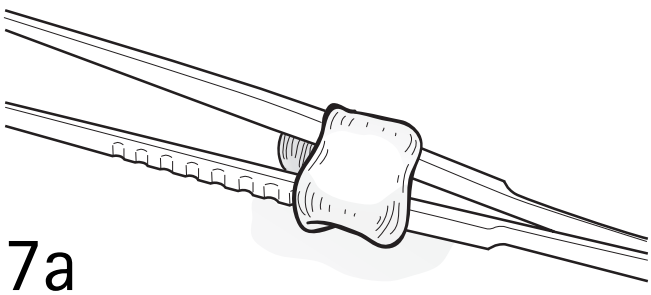
5c

Simultaneous Mitrofanoff procedure/absent appendix

SPLIT APPENDIX

6 When both a Mitrofanoff and MACE are required and the appendix is of sufficient length, it can be divided into two, provided the vascular anatomy is favorable. The MACE uses the in-situ technique as described above, and the distal end of the appendix is available for the Mitrofanoff.

6



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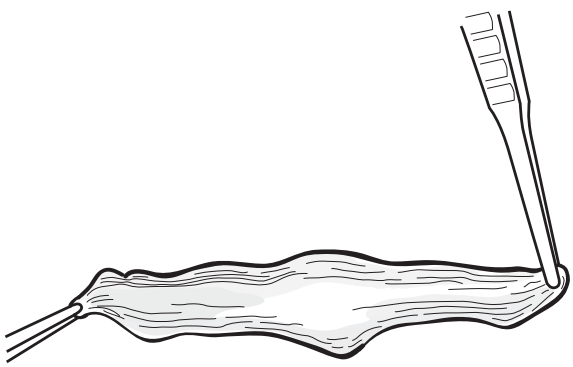

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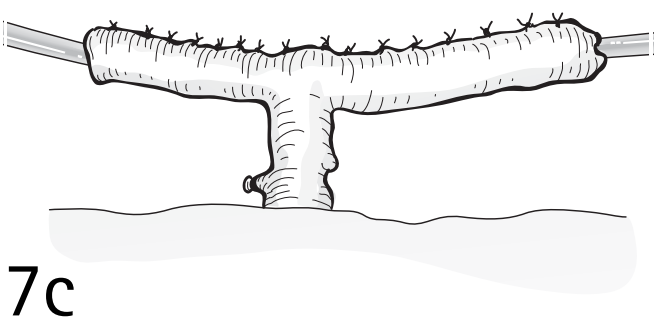


7c

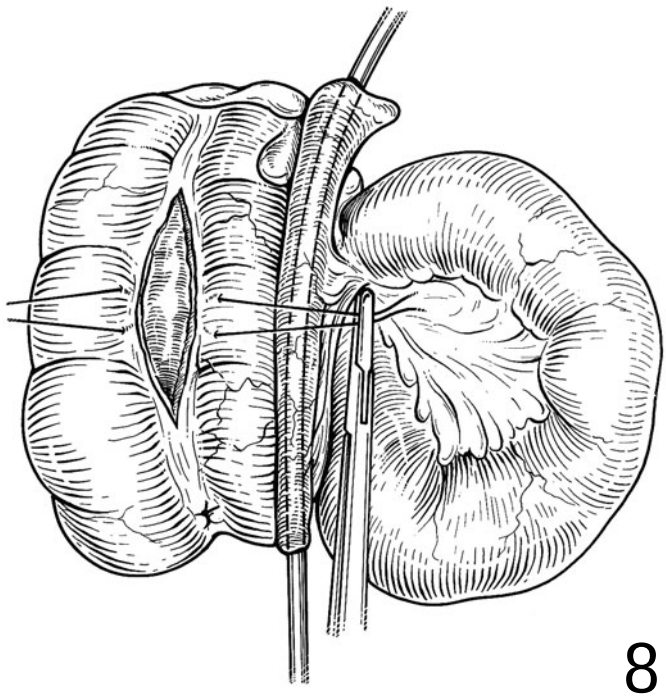
THE YANG–MONTI PROCEDURE

This technique can be used to create a conduit that can then be implanted into the colon at any site to create the MACE. It can be used when the appendix is absent and it is also used for a left-sided MACE.

7a–c A 2 cm segment of ileum is isolated on its vascular pedicle. Bowel continuity is restored by a standard end-to-end anastomosis. The ileum is opened along the antimesenteric border. It can then be seen that the valvulae coniventes are now running in a longitudinal direction along the length of the bowel. The bowel is then tubularized over a 12 Fr catheter by a single-layer, interrupted, extramucosal anastomosis using 6/0 PDS suture. One end is then implanted into a submucosal tenial tunnel in the colon and the other is brought to the skin as the stoma.

CREATION OF COLONIC SUBMUCOSAL TUNNEL

8 A tenia is stretched using proximal, distal, and two lateral stay sutures. The seromuscular layer of the tenia is incised with a scalpel down to the submucosa over a 5 cm length. The mucosa/submucosa is then freed from the overlying muscle using a combination of sharp and blunt dissection to leave an exposed strip of mucosa approximately 1 cm in width.



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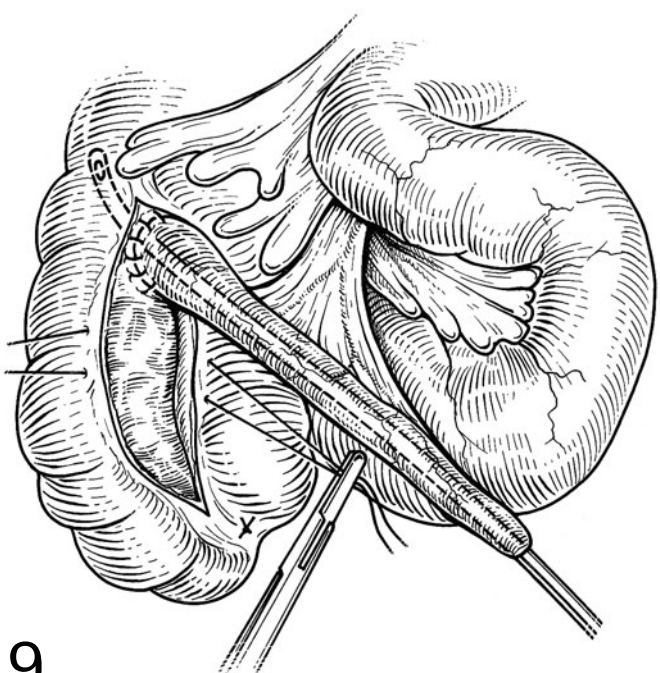


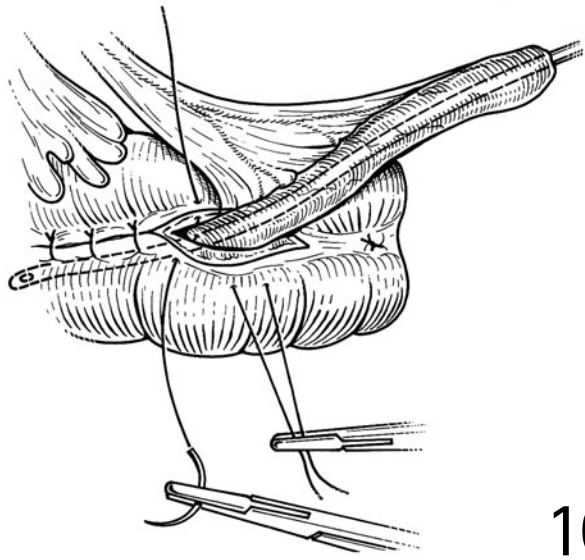
9

MONTI–MUCOSAL ANASTOMOSIS

9 A small hole is punched in the mucosa of the colon using artery forceps. This is usually placed at the distal end of the mucosal tunnel. The mucosa is anastomosed to the full thickness of the Monti tube using a 5/0 polyglycolic acid suture over the catheter in the conduit.

CLOSURE OF THE SEROMUSCULAR TUNNEL

10 The seromuscular wall of the colon is closed over the Monti tube using interrupted 4/0 polyglycolic acid sutures picking up partial thickness of the conduit wall to prevent it slipping out of the tunnel.



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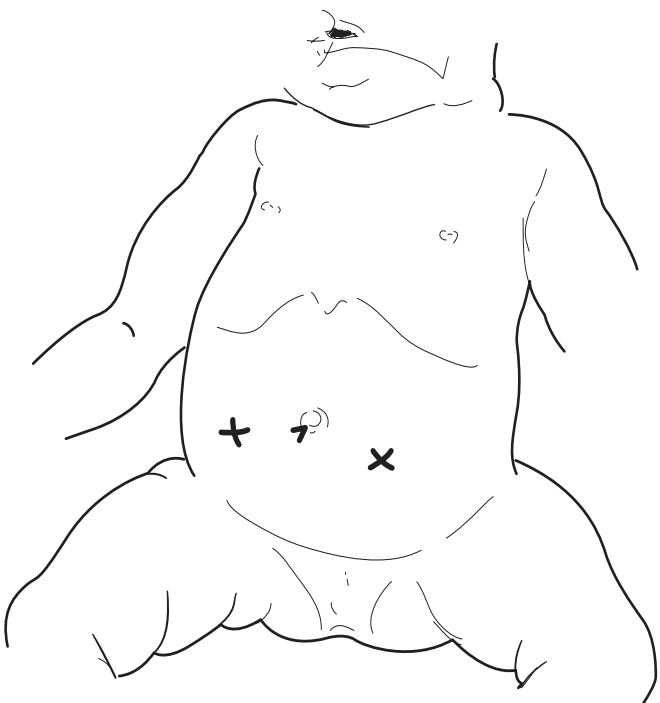


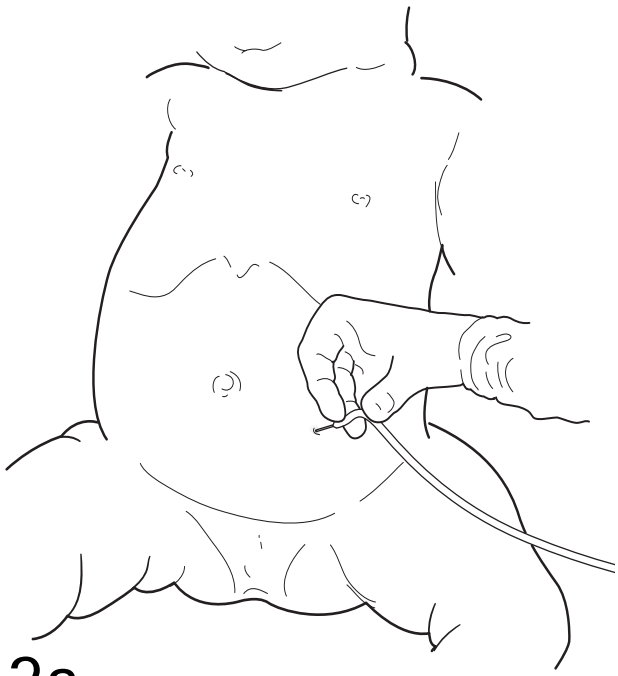
11

Laparoscopic MACE (LACE)

11 A 5 mm port is inserted at the umbilicus under direct vision. Two further 5 mm ports are inserted in both iliac fossae. The cecum is mobilized so the appendix can reach the umbilicus. The camera port is changed and the appendix is grasped with forceps and simply delivered through the umbilical port site, where a stoma is then created. The author does not usually create an antireflux valve during this procedure, and although leakage from the conduit is more common than when a valve is created, it is still not a common problem.

Percutaneous endoscopic colonic tube placement (PEC)

12a–e Following bowel preparation and under general anesthesia, a colonoscope is passed to the distal descending colon and the light can be visualized in the flank. A needle and thread is passed into the colon, grasped, and delivered through the anus. This is attached to a gastrostomy tube, which is pulled up into the colon till the flange on the tube pulls the colon to the abdominal wall. The tube is fixed externally to the abdominal wall and washouts can be commenced the following day. After a trial, if the MACE works, the patient has a choice of keeping the tube, changing to a button, or having a conduit constructed.



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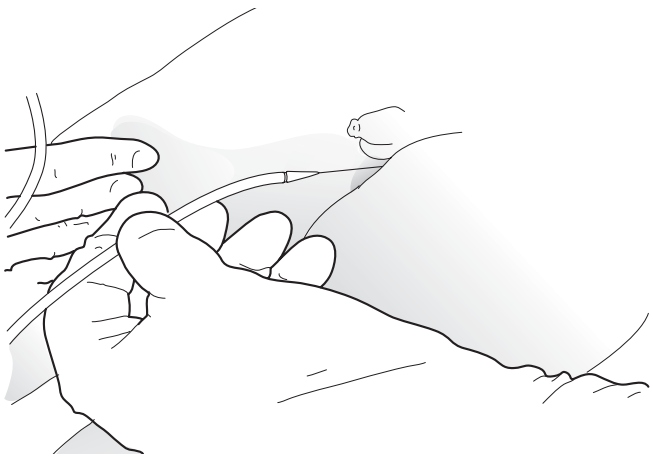

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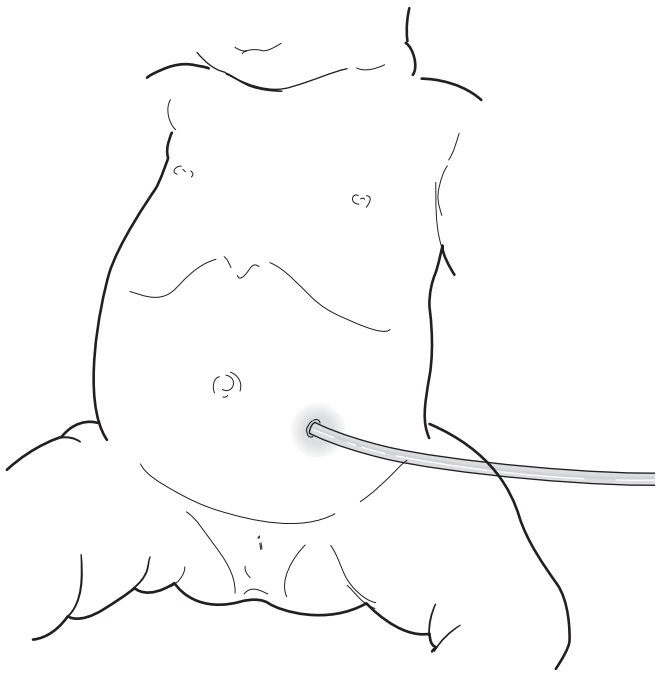

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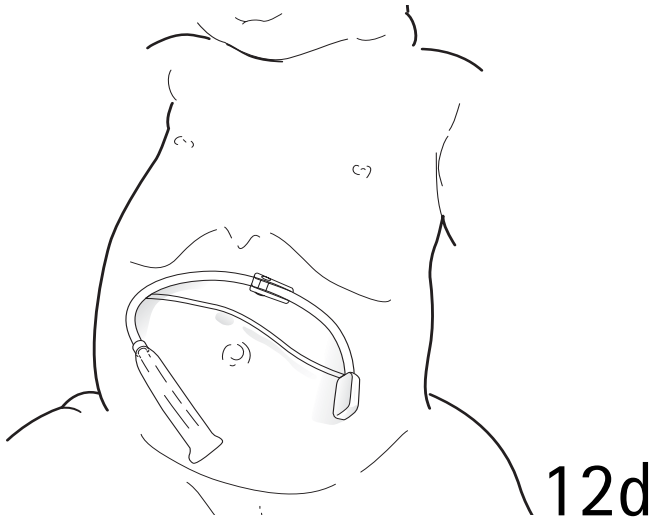

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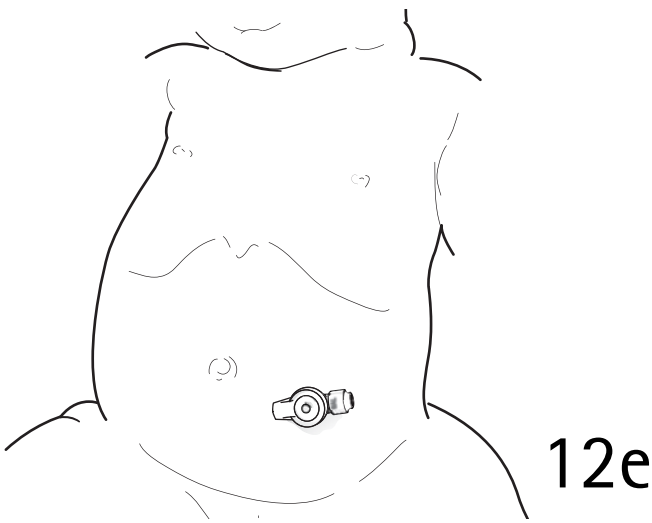


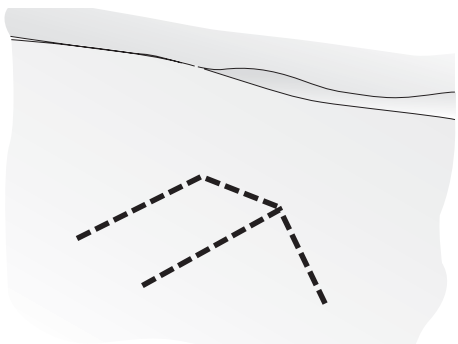
12e

Fashioning the stoma

ABDOMINAL VQC STOMA

13a–g Two skin flaps (V and rectangular) are created at the site of the stoma. A hole is created in the abdominal wall that is sufficiently wide to allow the conduit to pass through freely. The cecum or colon is sutured to the anterior abdominal wall to prevent tension on the stoma or volvulus of the bowel on the conduit.

The conduit is fish-mouthed and the apex of the V-flap is sutured into the defect using 5/0 Maxon sutures with the knots outside the catheterizing channel. The V-flap is gradually sutured into the defect until approximately 50 percent of the circumference of the conduit is complete. The rectangular flap is then sutured over the anterior circumference of the conduit until the anastomosis is complete. The resulting skin defect is closed in layers using 4/0 Maxon and 5/0 subcuticular polyglycolic acid sutures, resulting in a C-shaped wound (VQC stoma). A 12 Fr Silastic Foley catheter is left in situ for 4 weeks after the surgery prior to commencing catheterization.



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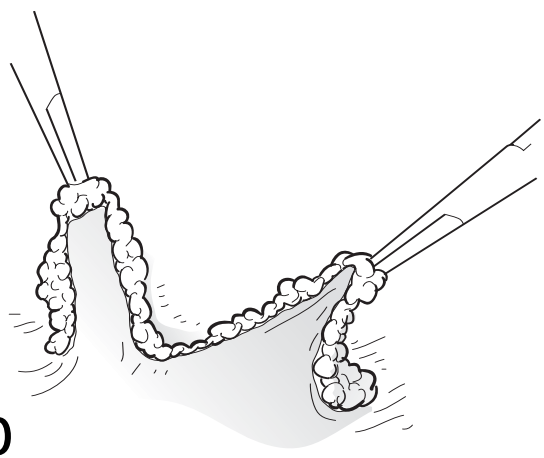

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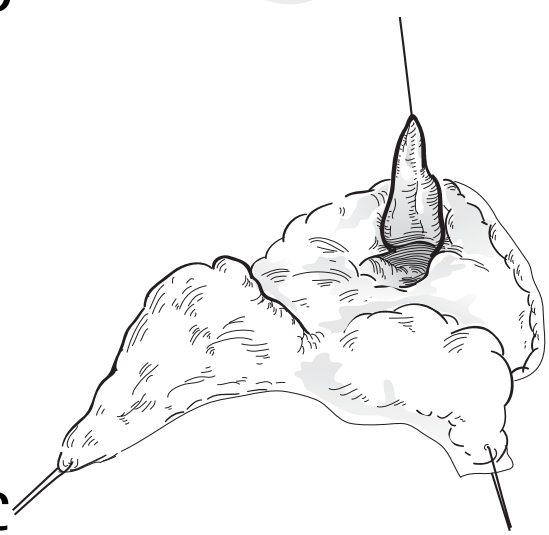

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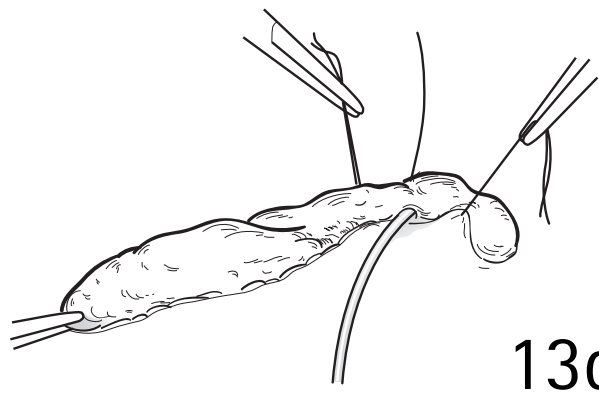

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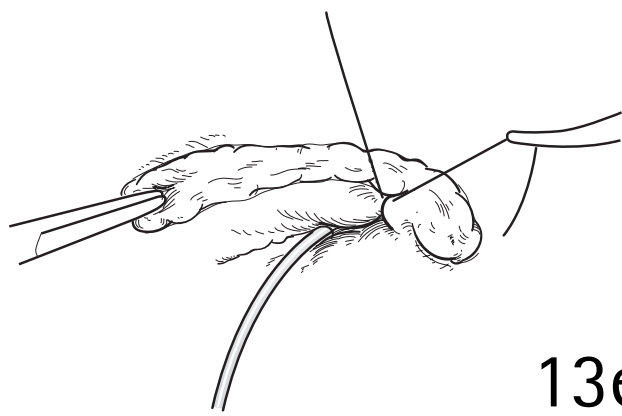

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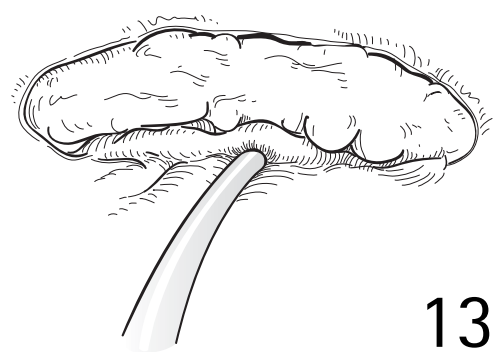

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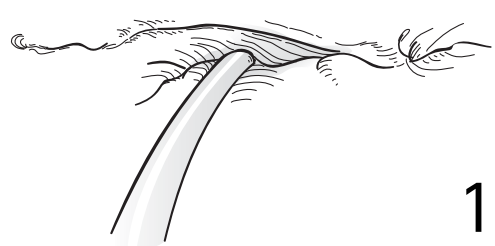
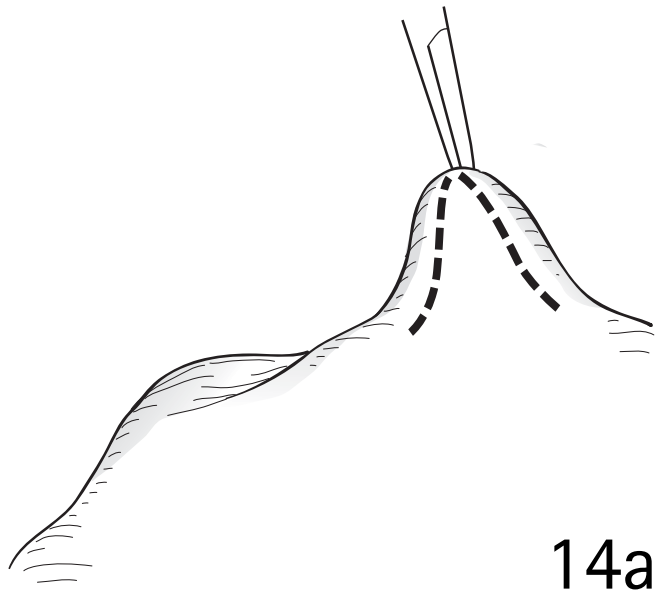


13g

UMBILICAL STOMA

14a–c The umbilicus is everted and a V-flap is created from the everted skin. This is sutured into the conduit as described above, and the remainder of the anastomosis is completed by suturing the conduit to the umbilical rim.



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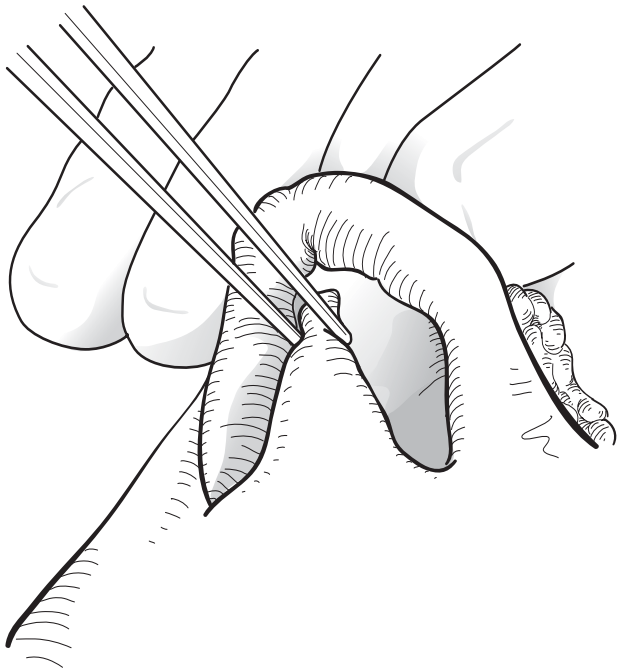

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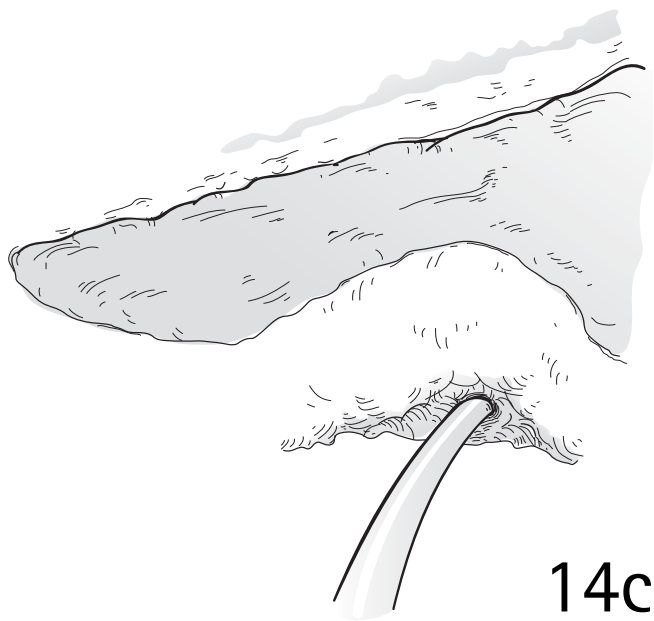

14c

POSTOPERATIVE CARE

Following an isolated MACE, enteral feeding can commence the following day, and the first washout can be administered when the patient has recovered from the adynamic ileus. Once the patient and carers are happy with the enema procedure, they can be discharged with the indwelling catheter, to return 4 weeks later to learn intermittent catheterization, which seldom takes longer than 48 hours. The stoma should be catheterized twice daily, whether or not an enema is to be administered, as this seems to reduce problems from stomal stenosis. A standard 12 Fr Nelaton catheter is used. The patient should be given some 8 Fr and 10 Fr catheters, because if catheterization becomes difficult, the smaller catheters can be used initially to help dilate the stoma. If severe stomal stenosis develops, dilatation under general anesthesia is recommended, following which a button can be left in situ for a period to reduce the risk of a further stenosis. ACE Stoppers are now commercially available in varying sizes, 10–14 mm diameters and 15–100 mm lengths (Medicina, Lancashire, UK), and these are very helpful for patients who experience ongoing catheterization problems. Occasionally, stoma revision is required, and this usually takes the form of a Y-V plasty.

ENEMA REGIMENS

There is no single correct enema regimen, and each patient develops an individual practice by trial and error. The author starts with 1 mL/kg of phosphate enema (Fletchers' phosphate, Fleet pharmaceuticals, Zaragoza, Spain) diluted to half-strength with an equal volume of tap water or normal saline. This is followed by a washout of tap water of between 10 mL/kg and 20 mL/kg. A daily enema is given for the first few months, but after that about half the patients use the washouts on alternate days or, rarely, even less frequently than that.

Initially many patients experience colicky abdominal pain, and this may be helped by reducing the concentration of the phosphate and the rate of enema infusion. If colic is persistent, the administration of mebeverine hydrochloride 30 minutes before the enema can help. Persistent pain may also be caused by constipation, and this can be managed by the administration of mineral oil via the ACE 4–6 hours prior to the washout.

If the enema does not produce a rapid result, the concentration of the phosphate can be increased in steps up to a full-strength enema, and in some patients this is used without a following washout. Most patients continue to use a washout, but if fecal leakage occurs between enemas, the volume can be reduced or increased and this usually resolves the problem.

Phosphate toxicity has been encountered, particularly in younger patients, and it is of vital importance that if there is no response from the enema after 6 hours, no further phosphate is administered until a result is obtained. Further washouts with tap water often help, but occasionally retrograde washouts are required.

ADMINISTERING ANTEGRADE COLONIC ENEMAS

Most patients use an infusion system such as a Kangaroo bag and pump set (Kendall, Tullamore, Ireland). The bag is filled with the required phosphate and infused over a 10-minute period. The bag is then refilled with water and infused over the next 15 minutes. Evacuation usually starts within 15 minutes and is complete 30 minutes later. As patients will spend a considerable time sitting on the toilet, the use of padded seat covers is recommended to reduce the risk of pressure sores.

COMPLICATIONS

The common complications and their management are discussed in the text. Uncommon complications include leakage of fecal fluid through the stoma, and if this occurs, the valve mechanism will need to be revised or a valve created, if this had not been done in the first instance.

OUTCOME

For patients with neuropathic conditions and anorectal malformations, the success rate of MACE procedures is 80–90 percent.

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Hirschsprung disease

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PRINCIPLES AND JUSTIFICATION

Advances in our understanding of the embryogenesis of Hirschsprung disease have come about over the past few years. Despite our increased knowledge of the disease, significant complications continue to be associated with this process. One must maintain a high degree of suspicion for the disease. Suction rectal biopsy and anal manometry studies have allowed for easier and less invasive methods of establishing the diagnosis; however, obtaining details of the infant's history is the most important first step. Failure to pass meconium within the first 48 hours of life, complaints of constipation and, finally, symptoms of enterocolitis should always be followed by a complete clinical examination for Hirschsprung disease.

The classic approach to the neonate diagnosed with Hirschsprung disease is to perform a leveling colostomy and to wait until 6–12 months of age to perform the definitive pull-through. This approach has changed dramatically over the past two decades, and transition to primary pull-through is now predominant. Use of laparoscopy to facilitate both the diagnosis and the pull-through procedure has also become common. The transanal approach is now used by a large proportion of pediatric surgeons, and is discussed in this chapter.

FULL-THICKNESS RECTAL BIOPSY

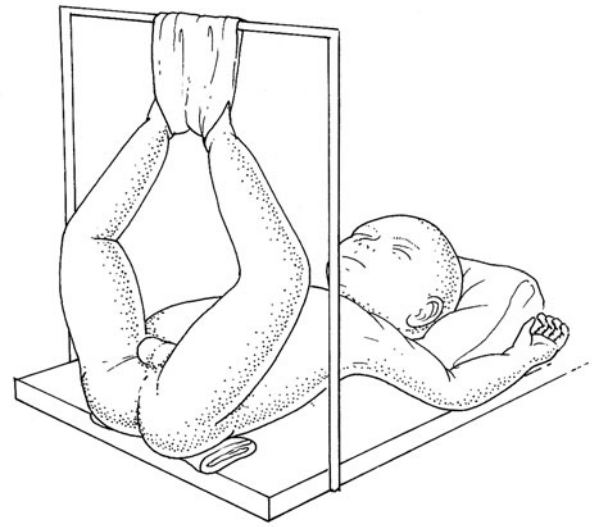
Suction rectal biopsy, because of its relative ease and low morbidity, has become the most established diagnostic tool for Hirschsprung disease. Nevertheless, full-thickness rectal biopsies are occasionally required, and the technique of full-thickness biopsy is presented here to assist surgeons who are not familiar with the procedure. Perhaps the most common indication for a full-thickness biopsy is the child who has undergone more than one indeterminate suction rectal biopsy.

PREOPERATIVE

No formal bowel preparation is required. The child's rectum is irrigated with saline or very dilute povidone-iodine solution. A sponge is placed into the proximal rectal vault to prevent stool from entering the operative field. One dose of preoperative antibiotics is given.

OPERATIVE TECHNIQUE

1 The patient is placed in the lithotomy position with the buttocks at the very end of the bed, supported with a folded towel. The feet are placed together (plantar surfaces adjoined) with a cotton roll, and both legs are suspended on an ether screen, or similar device, with the lower extremities flexed at the hips.



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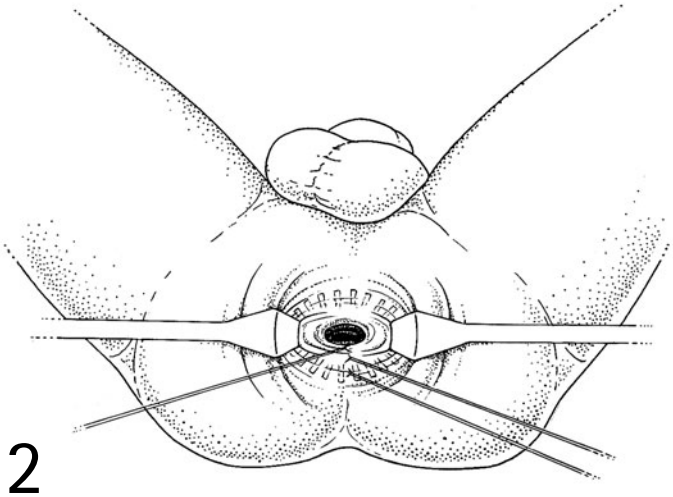
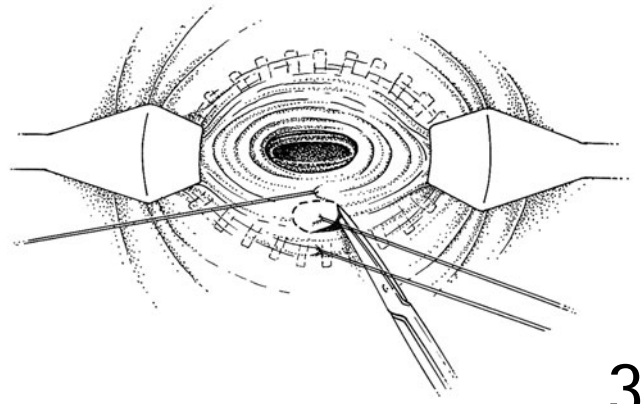


2

2 Digital dilatation is followed by placement of two narrow anal retractors. The posterior aspect of the dentate line is identified and marked with a silk suture (3/0), which is used for traction. Two additional silk sutures are placed on the posterior wall of the rectum at 1 cm and 2 cm proximal to the dentate line.

3 The surgeon's non-dominant hand holds the middle silk suture. Using sharp curved scissors, a full-thickness incision is made along the lower half of the rectal wall, between the dentate line and the middle suture. Once this is done, the scissors can be placed in the presacral space and gently spread. Bleeding can slightly obstruct the view at this point; however, by maintaining traction on the middle suture, the upper half of the rectum is incised with two smooth cuts of the scissors, each sweeping around one-half of the tissue suspended by the middle suture. The specimen is inspected and delivered off the table.

The rectal defect is closed in a single, running or interrupted layer with an absorbable suture (e.g., 4/0 polyglactin (Vicryl). Hemostasis is achieved fairly quickly once this suture has been placed.



3

LEVELING COLOSTOMY

Although a primary pull-through is preferred in most infants, use of a leveling colostomy is required in infants presenting with a severe episode of enterocolitis at the time of diagnosis. A colostomy is also required in those patients with a delayed diagnosis of Hirschsprung disease, where the colon has become overly distended and not amenable to a primary pull-through. Although a right transverse colostomy has been advocated by some surgeons as the initial procedure, the authors prefer a leveling colostomy. This allows for the determination of the aganglionic level at the time of the colostomy, facilitating the subsequent pull-through. In addition, placement of a leveling colostomy allows the proximal bowel to grow, which will stretch the mesentery and simplify the subsequent pull-through procedure. Finally, this colostomy can be closed during the pull-through, thus avoiding a third operation. Placement of the ostomy is just proximal to the transition zone. The incision is generally an oblique one in the left lower quadrant. If the level of aganglionosis is not readily apparent, this incision can be extended transversely across the midline.

PREOPERATIVE

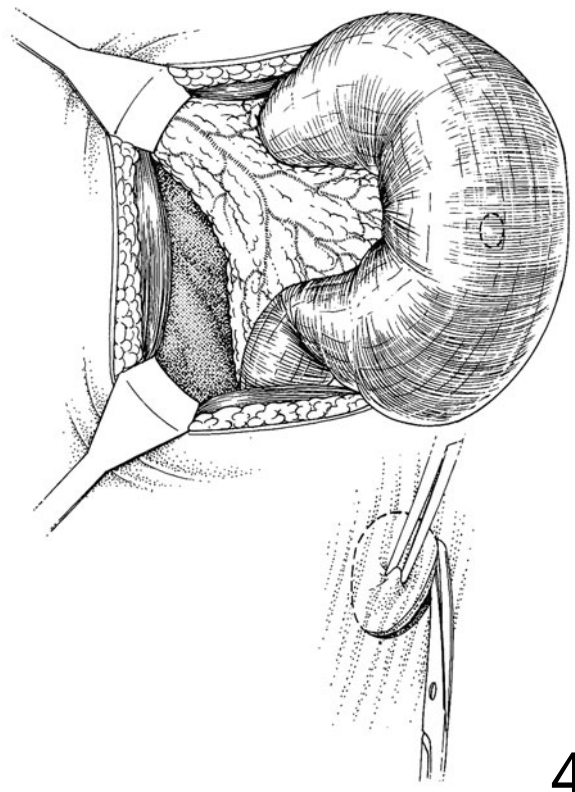
Essentially, only the diagnosis of Hirschsprung disease is needed. Most diagnoses can be suspected based on the history alone, but confirmation is required by histopathologic examination of a suction rectal biopsy. The infant should receive rectal washouts and be placed on broad-spectrum, intravenous antibiotics just prior to the incision, but no formal bowel preparation is required or effective.

OPERATIVE TECHNIQUE

4 Once the peritoneum is entered, an attempt should be made to define a gross transition zone. The bowel proximal to the transition zone is normally dilated and has a diffuse hypertrophy of the muscular layer with no clearly distinguishable tenea. In neonates, such a transition often may not be seen. If this is the case, a good starting point is just above the peritoneal reflection.

A pair of fine, sharp scissors is used to make an incision only through the seromuscular layers. The muscular layer, which is fairly thick, even in the aganglionic section, makes this dissection fairly easy. Blunt dissection is used to strip off the muscle. In general, a 1×0.5-cm biopsy specimen is taken and interrupted silk or polyglactin sutures are placed to close the biopsy site.

Each biopsy specimen is sent for frozen section, progressively moving more proximally until both ganglion cells as well as a loss of hypertrophied nerve bundles are seen. Hypertrophied nerve bundles, despite the presence of ganglia, indicate that one is still in the transition zone. Another biopsy specimen should be taken several centimeters more proximally.



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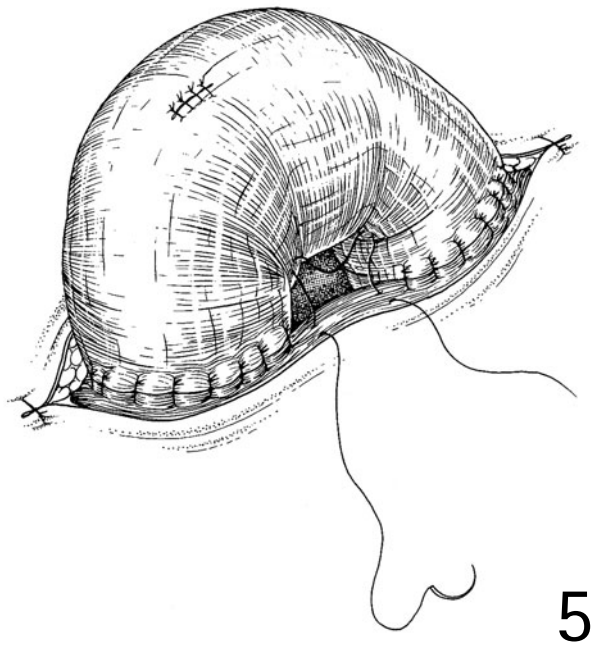
5 At this point, a loop colostomy can often be created at one of the normal biopsy sites. Because of the relatively large caliber of the bowel, stomal prolapse and peristomal hernias are common complications. It is extremely important to begin the colostomy by placing numerous fine polyglactin sutures both to the peritoneum as well as to the fascia. A stitch is placed between the proximal and distal loops of bowel, starting at the fascia, then to each limb of bowel, and finally back to the fascia.

If no transition zone is found and the first few biopsies are aganglionic, it is usually beneficial to perform an appendectomy. Aganglionosis of the appendix indicates the presence of total colonic Hirschsprung disease.



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POSTOPERATIVE CARE

The stoma usually begins to function within 24 hours, and feeding can begin shortly thereafter. It is occasionally helpful to perform intermittent dilatations of the proximal ostomy. These dilatations will prevent narrowing of the opening and allow the dilated proximal colon to return to normal size.

OPERATIONS

DUHAMEL

The Duhamel technique was advanced in 1956 to avoid the tedious pelvic dissection of the Swenson procedure, and to protect the nervi erigenti, which may be found lateral and anterior to the rectum. The procedure has undergone several modifications, the most important of which was by Martin and included the use of an automatic stapling device. It is fairly straightforward and continues to be popular today. Despite its relative simplicity, several key technical points must be followed.

As with other pull-through procedures, ganglionic bowel is brought down to less than 1 cm proximal to the dentate line. To preserve the autonomic nerve plexus to the genitourinary system, very little manipulation of the rectum is performed anteriorly.

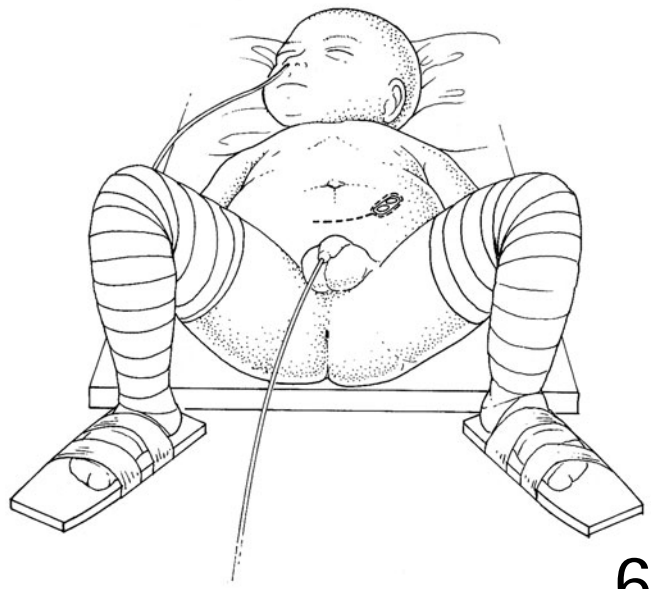
The child often has a leveling colostomy, which was placed several months previously. This serves to decompress the bowel and return it to normal caliber. The operation is generally performed when the child is 6–12 months of age with a weight of 10 kg. With the use of smaller endostapling devices, the procedure may also be performed primarily in the newborn period.

Preoperative

The child is admitted the day before the surgery for a mechanical bowel preparation as well as oral antibiotics. Care must be taken to give adequate rectal and colonic washouts, as stool is often inspissated in the distal rectum. It is necessary to do a rectal examination on the child before the pull-through to ensure that no residual stool is present. Preoperative antibiotics should be given.

Operative technique

6 A nasogastric tube is placed after induction of anesthesia. The child is placed in a supine position and prepared circumferentially from the abdomen to the feet. Stockinettes are placed around each foot and a Foley catheter is inserted in the bladder after the patient has been prepared and draped. Excellent exposure is obtained by assistants supporting and flexing the lower extremities at the hips during the anal anastomosis. Alternatively, the child can be placed in stirrups or on skis. A hockey-stick or oblique incision is made incorporating the colostomy (if present). The bowel is mobilized proximal to the former colostomy and the splenic flexure is brought down, if necessary, to ensure adequate length for the pull-through. In general, the ganglionated colon must reach the level of the perineum when drawn over the child's pubic symphysis with only modest tension. Occasionally, the mesentery is foreshortened and it is necessary to ligate the inferior mesenteric artery near the aortic root. By preserving the remainder of the arcades, the bowel should maintain its viability. The ureter is carefully identified and the peritoneal reflection between the rectum and bladder is incised. The distal rectum is mobilized for approximately 4 cm below the reflection. The colostomy site is removed with an automatic stapling device.



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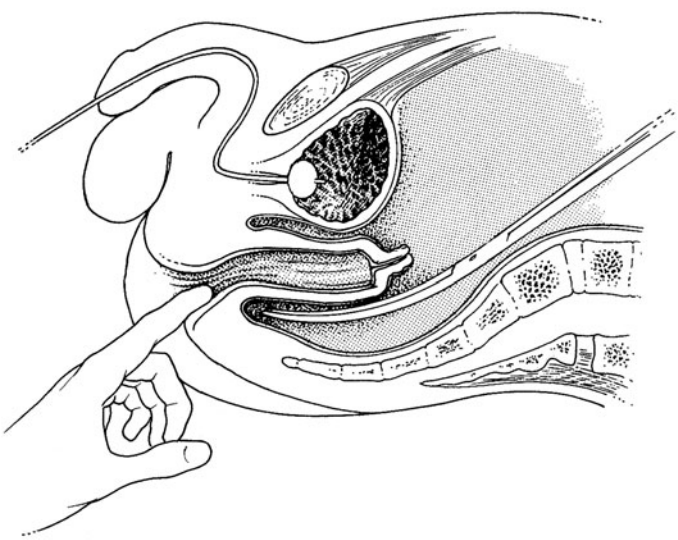

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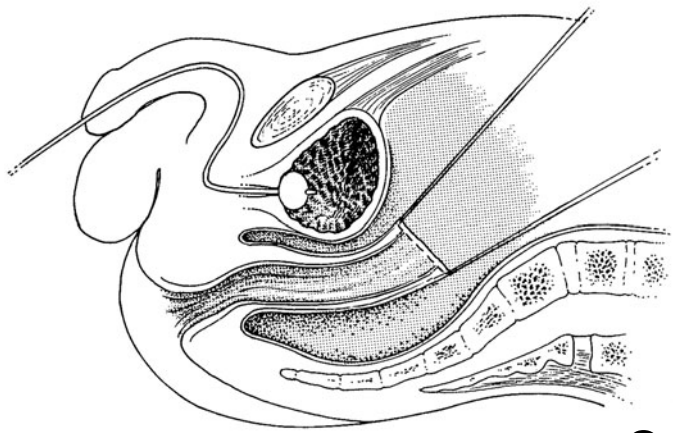
7 A retrorectal space is created, with dissection carried out directly in the midline. This dissection is carried down to the pelvic floor so that an assistant's finger can be felt when inserted no further than 1–1.5 cm into the anus. Dissection can be facilitated by a blunt clamp, but is also very easily performed with the index finger.

8 Once the retrorectal dissection has been completed, redundant aganglionic bowel is resected down to the peritoneal reflection with an automatic stapling device. Tacking sutures are placed on both left and right sides of the bowel so that it can be retracted anteriorly during the pull-through procedure.

The ganglionic bowel is labeled mesenteric and antimesenteric with a separate polypropylene and polyglactin suture. This allows the surgeon working on the pulled-through segment to maintain correct orientation of the bowel as it is pulled into the anus.

At this point, the surgeon's attention is directed to the perineum. If not placed into skis, both the patient's legs are drawn upward, allowing a clear view of the anus. Narrow anal retractors are placed and held in position by the two assistants still working on the abdomen. No separate field is created, which allows improved communication and keeps surgeon and assistant on the same operative field. The authors have not found it necessary to have two completely different setups. However, all the instruments used in the perineal portion of the procedure are treated as dirty, and gloves are changed at the end of the anastomosis.



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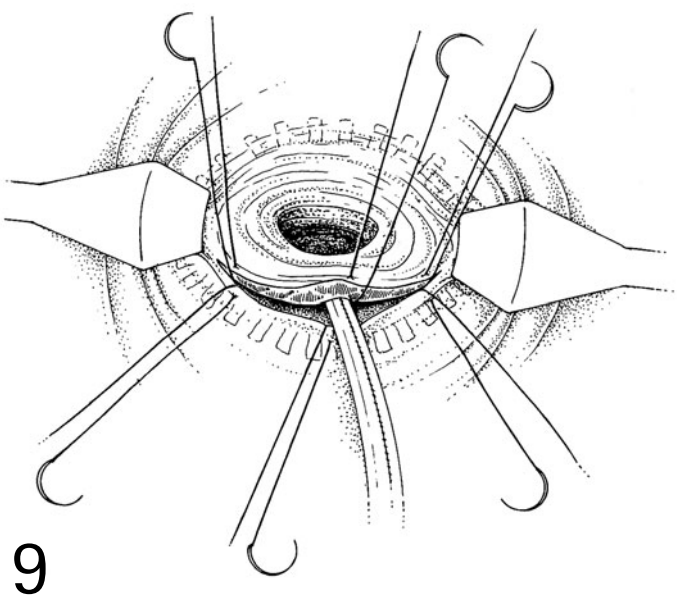

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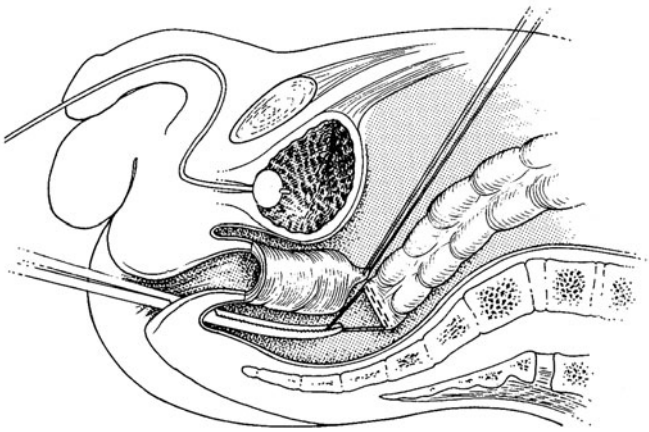
9 With the use of cautery, a full-thickness incision is made 0.5 cm proximal to the dentate line posteriorly. Care is taken to maintain this distance by curving the incision as one moves laterally in each direction. Three 4/0 undyed polyglactin sutures are placed on the inferior aspect of this incision, one in the midline and one each on the left and right sides. Three additional absorbable sutures (4/0 dyed polyglactin) are placed on the upper portion of this incision in similar positions. Each suture is held in position with hemostats. The different colored sutures prevent confusion of orientation once the ganglionic bowel is pulled through.

The surgeon operating on the anus inserts a long clamp into the retrorectal space towards the abdominal field.

10 The two tacking sutures on the distal ganglionic bowel are fed into this clamp and pulled down. The surgeon remaining in the abdominal field guides the bowel, and makes sure that it does not rotate as it is brought down.



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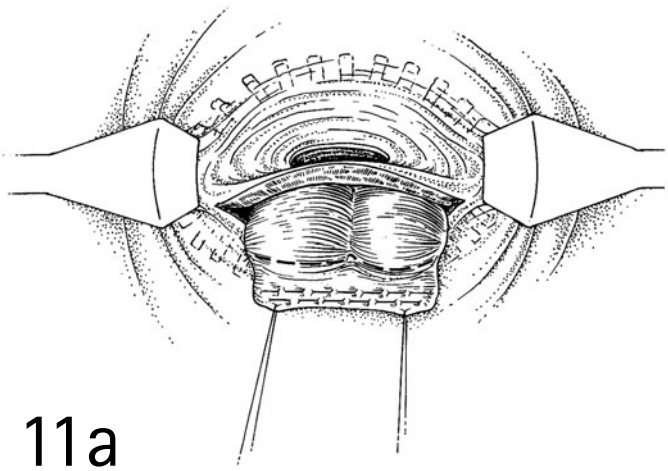
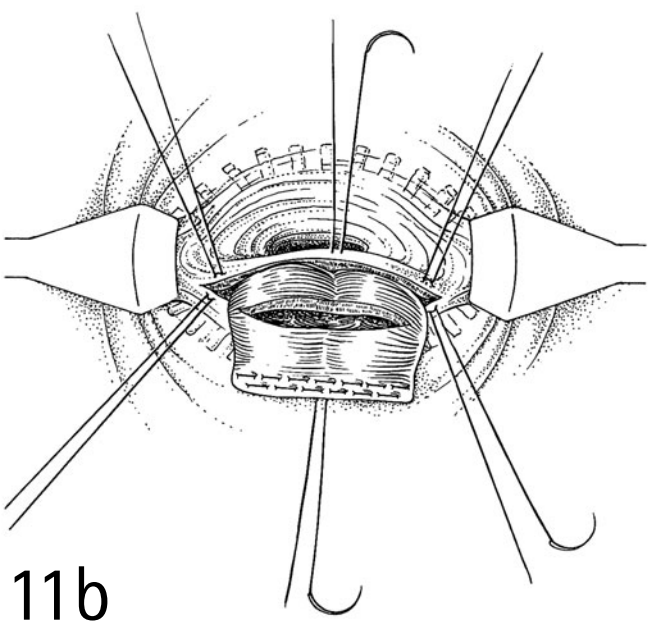
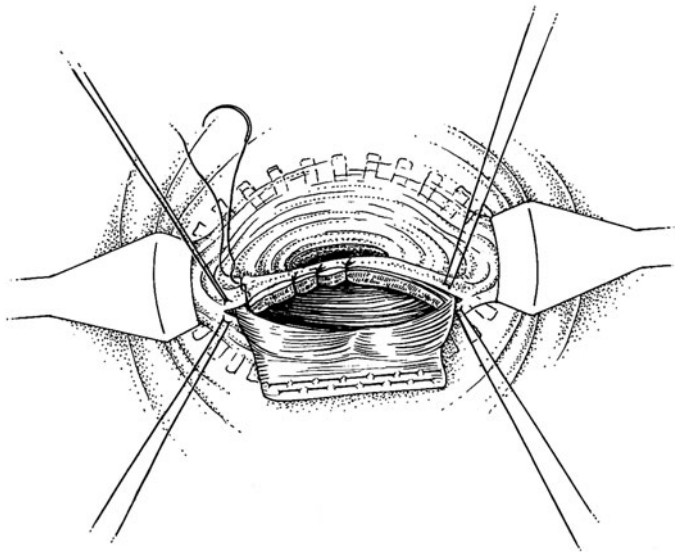


11a



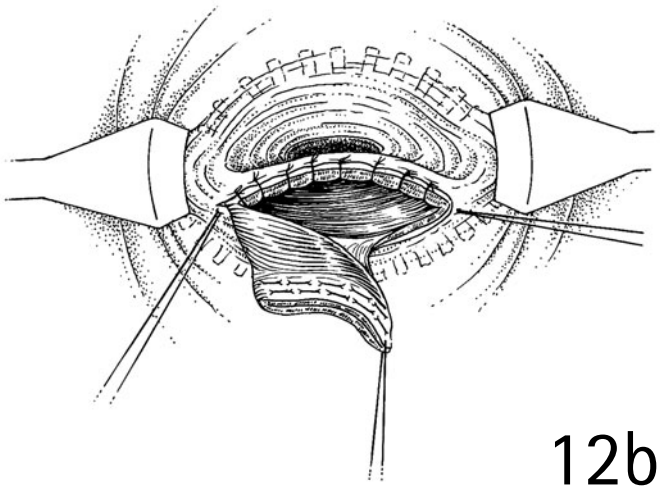
11b

11a,b Once the bowel is pulled through, the staple line is excised on the anterior half of the colon and a single-layered anastomosis is created, starting with the three previously placed polyglactin sutures. Care is taken with each stitch so that the anterior wall of the anus is not incorporated into any of the sutures.

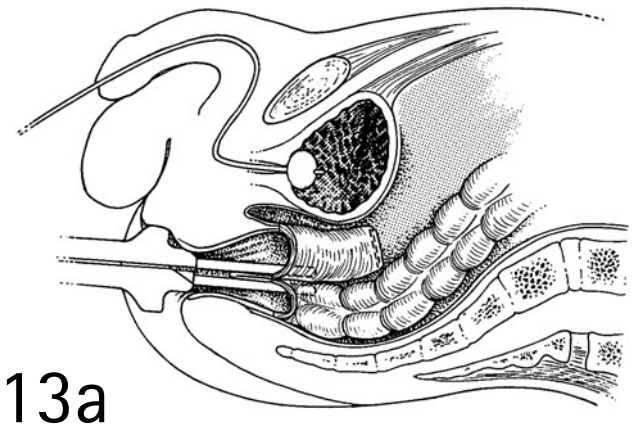


12a

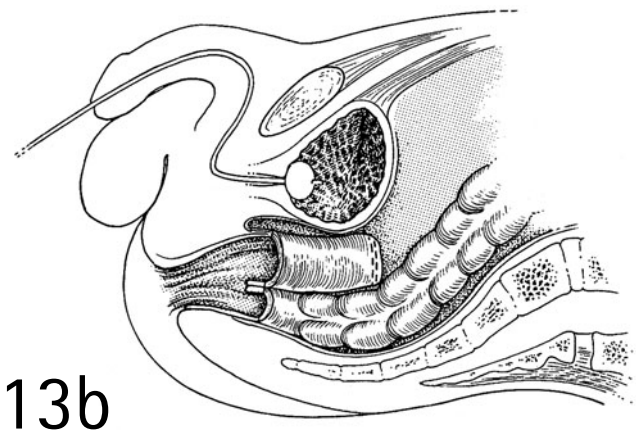
12a,b Once the anterior half of the anastomosis is completed, the remainder of the staple line is excised and the anastomosis is finished.



12b



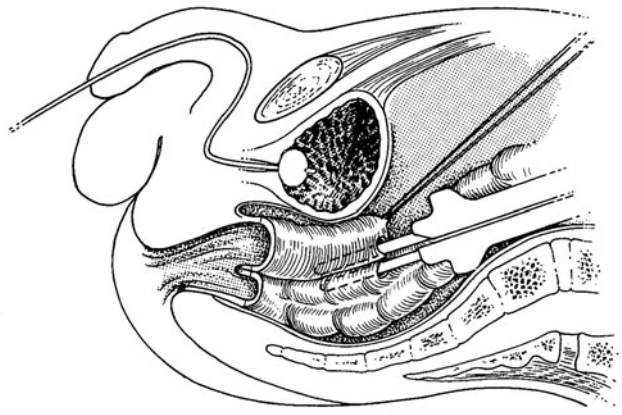
13a



13b

13a,b An automatic stapling device is placed with one arm into the native anal canal and the other into the neorectum. The stapler is fired directly in the midline. Hemostasis along the suture line is checked. In general, a long (80 mm) device is preferred; a smaller endostapler is used in newborns.

14 Often a complete anastomosis between the ganglionic and aganglionic bowel cannot be achieved with a single staple application from below. A second firing from the abdomen will be necessary. The staple line of the remaining aganglionic rectum is opened and a small enterotomy is made in the ganglionic colon at a similar level. The abdominal surgeon places a reloaded automatic stapler between the two limbs of bowel to complete the anastomosis. This last step is critical. In the past, a proximal spur left between the bowel segments caused the eventual formation of huge fecalomas. It is critical for the surgeon to inspect the anastomosis digitally and make sure that the two limbs of bowel are completely anastomosed. The operation from below is completed at this point.



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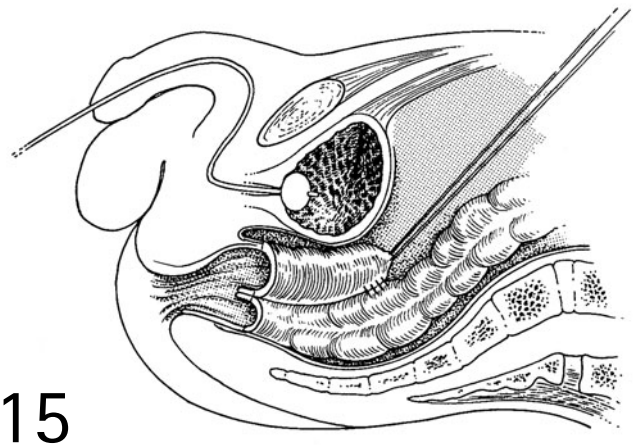


15

15 The anastomosis is completed by suturing the proximal end of the aganglionic rectum to the enterotomy in the ganglionic colon in two layers. The neorectum may or may not be reperitonealized, and the abdomen is closed.

ENDORECTAL PULL-THROUGH (SOAVE)

The Soave or endorectal pull-through was popularized by Franco Soave at the Institute G. Gaslini in 1955. The procedure was modified by Boley by performing a primary anastomosis at the anus, and then further modified by Coran. This procedure is the most popular one used at the authors' institute, and has been further modified to facilitate the suturing of the anal anastomosis. The operation is now most commonly performed in the newborn period, with the complication rate being identical to that seen with the standard two-staged approach. Over the past decade, the operation has been performed with laparoscopic assistance and via a transanal route.

As with the Duhamel technique, this procedure avoids injury to the pelvic nerves and, by remaining within the muscular wall of the aganglionic segment, important sensory fibers and the integrity of the internal sphincter are preserved. Although one imagines that leaving aganglionic muscle surrounding normal bowel could lead to a high incidence of constipation, this is not the case.

Preoperative

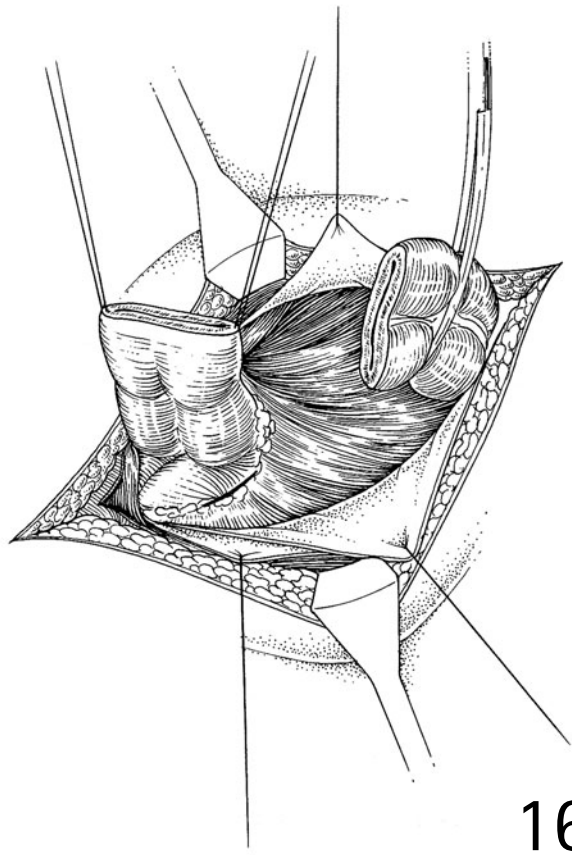
Even in the neonatal period, serial rectal washouts and digital dilatations of the rectum are performed before beginning the pull-through, the last of the rectal irrigations containing 1 percent neomycin. Intravenous antibiotics are given before the beginning of surgery and are continued for two doses after surgery.

Open operative technique

16 The child is placed in a supine position, with the buttocks brought to the end of the operating table, and propped slightly up with a folded towel. The legs are carefully padded and placed on wooden skis extending off the end of the table. A Foley catheter is placed and the entire field is prepared and draped. The operating table is placed in a slightly Trendelenberg position. A hockey-stick incision is made, incorporating the leveling colostomy (if present). The same type of incision is made, however, for infants undergoing a primary pull-through operation. The level of aganglionosis is established by frozen section.

Ganglionic bowel is mobilized proximally and then transected above the transition level with a stapling device. The distal colon is mobilized and resected to about 4 cm above the peritoneal reflection. Traction sutures are placed on either end of the distal bowel. The endorectal dissection is then started about 2 cm below the peritoneal reflection.

The authors have progressively shortened the length of the endorectal dissection, because longer lengths of muscular cuff may lead to increased bouts of constipation and enterocolitis.



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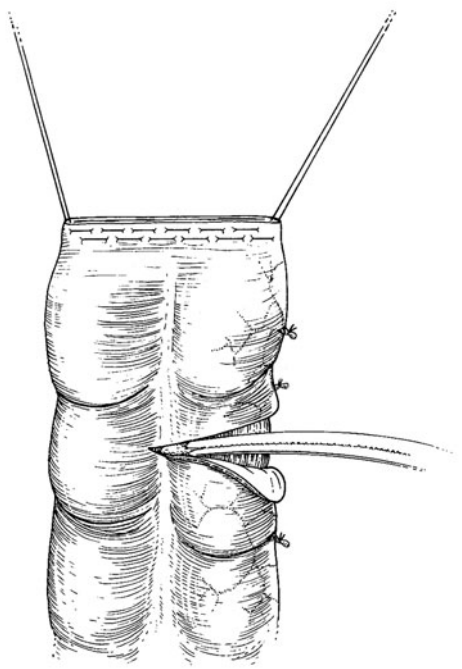
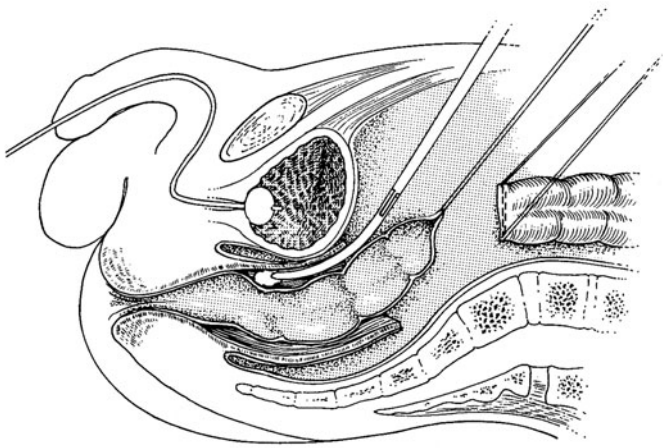


17

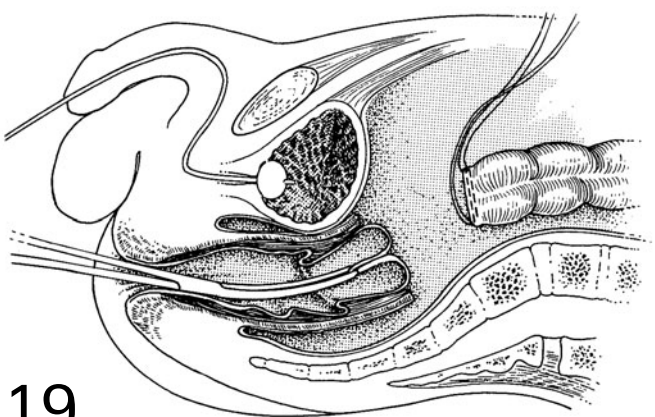
17 The endorectal dissection usually begins by completely clearing the serosa, mesentery, and fat over a 2 cm length of bowel. The seromuscular layer is incised with either sharp dissection or Bovie cautery. Once the submucosal layer is reached, the seromuscular layer is divided circumferentially using blunt dissection with hemostat or a Kitner dissector. In the neonatal period, a cotton-tip applicator is the most effective tool for this dissection.

18 After the plane is established, it is continued distally and facilitated by an assistant pulling upward on the already dissected mucosal/submucosal tube for countertraction. As the muscular cuff begins to develop, traction sutures are also placed in the muscle, one in each quadrant. Larger communicating vessels are coagulated; however, the majority of these are not cauterized during the dissection without significant blood loss, particularly in the neonatal period. Dissection is carried down to within 1.5 cm of the anal opening in older children and less than 1 cm in neonates.



18

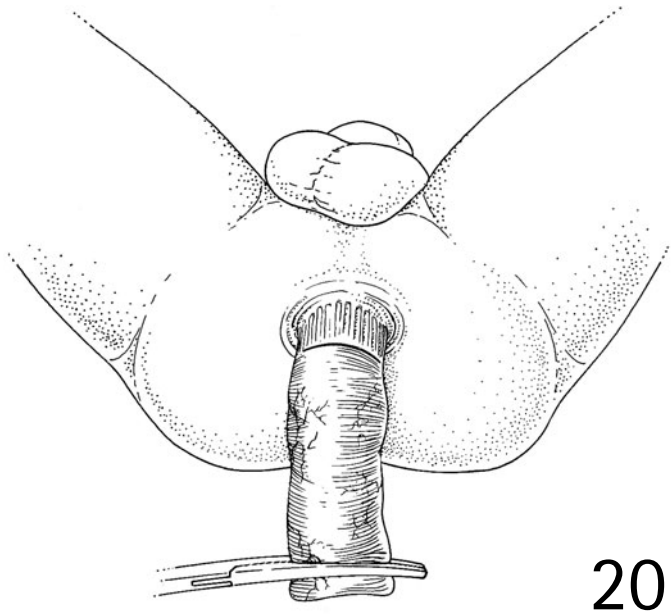
Some have advocated performing part of the endorectal dissection from the transanal approach, but the authors strongly advise against this if the dissection is started within the abdominal cavity. Once the endorectal dissection from above is started, it can proceed in a straightforward fashion. With appropriate traction and countertraction, the entire dissection can be performed in a child of almost any size (and adult).



19

19 One of the surgeons then moves to the foot of the table. Narrow retractors (phrenic or army–navy) are placed at the anal mucocutaneous junction and a ring or Kelly clamp is inserted into the rectum. An assistant at the abdominal field places the end of the mucosal/submucosal tube into the clamp. The segment is then everted onto the perineum. The end of the everted tube is placed in a clamp and held on traction by an assistant to facilitate the anastomosis.

20 The mucosal/submucosal tube is incised on the anterior half, 0.5 cm above the dentate line.



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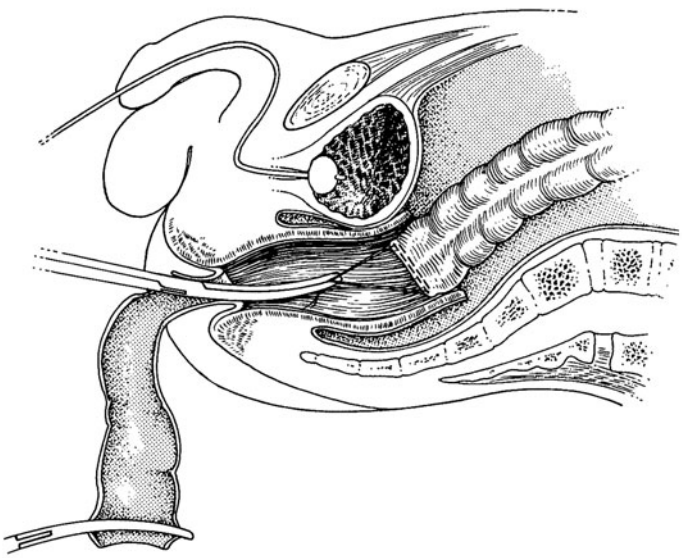
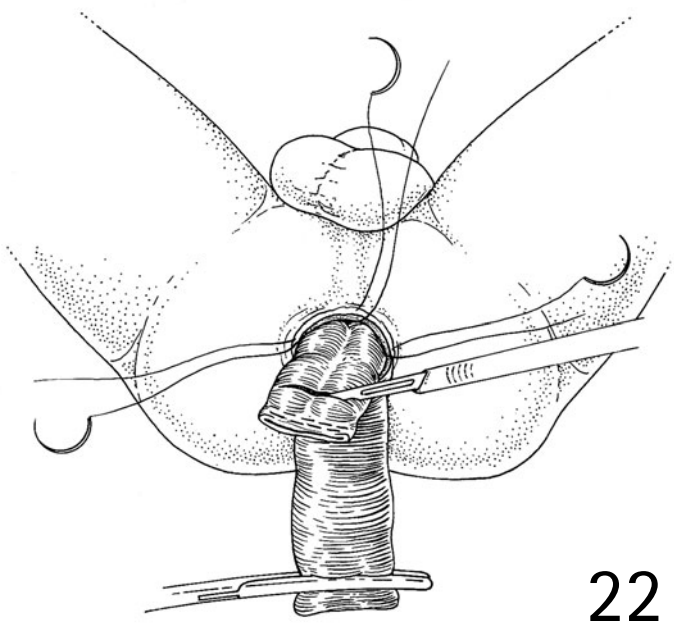


21

21 A Kelly clamp is inserted into this opening and the ganglionic bowel is brought down to this point by grasping the two previously placed traction sutures. Great care is taken not to twist the bowel as it is brought through the muscular cuff. As with the Duhamel procedure, different colored sutures on each side of the bowel are helpful in maintaining orientation.

22 The anterior half of the ganglionic colon is incised and anastomosed to the anterior half of the anus with a 4/0 polyglactin suture. The first sutures are placed at each corner and in the midline, followed by interrupted sutures in between.

The assisting surgeon can facilitate visualization of the two edges of the bowel by putting traction on these initial quadrant sutures as the operating surgeon completes each quadrant.



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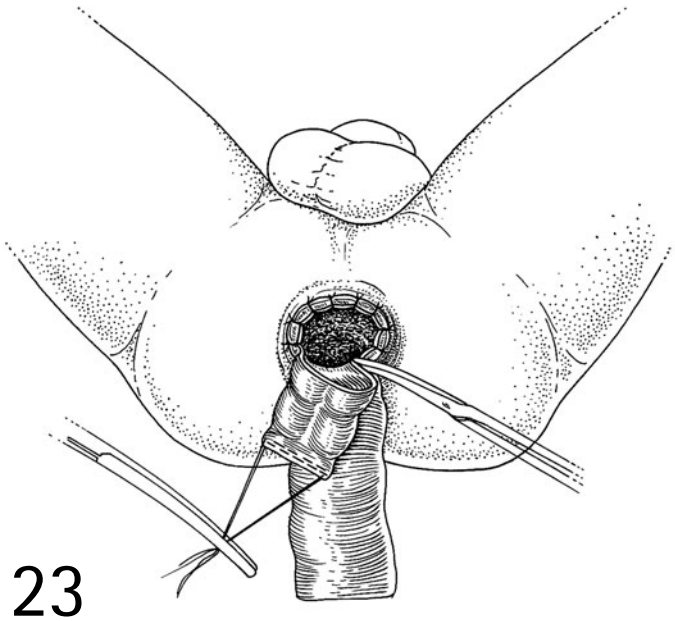
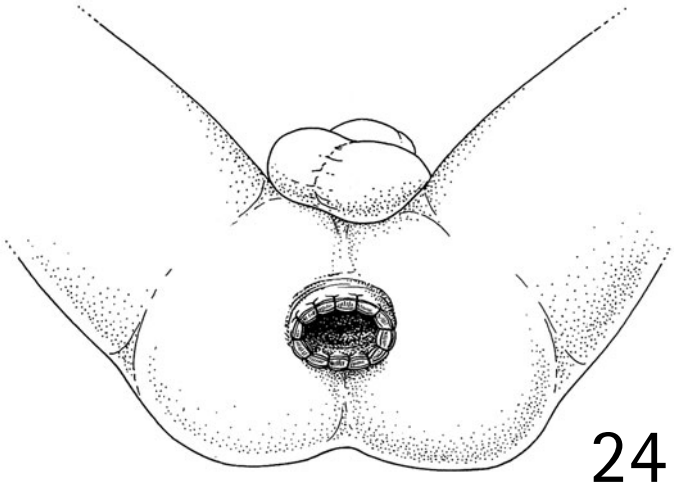


23

23 One-quarter of the remaining ganglionic colon and one-quarter of the everted mucosal/submucosal tube are opened. A suture is placed in the posterior midline and this quarter of the anastomosis is completed. Countertraction applied by an assistant on the everted tube will help with the exposure.

24 The final quadrant of the colon and tube are removed and the anastomosis is completed and inspected.

The colon is pulled slightly upwards to invert the neorectum back into its correct position. Rectal examination at this point should reveal a well-formed anastomosis 1.5–2 cm above the anodermal junction. Gloves are then changed and attention directed to the abdominal field.



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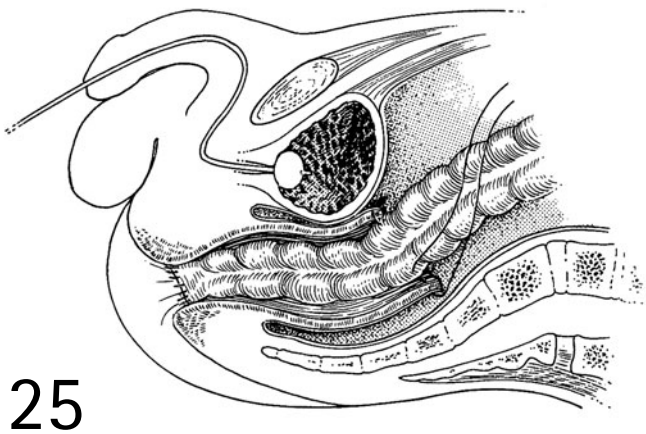
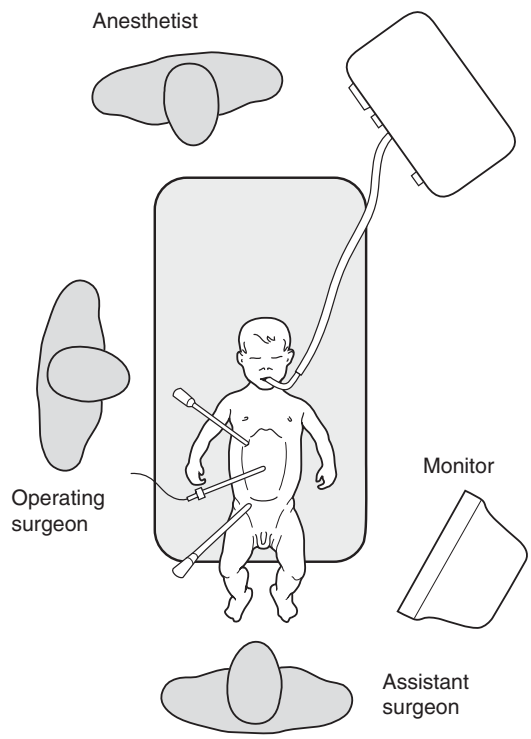


25

25 The pulled-through colon is attached with seromuscular bites to the muscular cuff to prevent the colon from prolapsing in the early postoperative period. No drain is placed, either through the anastomosis or from above in the muscular cuff, because there is rarely any significant oozing in the cuff (unlike the situation with ulcerative colitis in older patients).

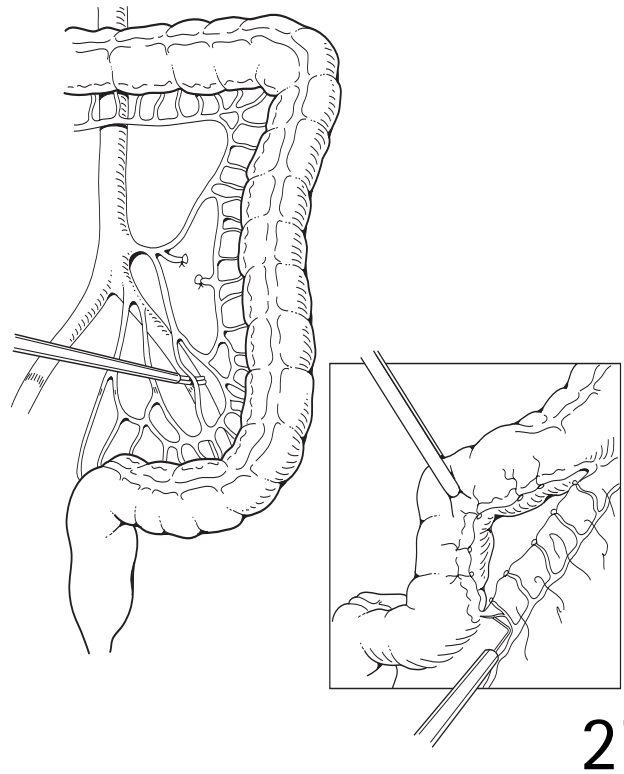
Laparoscopic approach

26 The basic operative principles are virtually the same with the laparoscopic approach. Trocar placement consists of an initial umbilical trocar, followed by trocars in the right upper quadrant, and the remaining one in the suprapubic area.



26

27 Dissection and leveling of the aganglionic segment are identical to the open technique. Blood vessels may be ligated with surgical clip appliers, or cautery for smaller vessels. Once mobilization is completed, the surgeon moves to the perineum, where a transanal dissection and anastomosis are performed (see below).



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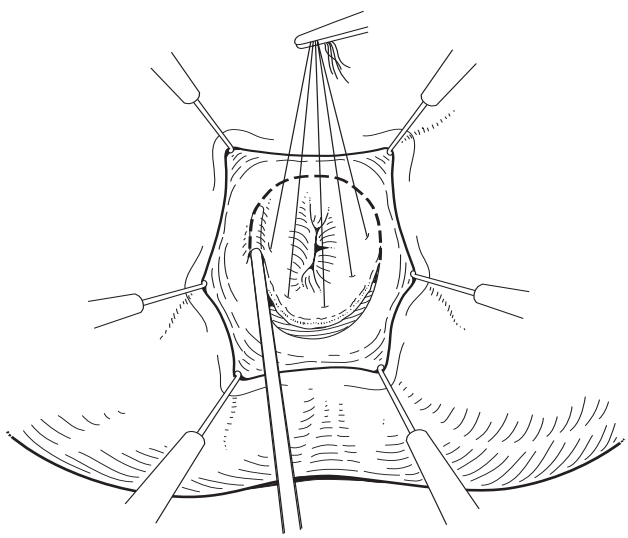
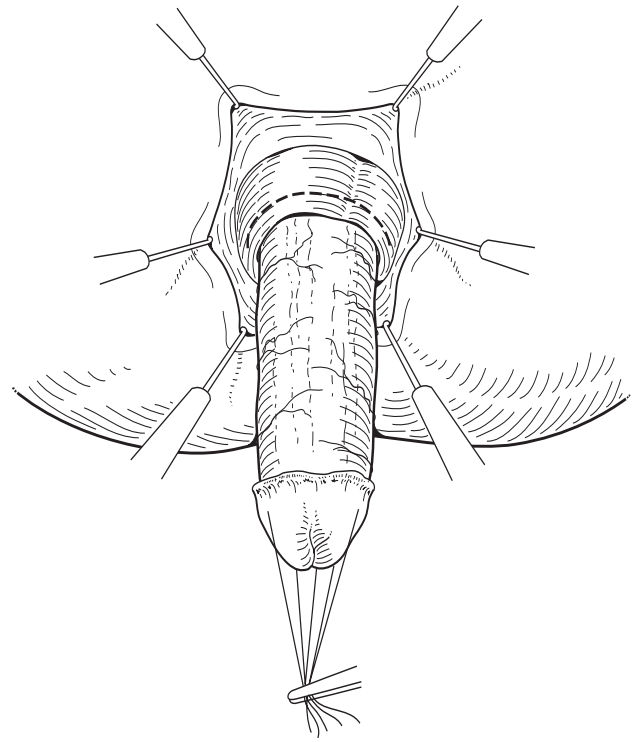


28

Transanal approach

28 The key element to beginning the case is placement of either a self-retaining retractor (e.g., Lone Star Medical Products, Houston, TX) or a series of sutures that retract the anal verge.

29 An incision 0.5 cm above the dentate line is begun with cautery in a circumferential fashion. Traction sutures are placed on the mucosa/submucosal tube, and dissection is carried proximally, primarily with blunt technique.



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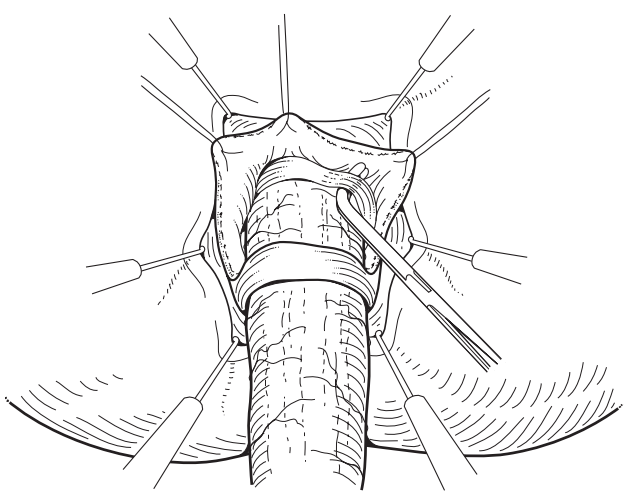
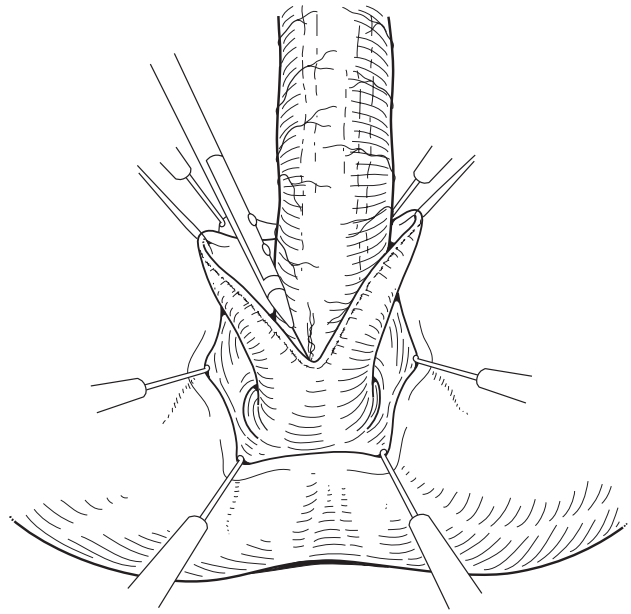


30

30 Once dissection is carried proximal to the peritoneal reflection, the muscular layer is entered, and the dissection becomes full thickness.

31 A key point is to split the muscular tube down to the internal sphincter in the posterior midline; this splitting prevents the muscular cuff from retracting and causing a relative obstruction. Although dissection of mesenteric vessels is easier with the laparoscopic approach, for rectosigmoid Hirschsprung disease, dissection of many of these vessels can be carried out via the transanal route. Biopsies are sent to determine the level of ganglionic bowel.



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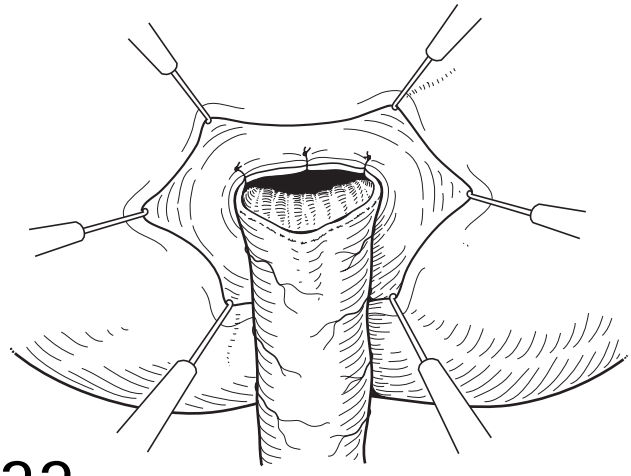


32

32 Completion of the anastomosis is very straightforward.

SWENSON PROCEDURE

This technique was originally described by Swenson and Bill in 1948 and was the first successful method of treatment for children with Hirschsprung disease. They based their technique on the principle that the diseased portion of the bowel was the aganglionic distal rectum, and that removal of this segment was necessary to allow for normal stooling. The initial incidence of postoperative enterocolitis was fairly high (early 16 percent and late 27 percent), and this was attributed to leaving too much aganglionic rectum. The procedure has since been modified by creating an oblique anastomosis, in which one resects virtually all of the posterior rectal wall (and some internal sphincter) while leaving 1.5–2 cm of anterior rectal wall.

The technique demands meticulous dissection of the rectum down to within 2 cm of the dentate line. If the dissection moves off the rectal wall, a significant incidence of injury to the genitourinary innervation may occur. Properly performed, the results with this procedure are quite good; however, because of the technical difficulties of the dissection, it has fallen into relative disfavor.

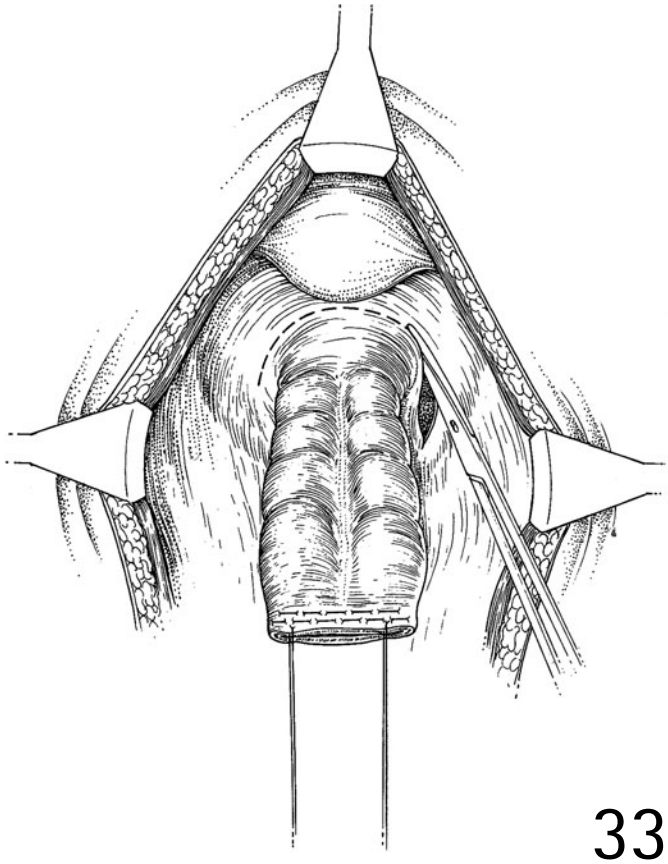
Preoperative

The child is admitted the day before surgery for a routine bowel preparation. Assessment and preparation are similar to those used in other pull-through operations.

Operative technique

33 The child is positioned in a fashion similar to that for an endorectal pull-through procedure. The incision was classically described as a left paramedian incision with takedown of the colostomy; however, the modified hockey-stick incision will work equally as well.

The redundant aganglionic rectum is excised and proximal ganglionated colon mobilized past the splenic flexure, if necessary, as in the other two techniques. At this point, the peritoneal reflection over the rectum is incised.



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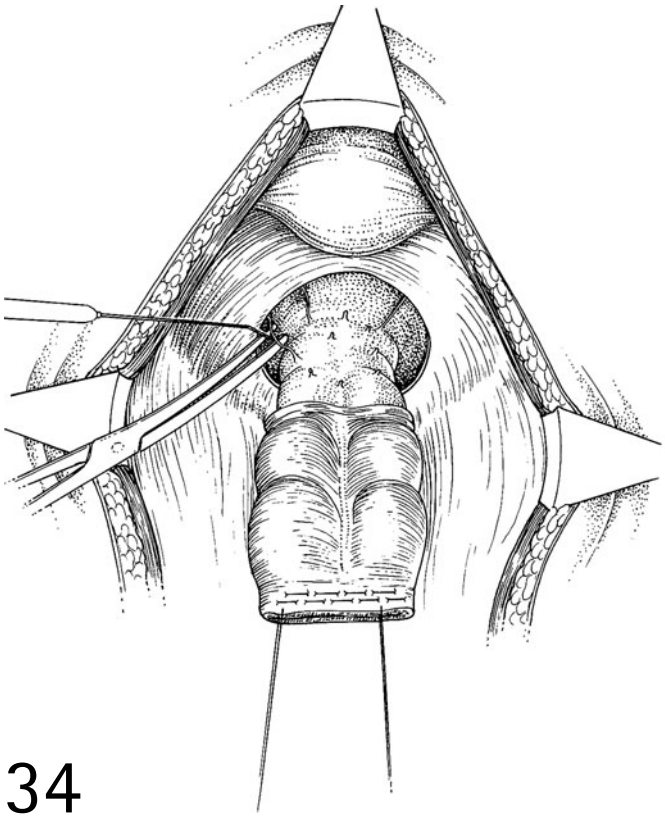
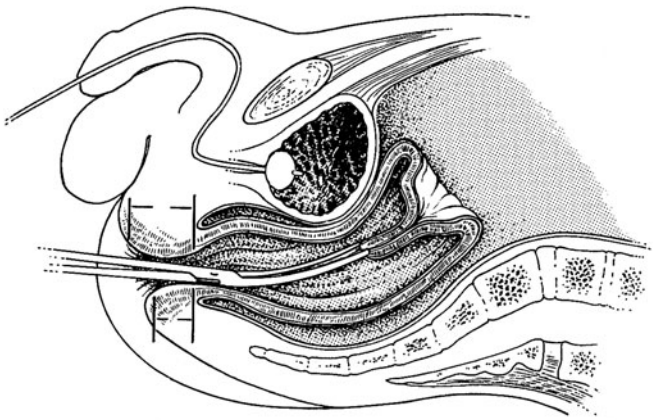


34

34 The operating surgeon then dissects the rectum caudally. This is a critical dissection, which demands the surgeon to stay directly on the bowel wall. Dissection is facilitated by the first assistant applying upward traction on the end of the aganglionic rectum. Multiple blood vessels enter directly into the bowel wall; each must be dissected out and can usually be coagulated. Dissection is carried down toward the anal verge, but is not carried as far anteriorly in order to avoid autonomic nerve injury.

35 The perineal part of the operation is then started. The rectum is everted with the use of a long clamp. The anterior half of the everted rectal wall is cut 1.5–2 cm proximal to the anodermal junction. The posterior wall will be no longer than 1 cm in length. A gently curved incision, which is shorter posteriorly, is thus created along the anterior half of the bowel.



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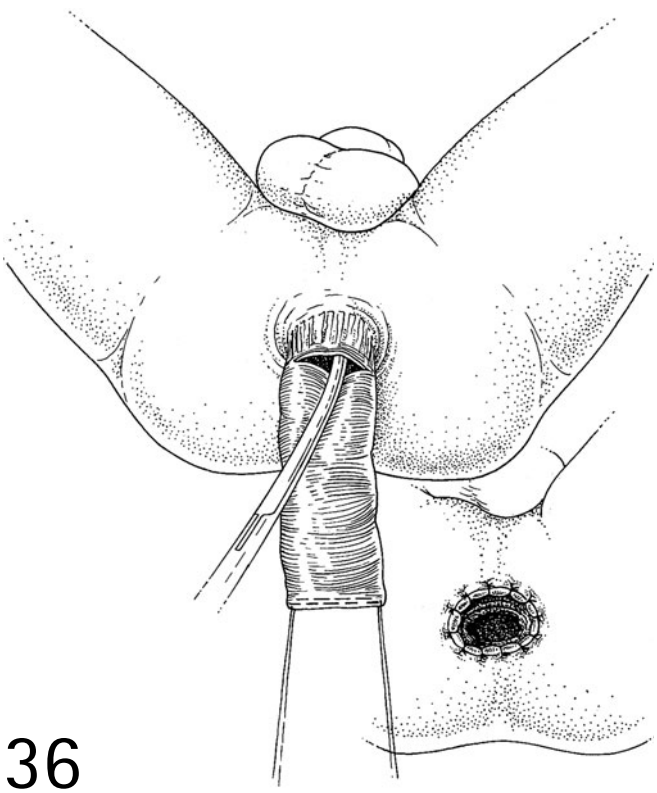


36

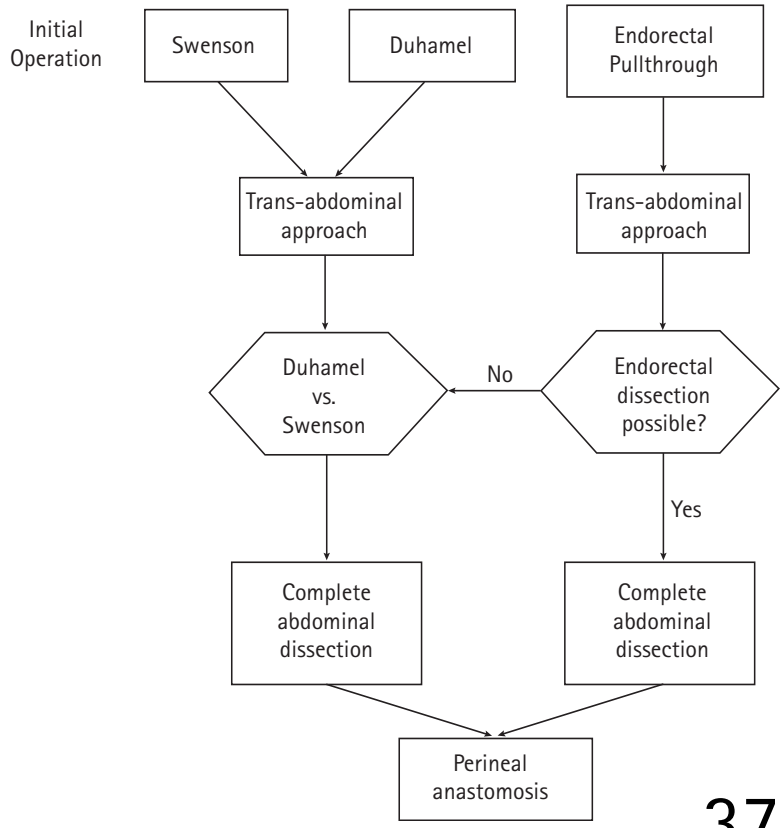
36 The ganglionic colon is pulled through and the anastomosis is virtually identical to that for the endorectal pull-through operation. Classically, silk sutures have been used, but the authors have utilized polyglactin sutures with good results.

The anastomosis is allowed to recede and is gently pulled upward from the perineum. Closure is essentially the same as for the previous procedures.

SECOND PULL-THROUGH

37 A child with an initial unsuccessful pull-through operation may present with severe constipation or significant incontinence. These children should undergo a thorough investigation of the details of the initial operation as well as a review of the pathologic specimens. In most cases, an appropriate pull-through has been performed and an aggressive bowel program needs to be instituted. It is essential to rule out a retained segment of aganglionic bowel. Should this be the case, a second pull-through operation will usually be necessary. Because of its relative ease, a Duhamel is generally the procedure of choice. The authors quite commonly perform an endorectal pull-through in patients who have previously undergone this procedure. For those patients who underwent an initial Duhamel procedure, a Swenson pull-through should be used.

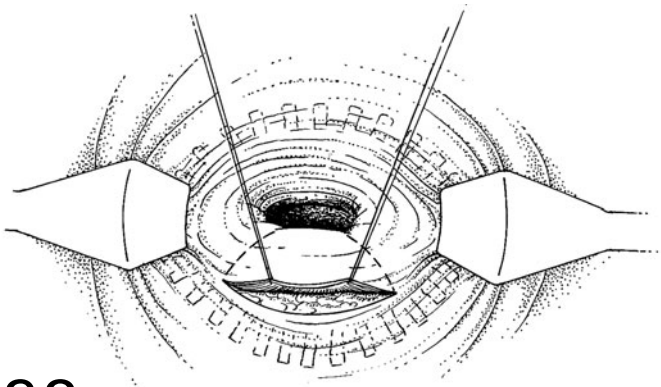
For the child with debilitating incontinence, a detailed history should be taken and an examination to rule out encopresis and to assess the degree of anorectal tone. Anal manometry is particularly helpful in this group, as a lack of normal muscular control is generally felt to be a poor indicator of a successful outcome for a repeat pull-through operation. An occasional child in this latter group may best be served by a diverting colostomy.



37

ANAL/RECTAL MYECTOMY

A child with short-segment Hirschsprung disease may be a candidate for an anal/rectal myectomy. The advantage of this procedure is that it avoids an abdominal operation. The procedure must be confined to very short-segment (5 cm or less) Hirschsprung disease. One must be certain that the myectomy is extended beyond the level of aganglionosis. Because performing an inadequate myectomy may adversely affect the outcome of a subsequent pull-through operation, a myectomy should be avoided if there is any uncertainty as to the level of disease. More commonly, an anal/rectal myotomy or myectomy is performed for a child who, following a pull-through procedure, has persistent enterocolitis symptoms, or who has difficulty stooling after a pull-through procedure. The most satisfying results with a myectomy are for those children with long-standing enterocolitis after a pull-through procedure.



38

Operative technique

TRANSANAL APPROACH

38 The child is placed in an identical position to that for the full-thickness rectal biopsy.

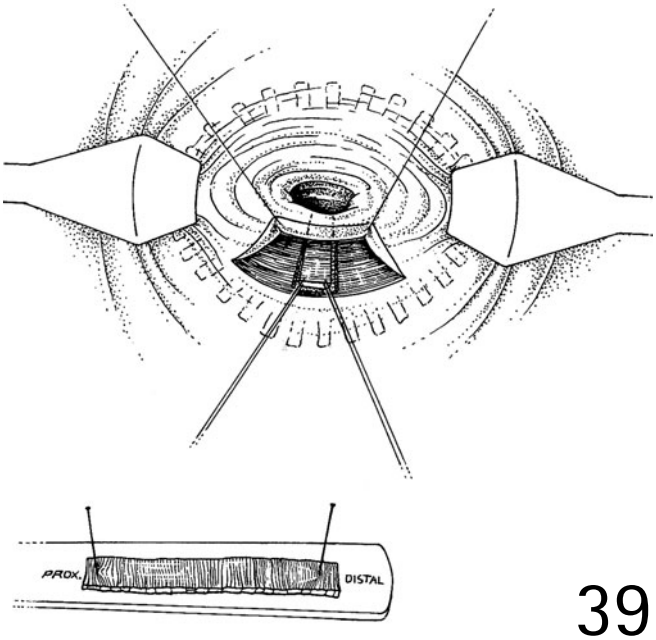
Digital dilatation is performed, and two narrow anal retractors are inserted and held by assistants. The posterior aspect of the dentate line is identified. A 2 cm transverse incision through the mucosa and submucosa starting 1 cm above the dentate line is made. Following this, a submucosal dissection is carried upward for several centimeters. The mucosal/submucosal flap is held up with silk sutures and can be extended proximally with vertical incisions on either side.

39 The myectomy is then performed by sharply incising the full thickness of the muscle layer and a 0.5–1 cm wide muscle strip is created in the midline using Bovie cautery. The initial strip removed should be at least 5 cm in length.

Before transecting this strip, two silk sutures are placed proximal to the point of transection so that, should further dissection be necessary, the proximal muscle will not retract beyond the view of the surgeon. The strip should be mounted on a tongue depressor with mounting pins or suture; proximal and distal orientation must be clearly depicted.

A frozen section confirming ganglion cells at the proximal margin must be obtained before the procedure can be terminated. The dissection can be carried out in this manner for approximately 8 cm. If the surgeon suspects that the level of aganglionosis is longer than this, it may be advisable to forego this approach.

Once a sufficient length of muscle has been removed, hemostasis is achieved with cautery. The wound is irrigated and closed with fine, interrupted, absorbable sutures approximating the mucosa/submucosal dissection.



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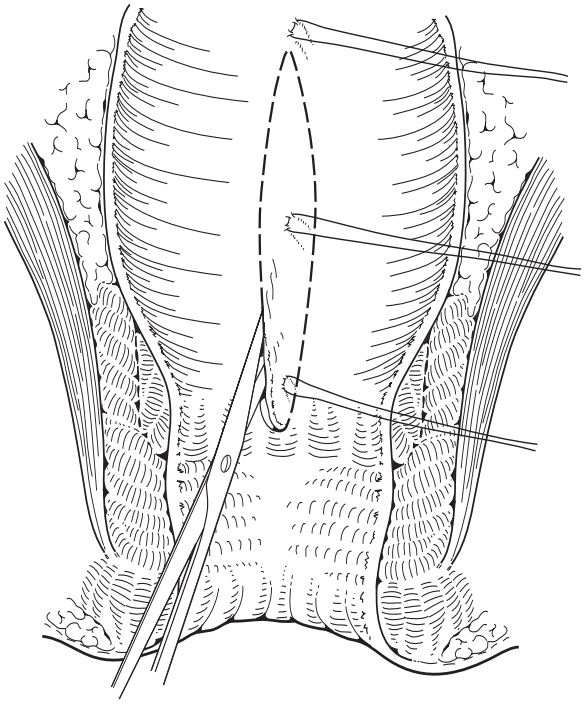
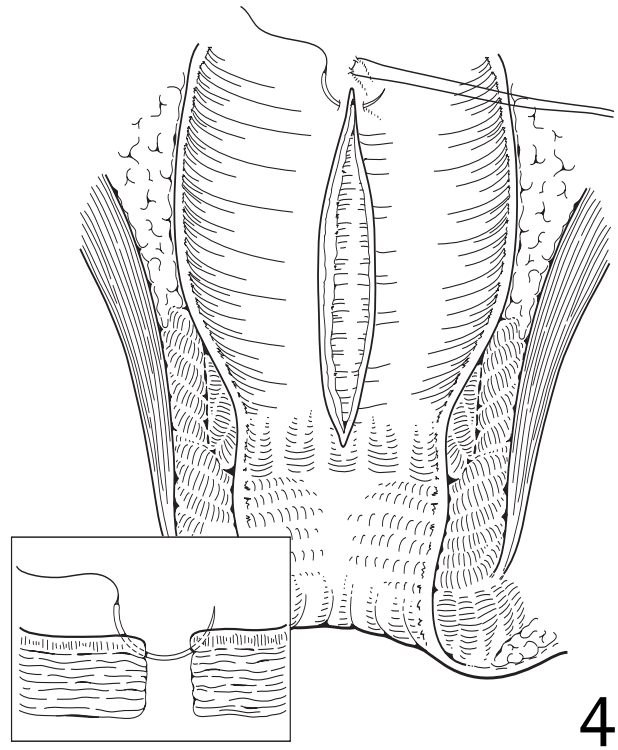


40

40 For patients who have had several procedures, obtaining a submucosal plane may be quite difficult and it is simpler to perform a vertically oriented, elliptical, full-thickness excision.

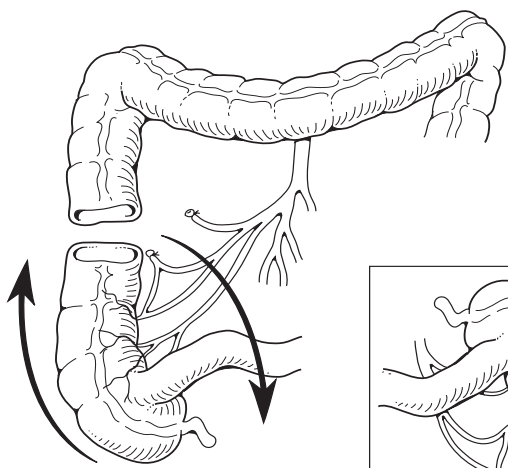
41 Closure is performed with a running absorbable suture that approximates only the mucosa and submucosa, leaving a defect in the muscular layer.



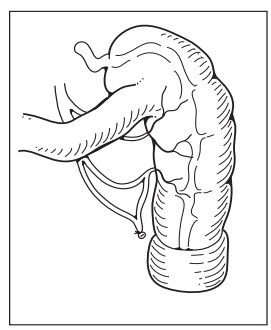
41

Right-sided aganglionosis

Pull-throughs in patients with extended colonic disease (to the right of the middle colic vessels) often create problems in maintaining the colon in the correct orientation, and are at greater risk for torsion of the pulled-through intestine. This problem was recognized by Duhamel early on.



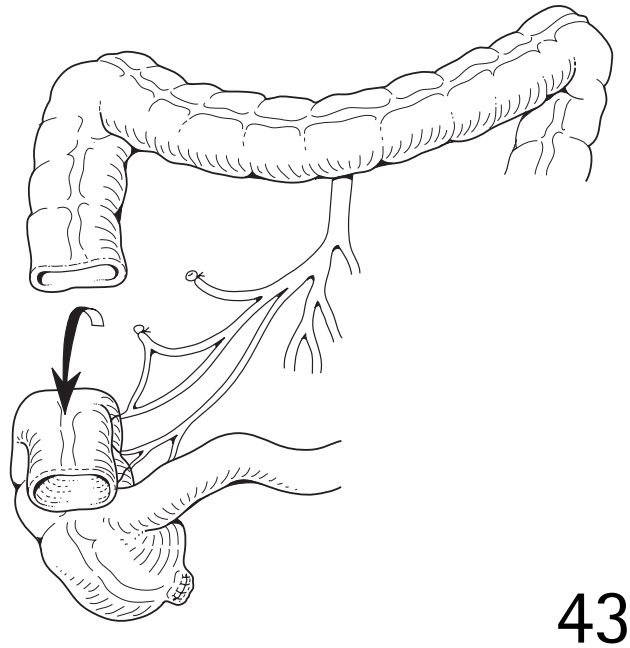
42a



42b

42a,b This problem may be best approached by rotating the right colon in a counterclockwise fashion, as shown below. Because the small bowel then exits the colon from the patient's left side, it is sometimes necessary to follow this with a complete release of the ligament of Treitz (i.e., create a malrotation).

43 Alternatively, the colon may be flipped anteriorly, with reasonably good results.



43

POSTOPERATIVE CARE

Following all of the pull-through procedures, the nasogastric tube is removed on the first postoperative day, and feedings are begun once gastrointestinal activity returns. The Foley catheter can be removed on the second postoperative day following the endorectal and Duhamel procedures because bladder innervation has not been affected by the pelvic dissection. However, after the Swenson pull-through or redo pull-throughs, the Foley catheter should be left in for 4–5 days because of the possibility that there may be some bladder atony from the dissection. Antibiotics are continued for 24 hours after surgery. Most patients are discharged by the third postoperative day.

Great emphasis is placed on examining the perineal region for the development of erythema or cellulitis, as this is an early sign of an anastomotic leak. Investigation of a leak consists of an enema contrast study or a computed tomographic scan with contrast medium. Although fairly uncommon, should a leak be found, the urgent placement of a diverting colostomy is needed.

For the endorectal pull-through, a gentle rectal examination with cotton-tip applicator is performed to ensure patency of the rectal anastomosis before discharge. Parents are given thorough instructions on perineal care and the potential development of enterocolitis. To avoid perineal excoriation, the parents are instructed to apply a thick coat of a zinc oxide-based ointment with each diaper change. If a significant rash develops, more intensified applications are used.

Digital or small Hegar calibrations are performed, starting on the third week after surgery. These are occasionally needed after the endorectal pull-through procedure, particularly in neonates. These dilatations are usually sufficient to prevent anastomotic stricturing.

Stool frequency is generally quite high (7–12 bowel movements per day) immediately after the pull-through operation, but this slowly decreases and is generally normal by 6–9 months.

Children should be followed up for several years. Occasionally some develop intermittent bouts of constipation, occasional soiling, and enterocolitis. Most problems with constipation or soiling can be managed with changes in diet or enema regimens. Episodes of enterocolitis are managed with oral metronidazole; however, severe cases will necessitate admission, intravenous antibiotics, and rectal washouts. Increasingly, a large number of patients are started on rectal washouts, beginning at 3 weeks post-pull-through. This acts as a prophylactic method of avoiding enterocolitis, and may be continued for the first few months following the surgery.

With regard to children with total colonic Hirschsprung disease, large volumes of stool and electrolyte losses can occur after either a modified Duhamel or an ileoanal endorectal pull-through. These losses must be replaced and are usually controlled with dietary changes and the addition of an opioid agent (e.g., loperamide). Perineal excoriation is very common in these infants and demands constant attention. Normalization of stooling will take much longer than in children with classic Hirschsprung disease, and these children may sometimes require parenteral nutritional support for several months before full enteral feeding can be initiated.

OUTCOME

There is controversy concerning which procedure yields the best results. Long-term outcomes with the Duhamel, Soave and Swenson techniques are basically similar, provided they are performed with meticulous technique. Depending on the anatomy and history of the patient, the surgeon should be familiar with all of these techniques, but should use one as the primary procedure. This is the only way that consistently good results can be achieved.

Of equal importance is the need to follow such patients over long periods of time, as many of the complications (e.g., enterocolitis, constipation, and urgency) may not become manifest until much later in the patient's life.

Acknowledgments

Illustrations by Paul Richardson and Shayne Davidson.

Illustrations 19, 21 and 24 are reproduced from Coran AG, Weintraub WH. *Surg Gynecol Obstet* 1976; **143**:277–82, with permission of the publishers.

FURTHER READING

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56

Inflammatory bowel disease

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HISTORY

The evolution of surgical procedures for inflammatory bowel disease has been one of trial and error. Based on previous successful and unsuccessful outcomes, a variety of procedures have been developed that allow for maximal preservation of bowel length as well as function. These include proctocolectomy with end-ileostomy, endorectal pull-through, and stricturoplasty. The pediatric patient with inflammatory bowel disease presents with additional growth, nutritional, and psychologic problems that may not affect the adult patient. All of these factors must be considered when determining the timing and type of procedure.

Over the past 20 years, the use of the endorectal pull-through for ulcerative colitis has become increasingly popular. The operation was initially a modification of the endorectal pull-through technique for Hirschsprung disease. The procedure has taken time to become accepted by the general surgical community, principally because of unfamiliarity with the endorectal dissection and the significant incidence of complications associated with many of the original cases.

PRINCIPLES AND JUSTIFICATION

Protocolectomy with end ileostomy

This has been the standard procedure for children with ulcerative colitis and multiple polyposis, but can also be utilized in severe Crohn's colitis. The abdominal colectomy portion of the procedure may also be performed in conjunction with an endorectal pull-through. For patients with ulcerative colitis, initial medical management is generally advised. In children with an acute exacerbation of ulcerative colitis, surgery is indicated in cases of severe hemorrhage or in those who fail to respond to intensive medical treatment after several weeks. The timing of an elective procedure is more difficult to establish. Some surgeons feel that resection is only indicated in those children who show mucosal atypia on colonoscopic biopsy. Detecting these changes is difficult because one cannot routinely biopsy the entire colon, and the incidence of carcinomatous changes increases as the duration of the disease increases. In fact, carcinoma can be found in many surgical specimens when only atypia was found on colonoscopic biopsy. Many surgeons therefore recommend a colectomy once the disease process has been present for 10 years. However, today many gastroenterologists recommend surveillance colonoscopy every year or every other year even if the disease has been present for more than 10 years. Surgery should be performed sooner if atypia is identified or in those children with significant growth failure or lack of sexual maturation, and in those on chronic high doses of steroids in whom significant changes and complications due to steroid use have occurred. In children with multiple polyposis, the timing can also be controversial. If all the polyps can be removed from the colon and the child is followed up every 6 months, surgery can be delayed at least until the child is past adolescence. If the polyps are too numerous to be removed, surgery should be performed earlier.

Endorectal pull-through

The endorectal pull-through is a curative procedure for patients with ulcerative colitis as well as colonic polyposis, while eliminating the need for a permanent ileostomy. An additional advantage of this procedure is the elimination of the extensive pelvic dissection outside the rectal wall, which can be associated with a significant incidence of injury to the nerves supplying the genitourinary system.

Stricturoplasty

Strictures secondary to Crohn's disease of the small bowel do occur in children. Conventional approaches to their treatment have consisted of complete resections or side-to-side bypasses of the involved area. Multiple resections of these strictured areas have not uncommonly led to the development of the short bowel syndrome. Bypass of significant areas of the bowel usually results in bacterial overgrowth. Over the past 10 years the use of stricturoplasty has therefore evolved. The technique has enabled patients to retain significantly greater lengths of bowel with adequate relief of the obstruction and is a modification of the Heineke–Mikulicz procedure for a pyloroplasty. Although the bowel length may appear shorter after the performance of multiple stricturoplasties, the functional length is the same. A few contraindications to this procedure exist, for example the occurrence of several small strictures very close together, which could be more simply managed with a resection, although it is not uncommon for a patient to have multiple stricturoplasties during one operation. If the stricture is associated with fistula or abscess formation, the bowel segment should be excised.

Ileocolectomy

Although ileocolectomy is commonly performed in the pediatric patient for Crohn's disease, the procedure is reasonably straightforward and an operative description is not included in this chapter.

PREOPERATIVE

Proctocolectomy with end ileostomy

The child is admitted the day before surgery and undergoes a complete bowel preparation. Caution should be observed in those patients with severe colitis or Crohn's disease. Overly aggressive laxatives may cause a perforation of the colon or an exacerbation of the disease process. In general, a balanced electrolyte solution should be slowly administered orally. Oral antibiotics and gentle enemas are given the day before surgery. An enterostomal therapist should mark the skin site of the ileostomy with indelible ink the day before surgery.

Endorectal pull-through

Before performing the procedure, the diagnosis of ulcerative colitis must be as firm as possible. If Crohn's colitis is present instead of ulcerative colitis, recurrent disease in the pulled-through ileum and fistula formation from this same segment of bowel are common. Repeat colonoscopy along with a series of small intestinal contrast studies should be performed to rule out Crohn's disease. If biopsies have been performed elsewhere, they should be re-read by pathologists at the center where the operation is to be carried out. Many of these children will have been on steroids during the previous year and will need stress doses of steroids in the perioperative period.

Stricturoplasty

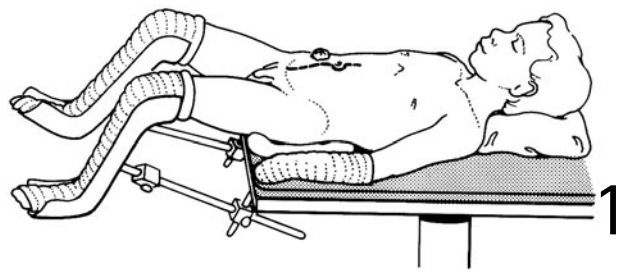
Suspicion of a stricture is usually initiated by the patient complaining of symptoms consistent with a partial bowel obstruction. Most patients complain of cramping abdominal pain, with obstipation and nausea. The diagnosis is made with a small intestinal contrast series. A thorough look for multiple strictures is necessary. No formal bowel preparation is needed, but intravenous antibiotics should be given.

OPERATIONS

Proctocolectomy with end ileostomy

POSITION OF PATIENT AND INCISION

1 The child is placed in the lithotomy position, with leg supports (without weight on the leg) for older children and skis for smaller children. Careful padding of the lower extremities is critical to avoid neurovascular injury. The procedure is best carried out through a large midline or left paramedian abdominal incision, which extends from the pubis to a few centimeters above the umbilicus.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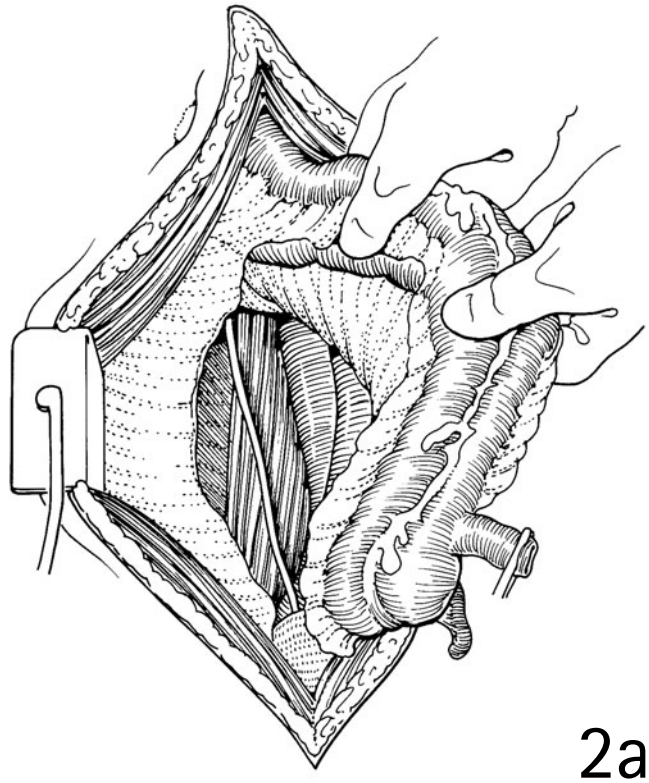
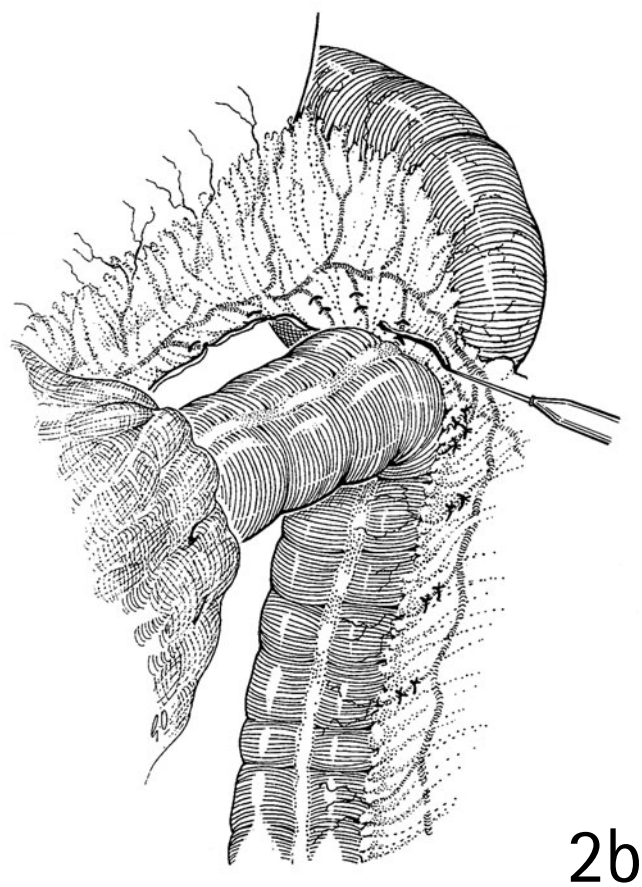


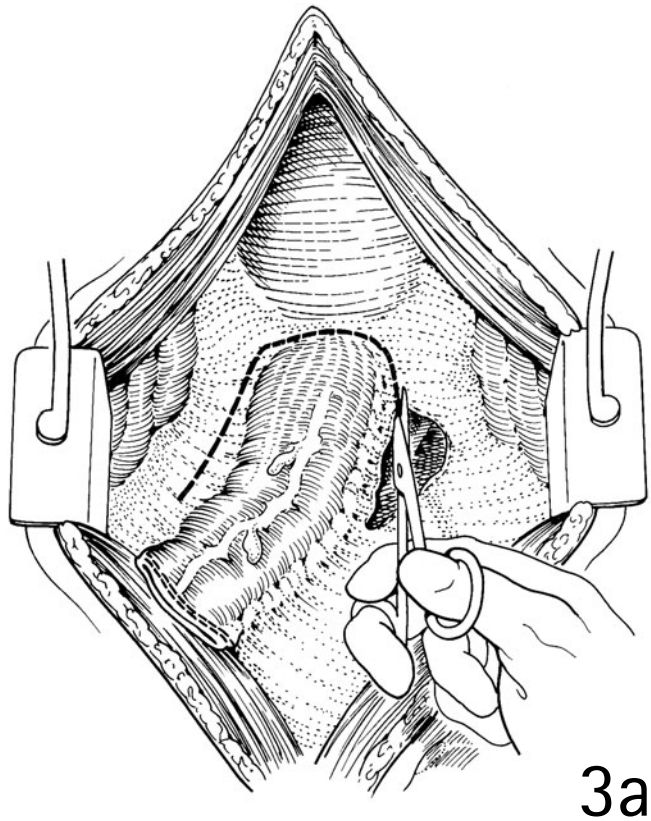
2a

MOBILIZATION OF COLON

2a,b The abdomen is explored and the right colon is mobilized by incising the line of Toldt and by dividing the terminal ileum approximately 1 cm from the ileocecal valve. Identification of the ureters is performed on both sides of the abdomen. The hepatic flexure and splenic flexure are then divided without putting traction on the spleen. The omentum is included in the transverse colectomy specimen.



2b



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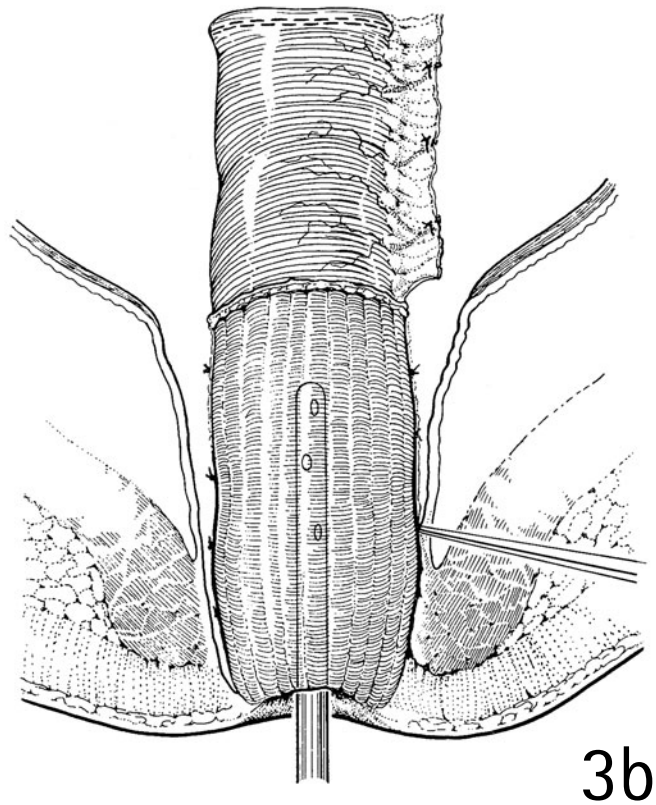
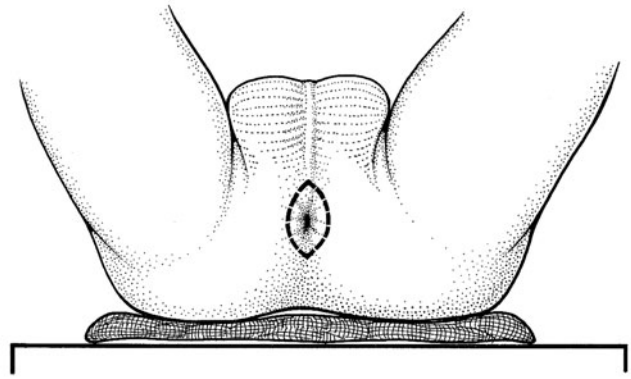


3b

3a,b The descending colon is mobilized in a similar fashion by dividing its retroperitoneal attachment. The mesentery of the colon is then divided and the vessels are suture ligated with 2/0 or 3/0 silk suture. The colon is then divided at the rectosigmoid junction with an automatic stapling device. The peritoneal reflection of the rectum is incised and the dissection is continued directly on the muscular rectal wall. Deviation from the rectal wall will increase the chances of injuring the pelvic autonomic nerves, with subsequent impotence or bladder dysfunction. This portion of the operation is facilitated by countertraction on the remaining proximal rectum by the assisting surgeon (b). Each individual vessel is grasped with forceps and either cauterized or ligated. Dissection should continue down as far distally as possible from the abdominal approach.

EXCISION OF RECTUM

4 The surgeon then moves to the foot of the table and an elliptical incision is made around the anus.



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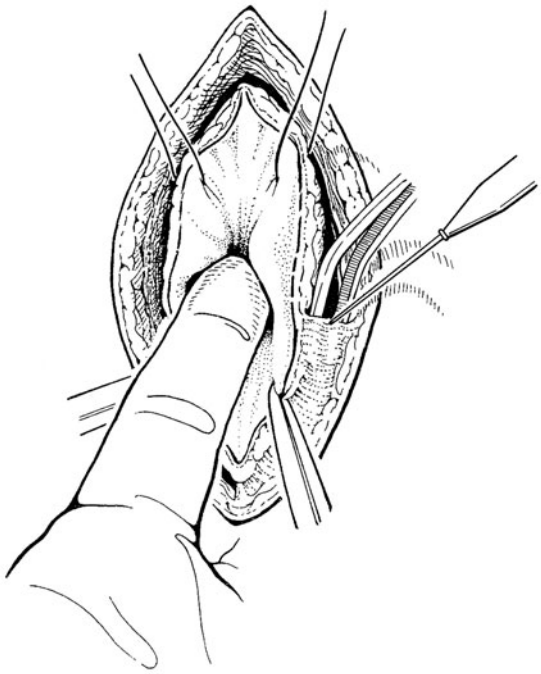
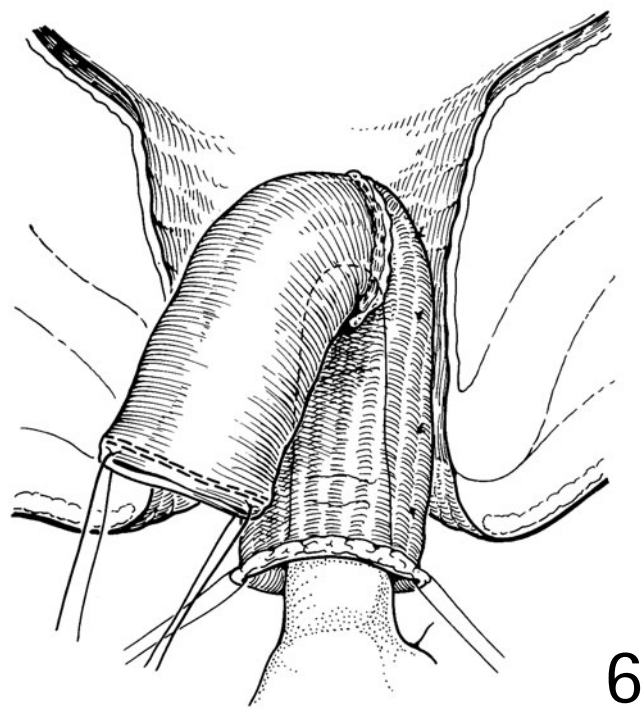


5

5 Dissection is carried upwards, remaining directly on the rectal wall. Skeletal muscles and the prostate should be avoided or considerable bleeding may arise.

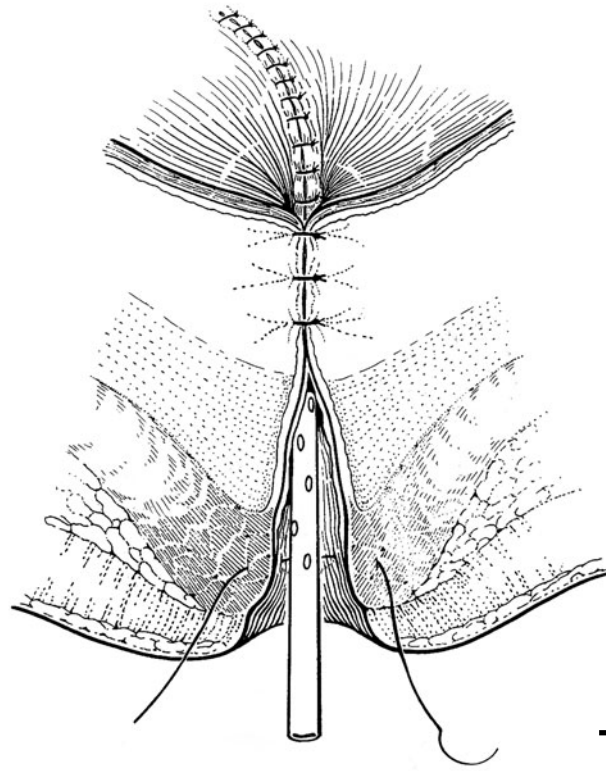
6 Dissection continues until the presacral space is entered posteriorly. The remainder of the excision is carried out with the proximal rectum everted onto the perineum. Sometimes, in a thin patient, the entire proctectomy can be performed transabdominally, with the actual excision of the anus from the abdominal approach. The peritoneum is approximated from the abdominal side.



6

TISSUE APPROXIMATION

7 The levator complex and subcutaneous tissues are approximated through the perineal wound. A small, flat, closed suction drain is placed into the perineal wound.



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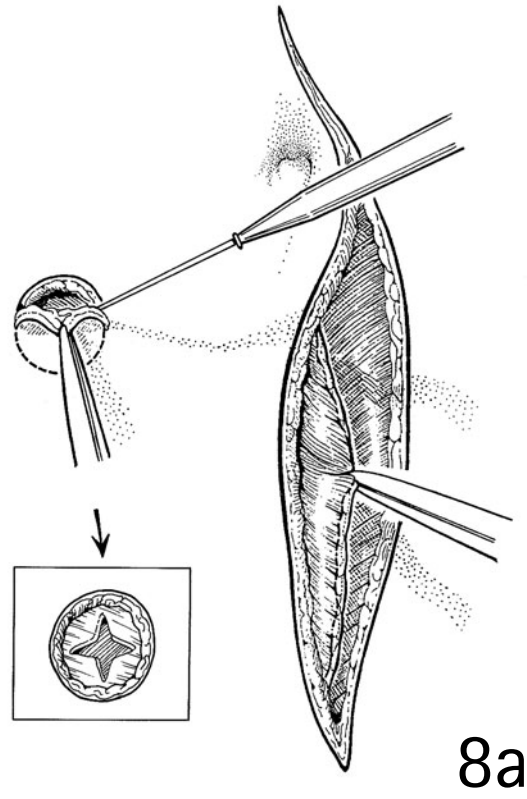
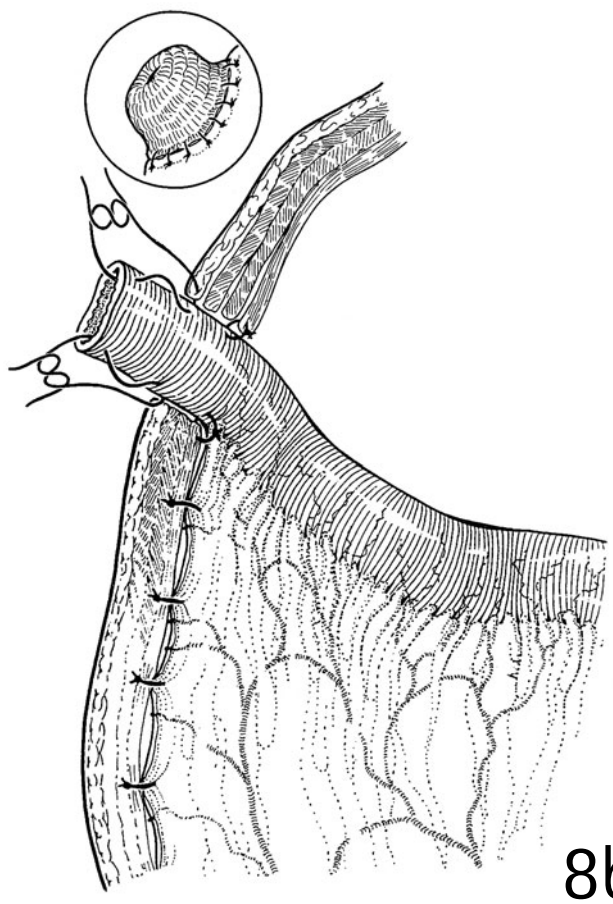


8a

FORMATION OF STOMA

8a,b The stoma is exteriorized at the previously marked site. The circle of skin about 1 cm in diameter is removed with cautery and the fascia is cruciated to allow the ileum to exit comfortably through it. The terminal ileum is prepared for creation of the ileostomy by ligating a few vessels along the most distal 3 cm of small bowel. The ileum is tacked to the peritoneum with interrupted sutures (silk or polyglycolic acid). Once the abdomen and skin are closed, a classic Brooke stoma is formed.



8b

Endorectal pull-through

POSITION OF PATIENT AND INCISION

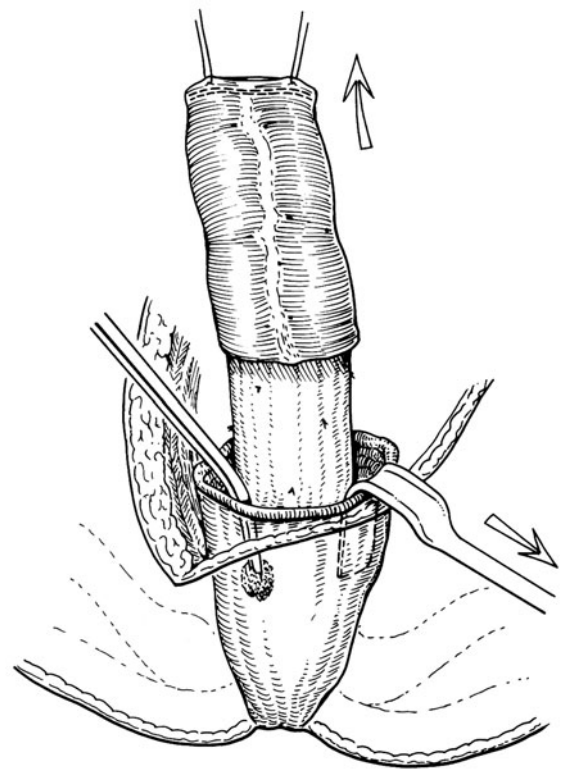
Positioning for this procedure is essentially identical to that for proctocolectomy; however, because of the longer procedure, very careful attention must be paid to padding of the lower extremities to avoid neurovascular injury. A long midline or paramedian incision is made from the pubis to several centimeters above the umbilicus. The abdominal colectomy proceeds as already described.

MOBILIZATION OF ILEUM

The ileum is mobilized proximally to allow for an adequate length for the pull-through. In general, length is rarely a problem with a straight pull-through. With the J-pouch, more time must be spent in gaining bowel length. Adequate bowel length can also be difficult when the patient has had a previous ileostomy, as the mesentery may be scarred and foreshortened. In general, the main branch of the ileocecal artery can often be spared and the more proximal arcades ligated in the distal ileum. It is important to preserve the distal vascular arcade to the end of the ileum. If the ileocecal artery must be ligated, a bulldog clamp should initially be placed on this vessel to determine if a significant length of bowel will be lost.

ENDORECTAL DISSECTION

The authors prefer to do the entire endorectal dissection from the abdominal approach. It is technically more difficult to develop the correct plane from the perineum and, although the initial portion of the dissection can be quite difficult in children with ulcerative colitis, once this correct plane is found the dissection proceeds fairly smoothly.



9

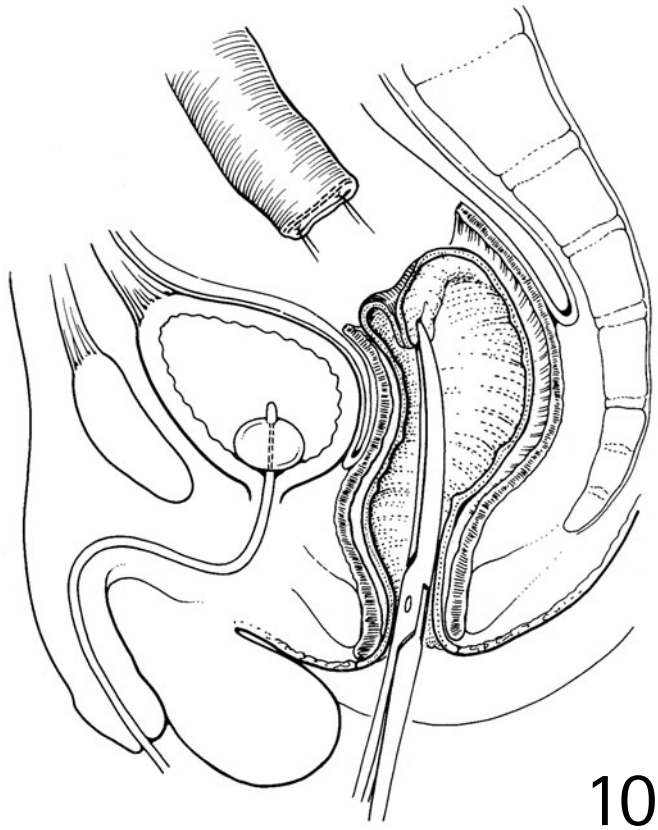
Countertraction of the dissected mucosal/submucosal tube, as well as traction in an upward and outward fashion on the muscular cuff, helps with the dissection.

The dissection involves greater blood loss than in cases of Hirschsprung disease. Any large penetrating vessels should be cauterized, but once the dissection is completed, bleeding usually stops spontaneously. If the correct plane is lost, the surgeon should turn to the opposite side of the bowel and work circumferentially until the dissection is distal to the area where the plane was lost. The level of the end of the dissection should be checked intermittently by feeling for an assistant's finger placed in the anus, just above the dentate line.

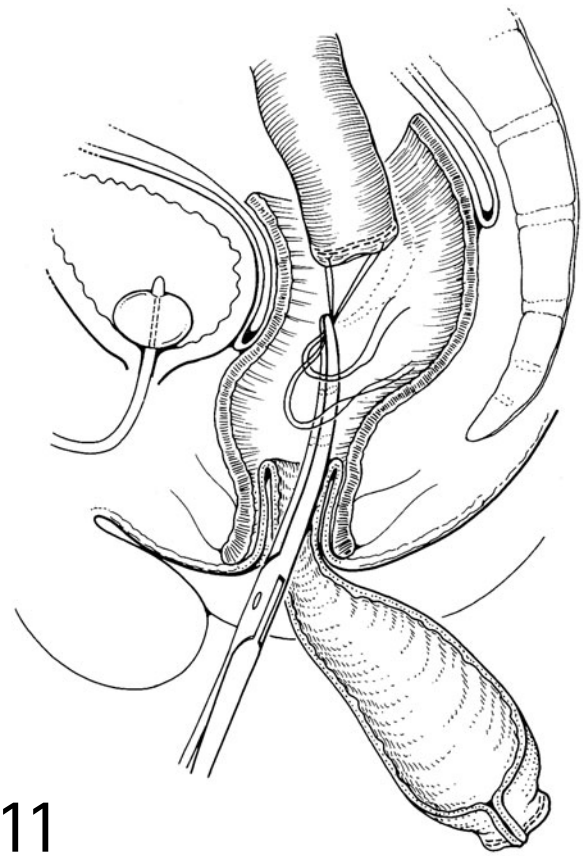
FORMATION OF RESERVOIR

The following description of the pull-through is applicable to all different types of ileal reconstructions (straight and reservoir procedures). Construction of the J-pouch, the lateral side-to-side ileal reservoir, and the S-pouch is subsequently described.

10 Once the endorectal dissection is completed, one of the surgeons moves to the foot of the table. Narrow retractors are placed at the anal mucocutaneous junction and a clamp is inserted into the rectum. An assistant working in the abdominal field places the end of the mucosal/submucosal tube into this clamp. The segment is then everted outside the perineum. The end of the everted tube is placed in a clamp and held on traction by an assistant to facilitate the anastomosis.

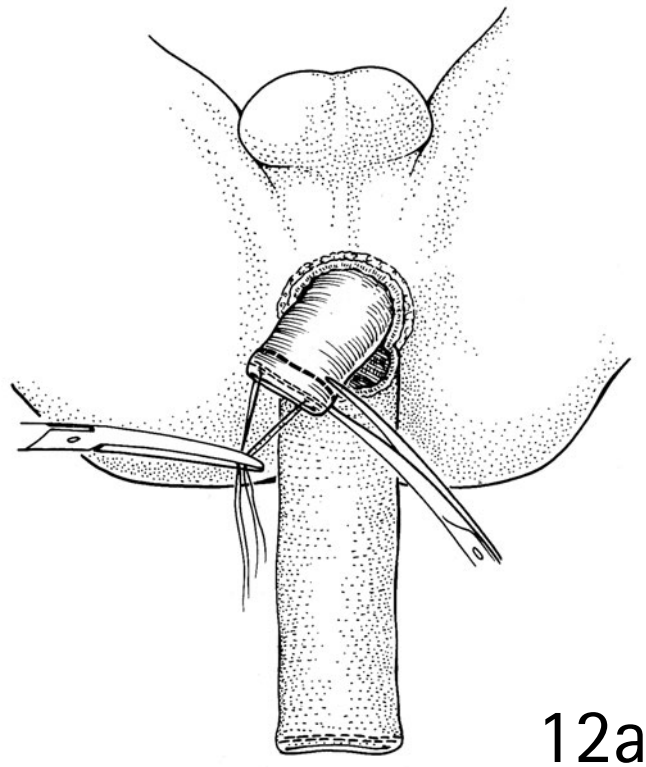


10

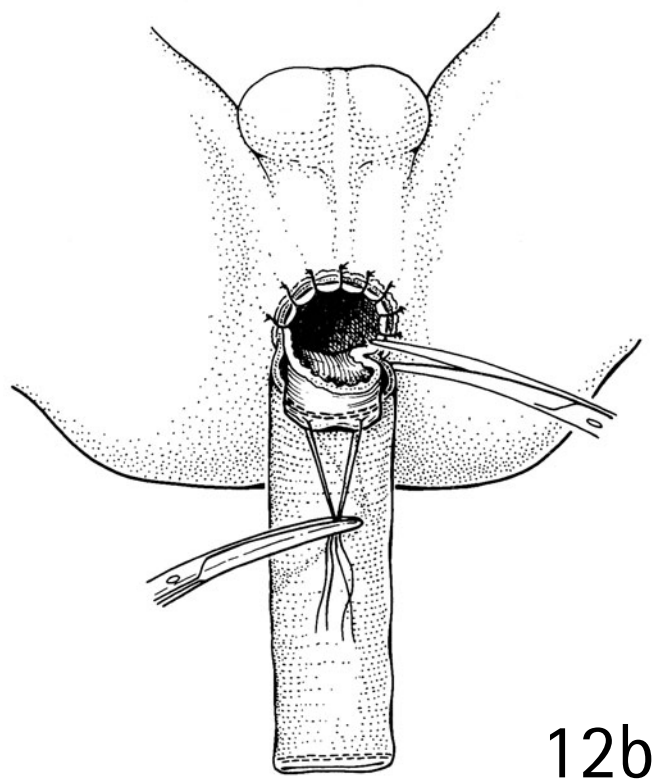


11

11 The submucosal/mucosal tube is incised on the anterior half, 1 cm proximal to the dentate line. A Kelly clamp is inserted into this opening and the ileum is brought down to this point by grasping two previously placed traction sutures. Great care must be taken not to twist the bowel as it is brought through the muscular cuff. Different colored sutures on mesenteric and antimesenteric sides of the bowel are helpful in maintaining this orientation.



12a

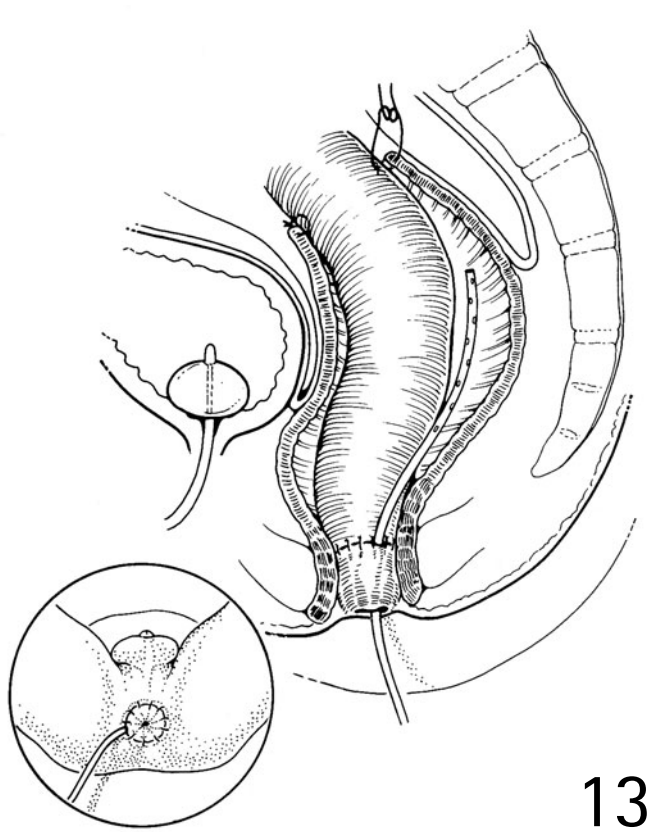


12b

12a,b The anterior half of the ileum is incised and is anastomosed to the anterior half of the anus with interrupted 3/0 or 4/0 absorbable sutures. The first sutures are placed at each corner and in the midline, and are followed by interrupted sutures placed in between.

13 One-quarter of the remaining ileum and the everted mucosal/submucosal tube are opened. A suture is placed in the posterior midline and this quarter of the anastomosis is completed. Countertraction, applied by an assistant on the everted tube and on the traction sutures on the ileum, will help with the exposure. The final quarter of the ileum and tube are removed and the anastomosis is completed and inspected. In many cases, bleeding is modest and no drainage is required. In those cases where a drain is chosen, it should be inserted before the last one or two sutures are placed using a thin, 3.5 mm closed suction drain between the rectal muscular cuff and the pulled-through ileum. The drain exits between the anastomotic sutures. It is placed most safely by advancing a small uterine sound retrograde through the anastomosis into the endorectal dissection. The drain can be secured onto this sound and then pulled through the anastomosis.

The ileum is pulled slightly upward, and this will invert the neorectum back into its correct position. Rectal examination at this point should show a well-formed anastomosis 1.5–2 cm above the anodermal junction. Gloves are then changed and attention is directed to the abdominal field. The pulled-through ileum is attached with seromuscular sutures to the muscular cuff to prevent it from prolapsing in the early postoperative period.

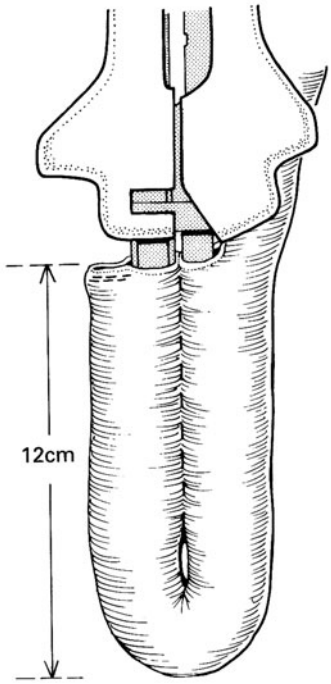


13

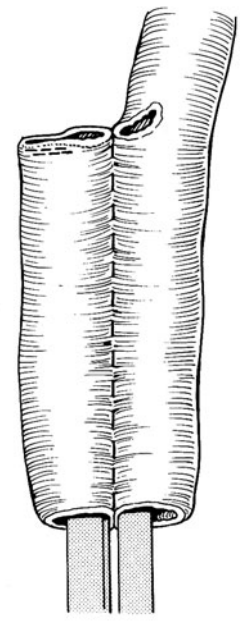
14a–c For the J-pouch, the ileum is folded back on itself for a length of 8–12 cm. The bowel is opened at the stapled end and at an adjacent position in the proximal ileum. An automatic stapling device is fired to create the anastomosis. A small section of ileum will remain separated at the apex of the J. The apex is opened and the stapler is fired in the opposite direction to complete the anastomosis. This open end is brought down for the anal anastomosis. It is critical that a septum is not left between the ileal limbs, as this will cause stasis, bacterial overgrowth, and a fecaloma.

Although the J-pouch is the most popular of all the reservoirs, some surgeons use other pouches, such as the lateral ileal reservoir, the S-pouch, and the W-pouch. The W-pouch is rarely used in adults or children and is not discussed here.

Increasingly, we have made the length of the J-pouch shorter (8–10 cm), in the hope of decreasing the incidence of pouchitis. In those patients where an 8 cm long pouch is created, a single firing (or at times two firings) of the stapler from the bottom of the pouch may be all that is required (b). This step eliminates the need (a) of opening the staple line; however, care must be taken to ensure the entire length of the J-pouch is opened between the two lumens of bowel.



14a



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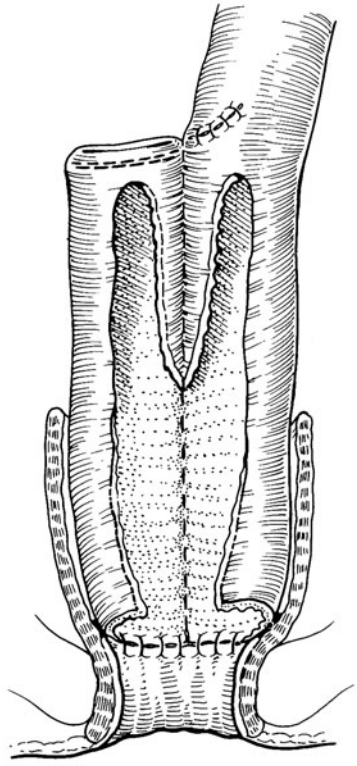


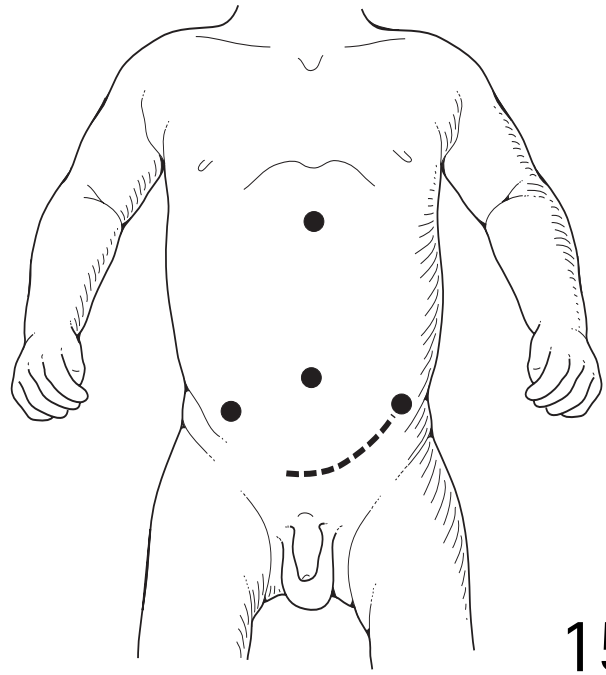
14c

LAPAROSCOPIC ASSISTANCE

The use of laparoscopy has been increasingly popularized in recent years. Two approaches have been advocated: complete laparoscopic proctocolectomy and endorectal pull-through, or a laparoscopic-assisted approach. The authors advocate the latter approach, as the total use of laparoscopy has been associated with a significant increase in operative time and blood loss.

15 The approach to the laparoscopic proctocolectomy consists of the placement of an initial umbilical port, followed by four 5 mm Innerdyne ports (Salt Lake City, UT), placed as shown here. The umbilical port initially contains the camera with a 30° telescope.

This is followed by an epigastric port and then left and right lateral ports. The colon, from the terminal ileum to the distal rectum, is mobilized and released from the peritoneal attachments and the splenic and hepatic flexures. Initial mobilization of the lateral attachments is facilitated by 'airplaning' the patient to the contralateral side, with traction on the colon, with a blunt bowel grasper through either the epigastric or the contralateral trocar sites, and using cautery scissors via the remaining trocar site. The ureters are identified early during this dissection. Depending on the site of mobilization, the camera and operating ports will vary.



15

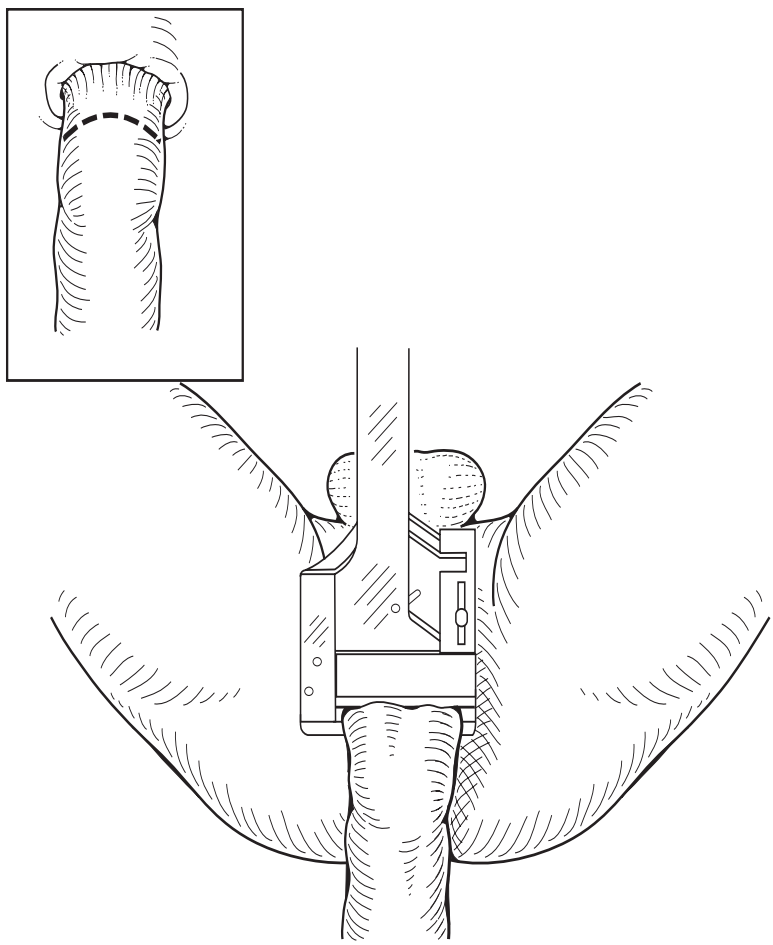
The more time-consuming aspect of the dissection is the mobilization of the omentum off the transverse colon. A pair of 5 mm ultrasonic scissors or the use of an endo-gastrointestinal anastomosis (endo-GIA) stapling device will help at this point in the dissection. Care is taken to identify and avoid the middle colic vessel. Alternatively, the omentum may be spared by retracting it superiorly and using electrocautery dissection between the stomach and colon.

Once the colon is fully mobilized, a low transverse suprapubic incision is made, predominantly on the left side of the midline (see dotted line in Illustration 15). The operating surgeon pulls the entire colon out through this incision and sequentially ligates the mesenteric vessels. The ileum is divided, and an optional ileal pouch may be created. The endorectal dissection is then performed.

ALTERNATIVE STAPLED ANASTOMOSIS

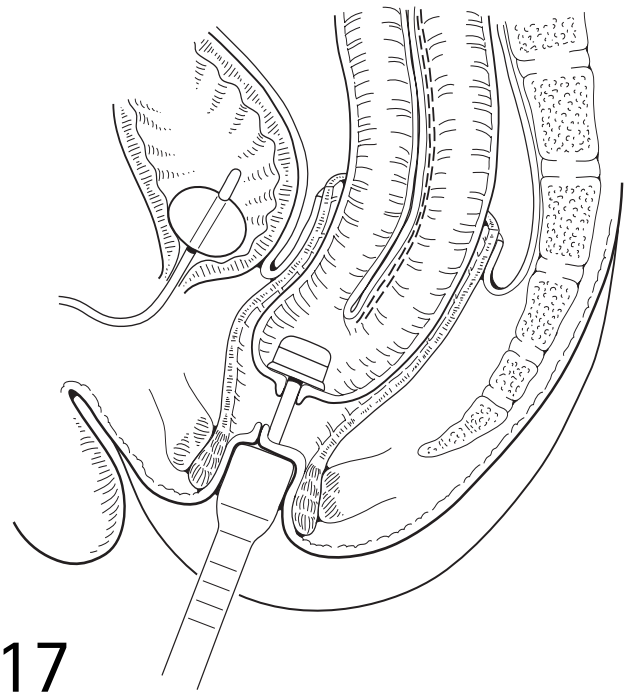
One of the major restrictions in performing a J-pouch pull-through is the difficulty in bringing down the end of the pouch sufficiently out of the anal canal to perform a hand-sewn anastomosis. Strategies of placing the patient in reverse Trendelenberg and extensive dissection of the mesenteric vessels may help; however, in some cases this may not be sufficient. A conventional stapled anastomosis has the limitation of leaving an excessive amount of rectum. The great advantage of the modification shown here is that the anastomosis of the pouch is performed within the anal canal, taking a tremendous amount of tension off the anastomosis.

16 The endorectal dissection is carried out in an identical fashion to the open technique and the mucosal/submucosal cuff is prolapsed out onto the perineum. At this point, a stapling device is used to staple and transect the bowel approximately 1 cm above the dentate line. In obese patients with a very deep gluteal cleft, an endo-GIA-type stapling device allows better positioning of the stapler.



16

17,18 The residual anorectum is placed back into the anal canal and the largest possible circular stapler is inserted. The assisting surgeon from the abdominal field places the anvil into an opening in the end of the J-pouch. Care is taken to ensure that this opening is away from mesenteric vessels, which may be injured during the subsequent anastomosis. A proline suture is placed in purse-string fashion and tied to secure the anvil. The abdominal surgeon then helps guide the J-pouch down toward the pelvis. At the same time, the surgeon on the perineal side turns the stapling device to fully advance the trocar through the mucosal/submucosal tube into the peritoneal cavity. Great care must be taken to ensure correct alignment of the two sides. Once this is done, the stapling device is fired and removed. Both tissue donuts are inspected and, in some patients, a sigmoidoscopy with air insufflation is done to assess the integrity of the completed anastomosis.



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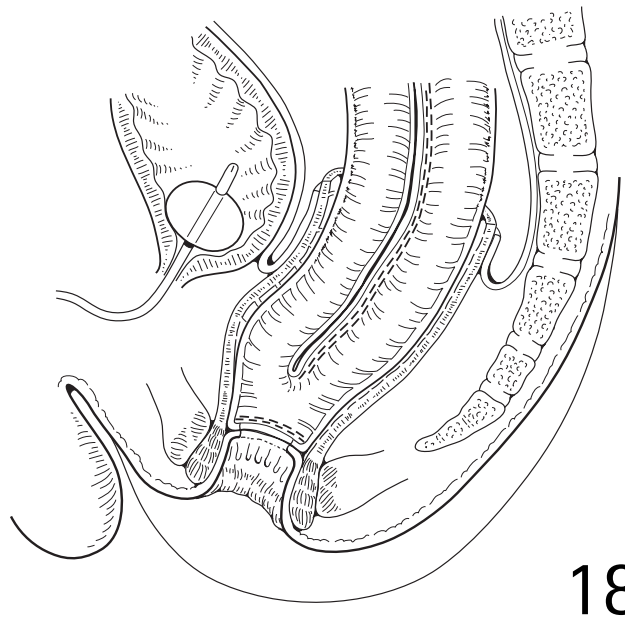


18

ADDITIONAL CONSIDERATIONS

The addition of a laparoscopic dissection will add time to the procedure. Thus the procedure may not always be desirable. Because of this increased time, care in patient positioning, as with all patients, is critical. In some cases, the authors elect to prepare the abdomen, buttocks, and entire lower extremities, with the legs placed in well-padded stockinettes. The legs are left supine and not placed in the stirrups until the ileoanal anastomosis is performed. The decision to perform a protective ileostomy is highly debatable. The advantage of eliminating the ileostomy is the ability to forego a subsequent surgery and the potential complications associated with an ostomy. The authors advocate performance of an ileostomy because of the extreme morbidity associated with a potential leak from the ileoanal anastomosis.

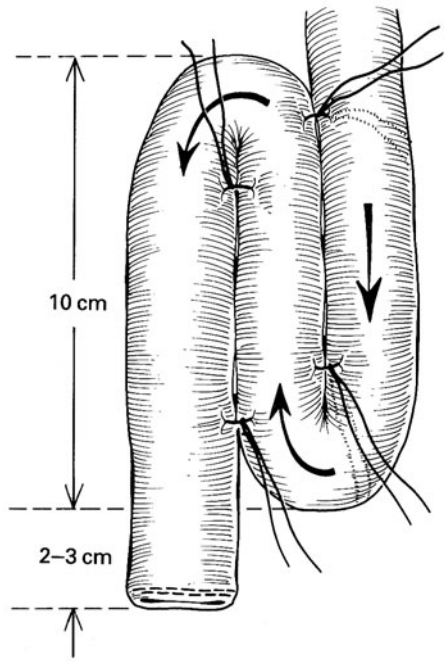
Using the stapled technique will save tremendous time in the operating room, but will almost inevitably result in a narrowing of the anastomosis, which will require two to three dilatations to alleviate.

S-pouch reservoir

19a–c The S-pouch is created in a manner very similar to the J-pouch, except that there are two *overlapping* limbs of ileum. Each limb is 10 cm long with a 2 cm spout, which is used for the ileoanal anastomosis.

For all types of pull-through operations, a loop ileostomy is placed proximally. Care is taken to place this in an appropriate location marked before the operation. The ileum is carefully sutured to the peritoneum to prevent prolapse and twisting of it at the site of the ileostomy.

An advantage of an S-pouch is that the end of the spout can easily reach the outside of the perineum.



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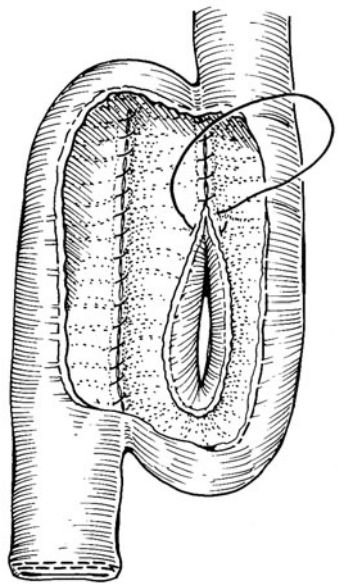

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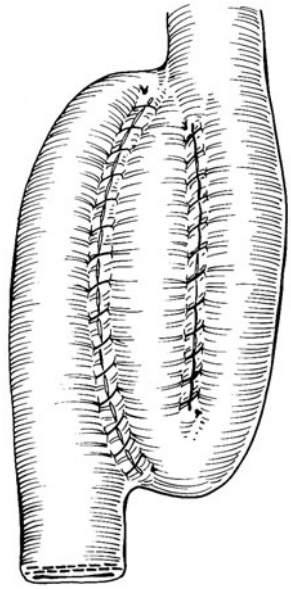
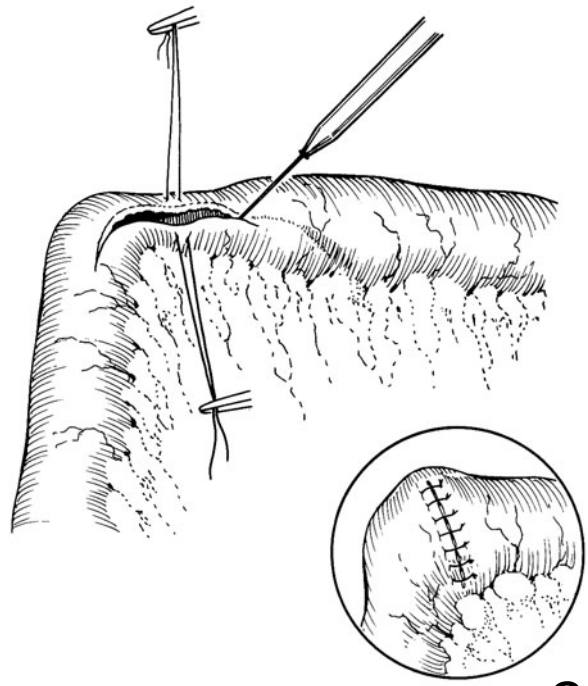


19c

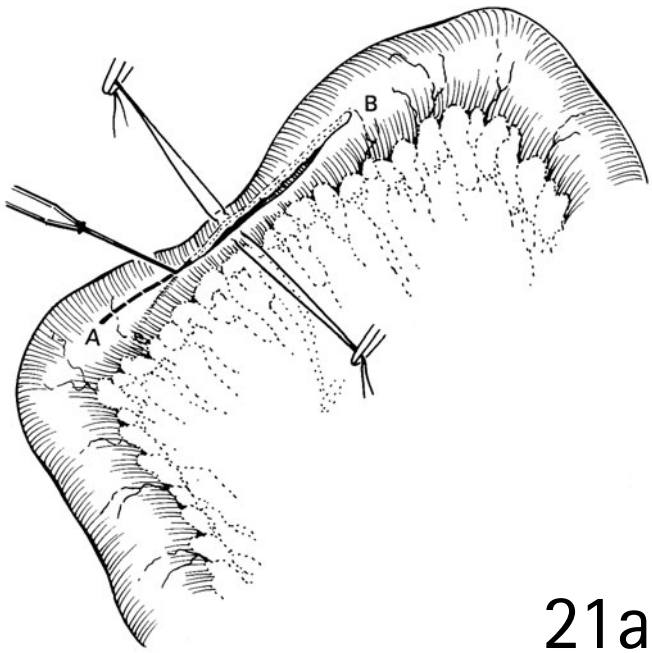
Stricturoplasty

20 The entire small and large bowel is inspected. Each stricture is identified and marked with a silk suture. A typical short-segment stricture is managed by placing traction sutures above and below the stricture on the antimesenteric surface of the bowel.



20

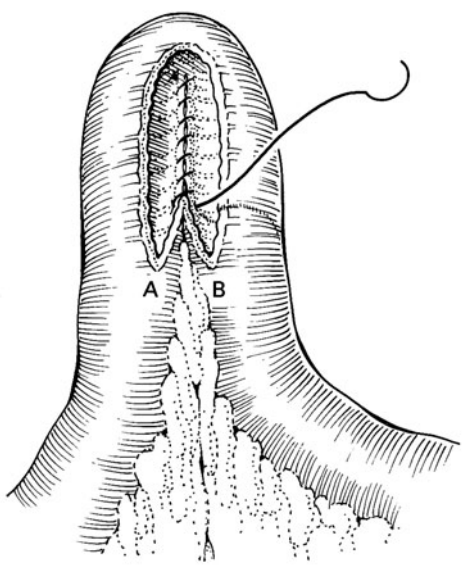
Using cautery, the bowel is opened longitudinally along the antimesenteric surface. It is then approximated transversely using interrupted 4/0 absorbable or non-absorbable sutures, and a second layer of Lembert sutures is placed.



21a

21a For strictures that are longer, the bowel is folded upon itself so that the stricture is at the apex and opened along a portion of both proximal and distal limbs.

21b,c The limbs are anastomosed using an inner layer of running 4/0 absorbable sutures and interrupted Lembert sutures on the outside.



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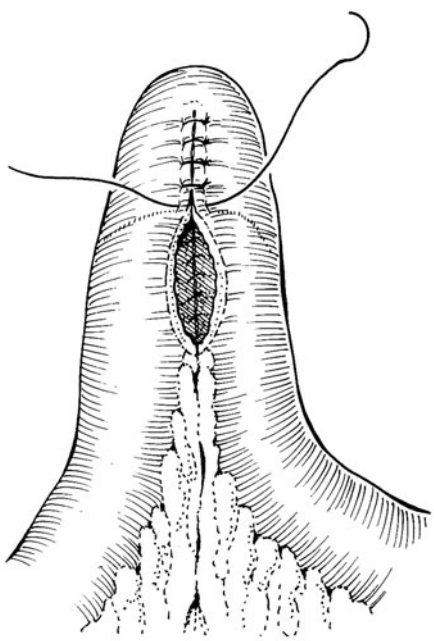

21c

POSTOPERATIVE CARE

Following proctocolectomy or pull-through, a nasogastric tube is maintained overnight and the child is not fed until gastrointestinal activity returns. The Foley catheter can usually be removed in 3 days, but occasionally is left longer if the pelvic dissection was difficult. An additional dose of steroids is given on the day of surgery, and then slowly tapered down over the next few weeks. Routine stoma care must be thoroughly learned by the family before discharge. The ileostomy is generally closed after 2 months.

Postoperative care after stricturoplasty is routine. Nasogastric decompression is used overnight, and feedings are started once gastrointestinal activity returns. The child is given supplemental steroid therapy in the perioperative period. If the child has been malnourished for a prolonged period of time, consideration should be given to perioperative parenteral nutrition.

COMPLICATIONS

Complications associated with the endorectal pull-through are not uncommon. Diarrhea, with 7–15 bowel movements a day initially after closure of the ileostomy, is expected and this should be explained to the child and family before the operation. Most patients slowly normalize their stooling pattern over the first few months after the pull-through. Major losses of fluids and electrolytes may occur at first, as well as excoriation of the perineum. Diarrhea is best controlled with loperamide hydrochloride as needed. Occasionally, Metamucil may be added. Cholestyramine is occasionally added to this regimen.

Pouchitis is a well-described complication of an ileoanal pull-through. The presumed etiology is stasis of stool in the ileal segment, with subsequent bacterial overgrowth. The incidence is around 25–50 percent if reservoirs are employed, but with a straight pull-through the incidence is lower (< 5 percent). Treatment of this condition consists of serial washouts of the pouch, sitz baths and oral metronidazole.

The incidence of adhesive obstruction after any type of surgery for ulcerative colitis is 20–25 percent, irrespective of the type of procedure done. In the authors' series, however, adhesive obstruction occurred in 10 percent of cases, with only half of these requiring an enterolysis.

Acknowledgments

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Rectal polyps

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PRINCIPLES AND JUSTIFICATION

Although the incidence of juvenile polyps is unknown, they are believed to occur in approximately 1 percent of all preschool children. Most appear in the first decade of life, with the peak incidence between 3 and 5 years. Polyps are solitary in 50 percent of cases, with the remainder having two to ten polyps. Forty percent of juvenile polyps are found in the rectum or sigmoid colon. The remaining 60 percent are found evenly distributed throughout the colon.

Juvenile polyps are also known as retention, inflammatory, or cystic polyps. Such polyps are generally considered hamartomas or a malformation in which normal colonic tissue has become arranged in a haphazard manner. Grossly, typical polyps have a glistening, smooth, spherical, reddish head and range from 2 mm to several centimeters in diameter. Polyps often have an ulcerated surface, which accounts for the rectal bleeding. A cross-section shows cystic spaces filled with mucus. Juvenile polyps are typically attached by a long, narrow stalk covered by colonic mucosa. This stalk predisposes the polyp to torsion, which results in venous congestion, surface ulceration, bleeding, and autoamputation.

Clinical features

Recent data suggest that juvenile polyps result from a structural rearrangement of the mucosa secondary to an inflammatory process. The initial event is probably ulceration and subsequent inflammation of the mucosa, leading to obstruction of regional, small colonic glands of the mucosa. The obstructed glands enlarge with mucous secretion and push up into the lumen. The fecal stream and peristalsis push the mass down the lumen, causing the stalk to elongate, resulting in the typical pedunculated appearance of the juvenile polyp. Ulceration of the surface or autoamputation leads to the bright red blood noted on presentation. Occasionally polyps prolapse through the anal canal and present as dark, cherry red protrusions at the anus. Although many of the polyps are within reach of a digital rectal examination, they may not be easy to feel due to their mobility. Clinical symptoms of intermittent bright red blood warrant an investigation.

The diagnosis and treatment of juvenile polyps require a combination of history, digital rectal examination, and colonoscopy. The historical shift of juvenile polyps to the more proximal colon and the concern for the presence of juvenile polyposis (more than five juvenile polyps), with its increased risk of malignancy, mandates that the entire colon be surveyed. Polyps in the rectum can be removed easily during anoscopy. More proximal surveillance needs to be done by pancolonoscopy.

Differential diagnosis

The differential diagnosis of juvenile polyps encompasses all of the causes of rectal bleeding in toddlers and children up to the age of 6 years. Anal fissures and rectal prolapse cause rectal bleeding but are easily distinguished from polyps on physical examination. Bleeding from a Meckel's diverticulum or duplication of the intestine usually causes more substantial blood loss than that from a polyp, and the blood usually commingles with the stool rather than coating it. Bleeding from an intussusception is accompanied by abdominal pain that is substantially worse than that seen with polyps. Inflammatory bowel disease is usually accompanied by diarrhea, which is not seen with polyps. Blood dyscrasias, such as Henoch–Schönlein purpura, should also be considered in the differential diagnosis. Rectal polyps occur in up to 15 percent of children with Peutz–Jeghers syndrome, and this condition should be considered.

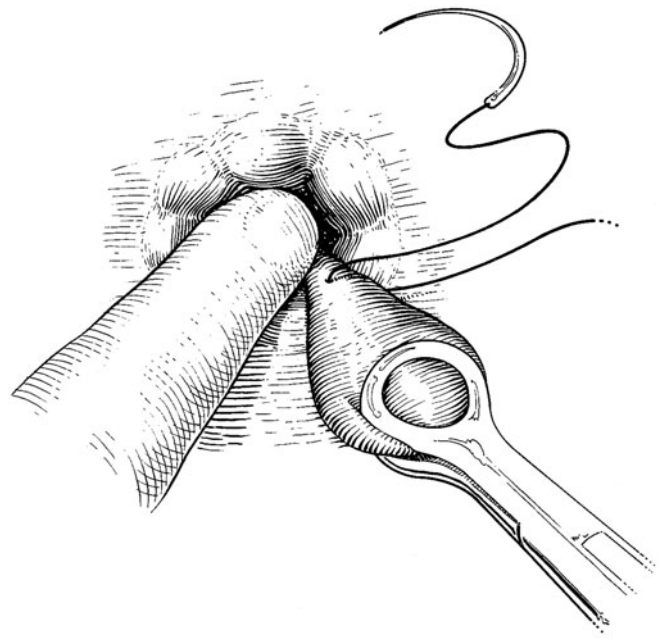
OPERATION

Anoscopy with polyp removal and pancolonoscopy can be performed as an outpatient procedure. Standard bowel preparation for a colonoscopy is performed by the patient's

parents at home prior to surgery. Suitable premedication is given and general anesthesia is induced. The child is turned into the left lateral position with the sacrum at the edge of the operating table. The anus and perineum are inspected and digital rectal examination is performed. Polyps in the anorectal area can usually be removed at this point if they are found at the anal verge (see Illustration 1).

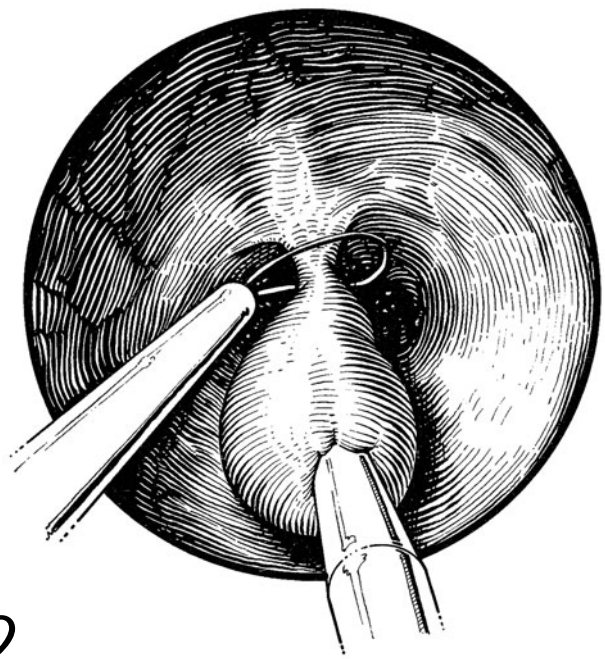
If a polyp is not identified on digital examination, an appropriately sized anoscope is lubricated and inserted. Since complete bowel preparation and cleansing have been performed, the presence of formed stool is unlikely. If stool is present, removal can be accomplished with a moistened cotton swab. When a polyp is identified deeper in the rectum, a chromic or Vicryl Endoloop is placed around the base of the stalk and cinched down. A second Endoloop is placed at a distance distally on the stalk, tightened down, and the stalk cut. This allows for accurate placement of sutures and easy removal of the polyp once cut (see Illustration 2).

After all polyps have been removed within the area of the anoscope or if no polyps are found in the most distant rectum, pancolonoscopy is performed. All polyps are removed endoscopically by the standard snare technique. The location of each polyp is recorded.



1

1 A polyp in the lower rectum or at the anal canal verge can often be brought out through the anus by digital examination. The stalk is then ligated by an absorbable suture and then cut. Often the stalk is torsed and the blood supply thrombosed. These polyps are occasionally avulsed during the maneuver to bring them out of the anal canal. Avulsion may cause minor bleeding, which will usually stop spontaneously or by applying pressure with a cotton-tipped applicator. Persistent bleeding can usually be controlled by the application of electrocautery.



2

2 Polyps further up in the rectum can be visualized using an anoscope or nasal speculum placed in the anal canal. An absorbable Endoloop snare is looped over the polyp and tightened down at the mucosal base. A second Endoloop is placed distally on the stalk and the stalk is then transected with scissors, electrocautery, or a scalpel. The polyp should be recovered for histologic examination, and this is easily accomplished by holding on to the distal Endoloop when the stalk is transected.

POSTOPERATIVE CARE

Most children can return home as soon as they have recovered from anesthesia. Parents should be warned that there may be some blood in the stool for a few days. The surgeon should follow-up with the parents, verifying the type of polyp found on histologic examination. The parents should also be counseled to return should new bleeding occur.

Complications

Perforation and hemorrhage can occur, but they are extremely rare complications that are usually self-limiting.

OUTCOME

Most children with juvenile polyps will not have a recurrence of the problem. Pancolonoscopy should be performed, however, for any child with recurrent bleeding. In children with two to four polyps, juvenile polyposis syndrome, with its increased risk of malignancy, should be considered. These children should undergo pancolonoscopy again a year after their initial polyp removal.

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Anal fissure and anal fistula

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ANAL FISSURE

HISTORY

The first description of the treatment of anal fissure was by Boyer (1825) in his *Traité des Maladies Chirurgicales*, and included the use of dorsal sphincterotomy without distinguishing the internal sphincter. Récamier described the use of anal dilatation in 1829. Since then, various methods have drifted in and out of vogue, including longitudinal incision of the base of the fissure (Lane, 1865), application of a sclerosant, anal dilatation and radical excision of the fissure (Gabriel, 1930), and dorsal sphincterotomy (Morgan and Thompson, 1956). Dorsal internal sphincterotomy was occasionally observed to cause fecal leakage, and the lateral internal sphincterotomy was developed by Parks (1967).

PRINCIPLES AND JUSTIFICATION

Anal fissure presents commonly to both general practitioners and surgical outpatient clinics, with peak incidence between 6 and 24 months. It is a tear to the squamous epithelial mucosa of the anal canal between the anocutaneous junction and the dentate line, and is thought to result from trauma by the passage of hard stool. Anal fissure is the commonest cause of rectal bleeding in infants. More significantly, it can cause pain on defecation, initiating a vicious cycle of pain, reluctance to defecate, development of a large and hard stool with further tears to the anal mucosa accompanied by more pain. Counseling after treatment is required to break the cycle of pain and apprehension.

Other contributing factors implicated in the formation of anal fissures are hypertonicity of the internal anal sphincter, anodermal ischemia, infection, and chronic constipation. Chronic anal fissures may indicate Crohn's disease.

The majority of children can be managed medically with dietary advice (increasing dietary fluids and fiber) and stool softeners. Unresponsive anal fissures can be treated with topical glyceryl trinitrate (GTN) paste applied to the lower anal canal. The GTN paste has been shown to reduce the resting anal pressure while increasing the anodermal blood flow. Doses of between 0.05 percent and 0.2 percent have been used in children. Headaches secondary to systemic absorption and vasodilatation have been reported in over half of patients. Future therapies whose efficacy is yet to be established include topical diltiazem (2 percent gel) and botulinum toxin injection.

Failure of the above medical treatments after approximately 2 months is an indication for surgical intervention. Anal manual dilatation and lateral anal sphincterotomy have both been shown to be effective. The former is usually preferred, reserving sphincterotomy for persistent cases.

PREOPERATIVE ASSESSMENT AND PREPARATION

Diagnosis of an anal fissure can be made by careful history and examination of the anus with gentle traction on the perianal skin to expose the lesion, most commonly found midline posteriorly. If the fissure is chronic, a sentinel tag may be visible at its base, representing epithelialized granulomatous tissue from chronic inflammation. No specialized investigations are required.

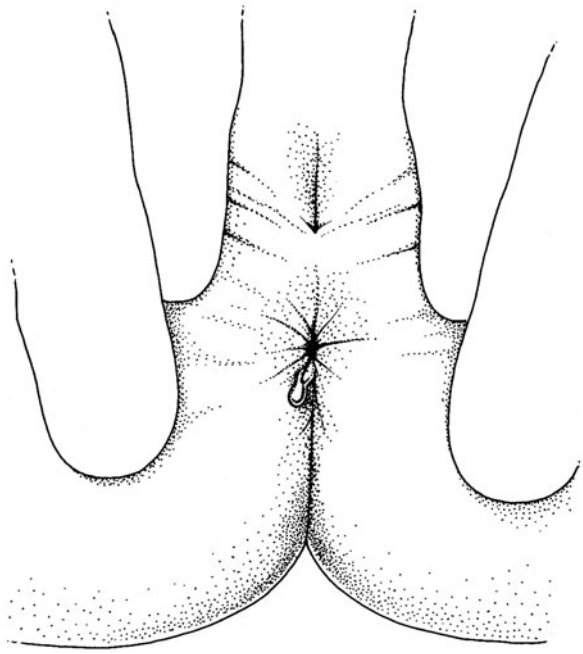
Anesthesia

General anesthesia is given. A caudal block is very useful for postoperative pain.

OPERATION

Anal dilatation

1 The patient is placed in the lithotomy position if an infant or in the left lateral position if a larger child.



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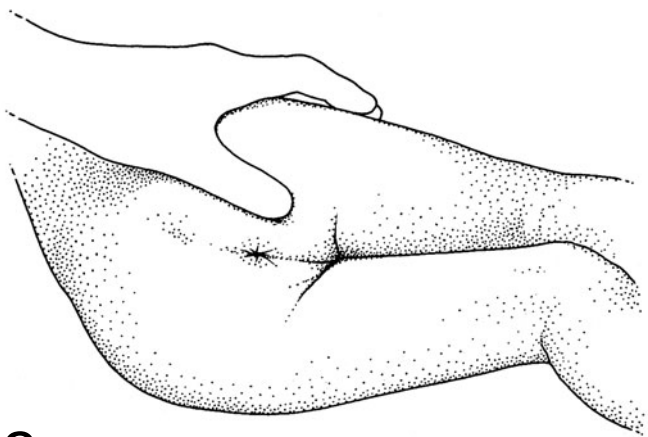
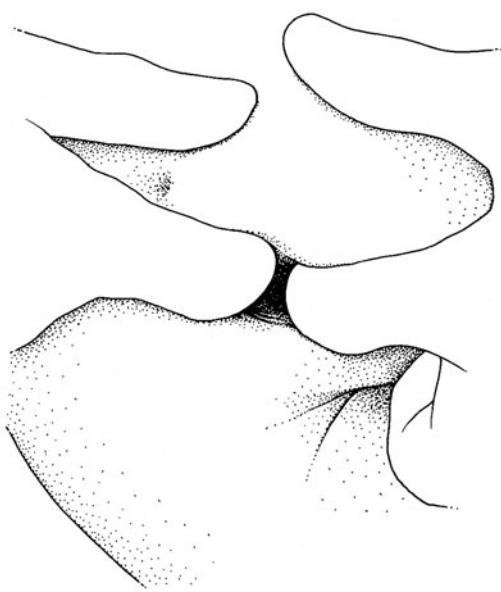


2a

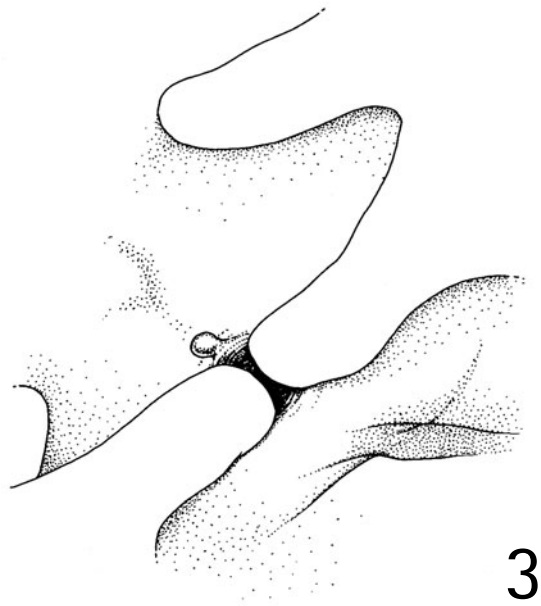
2a The anal region is inspected and a digital examination performed. If the fissure appears suspicious, a biopsy should be taken.

2b Dilatation is performed using the index fingers, with gentle traction with one digit posteriorly followed by the second digit in countertraction anteriorly.



2b

3 The hands are rotated; gentle traction is now applied laterally. If any hard feces are found, these can be manually evacuated. Following gentle dilatation, the mucosa may appear more engorged, but there should be no further mucosal tears or bruising.

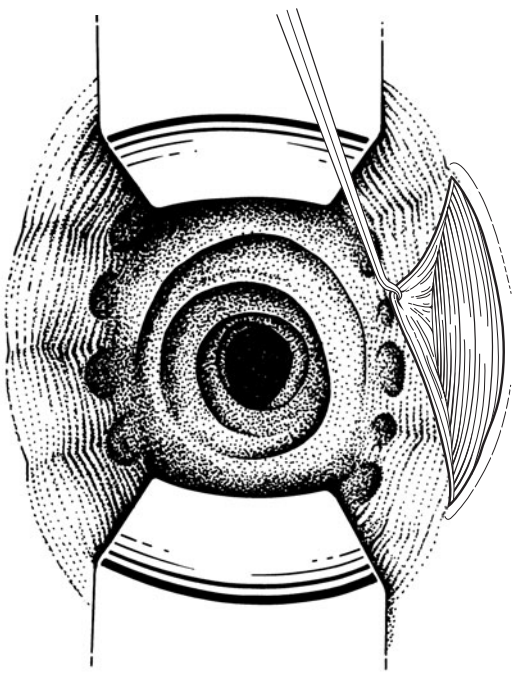


3

Lateral anal sphincterotomy

The patient is placed in the lithotomy position. An inspection of the anal region and digital examination are performed.

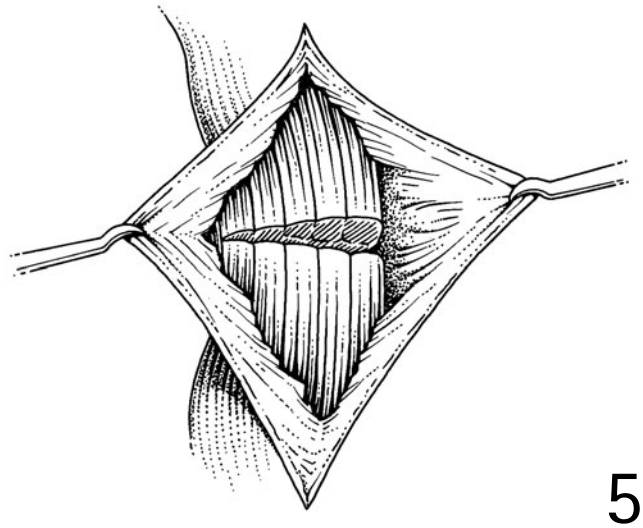
A rigid sigmoidoscopy is performed to confirm the diagnosis. A lubricated Park's retractor is then inserted and opened to expose the anal canal. The intersphincteric groove at the 3 o'clock position is palpated at the inferior border of the internal and external anal sphincters. This region is infiltrated with 1:250 000 epinephrine (adrenaline) with lidocaine (lignocaine) up to the dentate line and outside the internal sphincter.



4

4 A 2 cm curvilinear incision is made outside the anal verge, raising a flap of skin off the internal sphincter as far as the dentate line.

5 A similar flap is raised on the other side to expose the internal sphincter. The lower third of the internal sphincter is incised to the dentate line. The skin is closed. If the fissure appears suspicious, a biopsy should be taken. The sentinel tag may be removed, taking care that no damage to the external sphincter occurs.



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POSTOPERATIVE CARE

Patients should continue dietary modification and stool softeners. The child can be weaned off stool softeners once defecation is comfortable. Mild analgesia should be given as required.

OUTCOME

Complications may include some postoperative rectal bleeding and perianal pain. Local infection is rare. Incontinence has been reported in adults but not in children undergoing these procedures.

Data describing the long-term outcomes of the two surgical procedures are lacking. Anal dilatation usually provides prompt pain relief. Recurrence of symptoms has been reported in 5–30 percent of patients. More than one attempt may be required for permanent relief of symptoms. Lateral anal sphincterotomy has a reported recurrence rate of 0–10 percent.

ANAL FISTULA

HISTORY

Documentation of anal fistula dates back as far as Hippocrates (460 BC), who described treatment using a seton. Arderne (1307–1390), an English surgeon, described the application of a seton and provided the first report of laying open of the fistula in 1337. Further classification systems and theories of etiology were documented during the late nineteenth and early twentieth centuries. In 1976, Parks refined the classification system, which is still in use today.

PRINCIPLES AND JUSTIFICATION

Anal fistulae commonly present in infants as recurrent perianal abscesses, which sometimes discharge spontaneously. They are hollow tracts lined with granulation tissue, probably originating as an infected anal canal gland. Unlike in adults, the tracts usually run radially and straight from their anal canal origin to the site of the abscess. Anal fistulae occur most commonly in males and in infants.

Perianal abscesses are common in infancy and occur with equal frequency in males and females. Fluctuant abscesses should be incised and drained. Superficial non-fluctuant infections may be treated conservatively with Sitz baths. One-third of these will resolve without further treatment, but the majority will become fluctuant and require surgery. Almost 50 percent of all perianal abscesses develop a perianal fistula. A history of recurrent abscesses should alert the surgeon to possible underlying inflammatory bowel disease or chronic granulomatous disease and the child should be examined under general anesthetic.

PREOPERATIVE ASSESSMENT AND PREPARATION

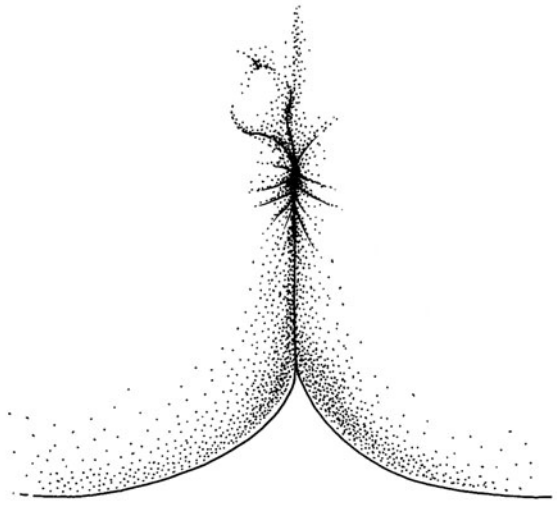
No special preoperative investigations are required.

Anesthesia

A general anesthetic is given with caudal block.

OPERATION

6 The child is placed in the lithotomy position. A careful inspection of the anal region is performed, looking for induration and signs of underlying pathology. Any suspicious lesions should be biopsied.



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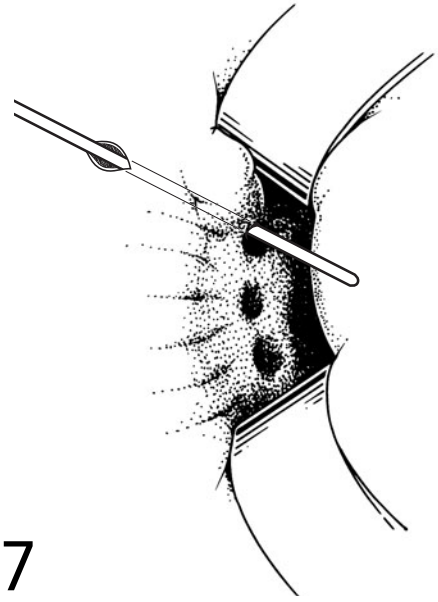
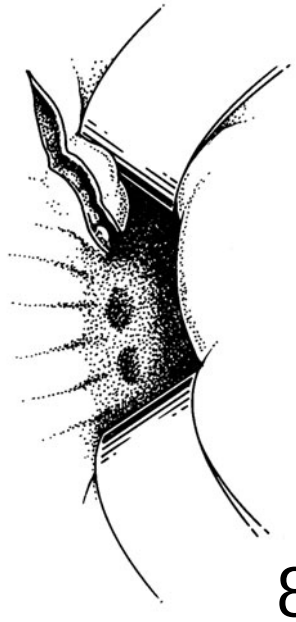


7

7 A lubricated Park's retractor is inserted to visualize the anal canal and the origin of the anal fistula. The abscess is incised and drained and a silver probe is gently inserted into the opening of the fistulous tract, taking care not to form a false tract. The opening in the anal canal is therefore visualized.

8 A knife or electrocautery is used to cut down onto the probe, laying the fistula open. If there is a lot of granulation tissue in the floor of the fistula or the abscess cavity, this may be curetted. To ensure the tract can drain freely, excess overhanging skin should be excised.



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POSTOPERATIVE CARE

Patients should be given stool softeners and dietary advice to reduce discomfort from trauma to the wound. They should also be advised to bathe carefully, using a shower jet to keep the wound clean.

OUTCOME

Wounds usually heal in 4–6 weeks. Recurrence should prompt further investigation into possible underlying disease.

FURTHER READING

Cohen A, Dehn TCB. Lateral subcutaneous sphincterotomy for treatment of anal fissure in children. *The British Journal of Surgery* 1995; **82**: 1341–2.

Festen C, van Harten H. Perianal abscess and fistula-in-ano in infants. *Journal of Pediatric Surgery* 1998; **33**(5,1): 711–13.

Nelson R. A systematic review of medical therapy for anal fissure. *Diseases of the Colon and Rectum* 2004; **47**: 422–31.

Simpson J, Lund JN, Thompson RJ, Kapila L, Scholefield JH. The use of glyceryl trinitrate (GTN) in the treatment of chronic anal fissure in children. *Medical Science Monitor* 2003; **9**(10): 123–6.

59

Colonoscopy

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PRINCIPLES AND JUSTIFICATION

Indications

Limited colonoscopy is extremely well tolerated by children of any age, pediatric colonoscopes being thinner than an examining little finger. Without either sedation or bowel preparation, it is possible to inspect, photograph, and obtain biopsy or other specimens from the rectosigmoid as part of the initial assessment of symptomatic patients. Since more extensive colonoscopy requires both full bowel preparation and some form of sedation, it is reserved for selected patients, usually those with failure to thrive or weight loss, chronic diarrhea, anemia, bleeding, and when there is radiologic abnormality (e.g., narrowed terminal ileal abnormality on small bowel follow-through) or a need for therapy (e.g., Peutz–Jeghers polyposis).

Colonoscopy, where it is readily available, now supplants barium enema as the colonic investigation of first choice in children, not only because it can be performed without irradiation, but also because high-quality double-contrast films are not usually obtained by pediatric radiologists and radiography is, therefore, less accurate, and also a tissue diagnosis of a mucosal lesion will be obtained. When indicated, it is feasible to perform both colonoscopy to the terminal ileum and gastroscopy to the duodenum at a single examination, a large proportion of the gastrointestinal tract thus being accessible to inspection, biopsy, or instrumentation in one procedure.

Contraindications

There are few contraindications to colonoscopy in sensitive hands with appropriate instrumentation. Examination is likely to be difficult and unrewarding in simple constipation or megacolon unless adequate preparative precautions are taken, and the diagnostic yield is extremely low in abdominal pain unaccompanied by features to suggest systemic illness. There is a risk of septicemia in marasmic, immunodepressed, or immunosuppressed subjects, who should receive appropriate antibiotics; prophylactic antibiotics should also be given in the presence of any cardiac lesion. The danger of septic peritonitis contraindicates colonoscopy in the presence of ascites. The availability of immersible instruments and appropriate solutions (glutaraldehyde peracetic acid 2 percent) means that full sterilization of the colonoscope is possible between examinations, preferably with an automated washing machine, and there should, therefore, be no possibility of transmission of infective agents.

PREOPERATIVE

Bowel preparation

A variety of bowel preparation regimens for children are now available that will produce a clean colon. The regimens are based either on large oral volumes (1–3 L) of balanced electrolyte polyethylene glycol or isotonic mannitol (5 percent) solutions or on aperients and laxatives such as senna and magnesium citrate/picosulphate solutions. However, many children will not drink an adequate volume of balanced electrolyte solutions. This results in either an inadequately cleansed bowel or the use of a nasogastric tube to ensure that an adequate amount is taken. The bowel preparation used successfully by the author for many years is shown in Table 59.1.

This regimen has given excellent results provided that the full dose is taken and that the child has plenty of fluids and is encouraged to use the toilet.

The following is an alternative protocol using a balanced electrolyte (Klean-Prep) solution.

- 5–10 kg body weight: first half hour 50 mL/h, then for 1 hour 100 mL/h, then at 200 mL/h until bowel is evacuated.
- 10–20 kg body weight: first half hour 100 mL/h, then for 1 hour 200 mL/h, then at 300 mL/h until bowel is evacuated.

Table 59.1 *Bowel preparation regimens prior to colonoscopy*

Age (years)	Picolax® powder (mg)	Senokot (mL)
0–1	None	10
1–2	None	20
2–5	2.5 first dose	
	2.5 second dose	30
5–8	2.5 first dose	
	2.5 second dose	40
8–12	5 first dose	
	5 second dose	50
> 12	10 first dose	
	10 second dose	60

- 20–30 kg body weight: first half hour 200 mL/h, then for 1 hour 300 mL/h, then at 500 mL/h until bowel is evacuated.
- > 30 kg body weight: first half hour 200 mL/h, then for 1 hour 400 mL/h, then at 600 mL/h until bowel is evacuated.

Any patient with constipation (e.g., some patients with cystic fibrosis/chronic constipation) should start bowel preparation 3 days before the procedure, or with Klean Prep the day before the procedure.

Sedation or anesthesia?

Premedication is useful for apprehensive children, for whom reassurance and explanation are often ineffective (trimeprazine and triclofos syrup orally, diazepam rectally, chlorpromazine or pethidine intramuscularly). In infants or older subjects, premedication should be unnecessary, assuming there is a friendly atmosphere and the parents are supportive. Infants can be managed with surprisingly little medication, even during the examination, and older children are frequently more interested and less embarrassed by the prospect of internal examination than adult patients.

Colonoscopic examination is, however, often uncomfortable or painful for a short period, as the instrument stretches the sigmoid colon mesentery or the visceral peritoneum, and adequate analgesia is therefore advisable to avoid traumatizing the child or parents. There is no contraindication to the use of light general anesthesia, but this is usually unnecessary and tends to make colonoscopy a more serious procedure, more difficult to organize, and less used; it also encourages heavy-handed instrumental technique. With appropriate intravenous medication, colonoscopy can be a routine day-case or side-room investigation. An indwelling venous cannula is inserted before the child is brought to the endoscopy room so as to avoid the trauma of venepuncture just before sedation for the endoscopic procedure. A combination of benzodiazepine (for amnesia) and opiate (for sedation and analgesia) is titrated by slow intravenous injection until the child is drowsy enough to accept introduction of the instrument through the rectum without protest. To avoid pain at the injection site, a lipid suspension of diazepam or water-soluble midazolam is preferred, and pethidine is diluted 1:5 with water. Initial dosage is based on midazolam 0.1 mg/kg and pethidine 0.5 mg/kg, but, according to results, larger amounts may be needed and 50–100 mg intravenous pethidine is not unusual without over-sedation in an anxious adolescent. Pethidine is favored for any incremental doses, since it is more effective in children and is reversible by naloxone. Flumazenil should be available to reverse benzodiazepines when required. Monitoring of heart rate and oxygen saturation by a portable oximeter during sedation and the procedure is mandatory. Appropriate use of oxygen as indicated by oxygen saturation level results in safer and more effective sedation.

Choice of instrument

In small children it is clearly preferable to have a suitable floppy 10–11-mm diameter instrument so as to pass small sphincters and variable colonic loops without undue stretching. In older children, an adult colonoscope may be more appropriate. The length of the instrument is not usually a limiting factor, pediatric colonoscopes being at least 130 cm long, whereas the cecum of a baby may be reached using only 50 cm of instrument and shortened back to 25–30 cm as the colon straightens. As colonoscopes are more flexible than gastroscopes, if a pediatric colonoscope is not available, an adult colonoscope is preferable to a pediatric gastroscope. With lubrication and slow dilatation, the anus of even a baby will accept an instrument of 14–15-mm diameter.

Is radiographic control needed?

Most examinations do not require radiographic screening control and the majority of colonoscopists never use it. In the learning phase and for the less experienced, however, the extra information given can be invaluable. If radiographic facilities are available, difficult procedures can be made quicker, safer, and less traumatic. Radiography will also help in the localization of biopsy sites or lesions found unexpectedly at colonoscopy. Irradiation should be kept to a minimum, an occasional brief image being sufficient to demonstrate the position of the instrument and to explain and resolve any looping of its shaft. The best compromise is to avoid the use of radiography in the majority of patients, but to have it available in case of need; if necessary, the patient can be transferred to an X-ray table with the colonoscope in situ.

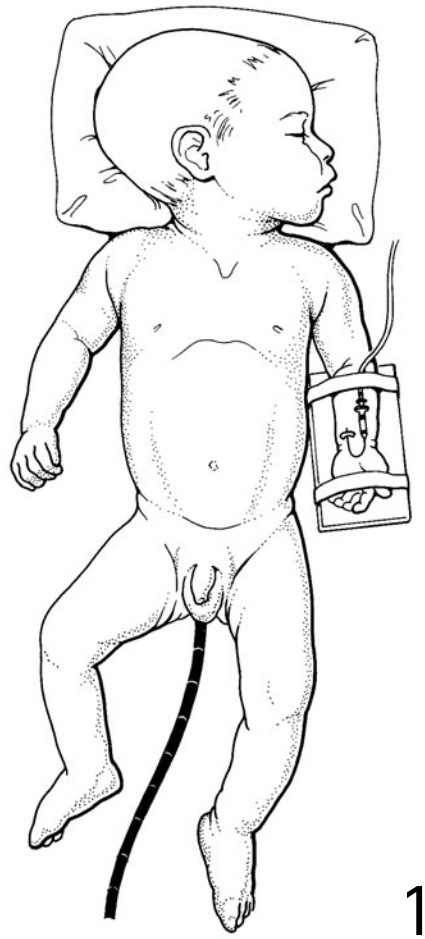
More modern developments using magnetic sensors and computer visualization of the shaft of the endoscope have rendered radiography redundant. However, as with radiography, most experienced colonoscopists do not need these aids.

OPERATIONS

Colonoscopy

POSITION OF PATIENT

1 Infants are usually examined supine and this position is also appropriate if general anesthesia is used; otherwise, most endoscopists commence with the patient in the left lateral position, and it is often possible to complete the examination without a change. If there are mechanical difficulties at any stage of the procedure, a change in position may alter the configuration of the bowel and facilitate examination. Changing to the right lateral position will make the splenic flexure less acute and can also help to drain fluid from the descending colon and facilitate air distension within it if the view is poor. In addition, the prone position sometimes aids passage through the hepatic flexure.



1

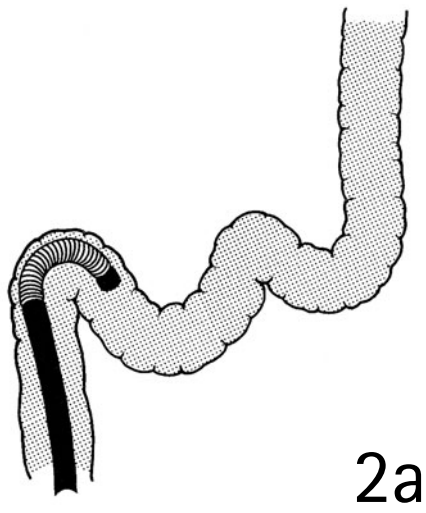
INSERTION AND PASSAGE THROUGH THE RECTOSIGMOID

The tip of the colonoscope and the perianal region are lubricated with jelly, the anus being dilated if necessary with either the finger or tubes of increasing diameter until examination is possible. On insertion, initially there may be no view because the tip is against the wall of the rectum. The instrument must be withdrawn slightly and air insufflated before a view is obtained, the tip then being angled and the instrument shaft rotated as necessary to follow along the lumen of the rectosigmoid.

In passing the many bends of the rectosigmoid, the object is to avoid distending or stretching the bowel so as to keep it short and pass almost straight to the descending colon. This is easier to suggest than to achieve, but is made more likely by observing the points set out below.

2a–c

- As little air as possible should be insufflated to see; excess air should be aspirated from time to time.
- The bowel lumen should be followed accurately.
- If the view is lost, even for a few seconds, the control knobs must be released and the colonoscope withdrawn a short distance – the lumen will automatically reappear.
- Blind pushing should be avoided, but on acute bends this may be necessary for a few seconds providing the mucosa continues to move and the general direction is known.
- If the tip will not angle round a bend, an attempt should be made to 'corkscrew' the instrument by pulling the shaft back straight and twisting it one way or the other.
- The colonoscope should be pulled back repeatedly after passing each bend and before starting each inward push. A straight colonoscope and a shortened colon will result.
- The instrument shaft should be held in the fingertips as far as possible – gripping in a clenched fist causes clumsiness.



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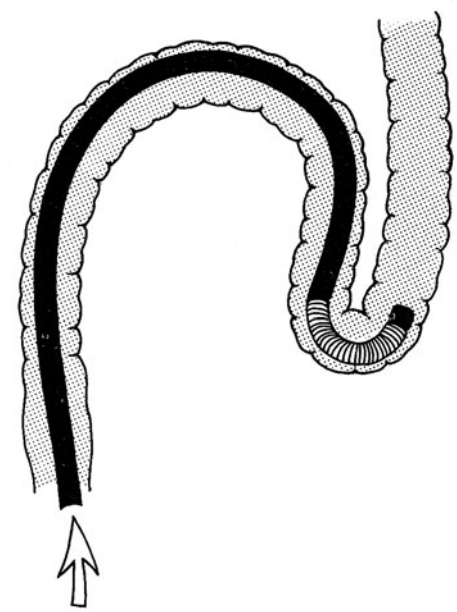

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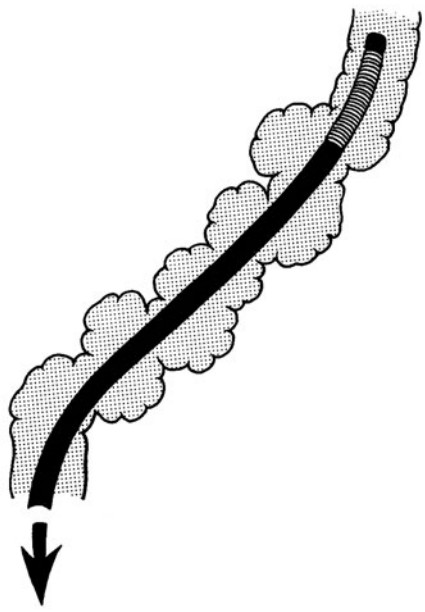


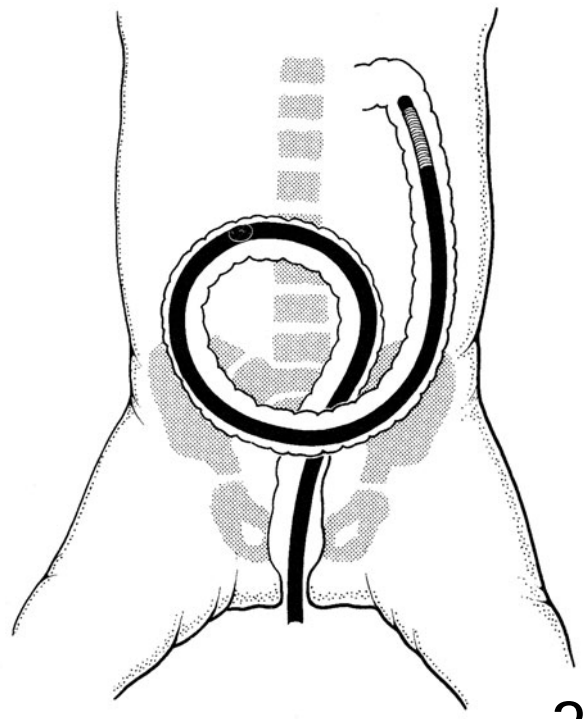
2c

SIGMOID N-LOOP: HOOK AND TWIST MANEUVER

The commonest situation on reaching the junction of the sigmoid and descending colon, in spite of all care, is for there to be an N-loop forming an acute tip angle which makes direct passage difficult or impossible. If the tip can be passed a short way around the bend, looking in to the retroperitoneal part of the descending colon, it can be held there without consciously hooking while the instrument is withdrawn 10–40 cm to reduce and straighten out the loop. Putting a clockwise twisting force or torque on to the shaft of the colonoscope while it is withdrawn will help to straighten out this loop and keep the tip in the descending colon.

SIGMOID 'ALPHA' LOOP

3 Often, if there is a redundant colon, a loop is obviously forming but the tip runs in easily without discomfort to the patient. This suggests that a spiral 'alpha' loop is forming (which can be confirmed if fluoroscopy is used). The correct thing to do is to continue pushing in as far as is comfortable for the patient, at least to the proximal descending colon and preferably to the splenic flexure. If there is little or no discomfort, the instrument can be pushed round into the transverse colon before attempting to withdraw it and straighten it out.



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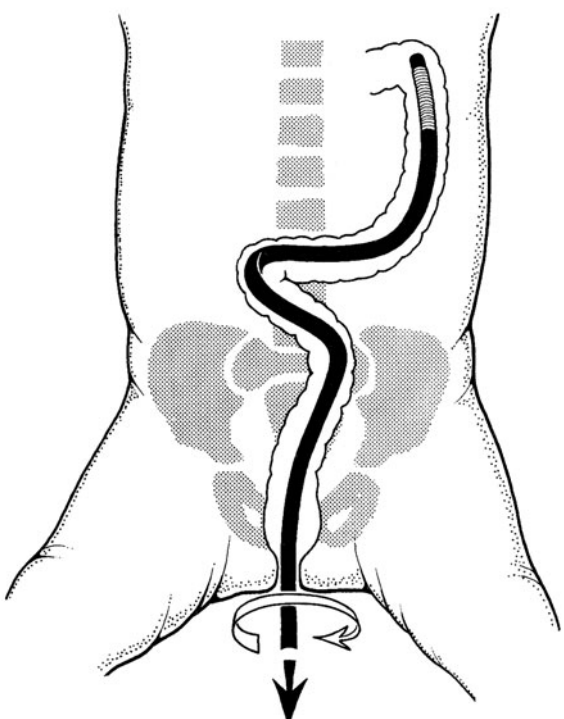


4

STRAIGHTENING OUT LOOPS

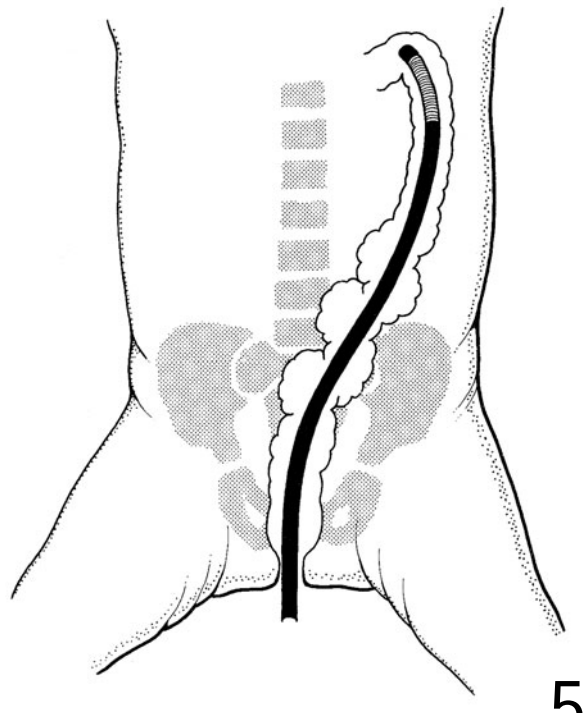
4 Having reached the upper descending or the transverse colon, the sigmoid colon loop should be removed, since loops create friction in the control wires and stress the instrument just as much as they stress the patient. To remove a loop, the instrument shaft should be withdrawn until the tip begins to slide past the mucosa or resistance to withdrawal is felt. Whilst pulling back, the application of twist, usually in a clockwise direction, will be found to stop the tip slipping back excessively and facilitate the straightening of the instrument.

In the young child, it is very likely that the colon will prove to be hypermobile, without conventional fixation of the descending colon and splenic flexure. In the 20–30 percent of patients who have a mobile colon, unpredictable and sometimes uncontrollable loops may form, which make it difficult or impossible to reach the proximal colon or terminal ileum. Such atypical loops (reversed 'alpha' loop, reversed splenic flexure) can sometimes be successfully removed by first pulling back to reduce their size and then twisting anticlockwise as the shaft is further straightened back.

SPLENIC FLEXURE: KEEPING THE SIGMOID COLON STRAIGHT

5 With the colonoscope straightened in the proximal descending colon *or* splenic flexure, some care may be needed to prevent the sigmoid loop reforming. Continued clockwise (or sometimes anticlockwise) twist on the shaft during reinsertion is often enough to keep it straight. Shaft insertion without tip movement, or losing the 1:1 relationship between shaft and tip, indicates looping. The instrument is immediately pulled back again and the assistant pushes into the left iliac fossa to resist the tendency for the sigmoid loop to rise up from the pelvis. In the splenic flexure, this tendency to re-loop in the sigmoid colon results because the hooked instrument tip impacts in the splenic flexure. A combination of the following small corrective measures will usually overcome this:

- the instrument shaft should be pulled back straight;
- hand/finger pressure should be applied by the assistant over the left iliac fossa;
- the instrument shaft should be twisted clockwise;
- if necessary, the instrument should be re-aimed toward the lumen, avoiding over-angulation;
- it should be pushed slowly inwards, continuing the clockwise twist.



5

Sometimes it is easier to reposition the child in the right lateral position to cause the splenic flexure to drop down and flatten out.

REDUNDANT TRANSVERSE COLON

The transverse colon may sometimes be pushed down by the instrument into a deep loop, which makes it difficult and painful to reach the hepatic flexure. Once again, the correct procedure is to withdraw the instrument to shorten this loop. If necessary, withdrawal may need to be repeated several times, the instrument advancing a few centimeters on each withdrawal ('paradoxical movement') until the loop is straightened. Keeping the colon deflated also helps to shorten the hepatic flexure region, making it easier both to reach and to pass. In addition, an assistant pushing the transverse colon upwards and straightening this loop out is often helpful.

Difficulty in the transverse colon is often due to recurrent looping in the sigmoid colon, and the best corrective measures are abdominal pressure in the left iliac fossa and gentle clockwise twisting during reinsertion.

PASSING THE HEPATIC FLEXURE

Having reached and deflated the hepatic flexure, and angled acutely around it into the ascending colon, the transverse loop may remain and make it difficult to pass the rest of the instrument into the ascending colon. By once again withdrawing the colonoscope and straightening out this loop it becomes easier to pass. Deflating the ascending colon by aspiration and simultaneously steering carefully to avoid haustral folds will often cause the colonoscope to descend spontaneously towards the cecum.

REACHING THE CECUM

Reaching the cecal pole can be facilitated by change of position (supine or prone), deflation, abdominal pressure, and

clockwise twist on the straightened instrument; aggressive pushing usually only results in looping. The colonoscope is seen to have reached the cecum when the bulge of the ileocecal valve is seen or, 2–5 cm beyond it, the appendix orifice is identified. Brilliant transillumination in the right iliac fossa is usually apparent at this point. The depth of insertion of the straightened instrument is variable according to the age of the patient: 70–80 cm in a teenager, down to 25 cm in a small infant. During withdrawal, the splenic flexure or descending colon is found at appropriately shorter distances. During insertion, in mobile colons and if any loops have been formed, these distance rules may not apply, but if the room is darkened, transillumination will show the position of the instrument tip.

To enter the terminal ileum it is necessary first to identify the bulge of the ileocecal valve, which may bubble or gush on deflation. The instrument tip is then pushed in just proximal to the bulge, angled in towards it, and slowly withdrawn until a 'red-out' indicates embedding into the valve region, at which point air is insufflated to attempt to distend the ileum. Ileal mucosa is characteristically granular or nodulated by lymphoid hyperplasia, in contrast to the shiny surface and vascular pattern of the colon.

In infants under 1 year of age, entry into the ileum may be impossible, either because the orifice is too narrow or because the dimensions of the cecum are too small to allow the instrument to make the necessary right-angle turn.

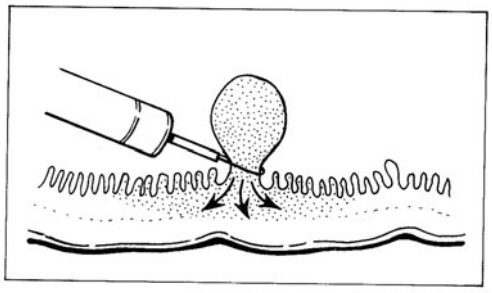
EXAMINATION

The colon is visualized to some extent during insertion of the instrument, but active examination, biopsy, or polypectomies are normally undertaken during withdrawal because the instrument is then straight and easy to maneuver, the view is better, and the patient is more comfortable. At all stages during the examination, but particularly during withdrawal, it is best for the endoscopist to control the instrument, using a one-handed technique. Very active maneuvering of the controls, with rotation and to-and-fro movements of the shaft, allow a good view to be obtained of nearly all areas, although around acute bends and convoluted folds there may be some blind spots.

Colonoscopic polypectomy

The principles of colonoscopic polypectomy are identical to those for proctosigmoidoscopic polypectomy, but it is particularly important that full coagulation of polyp stalk vessels is achieved before transection, as any hemorrhage is difficult to control endoscopically. Most polyps in pediatric practice are hamartomatous, thin-stalked, and easy to coagulate. If a thick stalk (1 cm or more) is to be snared, it may be wise to inject it with epinephrine (adrenaline) (1 mL of 1:100 000 solution), using a long Teflon sclerotherapy needle before applying the polypectomy snare.

6a–e Endoscopic snare wires are characteristically thick, to guard against cutting too fast, but care should be taken not to apply excessive mechanical pressure before adequate electrocoagulation has occurred, otherwise 'cheese cutting' of an uncoagulated stalk may occur with consequent hemorrhage. A low-power coagulating current (15–25 W) is employed until local whitening or swelling of the stalk indicates adequate coagulation, at which point tight strangulation should result in severance of the head. If bleeding does occur, the stalk remnant can be quickly re-grasped with the loop and strangulated for 15 minutes, after which bleeding will not normally recur. The correct position of the snare is shown in a. Care must be taken to avoid contact of the polyp surface with the opposite wall leading to burns from dissipation of the current (b). Burns may also result if the active electrode or metal components of the colonoscope tip are in contact with the local tissue (c and d) or if the electrode is in contact with a pool of fluid (e).



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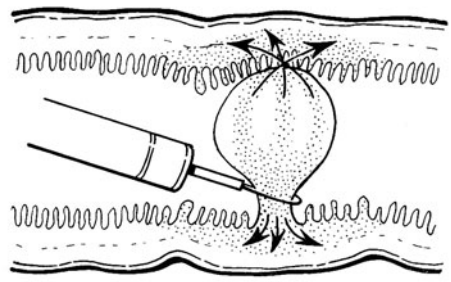

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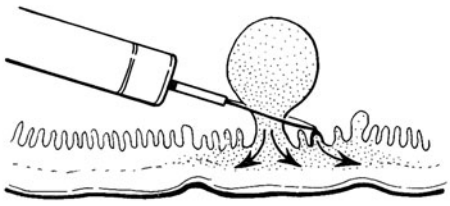

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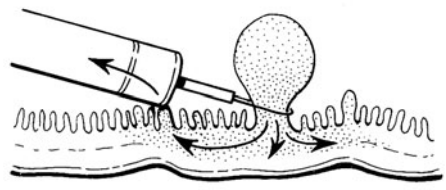

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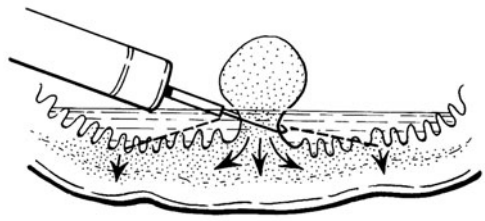

6e

Small polyps up to 6–7 mm can be destroyed using plastic, insulated 'hot biopsy' forceps, which simultaneously obtain a small biopsy specimen. The smallest hamartomatous polyps (1–3-mm diameter) can be numerous and frequently disappear spontaneously, so that it may be safer to ignore them. Retrieval of the larger polyps can be achieved by grasping them with the polypectomy snare or by aspirating them on to the tip of the instrument, which risks missing any other polyps during withdrawal unless the instrument is reinserted. Small polyps may be retrieved by aspirating them through the suction channel into a bronchial mucus trap placed in the suction line. Large numbers of polyps in patients with polyposis can be washed out after suction by passing the colonoscope proximal to them and infusing 500 mL saline into the colon through the suction channel, followed by a phosphate enema or stimulant suppository after the instrument has been withdrawn.

Other therapeutic maneuvers

Electrocoagulation of telangiectases or cavernous hemangiomas (blue rubber-bleb nevus syndrome) is easy through the colonoscope. The use of laser photocoagulation for this purpose has been described but is probably unnecessary, since careful local electrocoagulation with hot biopsy forceps or judicious scleropathy of raised lesions, repeated as necessary, gives excellent results. Strictures, particularly anastomotic strictures after resection of Crohn's disease, can be successfully dilated with transendoscopic balloon dilators. The colonoscope can be used to introduce guidewires, tubes, and other devices to any point in the colon, although this is rarely indicated in pediatric practice. The use of submucosally injected Indian ink can be useful as a long-lasting marker. Surface irrigation with colorant (1:4 dilution of washable blue fountain pen ink is convenient) can be helpful in demonstrating the smallest lesions in conditions such as familial adenomatous polyposis.

POSTOPERATIVE CARE

In most cases, no special care is needed after colonoscopy, apart from a short period of rest until the after-effects of sedation or anesthesia wear off. Food and drink can be taken immediately. When the patient appears and feels well, normal activities can be resumed, many examinations being performed on a day-case basis.

Follow-up is probably unnecessary after colonoscopic polypectomy if only one to three juvenile polyps are present in the colon; larger numbers may suggest the possibility of juvenile polyposis, which mandates follow-up because of the association with dysplastic foci. For subjects with Peutz–Jeghers polyposis, colonoscopy is normally repeated every 2 years, often combined with gastroscopy as 'top and tail' endoscopy.

Complications

In normal children, the elasticity of the colon means that the theoretical risk of bowel perforation during insertion of the instrument has not been observed in pediatric practice (in contradistinction to adult colonoscopy). The presence of severe acute inflammatory bowel disease with peritonism or deep ulceration, however, contraindicates examination because of the increased possibility of perforation; if unexpectedly severe ulceration is seen during colonoscopy, it is wise to terminate the procedure as early as possible and to avoid excessive air insufflation. Even in a normal colon, if the procedure proves technically difficult, common sense and humanity may nonetheless recommend abandonment of an examination; the percentage of failures to reach the cecum varies from 5 percent to 50 percent of all colonoscopies according to the skill and motivation of the examiner and whether or not there has been previous intra-abdominal surgery or sepsis. Neonatal examination is the most difficult. The highest percentage of complications occurs following therapeutic maneuvers such as snare polypectomy, when both perforation and bleeding have been reported.

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60

Rectal prolapse

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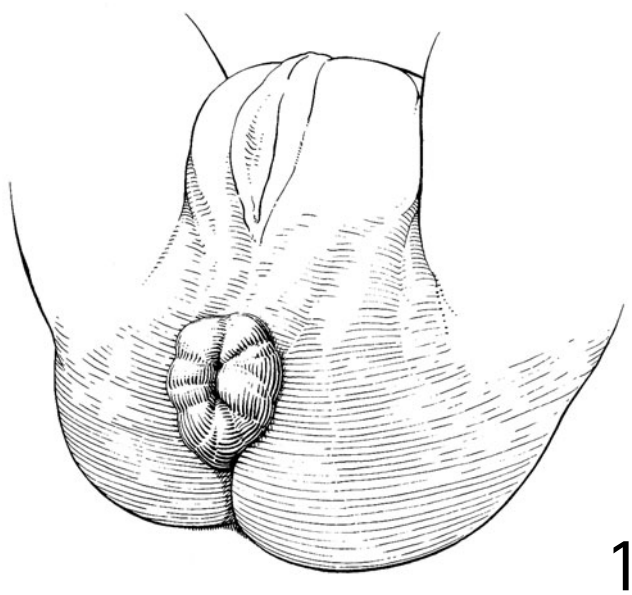
PRINCIPLES AND JUSTIFICATION

Rectal prolapse is a relatively common problem in young children, with a peak age of incidence of 1–3 years. In this age group, most cases are idiopathic and frequently self-limiting. Prolapse is also associated with tenesmus and excessive straining at stool associated with diarrhea, constipation, parasitic worms, and rectal polyps. Children with neuromuscular problems such as menigomyelocele or exstrophy of the bladder often have rectal prolapse. There is an increased incidence of rectal prolapse in children with cystic fibrosis associated with tenacious stool, chronic cough, and loss of perirectal fat.

Appearance

1 Most commonly, the prolapse is incomplete, limited to 2–3 cm of mucosa protruding from the anus and classically displaying radial folds.



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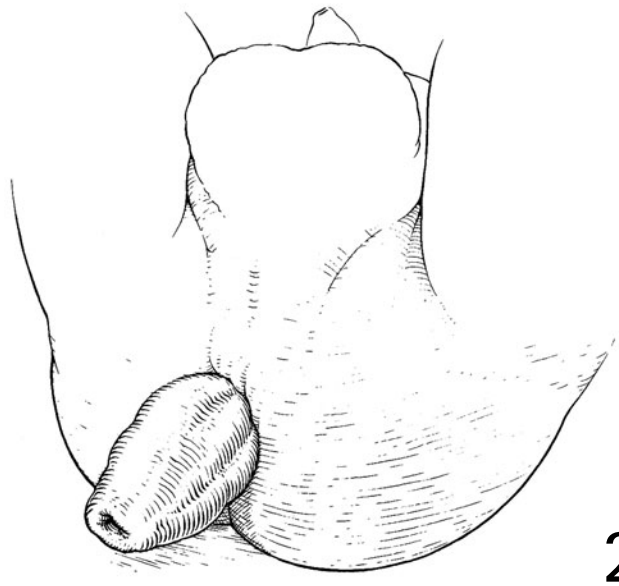
2 Complete, full-thickness rectal prolapse is more unusual. The mucosal folds in the acute complete case are circumferential. However, in both types the mucosal definition is lost with time as the mucosa becomes edematous, smooth, and featureless. The size and palpable thickness of the wall will differentiate the two types.



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Rectal prolapse may present initially as a pouting rosette of rectal mucosa typically occurring following staining at defecation. The prolapse may reduce spontaneously or require manual reduction. Prolapse may also present as mucosal bleeding.

Assessment

The assessment of children presenting with rectal prolapse should include a general history and physical examination to exclude associated etiological factors. Initial investigations should include a sweat test or gene probe to exclude cystic fibrosis. Stool analysis should also be performed.

NON-OPERATIVE MANAGEMENT

The initial management of rectal prolapse is non-operative and aims to facilitate normal stooling without excessive straining by: (1) prescribing a laxative (e.g., lactulose); (2) encouraging a high-fiber diet; (3) encouraging regular, prompt defecation from a sitting, not squatting, position.

Additional support to the perianal region during defecation has been recommended in the past; however, there is no evidence that this prevents recurrence. The parent may provide support at defecation by placing their hands under the child's buttocks, fingers just inside the ischial tuberosities beside the anus. More prolonged support can be given by strapping the buttocks together. The authors do not recommend the use of external support as a definitive treatment.

Operative management is indicated when conservative measures have repeatedly failed to prevent recurrent prolapse.

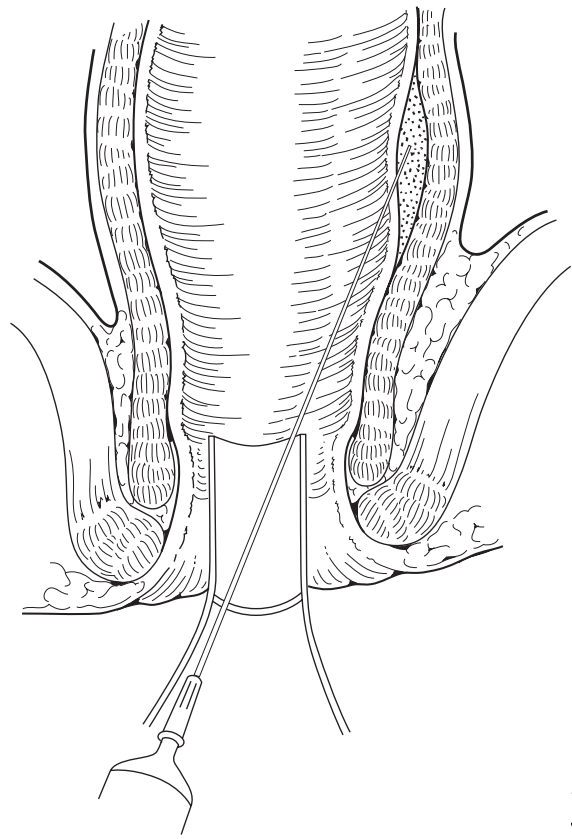
PREOPERATIVE ASSESSMENT AND PREPARATION

Under general anesthesia, an initial rectal examination and proctoscopy are performed to exclude rectal polyps. If the rectum is loaded with hard stool, this should be evacuated.

OPERATIONS

Injection of mucosal prolapse

3 A proctoscope of appropriate size is gently introduced into the lower anorectal region. A long, 23-gauge needle is placed under vision into the submucosal plane of the lower rectum approximately 4 cm from the anal verge. Then 1–2 mL of 5 percent phenol is injected into each of the four quadrants. A bulge at the injection site or blanching of the mucosa will indicate that sufficient sclerosant has been injected.



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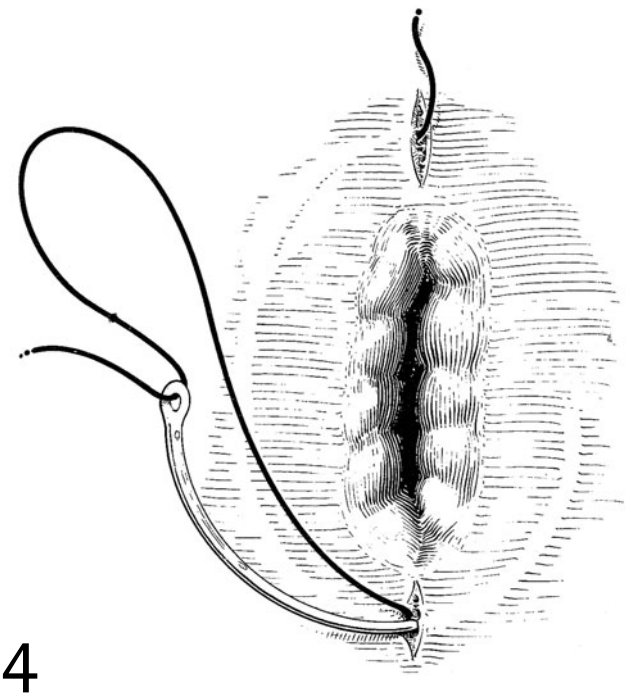

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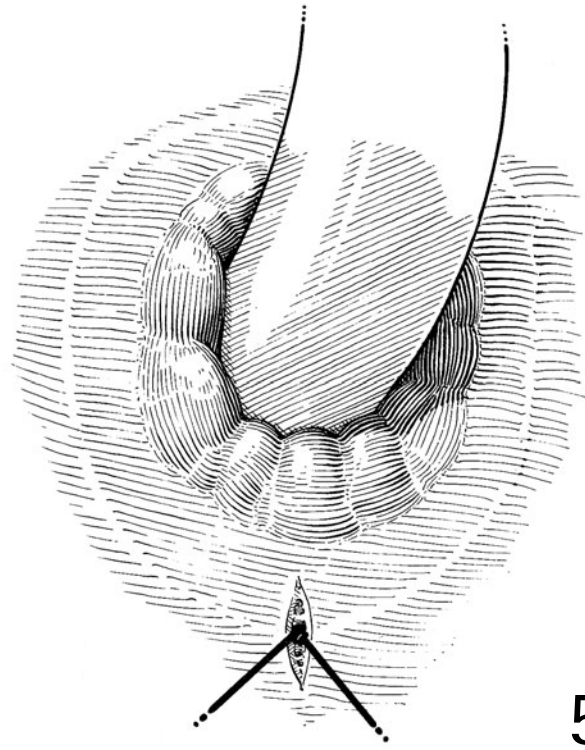
Thiersch operation (modified)

4 The child is placed in the lithotomy position and the perianal area is prepared and draped. Two small incisions are made 2 cm from the anal verge at 12 o'clock and 6 o'clock. A length of absorbable (e.g., 0 caliber polydioxanone [PDS]) suture material is threaded from the posterior incision to the anterior incision around the anus, just deep to the external sphincter muscle. The suture is continued from anterior to posterior so that eventually a ring is placed around the anus.

5 With an assistant's finger or a Hegar's dilator held inside the anal canal, the suture is pulled and tied inside the posterior incision. Absorbable sutures are used to close the two incisions. Thiersch's procedure acts by narrowing the anal orifice and thereby mechanically supporting the prolapse.



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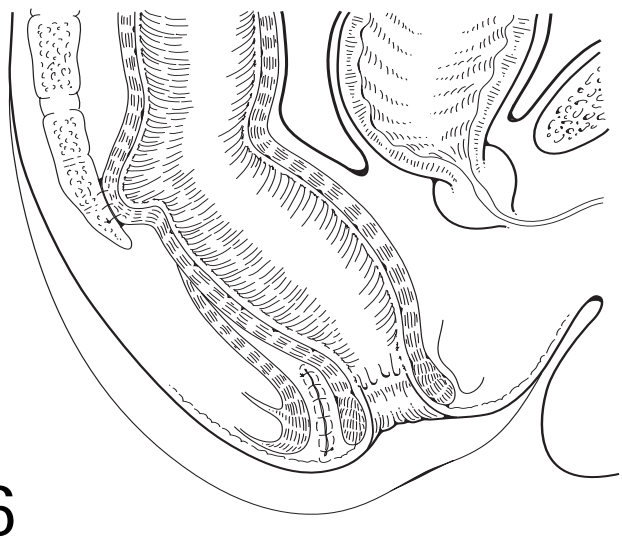

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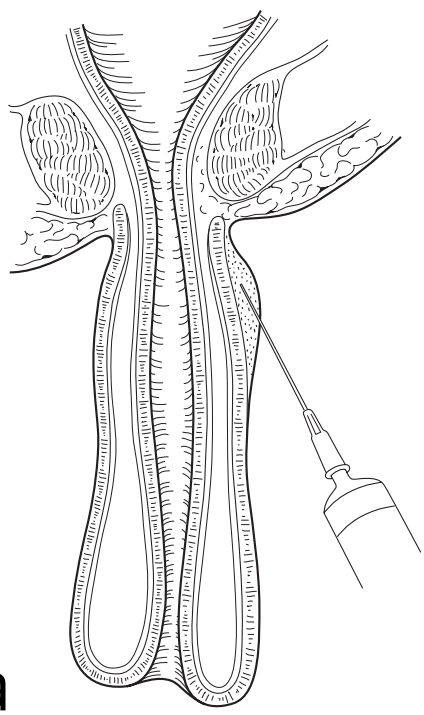
Posterior plication

6 The patient is placed in the jack-knife position. A midline skin incision is made from the coccyx and extended halfway to the anus. This incision is deepened towards the coccyx, which, if the distance from the coccyx to the anus is short and the operating field limited, may be excised. The parasagittal fibers and levator muscle are divided exactly in the midline using cautery, taking care not to incise the muscle complex. The rectum is then dissected free for two-thirds of the circumference and up to 10–15 cm vertically. Three or four permanent seromuscular sutures (3/0 or 4/0 polypropylene [Prolene]) are then placed in a longitudinal, U-shaped, mattress pattern. When these sutures are pulled together, the redundant rectum is drawn together. A further set of sutures may then be passed through the last segment of the sacrum and tied on its surface. The muscle layers are then approximated and the wound closed.

Transanal mucosal sleeve resection

7a–d The patient is placed prone in the jack-knife position. The prolapse is gently drawn out and four quadrant traction sutures are placed through the submucosa at the apex. Epinephrine solution (1:200 000) may be injected to separate the mucosal and submucosal from the muscular layers, defining the plane of dissection (a). A circumferential incision is made through the mucosal and submucosal layers approximately 1 cm proximal to the pectinate line, and blunt dissection is used to strip this layer from the underlying muscle (b). The denuded muscle layer is gradually reduced into the pelvis while the traction sutures are used to pull the mucosal sleeve in the opposite direction. When the submucosal has been separated from the entire length of the prolapse, it is divided longitudinally into two halves (c). As the sleeve is incised circumferentially, single absorbable sutures are placed to approximate the edges of the proximal and distal mucosal cuffs. Traction is maintained on the sleeve until the resection is complete; the sutures are then cut and the anastomosis retracts into the pelvis (d).



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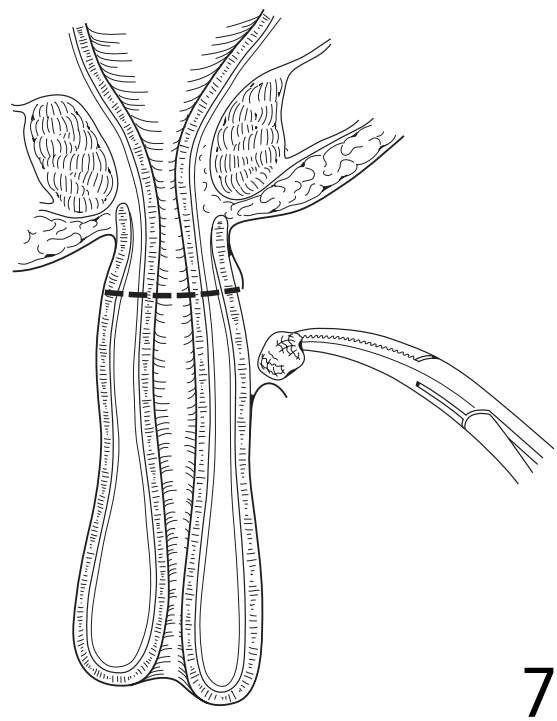

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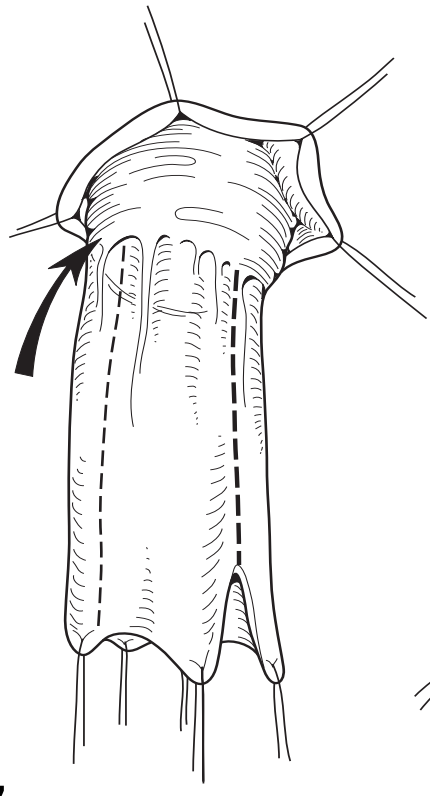

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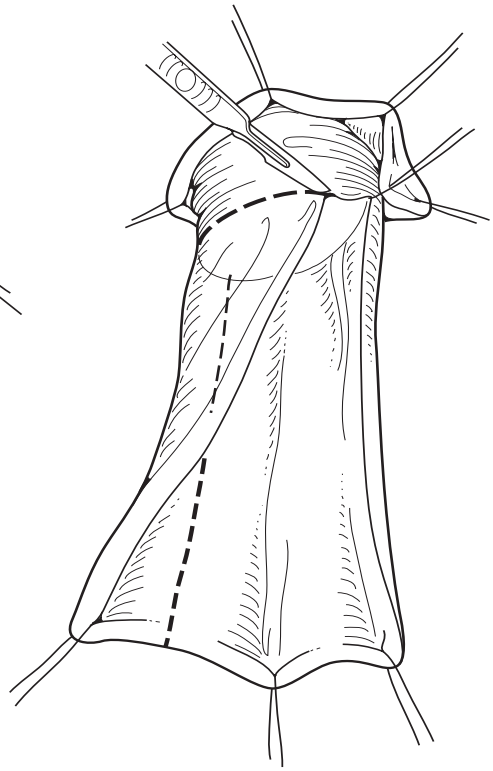

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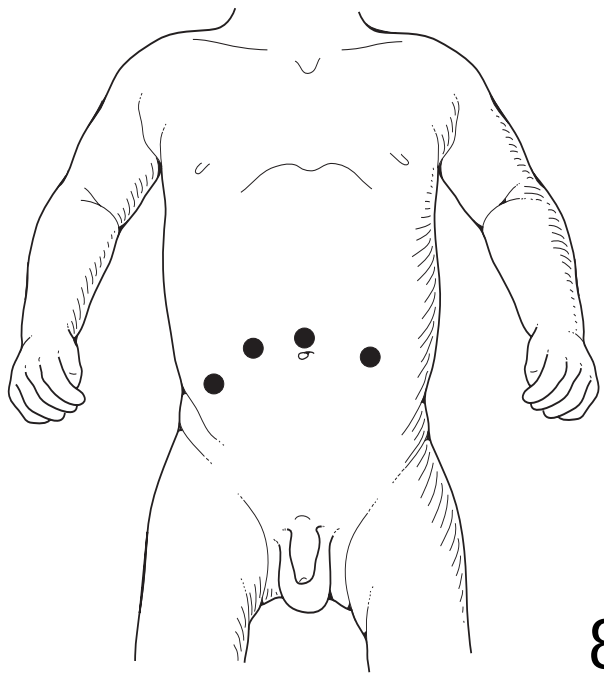
Laparoscopic abdominal rectopexy

Many open abdominal procedures previously advocated for severe recurrent rectal prolapse are now performed using laparoscopic techniques, such as the suture rectopexy and the modified sling rectopexy using a polypropylene mesh to secure the rectum.



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8 The patient is placed in the lithotomy position. A nasogastric tube and urethral catheter are placed. A pneumoperitoneum is established under direct vision by placing a Hasson cannula. A 5 mm 0° laparoscope is passed to inspect the intra-abdominal contents. Three further 5 mm trocars are then inserted under direct vision, two in the right paraumbilical region and one in the left paraumbilical region. After identification of the floppy rectosigmoid, the rectum is grasped and mobilized from the presacral fascia down to the pelvic floor. After identifying the iliac vessels and the ureter, two 3/0 non-absorbable sutures are placed bilaterally to fix the rectum in position. The trocar sites are then closed.

POSTOPERATIVE CARE

Mucosal injection and Thiersch suture can be performed as a day-case procedure. Regular bowel habit is encouraged, as in non-operative management. Stool softeners may be advocated for 3–6 months, with advice to avoid sitting on the toilet for long periods.

Complications

All of the above procedures may be associated with infection and perianal abscess formation. Usually these complications resolve fully with conservative treatment, including antibiotic therapy; occasionally incision and drainage may be required. Rarely, serious scarring and stricture formation may result, causing deformity of the rectum and leakage of mucus or fistula formation. The Thiersch suture may cause stool retention and fecal impaction if tied too tightly, in which case suture removal should be performed. Disruption of the skin wounds and exposure of knots may occur if they have not been buried sufficiently.

OUTCOME

Most patients presenting with a simple mucosal rectal prolapse respond to conservative, non-operative management. For recurrent rectal prolapse, the authors recommend the approach outlined in Figure 60.1.

Following injection of sclerosant, recurrence occurs in 10–20 percent of cases. In these cases, injection therapy may be repeated 4–6 weeks later. Many different treatments have been suggested for those persistent or severe cases that are resistant to injection therapy. Encircling procedures, abdominal rectopexies, and abdominal–perineal bowel resections and have a recurrence risk of approximately 25 percent. Posterior sagittal and transanal procedures have a higher success rate, of between 80 and 100 percent.

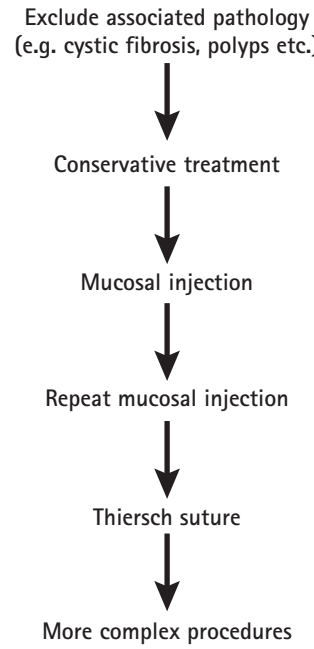

Fig. 60.1 Management of recurrent rectal prolapse.

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Colostomy: formation and closure

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HISTORY

Alexis Littre (1658–1726), the Parisian anatomist, is credited with being the first to propose a planned colostomy, or artificial anus. This occurred in 1710 during an autopsy on a child with anal atresia, when he explained how, after making an incision in the abdomen, it would be possible 'to bring the superior part of the bowel to the abdominal wound, which should never be closed and which would perform the function of the anus.' According to Scharli, it was not until 1776 that this concept was applied, when a colostomy was performed by Pillore of Rouen on an adult with rectal carcinoma. Dubois in 1783 is said to have been the first to construct a colostomy in an infant, but the patient, who had anal atresia, died after 10 days. The first long-term survivor was an infant, also with imperforate anus, who had a colostomy made by Duret in 1793 and was still alive 45 years later. During the nineteenth century, the procedure was introduced at centers across Europe and various modifications were developed, including loop colostomy over a rod by Maydl (1888). Operation 'a deux temps,' reported in 1885 by Davies-Colley of Guy's Hospital, London, consisted of suturing the bowel to the skin followed by delayed opening once the wound edges had sealed; this was an important development in an era when infection was the major cause of morbidity. The Hartmann procedure, described in 1923 for use following rectal resection for carcinoma at a time when anastomosis of the colon to the rectum was still dangerous, is still widely used in pediatric surgery.

Colostomy closure, too, was a dangerous procedure with a high risk of leakage and fecal peritonitis. This led to the introduction of techniques to minimize the risk of peritoneal contamination, such as extraperitoneal closure. With the Mikulicz technique, the common walls of a double-barrelled colostomy are crushed using an enterotome or forceps, and the resulting fecal fistula is closed later, leaving the suture line extraperitoneal if desired.

Today, colostomy formation and closure are safe operations provided basic surgical principles are adhered to, but the risk of potentially serious complications is ever present and must not be underestimated.

PRINCIPLES AND JUSTIFICATION

A colostomy is used to divert the fecal stream from the distal colon and rectum, and may be temporary or permanent. A temporary colostomy may be a primary procedure in the management of a congenital anorectal anomaly, distal obstruction as with Hirschsprung's disease, or an injury, severe inflammatory condition, or infective disease of the distal colon, rectum, or anus. It may also be a secondary procedure to protect a distal anastomosis. A permanent colostomy is required following radical excision of the anorectum for disease or debilitating fecal incontinence, for example due to anal sphincter dysfunction or colorectal dysmotility.

The following types of colostomy are in common use.

- *Divided colostomy*: this has the advantage of complete fecal diversion and a low risk of prolapse or retraction. The colon is transected and the distal end is exteriorized as a mucous fistula and either placed adjacent to the proximal stoma so that both stomas can be enclosed within the colostomy bag, or the stomas are widely separated by placing them at opposite corners of the incision so that the proximal but not the distal stoma lies within the colostomy bag in order to prevent feces entering the distal colon. Alternatively, the distal stoma is closed and placed within the abdominal cavity (Hartmann procedure) (see illustration 10).

- *Loop colostomy*: a loop of colon is exteriorized and opened, but not divided. The procedure has the advantages of being simple and quick to perform and is of particular value in seriously ill patients, but it carries a higher risk of prolapse and retraction. The extent of fecal diversion is variable, which is a disadvantage in the presence of a complete anal occlusion, when the rectum cannot be washed out, or with an anorectal fistula, where there is a risk of recurrent urinary tract infection from rectal organisms.

Siting the colostomy

An elective colostomy is usually sited either in the proximal transverse colon or in the sigmoid colon, where the colon is readily accessible and sufficiently mobile to be exteriorized without tension. The sigmoid colostomy has the advantage that there is a greater length of proximal colon available for fluid and electrolyte absorption, and the stools are thicker.

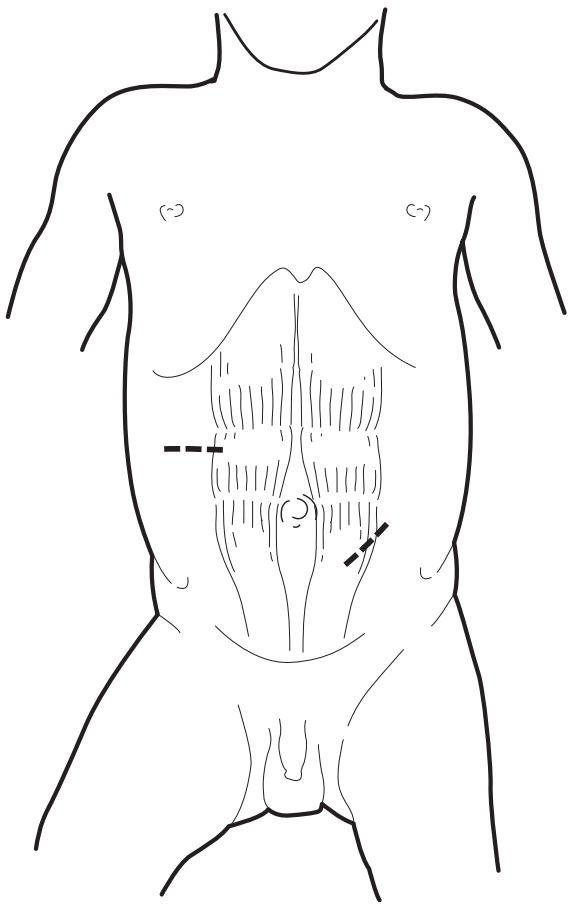
Optimal placement of the colostomy is important and will depend on the clinical situation. The colostomy bag must fit comfortably over the proximal stoma and, to avoid leakage, the surrounding skin surface must be flat, avoiding bony prominences, to ensure watertight adherence of the bag. Patients who manage their own colostomy, particularly those who are wheelchair bound, must be able to access the stoma easily; this requires preoperative planning with a stomatherapist.

PREOPERATIVE MANAGEMENT

Colostomy formation is a major operation and the patient's condition must be optimized before operation. Nasogastric drainage and intravenous fluid therapy are indicated in the presence of obstruction. Prophylactic antibiotics are given preoperatively.

Anesthesia

General anesthesia is employed with muscle relaxation.



1

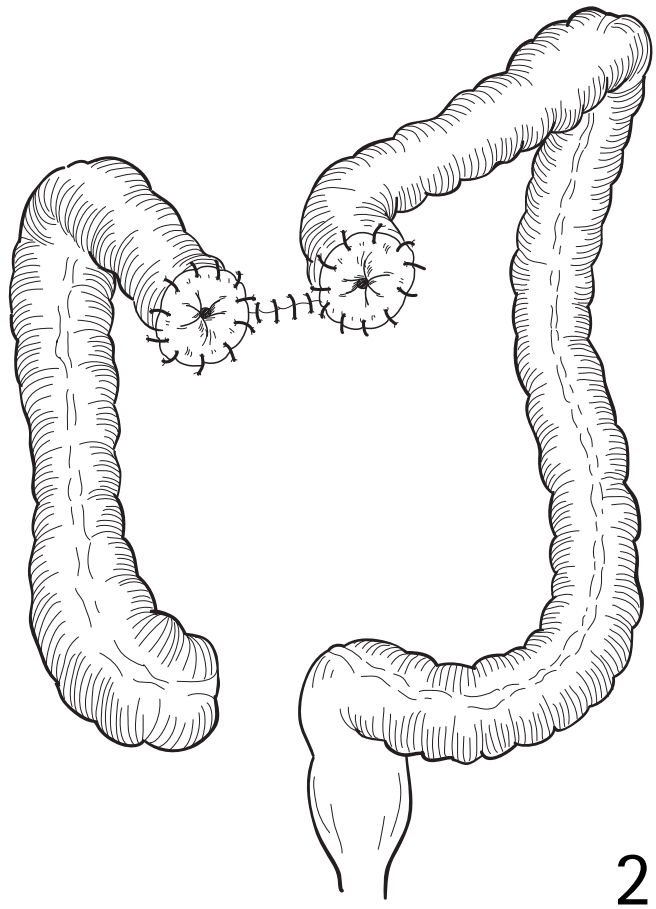
OPERATIONS

Divided colostomy

1 For a *transverse colostomy*, a right upper transverse incision is planned so that a colostomy bag can be placed on a flat surface away from the costal margin. An oblique muscle-cutting incision is used for a *sigmoid colostomy*, centered about midway between the umbilicus and the left anterior superior iliac spine; it may be extended if necessary by curving the lower end medially.

After incising the skin, the medial part of the incision includes the rectus sheath and rectus muscle; laterally the abdominal wall muscles are divided (see Chapter 32). The peritoneal cavity is entered and the colon identified and exteriorized. This may be difficult when the colon is severely distended and a rectal washout is not possible, as in the newborn infant with an anorectal malformation, in which case the colon can be decompressed at operation by direct needle puncture at the site of the proposed colostomy.

2 A transverse colostomy is placed towards the right side of the transverse colon. If the sigmoid colon is distended, it may lie in the upper abdomen and must be distinguished from the transverse colon by the presence of the fatty taenia coli, which are not found on the transverse colon.



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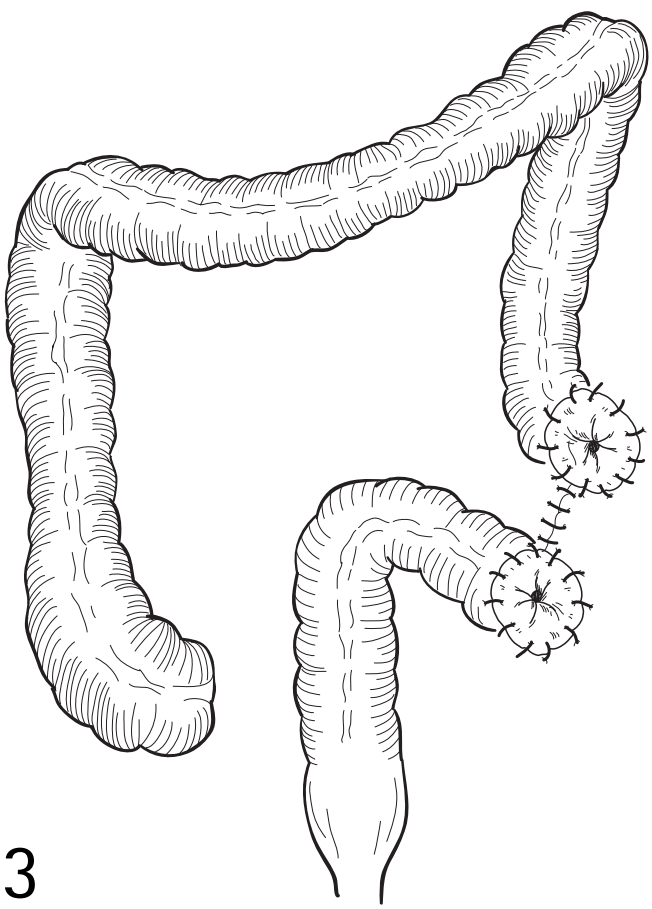
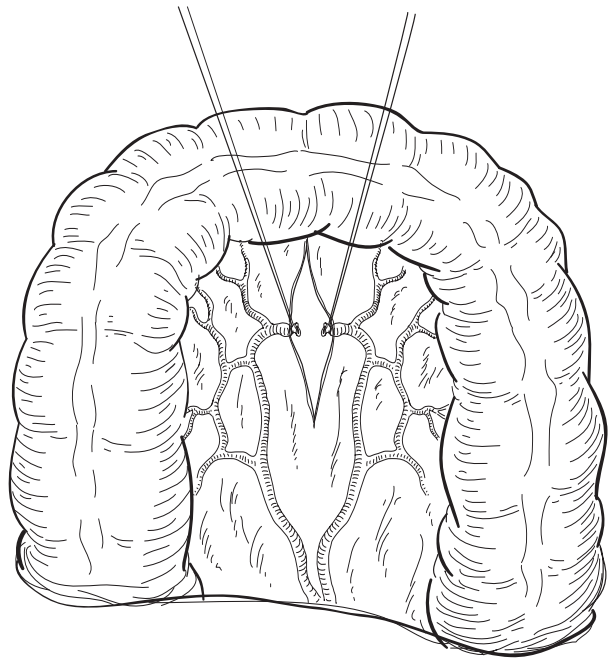


3

3 A sigmoid colostomy is usually placed near the apex of the sigmoid colon, but the underlying abnormality must be taken into consideration. For some anorectal anomalies the stoma is placed more proximally so as not to compromise the subsequent pull-through procedure, whereas for Hirschsprung's disease the stoma is sited proximal to the transitional zone as determined by intraoperative seromuscular biopsy. Note that for cloacal anomalies a transverse colostomy is recommended to allow sufficient distal colon for vaginal reconstruction.

4 At the proposed point of transecting the colon, the marginal artery is divided between ligatures and smaller branches to the colon are coagulated using bipolar diathermy.



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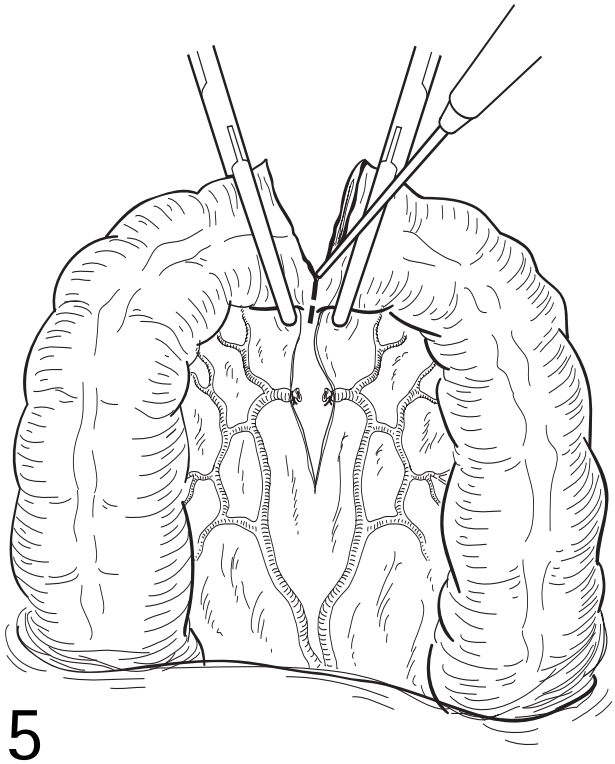
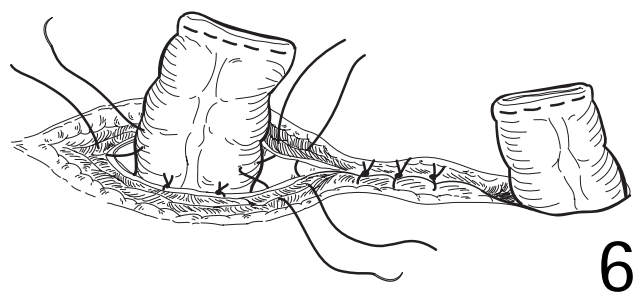


5

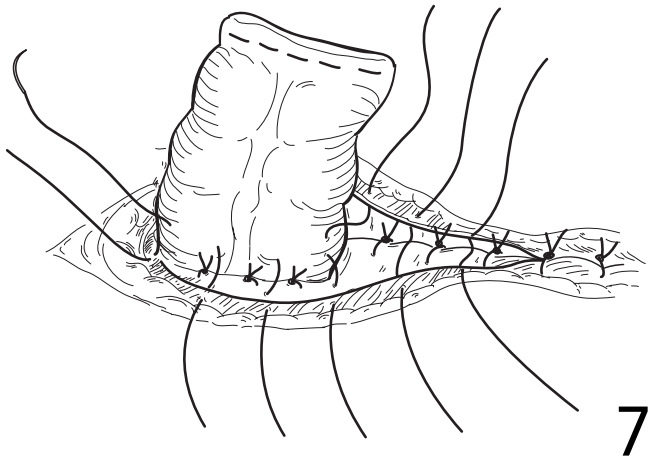
5 Artery forceps (or bulldog clamps in the neonate) are placed across the colon, which is then divided using a scalpel or diathermy. Alternatively a GIA stapler may be used.

6 The proximal and distal limbs of the colon are positioned at opposite corners of the incision, with sufficient distance between them to enable a colostomy bag to enclose the proximal stoma. Each limb is anchored to the abdominal wall with interrupted absorbable sutures in layers. The first layer is to the peritoneum and transversalis fascia; these sutures must be sufficiently close together to prevent loops of small intestine from prolapsing between them and should not penetrate the colonic mucosa, as this may result in a fecal fistula.

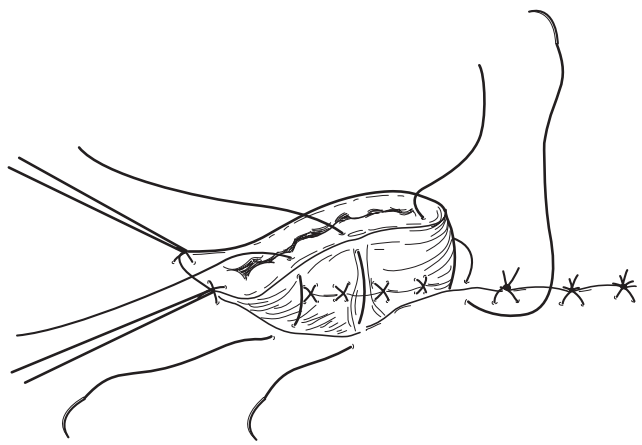


6

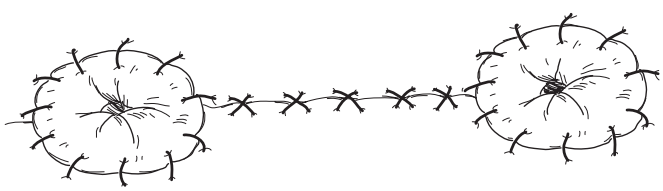
7 For the second layer, the colon is sutured to the surrounding external oblique muscle aponeurosis, taking care not to constrict the colon. The abdominal wall layers are approximated between the two limbs of the colostomy.



7

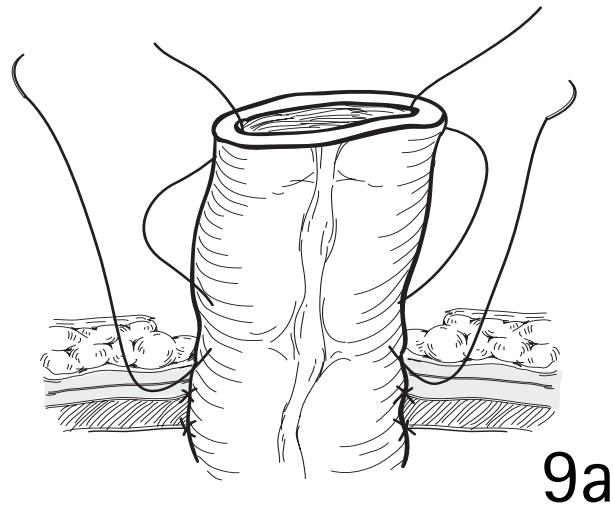


8a

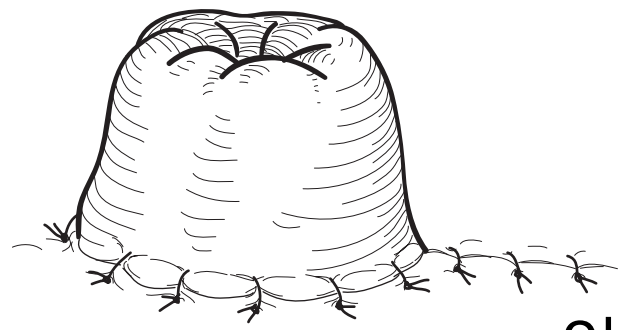


8b

8a,b Each limb of the colostomy is then opened. Full-thickness, interrupted sutures approximate the cut edge of the colon to the adjacent edges of the skin incision.

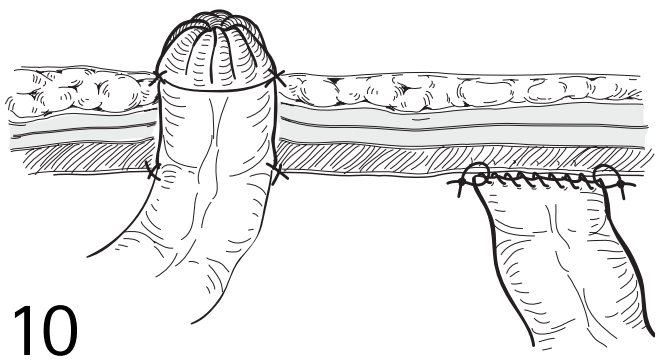


9a



9b

9a,b If required, the proximal stoma may be constructed with a spout that will protrude into the colostomy bag. Three-point interrupted sutures are placed through the abdominal wall muscle, the seromuscular wall of the colon and the cut edge of the colon to evert the end of the colostomy.

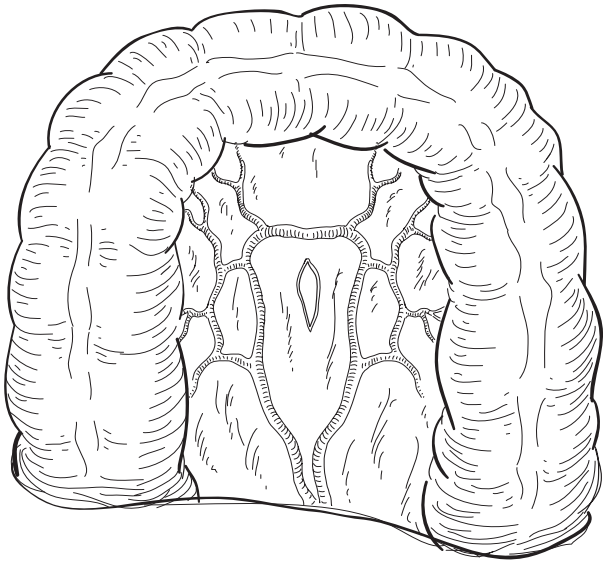


10

10 Where there is no obstruction distal to the colostomy, the distal limb may be closed transversely in two layers and placed within the peritoneal cavity (Hartmann procedure). To enable the blind end to be located easily for subsequent reconstruction, it is anchored to the undersurface of the peritoneum when the wound is closed.

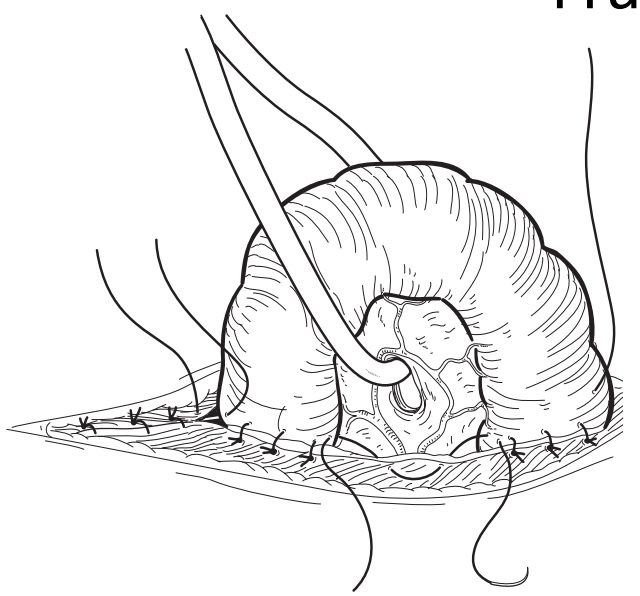
Loop colostomy

A muscle-cutting incision is used, as for a divided colostomy, but the shape of the skin incision will depend on whether the exteriorized colon is to be supported by a catheter tube, in which case a standard straight skin incision is used (see Illustration 1), or with a skin flap, for which an inverted-V incision is made (see Illustration 13).

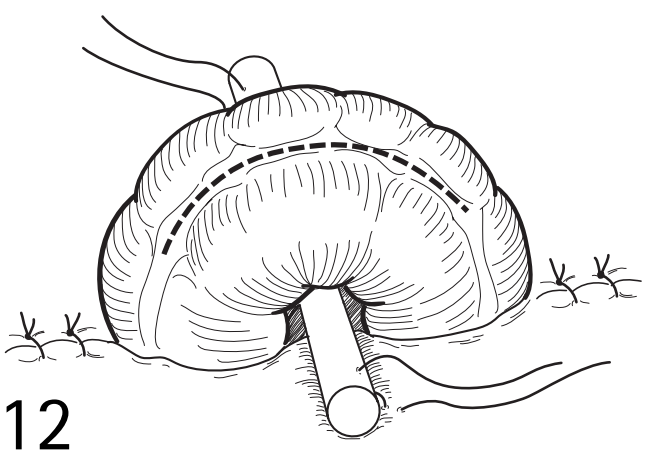


11a

11a,b The colonic loop is exteriorized and elevated using a soft catheter passed through a window in the mesentery at the point of the proposed stoma. The mesenteric artery is not divided. The two limbs of the colon are secured to the abdominal wall in layers using interrupted sutures as described for a divided colostomy. The space between the two limbs is closed with interrupted sutures to prevent the small intestine prolapsing between them, taking care to avoid damaging the mesenteric blood vessels.



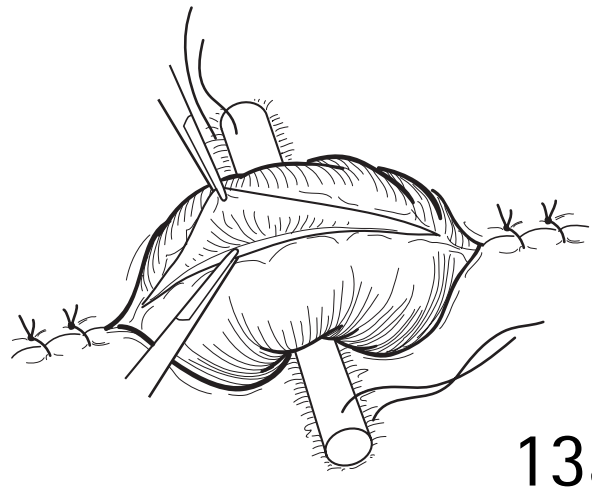
11b



12

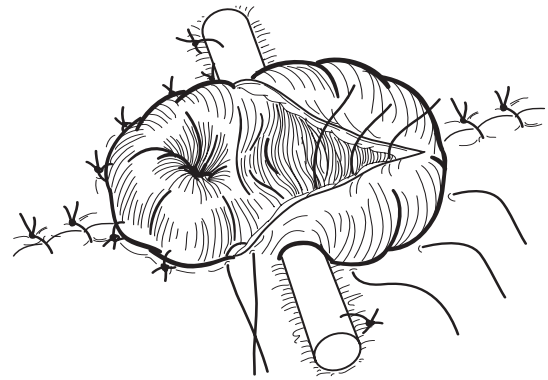
12 The catheter is shortened and anchored to the skin on either side of the loop of colon using non-absorbable sutures. The catheter must be short enough to be enclosed by the colostomy bag. The stoma is opened through a longitudinal incision at the apex of the colonic loop.

First a short incision is made so that the edges of the colon can be lifted to avoid damaging its closely opposed opposite wall, particularly if diathermy is used to incise the colon.

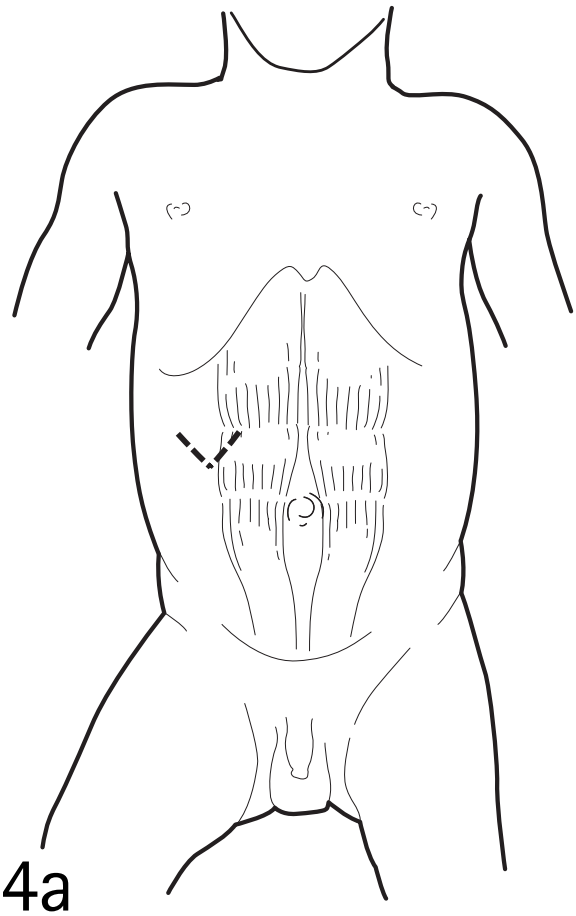


13a

13a,b The edges of the stoma are everted and sutured to the skin incision A Hegar dilator is used to confirm that the stoma has not been constricted.

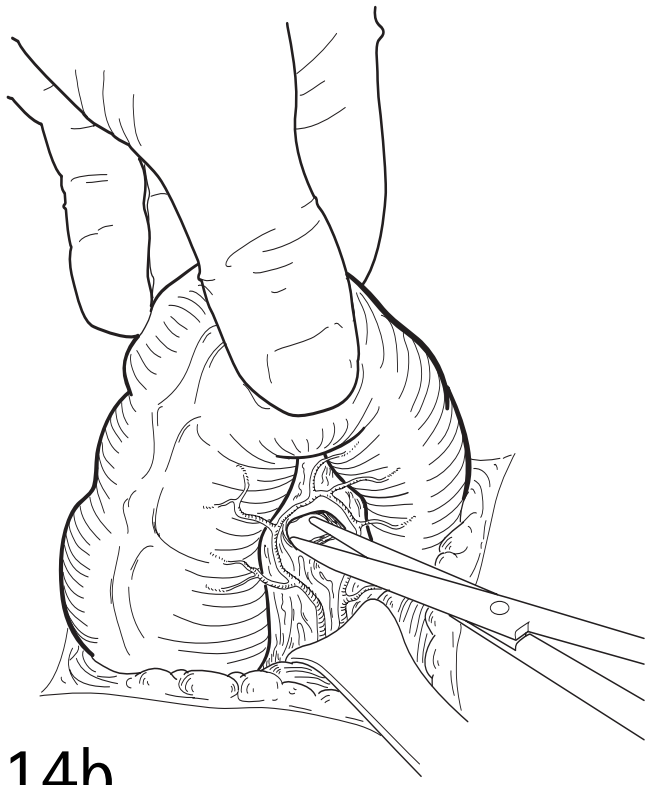


13b

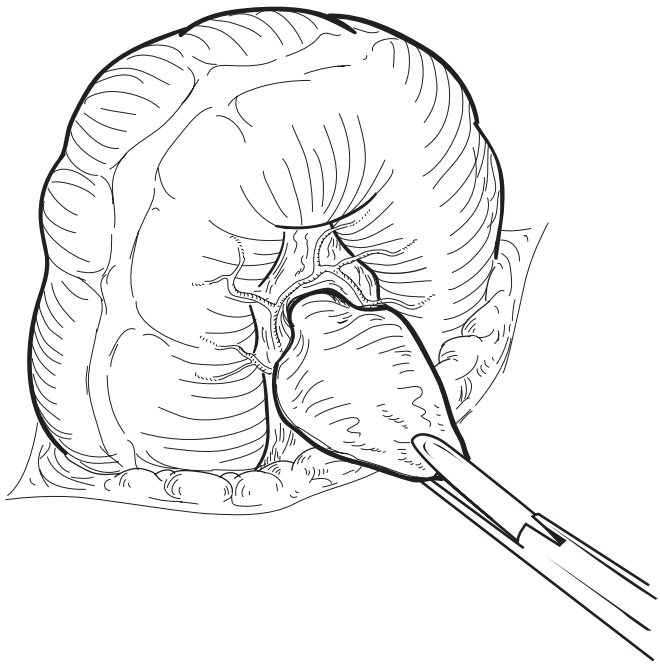


14a

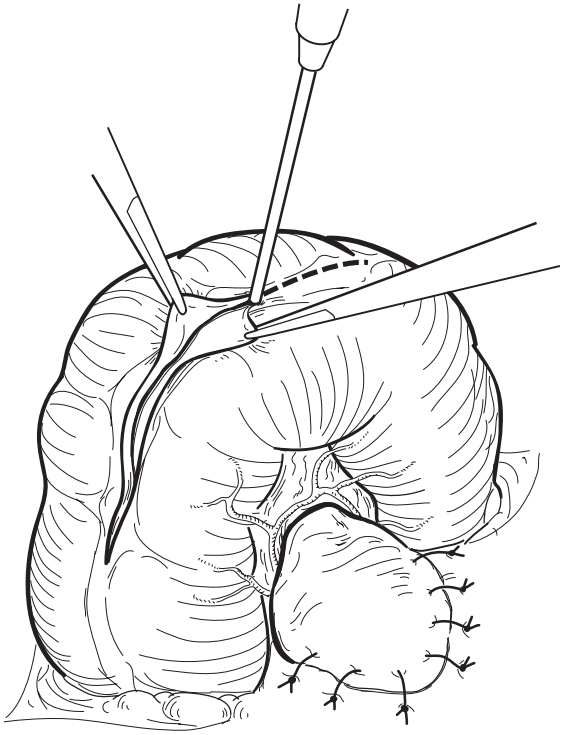
14a–e If a skin flap is used to support the colostomy, the flap is drawn through the window in the mesentery and sutured as illustrated.



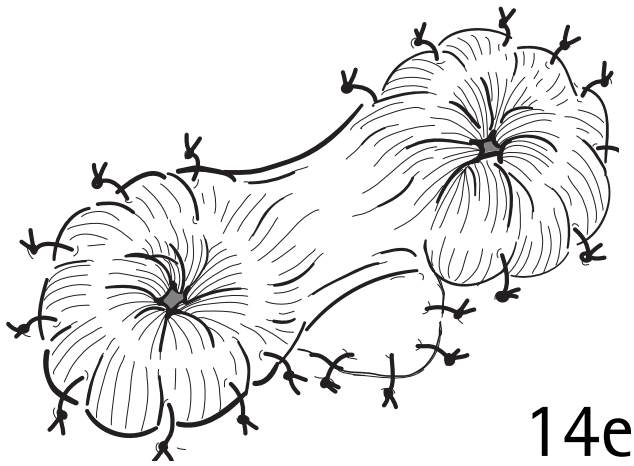
14b



14c



14d



14e

Intraoperative colostomy

If a colostomy is required following intestinal resection, it should ideally be brought out through a separate incision. An exception is when the remaining intestine is fragile and compromised (as in neonatal necrotizing enterocolitis) and exteriorizing the colonic loops through separate incisions would be traumatic and risk further loss of valuable intestinal length. In this situation, the colonic loops may be brought out at either end of the main abdominal incision and carefully anchored to the abdominal wall. It is not necessary to evert and 'mature' the stomas, as this will occur spontaneously.

COMPLICATIONS

Following colostomy formation, colostomy-related complications have been reported in up to 32 percent of patients in the United Kingdom. Constriction of a colonic loop as it traverses the abdominal wall may result in ischemia or obstruction of the stoma; usually this problem is transient, but occasionally dilatation of the stoma or revision of the colostomy may be required. Prolapse from either loop may be troublesome, requiring repeated manual reduction and, occasionally, stomal revision. Retraction of a stoma may also require revision. Leakage of stool onto the skin may cause painful peristomal excoriation and skin dehiscence; this usually arises because the colostomy bag is not properly adherent to the skin, and requires meticulous attention to skin care using specific protective preparations under the supervision of a stomatherapist. The incidence of skin excoriation and related problems is over 30 percent where specialist care and appropriate resources are not available. Parastomal hernia occurs as a result of small intestine herniating between the sutures anchoring the colostomy to the abdominal wall. Diversion coloproctitis, characterized by a mucopurulent rectal discharge, bleeding, and tenesmus, is a late effect of colonic diversion with characteristic endoscopic and histological features.

Colostomy closure

PREOPERATIVE PREPARATION AND ANESTHESIA

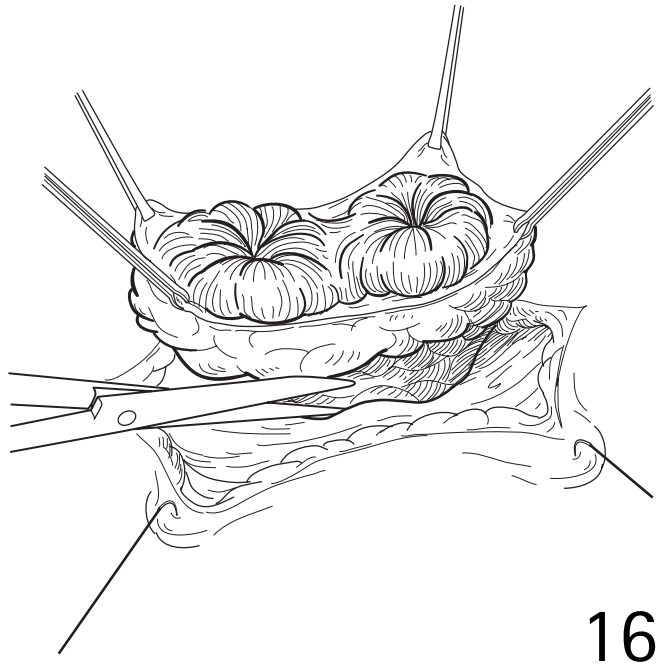
Prior to closing the colostomy, the integrity and patency of the bowel distal to the stoma must be confirmed by preoperative contrast radiology and, if appropriate, by digital rectal examination at the time of operation. Bowel preparation to empty the colon and rectal irrigation to evacuate residual stool may be necessary.

General anesthesia is employed with muscle relaxation. A nasogastric tube is inserted. Prophylactic broad-spectrum antibiotics are administered at induction of anesthesia. Any colostomy bags are removed, and residual adhesive paste is removed from the skin with solvent.

15 The incision encircles the two stomas and intervening skin scar. The skin should be incised in stages, as there may be brisk arterial oozing from the margins of the incision.

15

16 The incision is deepened to expose the external oblique aponeurosis and the margins of the opening in the abdominal wall are identified. With careful dissection using a combination of scissors and bipolar diathermy, a plane is developed between the colonic loop and the abdominal wall to enter the peritoneal cavity, dividing the anchoring sutures and inevitable scarring without breaching the colonic wall or damaging adjacent adherent loops of small intestine. This may be a difficult process and patience is essential. With a divided colostomy, the incision between the stomas is opened completely.



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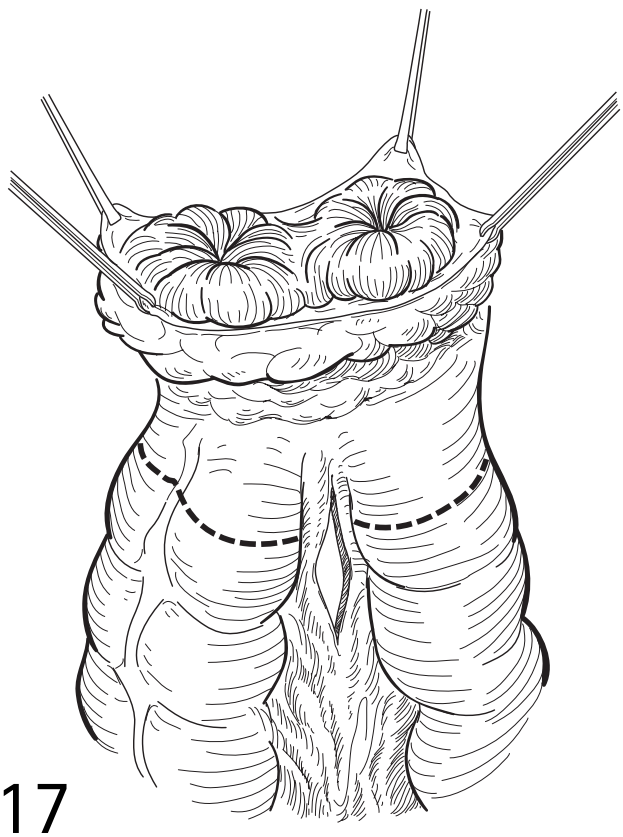
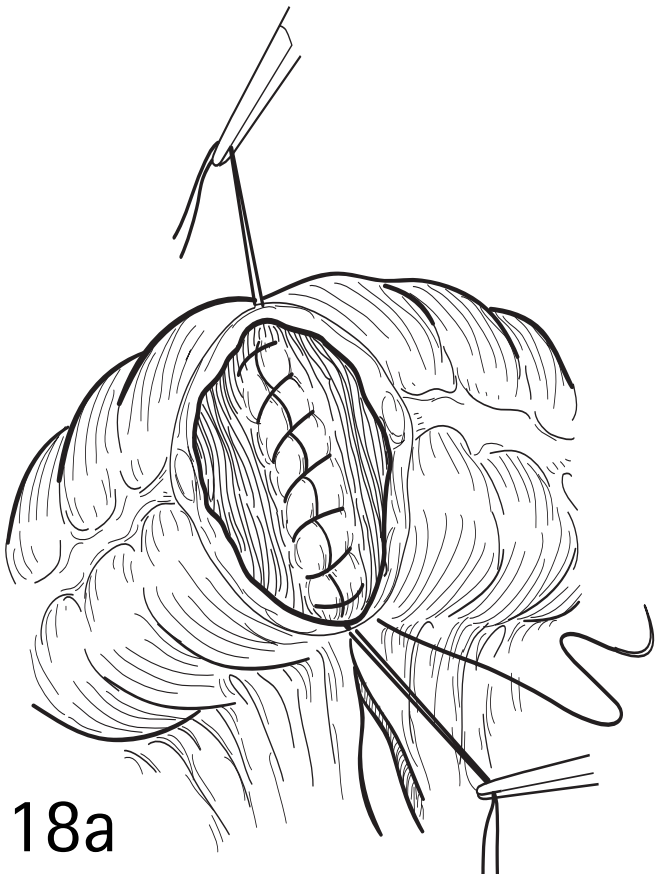


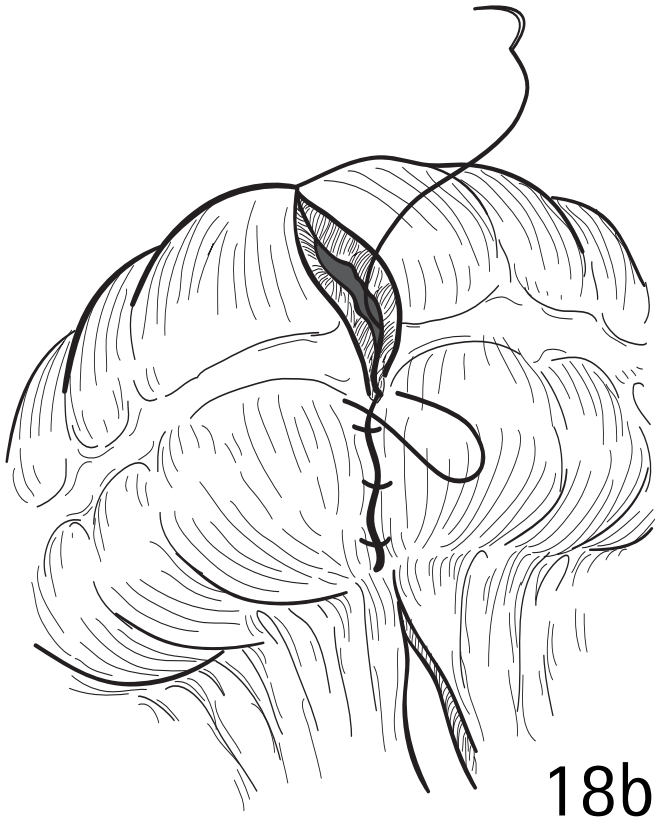
17

17 Once the two limbs of the colostomy have been mobilized sufficiently to allow them to be exteriorized, the stomas are resected back to healthy bowel and approximated by end-to-end anastomosis.

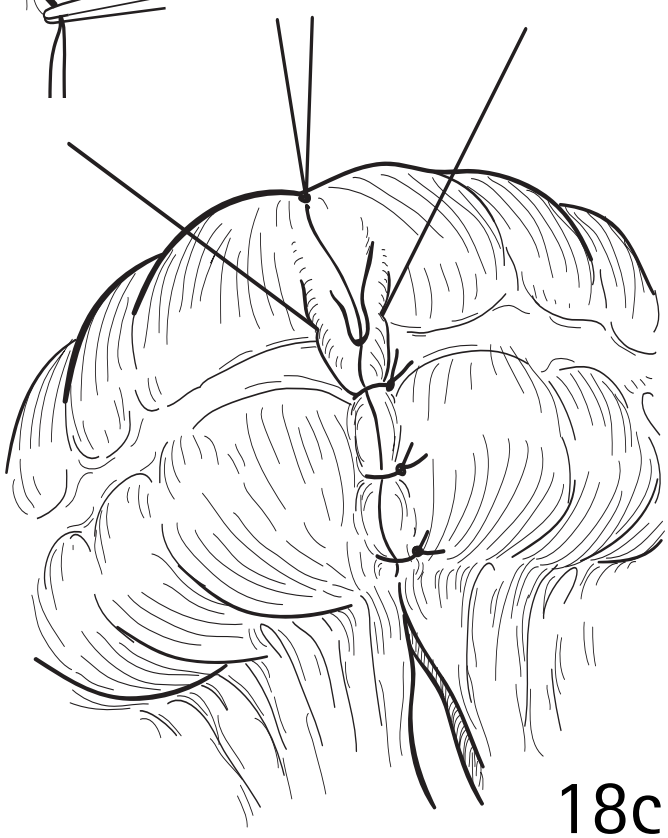
18a–c A two-layer end-to-end colonic anastomosis is suitable for the older child. In the neonate, a single-layer seromuscular suture is often preferred.



18a

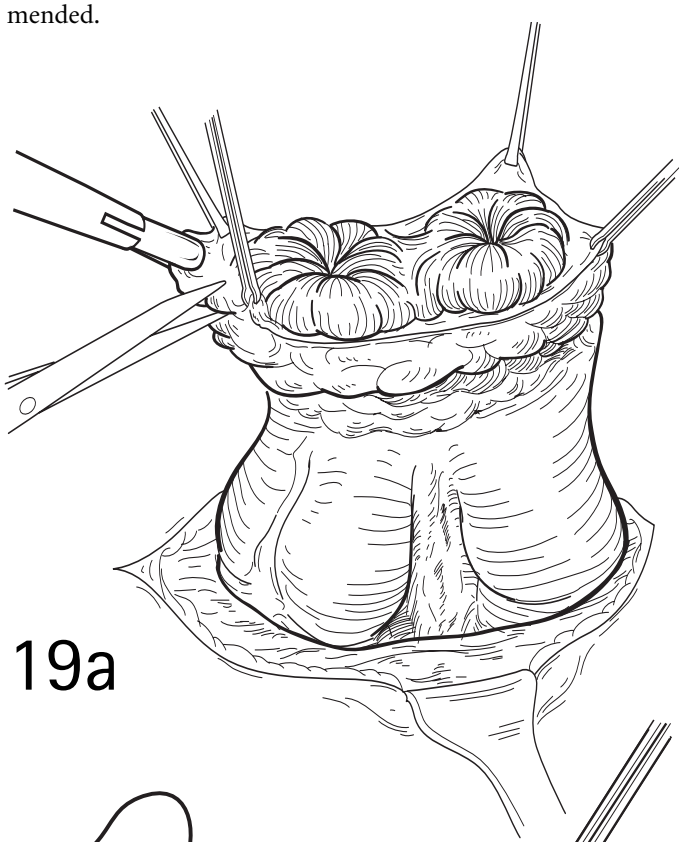


18b

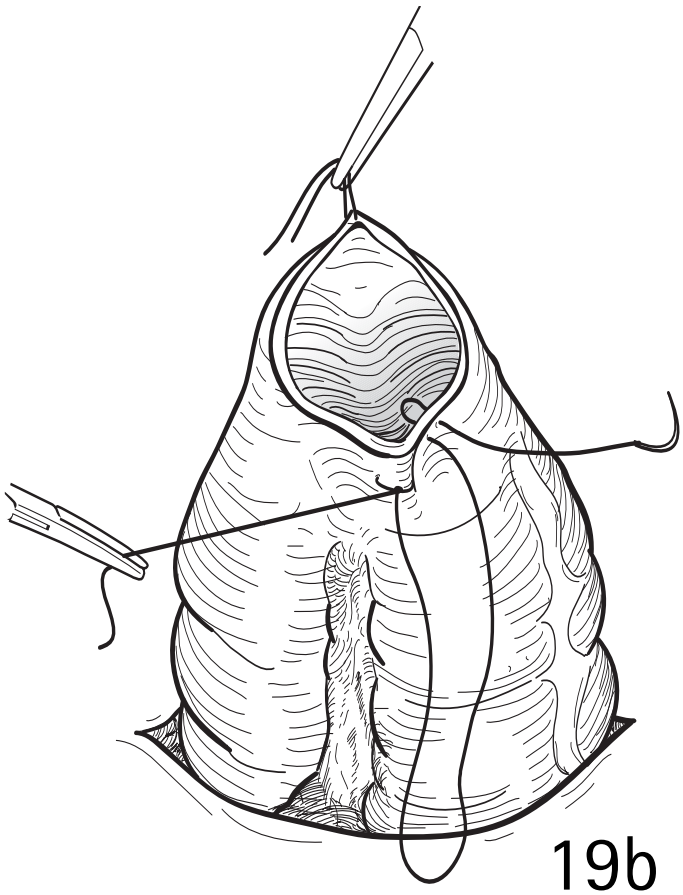


18c

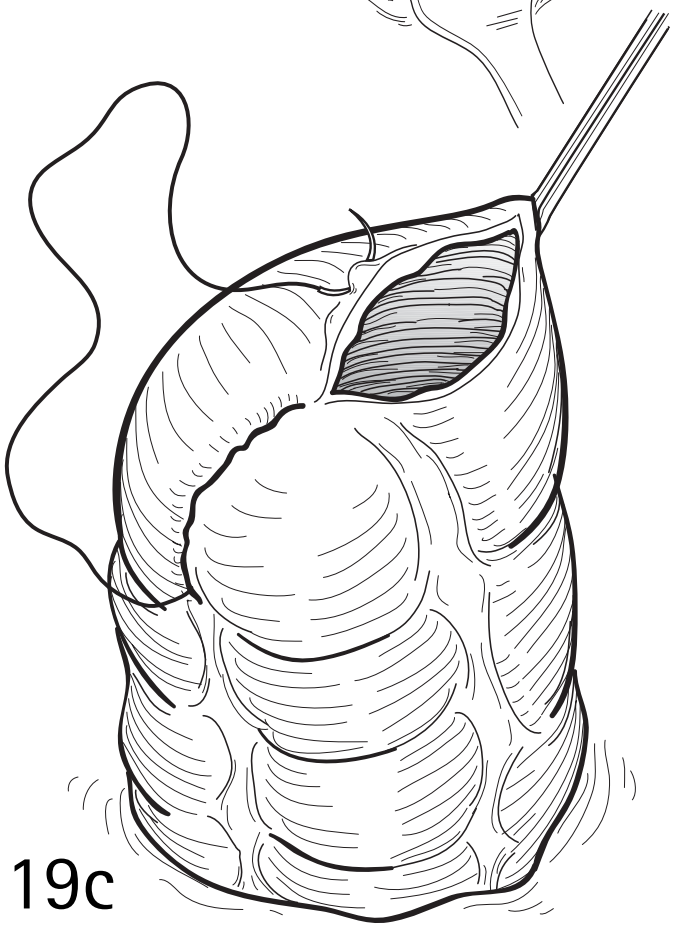
19a–d In the older child, a loop colostomy may be closed without resecting the stoma. After mobilizing the colostomy, the edges are trimmed and the colon is closed transversely (see illustrations 18b & c). In the infant, resection and end-to-end anastomosis are recommended.



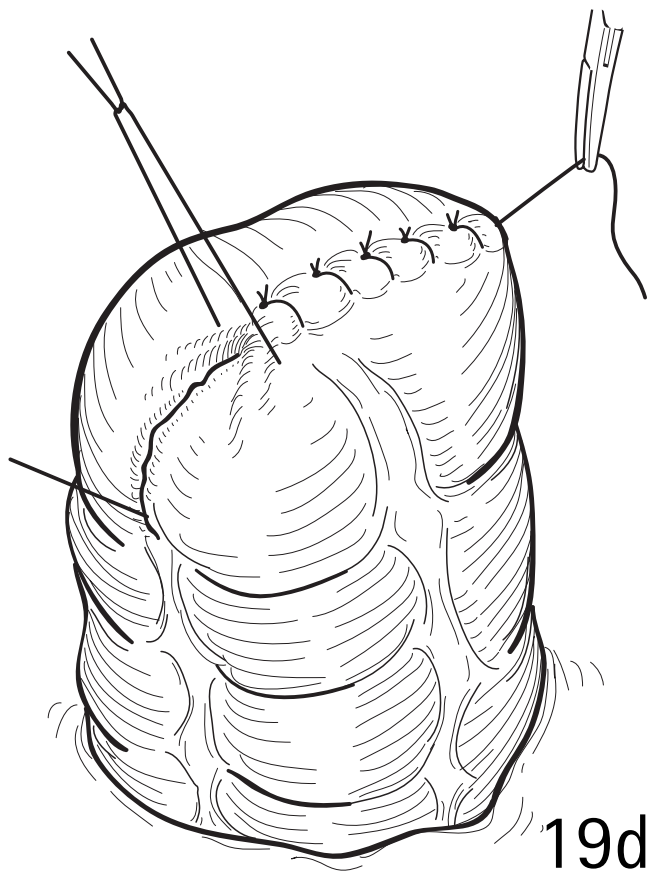
19a



19b

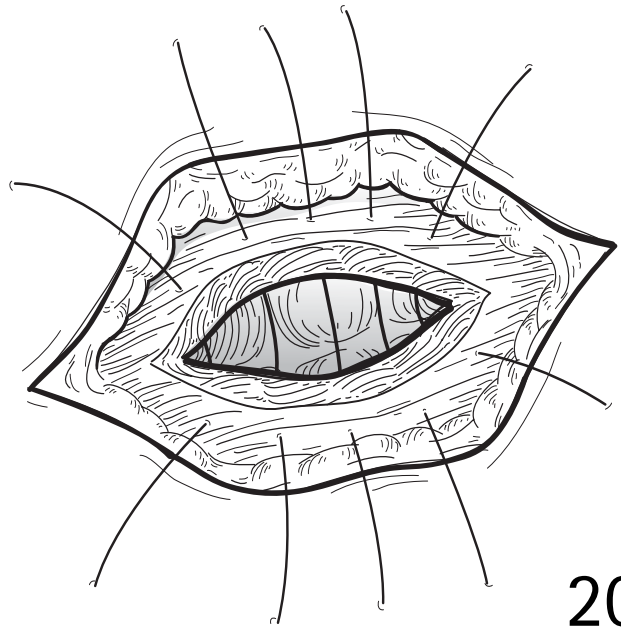


19c



19d

20 The abdominal incision is closed in layers. Interrupted, full-thickness sutures are used for the peritoneum and muscle layers if the tissue planes are not distinct. If regional analgesia has not been used, the wound is infiltrated with local anesthetic.



20

POSTOPERATIVE CARE

Nasogastric drainage and intravenous fluids are continued until gastric drainage diminishes and an adequate oral fluid intake is tolerated. Enteral feeds are increased cautiously until flatus or stools are passed. Systemic analgesia is maintained as required.

COMPLICATIONS

Early complications following colostomy closure include wound infection, the risk of which is reduced by prophylactic antibiotics, and anastomotic leak. The latter is uncommon provided the bowel is healthy and well vascularized, there is no tension on the anastomosis, and no distal bowel obstruction. Late stenosis at the suture line is also uncommon if the colon is healthy. Adhesive small bowel obstruction has been reported in 6.5 percent of children following colostomy closure, and a mortality rate of less than 1 percent.

FURTHER READING

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62

Bowel-lengthening procedures

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INTRODUCTION

The short bowel state is characterized by inadequate absorption from insufficient mucosal surface area. It follows extensive small bowel loss from antenatal or postnatal volvulus complicating malrotation or gastroschisis, and postnatal bowel loss or extensive resection for necrotizing enterocolitis. Other less common causes include long segment Hirschsprung's disease extending high into the jejunum, and vascular accidents (embolism), tumors, or injury. The impact of extensive bowel loss reflects not only on the absorptive capacity of the small bowel, but also on gut-associated immunity, such that short-gut patients are at greater risk of gut-related infection and associated liver injury. Long-term survival will depend on the natural intestinal adaptation response within the residual 10–30 percent of absorptive small bowel, the presence of the ileocecal valve, and a greater length of colon.

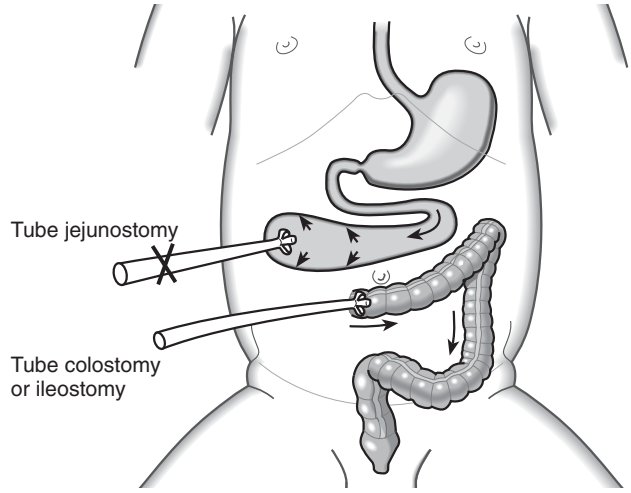
Antenatal bowel loss presents as atresia with the typical obstructed, massively dilated proximal segment, and the defunctioned undeveloped distal bowel, usually the distal colon. End-to-end anastomosis between the two segments is followed by a failure of propulsion with stasis, sepsis, and portosystemic bacterial translocation from the proximal loop. Liver dysfunction follows gut-related sepsis, and is exacerbated by hyperalimentation, parenteral nutrition-related toxicity largely from plant phytosterols in lipid preparations, and possibly from a lack of small-bowel-related 'hepatoprotective factors'. In a high percentage of short-gut children, rapidly progressive liver injury leads to cholestasis and hepatocyte loss, with end-stage liver failure within a few months. However, liver dysfunction may be reversible if bowel adaptation is sufficient to shift the balance toward enteral nutrition and better gut-associated immunity. Bowel reconstructive procedures are designed to reduce the stasis and sepsis within the poorly propulsive dilated segment and to enhance the intestinal adaptation process toward enteral autonomy.

The child with short bowel: initial surgery

The child with short bowel presents a difficult, complex, multifaceted problem that requires close cooperation between the local team and a designated 'intestinal failure center' combining particularly pediatric gastroenterology and nutrition, autologous gastrointestinal reconstruction, and liver and bowel transplantation. Immediately at diagnosis, a 'management plan' is jointly developed, initially concentrating on survival and growth, liver protection, and preservation of venous access. Once stable, the child is best managed within his or her family and social environment, being hospitalized only for specific assessments or procedures.

1 At first surgery, all available bowel is preserved, and minimal, if any, bowel reconstruction is undertaken. A large-size Malecot catheter (16 Fr) is placed in the distal-most jejunum, and is brought out on the abdominal wall as a tube jejunostomy.

This allows free drainage of the dilated bowel, thereby avoiding stasis, sepsis, and bacterial translocation. It is then also possible to commence oral (enteral) feeding, thus stimulating the child to become 'food-wise'and inducing small-bowel mucosal adaptation. Intermittent occlusion of the Malecot jejunostomy tube allows 'controlled bowel expansion,' developing more autologous bowel for subsequent planned reconstruction. A smaller size Malecot catheter (10 Fr) is placed in the distal bowel (usually colon) to allow for recycling of jejunostomy losses, to maximize absorption, and to stimulate adaptation also in the distal colon.



1

Autologous gastrointestinal reconstruction (AGIR)

Table 62.1 highlights the surgical techniques now available to enhance absorption and adaptation in the residual autologous bowel. These may be relevant alone or in various combinations.

Such procedures are not an end in themselves, but should form part of a structured management plan specific to a particular child. Thus the bowel reconstructive plan may commence with a period of jejunostomy tube drainage of the dilated loop. The Malecot tube is then clamped for graded intervals to increase mucosal contact time for absorption and to induce bowel expansion, thereby creating more autologous tissue for eventual reconstruction. This may take the form of longitudinal intestinal lengthening and tailoring (LILT), possibly combined with reversed antiperistaltic segments or colon transposition. In the event of bowel redilatation after LILT, a serial transverse enteroplasty (STEP) reduces bowel diameter to assist propulsion and further increases length to aid absorption.

Bowel-lengthening procedures

Bowel-lengthening procedures aim to reduce the diameter of dilated bowel without loss of absorptive mucosa, to establish effective propulsion, and to use the tailored bowel to create additional isoperistaltic length for increased mucosal contact and enhanced absorption. Since publication of the original technique in 1980, experience with LILT has been increasing. Results have been variable and largely dependent on the extent and quality of the residual bowel, and the condition of the child at the time of surgery. The procedure does not 'guarantee a cure,' but has often been followed by enteral autonomy or by a significant reduction in parenteral nutrition requirements.

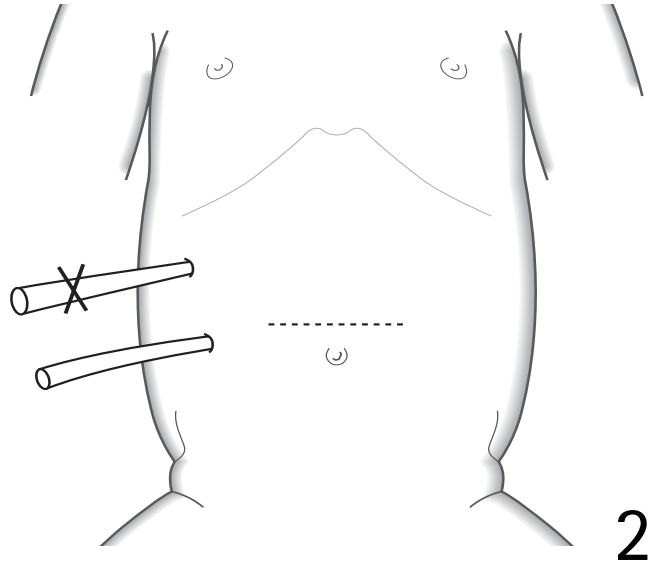
Table 62.1 *Procedures for bowel reconstruction*

Delay transit and increase contact time	Reversed antiperistaltic segments Prejejunal or pre-ileal colon transposition Intestinal valves Intermittent occlusion tube jejunostomy
Improve propulsion	Antemesenteric tailoring Plication (de Lorimer, Harrison)
Bowel expansion	Tube jejunostomy for 'controlled occlusion–recycle' (Bianchi) Nipple valve (Georgeson)
Bowel lengthening	Longitudinal intestinal lengthening and tailoring (LILT) (Bianchi) Serial transverse enteroplasty (STEP) (Kim et al.)
Antemesenteric blood supply	Isolated bowel segment Iowa model (Kimura) Composite bowel loops (Bianchi)
Sequential and combined techniques	Isolated segment + LILT (Georgeson) LILT + reversed segment (Bianchi) LILT + STEP (Kim et al.)

OPERATION

Longitudinal intestinal lengthening and tailoring (Bianchi 1980)

2 The abdomen is opened through an already existing scar or through a transverse supraumbilical incision, and the whole of the small and large bowel is dissected free of adhesions and exteriorized. During this phase, careful attention is given to avoiding damage to the mesenteric vessels supplying the dilated bowel. It is often easier to take down a previous anastomosis between the dilated proximal bowel (to be lengthened) and the distal bowel (often the colon). The bowel diameter and bowel length, measured along the antemesenteric border, are recorded. Bowel division may be undertaken using a manual technique or the endoscopic GIA stapler. The author prefers, and recommends as safer, a manual division-and-suture technique.



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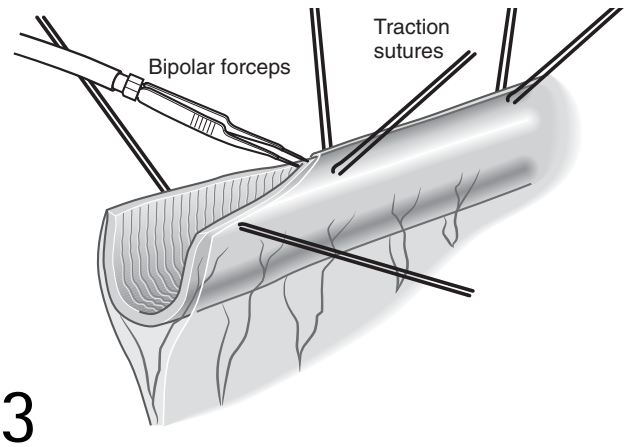
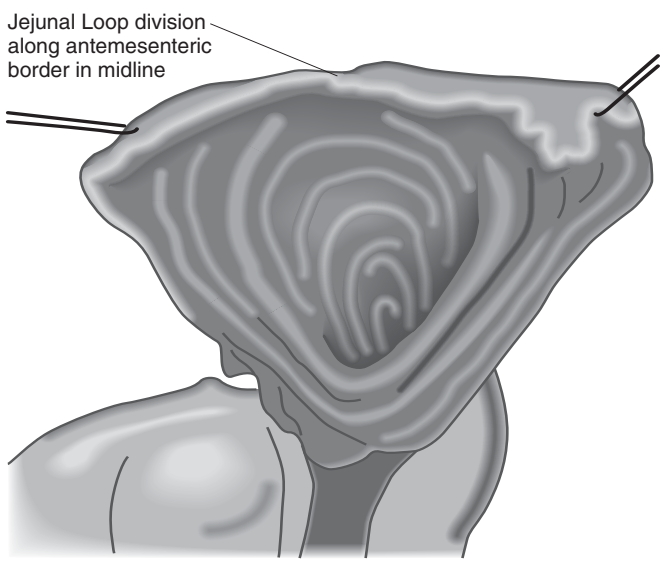


3

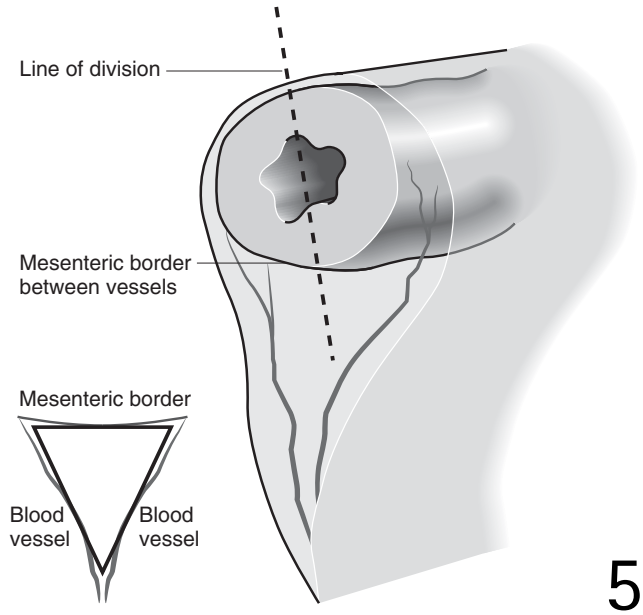
3 Traction sutures are placed along the antemesenteric border to the right and left of the midline at about 10 cm intervals, and the bowel is drawn upwards and outwards against the base of the mesentery.

4 With the 'cutting wave' of the bipolar diathermy, the bowel is divided longitudinally for a comfortable working length, passing between the traction sutures along the antemesenteric border to the right and left of the midline for about 10 cm.



4

5 Outward and upward traction on the opened bowel loop against the base of the mesentery gives access to the blood vessels between the leaves of the mesentery. This natural plane is developed by blunt dissection such that the mesenteric border of the bowel of approximately 1 cm width between the vessels forms the base of an inverted triangle. The mesenteric border is divided longitudinally in the midline with cutting bipolar diathermy, passing between the blood vessels, which can be safeguarded.



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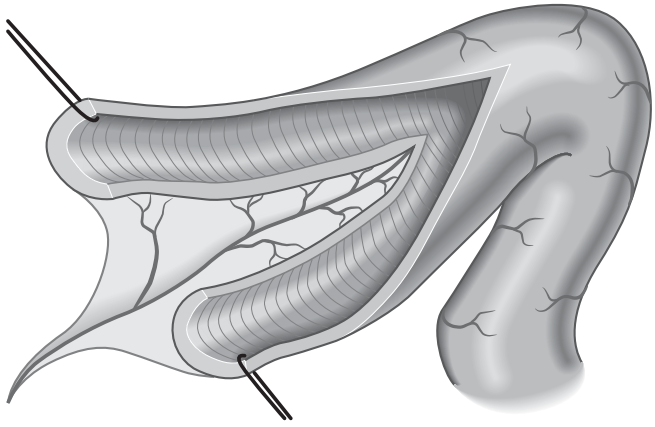
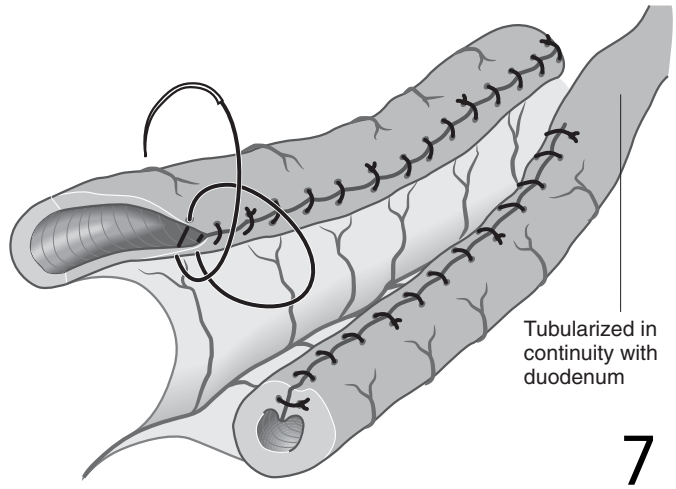


6

6 Stages 3, 4, and 5 are repeated moving proximally until the dilated bowel is divided into two fully vascularized hemisegments.

7 One hemisegment is completely detached by division along the lateral wall. The other hemisegment is tubularized, in continuity with the duodenum, with a continuous horizontal inverting mattress suture of 5/0 absorbable material, tying a securing knot every fourth throw. The sutures are placed some 2 mm from the cut edge of the bowel, turning the edges into the lumen. It is important to avoid injury to consecutive blood vessels, which are more clearly seen when using a manual suture technique as compared to the stapler.

The second hemisegment may be tubularized and then anastomosed isoperistaltically to the first. Alternatively, to ensure a safer hemiloop anastomosis, the second hemisegment, while still open, is anastomosed to the end of the first, and is then tubularized in continuity.



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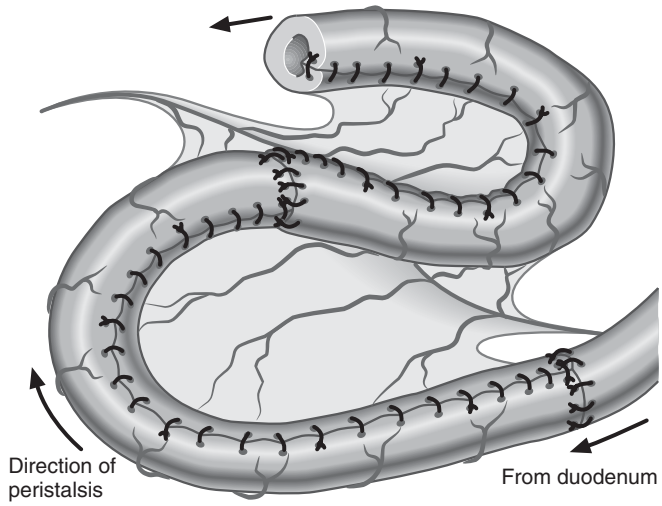
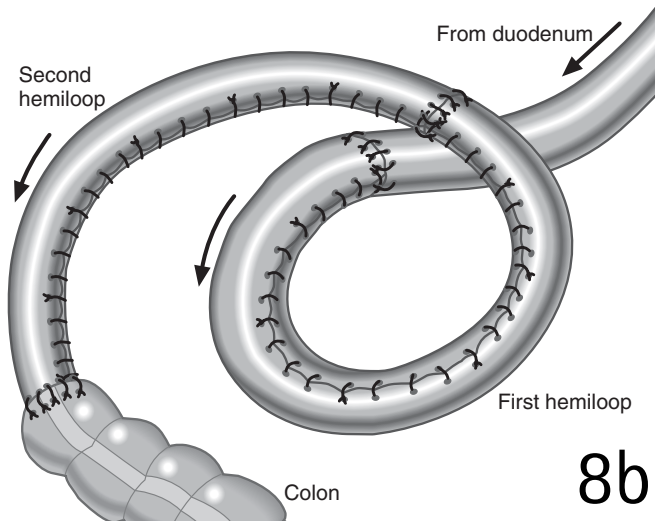


8a



8b

8a,b Isoperistaltic anastomosis requires apposition of opposite ends of the hemiloops. This can be performed in the shape of a Bianchi-S (Illustration a) with the bowel lying over the mesentery, or as an Aigrain Spiral (Illustration b) with one loop passing beneath the other. The distal end of the second hemiloop is anastomosed to the distal bowel (often the colon) to establish bowel continuity to anus.

The abdomen is closed in layers, and abdominal drainage is optional. Blood loss is not usually significant; however, fluid and colloid losses may be appreciable and require appropriate replacement intraoperatively and postoperatively.

POSTOPERATIVE CARE

The child is managed with nasogastric aspiration, antibiotics, and intravenous alimentation until return of bowel function, often by the third to fifth day, when oral/enteral feeding is slowly reintroduced. Initial delay in passage through the lengthened bowel and intermittent vomiting are not uncommon and resolve rapidly. Parenteral nutrition is progressively reduced over several weeks in line with improving absorption.

Complications

There has been no operative mortality, and morbidity, which has been minimal, has been largely related to stenosis at the hemiloop anastomosis, occasional external fistula formation from suture line disruption, and the rare event of hemiloop loss from compromised blood supply during the surgery. Over the longer term, areas of significant stenosis and recurrence of bowel dilatation with stasis and sepsis may require further surgery. Despite the apparent lack of morbidity, bowel-lengthening procedures have the potential for serious complications and should not be undertaken lightly. Inappropriate and ill-timed application may compromise the child's only chance for enteral autonomy on autologous bowel and may precipitate the need for bowel or liver/bowel transplantation.

LONG-TERM OUTCOME

A period of at least 3–24 months is often necessary for steady progress to enteral autonomy. However, intestinal adaptation often continues for several years, and referral for bowel transplantation should not be hasty. Once enteral autonomy has been achieved, it is likely to be sustained, with relatively

normal growth and development. Loss of the ileum with its specific binding sites will necessitate lifelong vitamin B_{12} supplements. Interruption of the enterohepatic circulation and loss of bile salts into the colon generate abnormal bile with an increased incidence of gallstones, and a greater absorption of free uric acid with the formation of renal calculi.

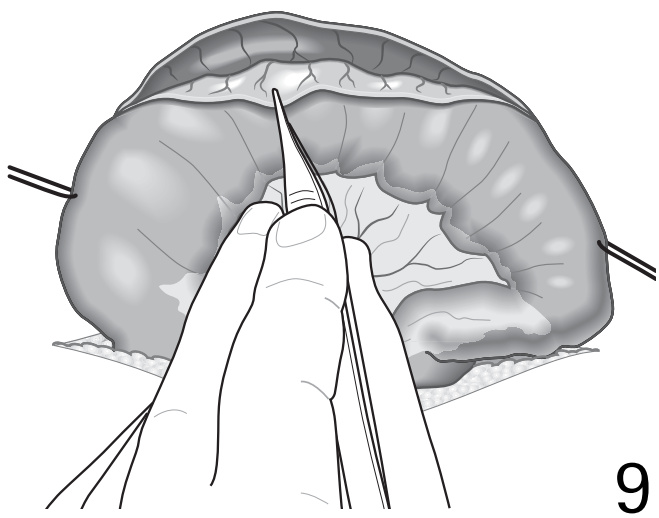
Failure to establish enteral autonomy will lead to consideration of 'life with parenteral support' or bowel transplantation, largely determined by quality of life and the ability to sustain parenteral feeding (venous access). Bowel transplantation still carries significant short-term and, particularly, long-term hazards and should not be regarded as the 'easier' primary option, but rather as a backup when all prospects for enteral autonomy on autologous bowel have been exhausted.

OTHER BOWEL-LENGTHENING PROCEDURES

The isolated bowel segment 'Iowa' models (Kimura 1993) and the composite bowel loop (Bianchi 1995)

These procedures import a new blood supply to the antemesenteric border of the dilated bowel by grafting the bowel to the liver and abdominal wall (Kimura – isolated bowel segment), or to a vascularized muscle flap from the greater curve of the stomach or a mucosally denuded colonic muscle patch (Bianchi – composite loop).

9 A longitudinal seromuscular myotomy incision is made in the midline along the antemesenteric border of the dilated bowel and the seromuscular layer is peeled back to expose the submucosa.



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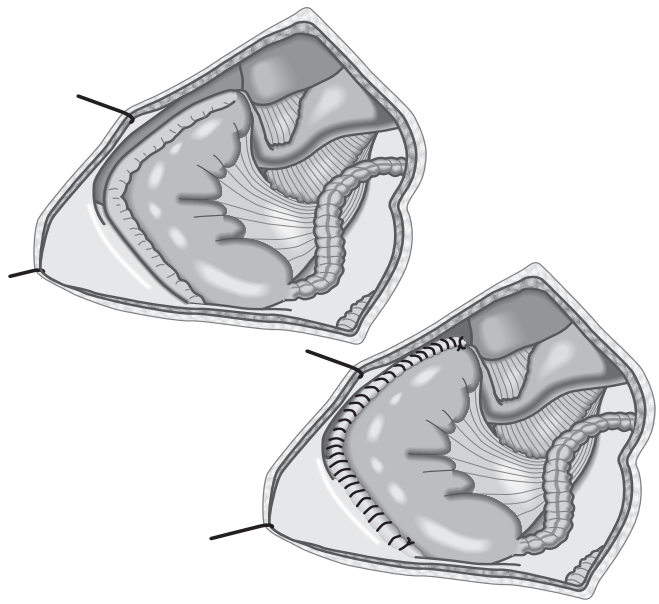
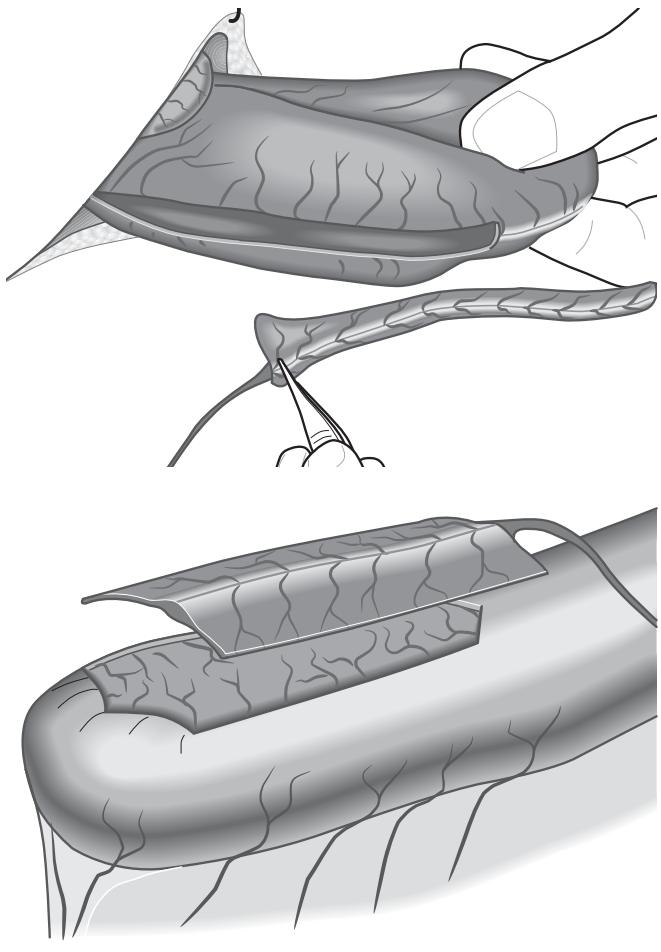


10a

10a,b For the Kimura procedure the exposed submucosal surface is grafted to the undersurface of the liver and the abdominal wall (Illustration a). In constructing a composite loop, a 2–4 cm wide mucosally denuded, vascularized gastric muscle flap based on the right gastroepiploic artery is raised from the greater curve of the stomach and is grafted to the submucosal surface of the dilated jejunum (Illustration b). A similar vascularized, mucosally denuded colonic muscle patch can be prepared by removing the mucosa along the submucosal plane.



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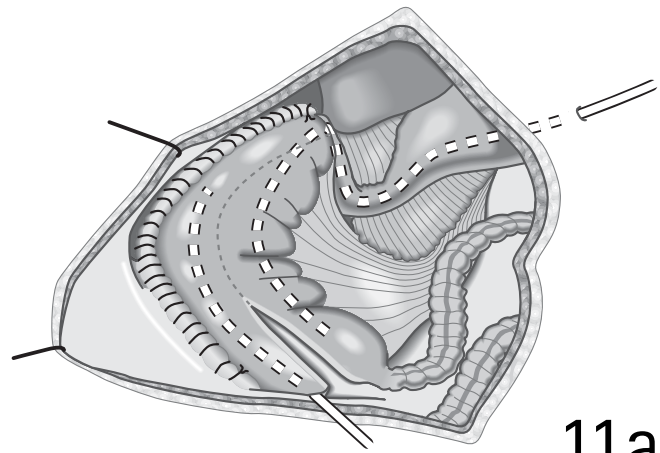

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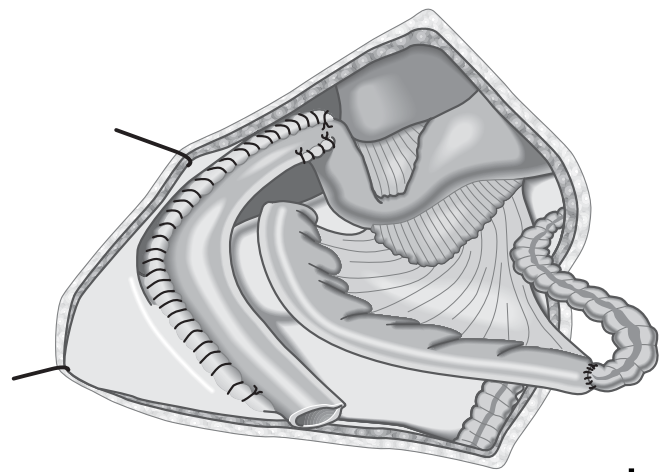

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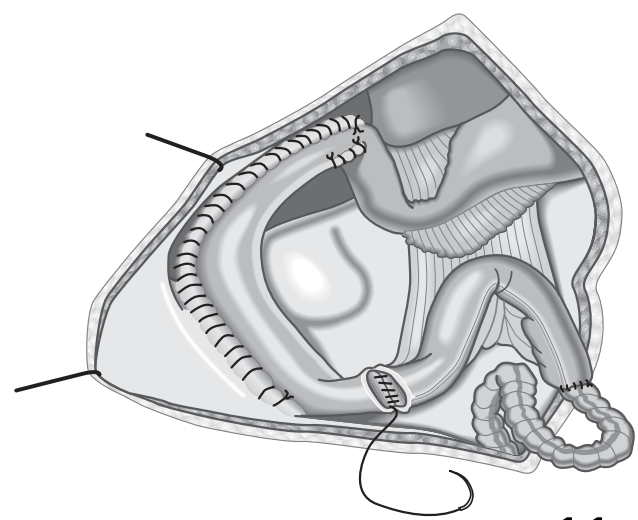
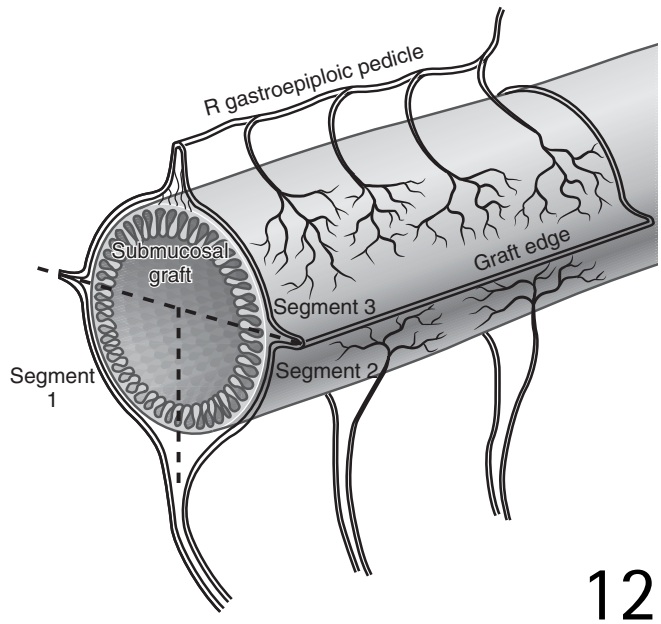


11c

11a–c After 12 weeks to allow neovascularization, the bowel loop is divided *horizontally*, creating an isolated antemesenteric segment vascularized from adhesions to the liver and abdominal wall (Kimura isolated segment – Illustration a) or from the gastric or colonic muscle flap (Bianchi composite loop – Illustration b). Isolated segments are tubularized and anastomosed in continuity with the distal bowel (Illustration c).

12 A combination of the conventional Bianchi LILT and the Kimura isolated bowel segment (Georgeson sequential lengthening 1994) or the Bianchi composite loop allows the development of a third length of bowel. Clinical application has been limited, and these procedures should be applied with caution and only within specialized units experienced in the advanced management of the short bowel state.



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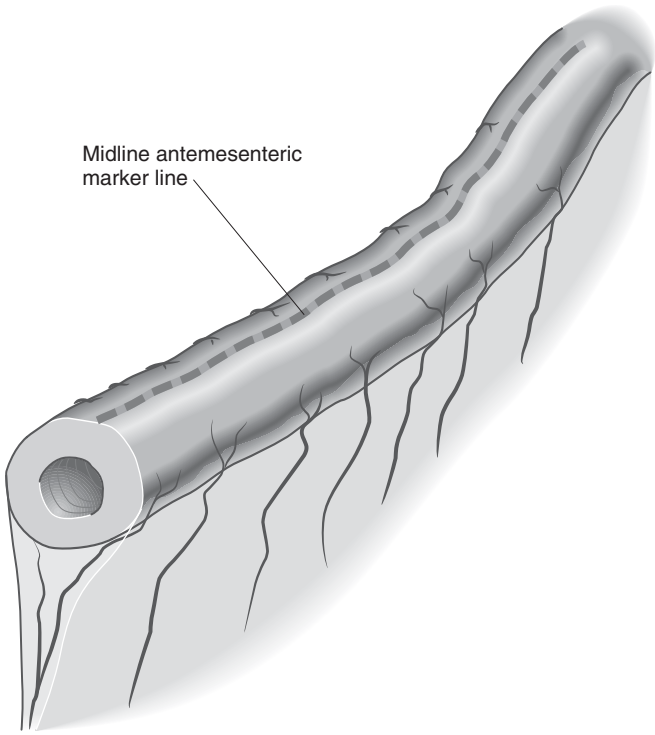
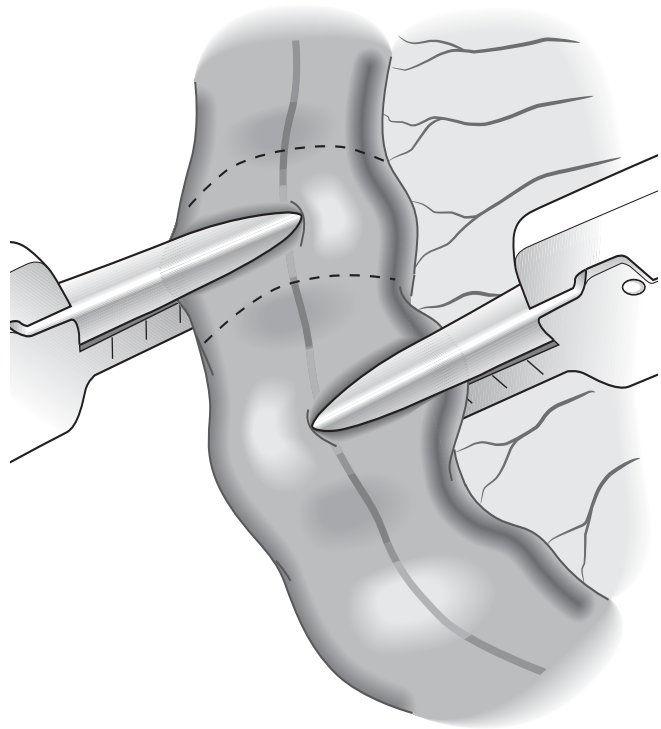


13

Serial transverse enteroplasty (Kim et al. 2003)

13 The whole of the dilated small bowel is mobilized on the mesentery, and bowel continuity is maintained. Bowel length along the antemesenteric border and bowel diameter are recorded. Following Kim et al. (2003), a marker line is drawn longitudinally along the midline on the antemesenteric border of the dilated bowel to act as an orientation guide when closing the endoscopic GIA stapler.

14 Starting at the proximal end of the dilated loop, the larger arm of the stapler is inserted through a small mesenteric defect between consecutive blood vessels. As the stapler is closed, the mesenteric and antemesenteric surfaces of the bowel are apposed, keeping the antemesenteric marker line strictly in the midline. It should be noted that the stapler lies perpendicular to the mesentery (the long axis of the bowel) and that *the stapled cuts are strictly transverse* to ensure adequate blood supply to the bowel. A 2 cm gap is left uncut beyond the end of the staples (guide-mark on short arm of stapler), which will represent the final diameter of the reconstituted bowel loop.



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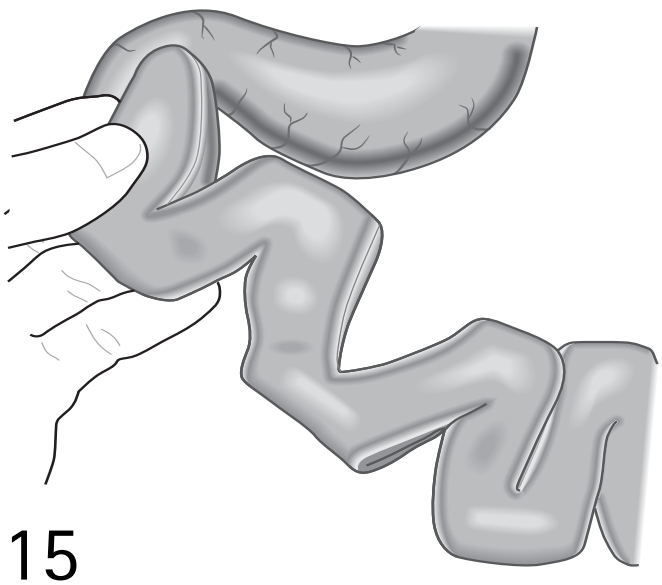

15

15 The next cut is taken some 2 cm distally and *the stapler is inserted from the opposite side*. The procedure is carried on serially down the dilated loop until all of the dilated bowel has been divided, creating a lengthened bowel of 2 cm luminal diameter.

COMMENT

Serial transverse enteroplasty is a relatively new procedure, having been introduced clinically by Kim et al. in 2003. The procedure is straightforward and attractive and may be applied as a primary form of bowel lengthening, although it may have even greater advantage as a secondary procedure in creating additional bowel length following a previous Bianchi LILT. Present experience suggests that the procedure is relatively safe, and complications from staple-line dehiscence or interference with bowel blood supply have not as yet been recorded. Early reports suggest that isoperistalsis is maintained and that there is improvement in overall absorption. Present experience is limited and the long-term outcome is as yet unknown.

FURTHER READING

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63

Management of portal hypertension

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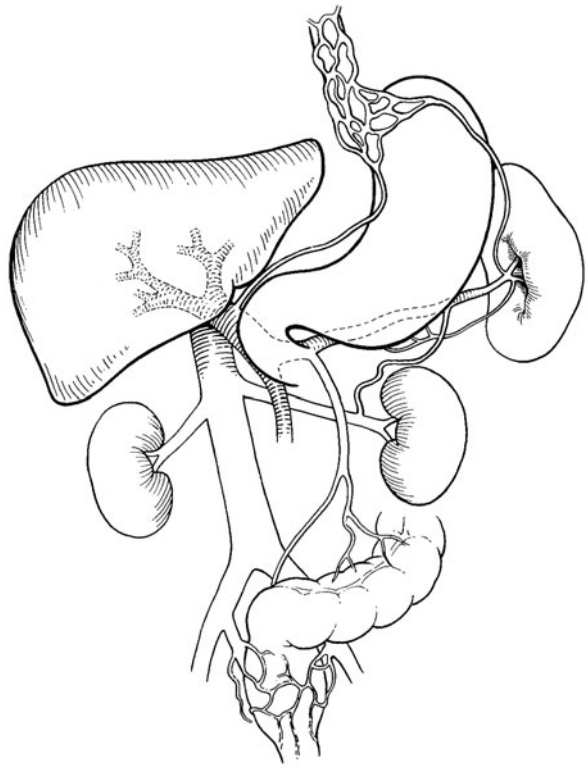
The treatment of portal hypertension in both children and adults remains the subject of considerable controversy. Therapeutic options now include a broad range of pharmacologic, endoscopic, radiologic, and surgical procedures. The etiology of portal hypertension in children is different from that in adults, however. In adults, the vast majority of cases are caused by cirrhosis (usually due to alcohol abuse), whereas in children portal hypertension is due to portal vein thrombosis (PVT) in 30 percent of cases. When cirrhosis is the cause in children, it is usually secondary to biliary atresia. The differing etiologies of portal hypertension compel a distinct management approach.

1 The main clinical manifestation of portal hypertension is the same, namely esophageal variceal hemorrhage. Esophageal varices develop as a consequence of increased resistance to flow through the portal circuit. In an attempt to increase outflow from the splanchnic circulation, collateral vessels dilate. Many collateral pathways develop, including (1) left gastric (coronary) vein and short gastrics to esophageal veins and thence to azygous/hemiazygous veins in the thorax; (2) superior hemorrhoidal veins to the middle and inferior hemorrhoidal veins and ultimately the inferior vena cava (IVC); and (3) umbilical vein to superficial veins of the abdominal wall and superior/inferior epigastric veins. The most clinically important collaterals are the submucosal esophageal varices, which have a propensity to rupture as a consequence of increased pressure within the varix secondary to coughing or straining or from ulceration secondary to esophagitis.



1

HISTORY

The management of esophageal bleeding has evolved dramatically over the last two decades. The use of surgical techniques employing a direct attack on the bleeding varices and portosystemic shunt procedures have been increasingly supplanted by the use of endoscopic means (sclerotherapy and banding) and transjugular intrahepatic portal systemic shunts (TIPS). The success of liver transplantation has also dramatically altered the way in which we approach those patients with end-stage liver disease. Most patients with cirrhosis as a cause of portal hypertension are now listed for liver transplantation and bleeding from esophageal varices is managed by non-surgical means.

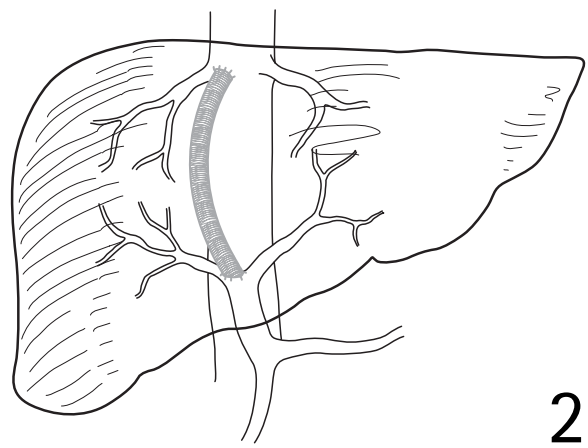
PRINCIPLES AND JUSTIFICATION

Bleeding from esophageal varices typically occurs suddenly, without warning, as massive hematemesis. Occasionally, the bleeding is more insidious, with melenic stools as the first sign. When the cause of portal hypertension is portal vein thrombosis, the onset is typically in a previously healthy young child, by the age of 6 years in 80 percent of cases. Other stigmata of liver disease are usually absent, except for an enlarged spleen. The hemorrhage usually stops spontaneously, partly because coagulation is normal.

Conversely, in those children with portal hypertension caused by cirrhosis, the liver disease is usually known about and physical findings of chronic disease are obvious (e.g., jaundice, ascites, splenomegaly, spider angiomas, caput medusa). Esophageal variceal bleeding in this setting should not be surprising or unexpected.

The choice of management technique is highly dependent on the etiology and on the experience and expertise available. In general, bleeding in patients with PVT is managed by endoscopic techniques or by portosystemic shunts. Bleeding in patients with end-stage liver disease is managed by methods that do not interfere with subsequent liver transplantation, such as endosclerosis/banding or radiologic techniques (TIPS). Shunts in these patients have a high morbidity rate and can make liver replacement very difficult or impossible. Non-shunting surgical procedures (ligation of varices, esophageal transaction, Sugiura's procedure) have largely been abandoned and are not recommended as primary surgical therapy. The only role for these procedures may be in cases where shunts and transplantation are not possible and endoscopic methods have failed.

2 Recently, the use of non-operative shunts for adults has been gaining popularity. Transjugular intrahepatic portosystemic shunts have not yet been widely applied to children, but they avoid some of the surgical complications and should not interfere with future liver transplantation. Complications include those arising from the technical aspects of the procedure (bleeding and hepatic capsule perforation), those arising from the shunt itself (encephalopathy), and the development of shunt stenosis, occlusion, and infection. Continued shunt surveillance and periodic dilatation are necessary to maintain patency. Therefore, TIPS is generally used as a bridge to liver transplantation and should be considered for short-term use only.



2

PREOPERATIVE ASSESSMENT AND PREPARATION

The first priority in the treatment of gastrointestinal bleeding is volume replacement and stabilization. Two large-bore intravenous catheters should be placed for volume repletion. Until blood is available, crystalloid and colloid solutions can be judiciously administered. Red blood cells and fresh frozen plasma should be used to replace shed blood and correct coagulopathy as needed. The goal of resuscitation is to restore tissue perfusion, which is best judged clinically by urine output monitored by Foley catheter placement. Central venous pressure is also helpful to judge volume status, as over-resuscitation can be detrimental by increasing portal venous pressure. Somatostatin and octreotide can be helpful in acute variceal hemorrhage to reduce portal pressure and promote the cessation of persistent bleeding.

Routine laboratory parameters (bilirubin, albumin, and prothrombin time) are used to assess the patient's liver function. Other liver enzymes (transaminase, alkaline phosphatase, gamma glutamyl transferase) are helpful to suggest liver injury or hepatitis. Viral serologies are determined if indicated by history or preliminary evaluation.

Once the patient is stabilized, upper intestinal endoscopy

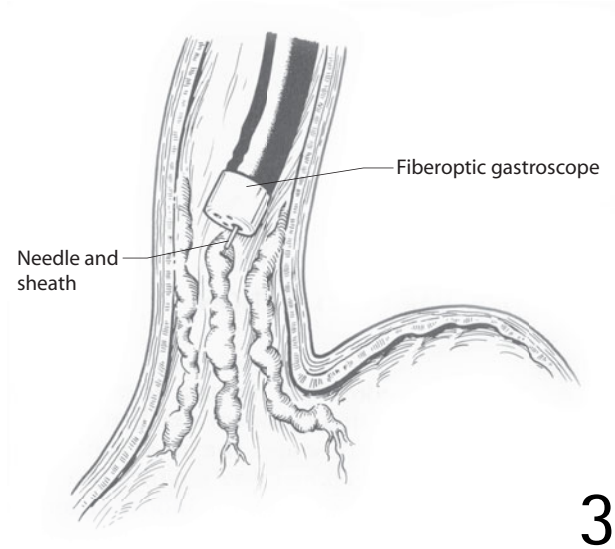
should be performed to assess the source of bleeding and potentially to provide therapeutic interventions. Even in patients with cirrhosis, up to 50 percent of upper gastrointestinal bleeding is from sources other than varices (gastritis, peptic ulcer disease, Mallory–Weiss tears).

As part of the work-up, portal venous anatomy should be assessed in all patients by Doppler ultrasound. Ultrasonography should show the main portal vein, superior mesenteric vein (SMV), splenic vein, and intrahepatic veins. Although suitable to demonstrate patency and flow, patients being considered for shunting should be also evaluated by angiography. Both the arterial and venous anatomy should be visualized. Injection of the superior mesenteric artery and splenic artery with venous phase images will identify the necessary major splanchnic veins, and venography of the IVC and left renal vein is required, as these veins will be used for various shunting procedures. Magnetic resonance angiography (MRA) is now a non-invasive alternative to traditional angiography. The surgeon should work closely with the radiologists to ensure that the necessary anatomy is clearly demonstrated.

OPERATIONS

Endosclerosis

3 Originally described using a slotted, rigid esophagoscope, most sclerotherapy is currently performed with a flexible endoscope. The advantages of flexible endoscopy are the superior optics and the ability to visualize the stomach and duodenum in order to assess further the source of hemorrhage. Most of these procedures are performed under general anesthesia to protect the airway during the operation. A variety of sclerosing agents have been used in children, including 5 percent ethanolamine, 1 percent tetradecyl sulfate, and 5 percent sodium morrhuate.



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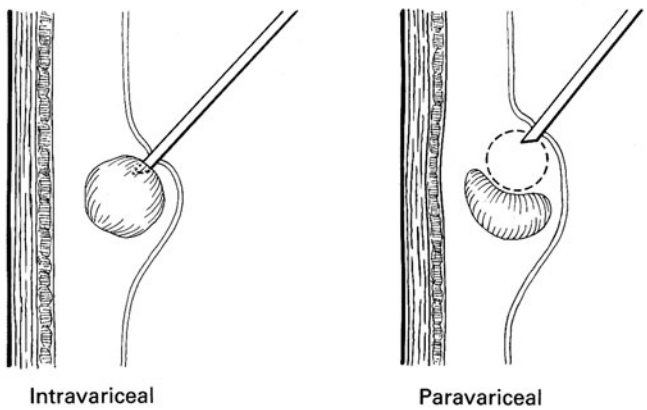

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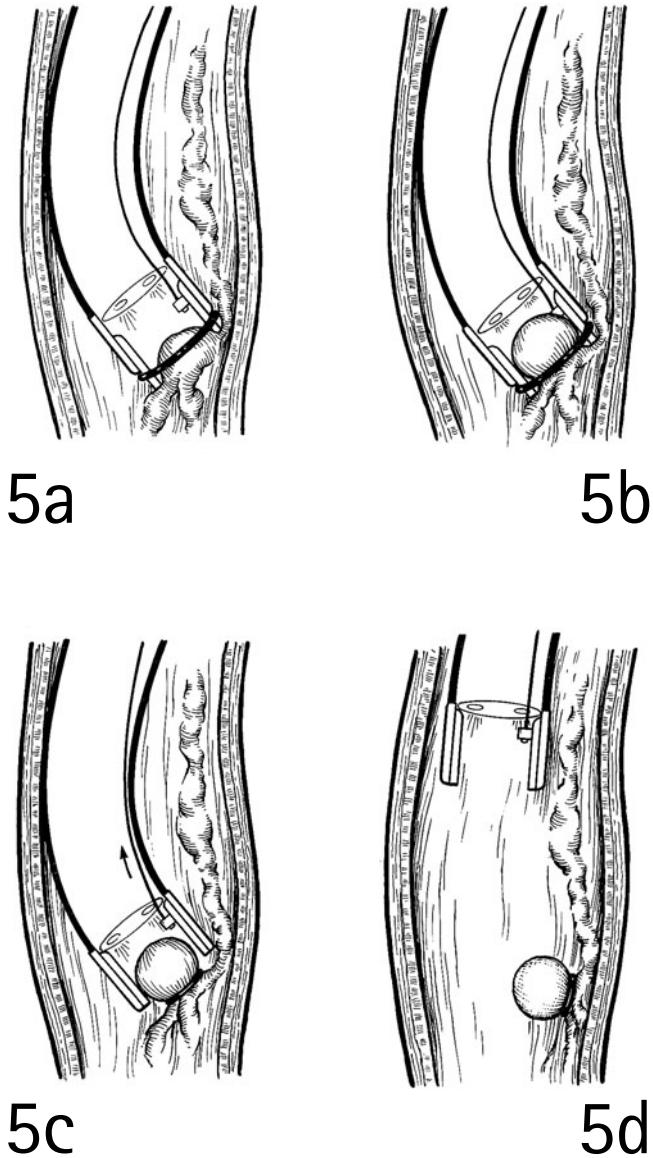
4 Both intravariceal and paravariceal techniques have been reported. We prefer direct intravariceal injection because it is associated with better control of acute variceal hemorrhage. Usually no more than three varices are injected at each session, and sclerosis is repeated every few weeks until the varices are obliterated. The interval between endoscopic sessions can then be gradually lengthened up to annual surveillance examinations.

Variceal banding

5a–d Endoscopic variceal band ligation, first reported in 1989, utilizes mechanical ligation and strangulation of varices with elastic O-rings. The device consists of two fitted cylinders attached to the tip of a standard flexible endoscope. The inner cylinder has the small elastic O-ring stretched over the end, which is released as the inner cylinder is drawn into the hood by a tripwire running through the biopsy channel of the endoscope. When the varix to be ligated is identified, it is drawn into the cylinder by applying suction. The O-ring is released by pulling on the tripwire, resulting in strangulation of the varix and thrombotic obliteration of the submucosal venous channels. Usually one to three elastic band ligatures are placed at each session. The bands and varices slough off after 5–7 days. The procedure is repeated, similar to endosclerosis, until the varices are obliterated, and the frequency is gradually reduced to annual surveillance.



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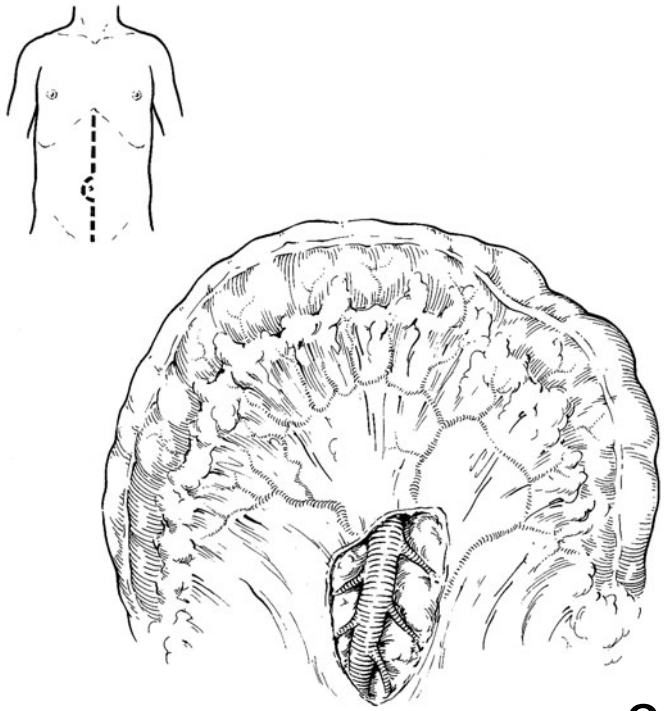
Mesocaval shunt

The classic mesocaval shunt, described by Clatworthy, was designed specifically for children in whom the portal vein is not usable because of PVT. It is easier to perform than some other shunts in children because the veins used are larger.

6 A midline incision is preferred to avoid interruption of venous channels in the abdominal wall that serve as important collaterals after division of the IVC. The transverse colon is retracted cephalad and the small intestine is retracted inferiorly. A vertical incision is made in the mesentery of the small intestine over the SMV, which lies to the right of the arterial pulsation. Identification of the SMV can be aided by following the veins in the transverse mesocolon. Once located, the SMV is dissected free for about 5 cm below its passage behind the pancreas.



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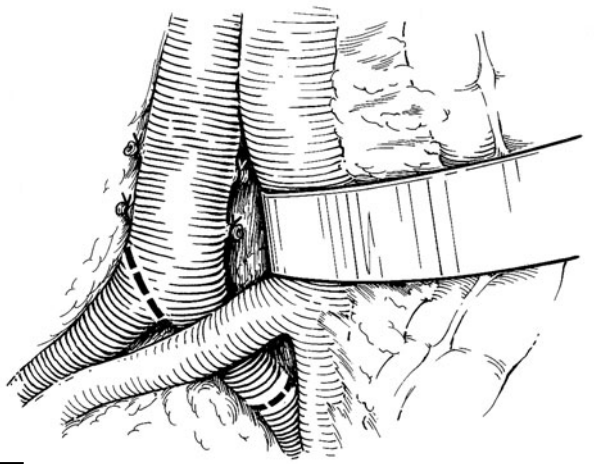

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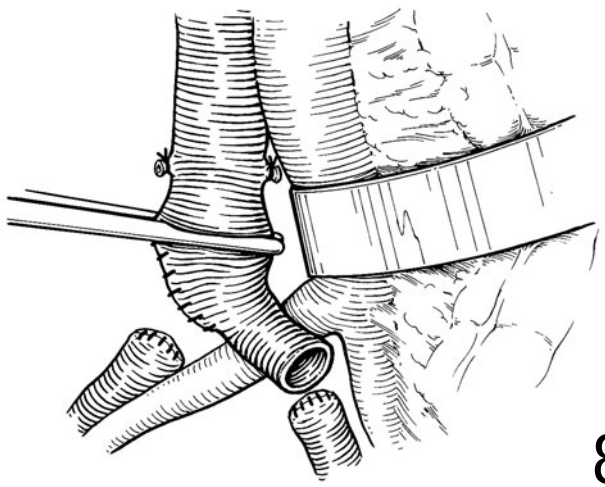
7 Next, the IVC is exposed by mobilizing the right colon. The duodenum is reflected medially by a Kocher maneuver to expose the junction of the renal veins and IVC. The IVC is freed from the renal veins to the bifurcation. Individual lumbar veins are ligated. A tunnel is made in the posterior mesentery, which is often thick and edematous, to reach the SMV.

8 At this stage, a critical decision must be made about the length of the IVC needed to reach to the SMV. Often it is advisable to gain additional length by dividing the left iliac vein at some distance below the bifurcation of the IVC. The right iliac vein is over-sewn flush with the bifurcation. The cava/iliac vein is then brought through the mesenteric tunnel and fashioned to appropriate length.



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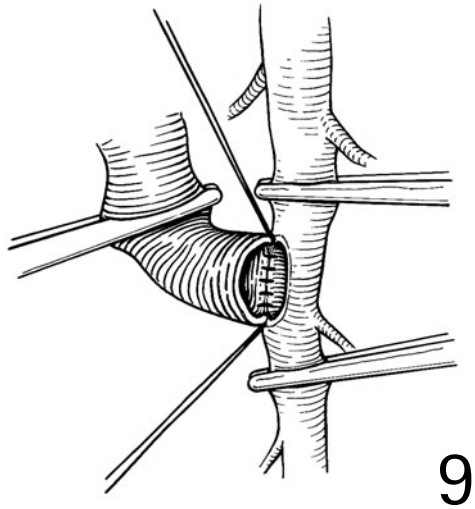
9 A small ellipse is cut in the SMV before anastomosis with fine, continuous monofilament suture (6/0 polypropylene). Special attention to alignment of the venotomy and anastomosis is necessary to prevent kinking. The retroperitoneum and the mesentery are re-approximated with a few absorbable sutures.



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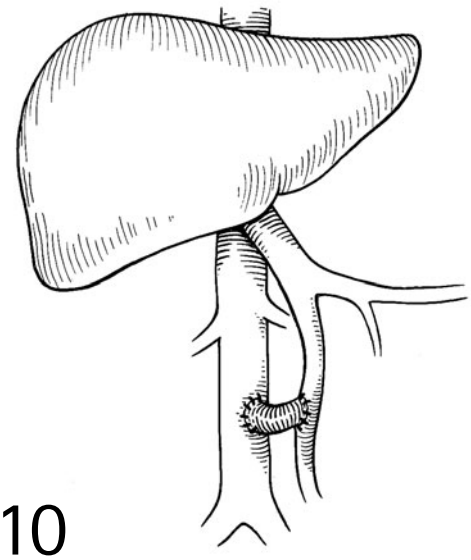

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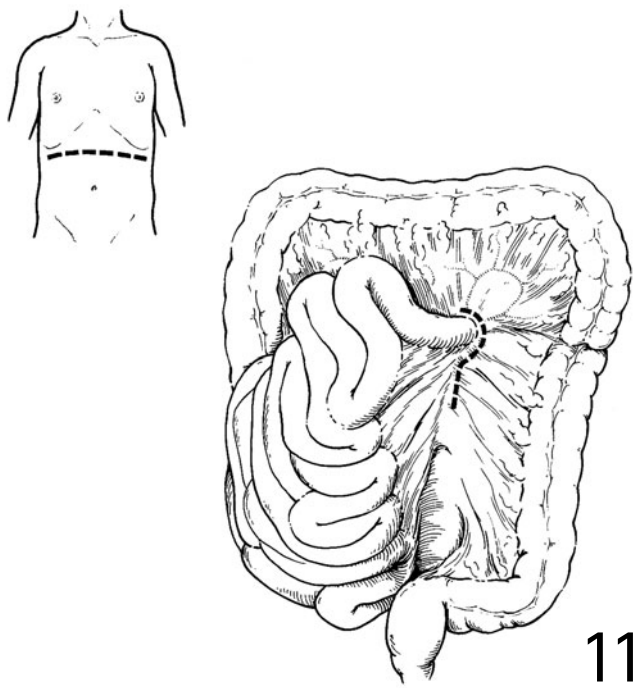
10 Because Clatworthy's classic mesocaval shunt requires division of the IVC, it can result in swelling of the lower extremities. Although usually temporary, an alternative procedure avoids this disadvantage by utilizing an interposition graft between the SMV and the IVC. Synthetic vascular grafts have been used, but internal jugular vein offers the best chance of long-term patency. For wound closure, the abdomen is closed in a watertight fashion, without drains.

Side-to-side splenorenal shunt

11 A wide transverse upper abdominal incision is made. The transverse colon is retracted cephalad and the small intestine to the right to expose the base of the transverse mesocolon and duodenum. The ligament of Treitz is incised and the inferior mesenteric vein divided at its junction with the splenic vein. This allows the duodenojejunal junction to be swept cephalad and to the right. The operation is often modified to a splenectomy and central end-to-side splenorenal shunt in patients with very large spleens.



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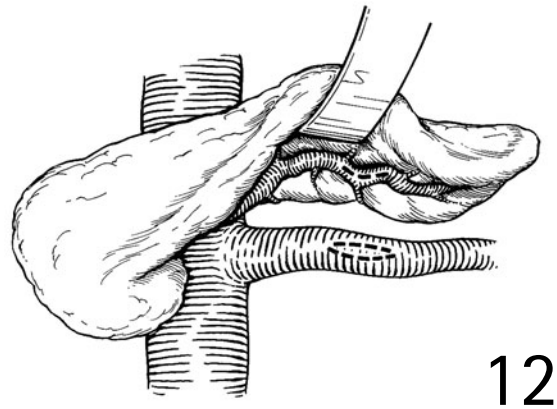
12 The left renal vein is exposed from the kidney hilum to the IVC. The left gonadal and left adrenal veins are ligated and divided. At the base of the transverse mesocolon, the inferior edge of the pancreas is dissected transversely. In longstanding portal hypertension, the retroperitoneum can be thick and edematous with numerous small collateral veins. With cephalad traction on the transverse mesocolon, the pancreas is rotated along its long axis to expose the splenic vein. The splenic vein is dissected out for a distance of 4–5 cm by ligation of small tributaries. Numerous small pancreatic veins must be meticulously ligated and divided to provide sufficient mobility of the splenic vein.



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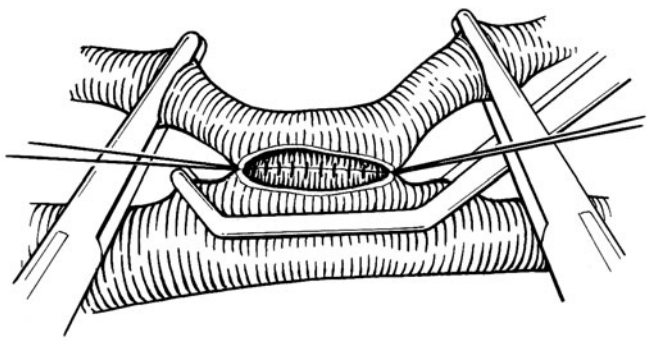

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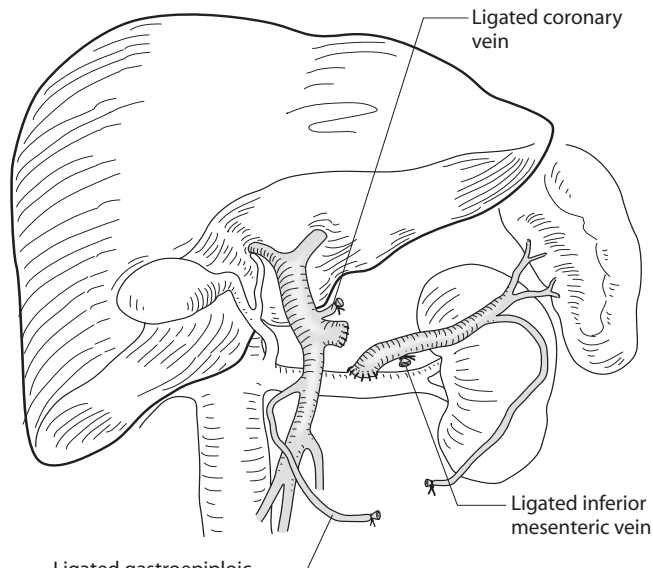
13 Vascular clamps are placed on both the splenic and the left renal veins. The splenic vein is opened transversely in its most dependent portion, extending into the stump of the inferior mesenteric vein if necessary, to create a larger anastomosis. The left renal vein is opened in a similar way. The anastomosis is performed with loop magnification using fine monofilament suture (6/0 polypropylene). The posterior wall is completed first from the inside, then the anterior wall. The completed anastomosis should be from 1.5 cm to 2.5 cm in length. The retroperitoneum is closed with absorbable sutures and the incision is closed in layers without drains.

Distal splenorenal shunt

14 The distal splenorenal shunt (Warren's shunt) is a selective shunt. The portal circulation is divided into two components: one maintains antegrade portal flow toward the liver via the SMV, and the other shunts flow away from the esophageal varices to the short gastrics then through the splenic vein into the renal vein.



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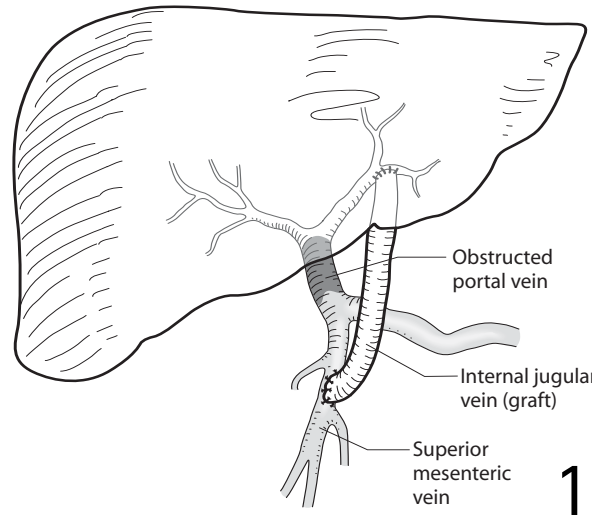
Through a generous midline or bilateral subcostal incision, the body of the pancreas is mobilized in a fashion similar to that used for a central splenorenal shunt. Once mobilized, the splenic vein is divided just before its junction with the SMV. The distal splenic vein is then swung inferiorly and anastomosed with fine vascular suture to the left renal vein without angulation. Another essential step in the performance of Warren's shunt is division of the left gastric (coronary) vein, the umbilical vein, and the gastroepiploic arcade. This completes the division of the portal circulation. The wound is closed in layers in a watertight fashion.

Mesentericoportal shunt

15 The last type of shunt that deserves mention is an SMV to intrahepatic left portal vein shunt (the Rex shunt). The shunt was originally developed for the treatment of portal vein blockage following liver transplant, but has been increasingly used for PVT in the absence of a transplant, with good success. Uniquely, this shunt restores flow from the portal circuit back to the liver via the intrahepatic left portal vein. This vein remains patent in about two-thirds of patients with cavernous transformation of the portal vein.



Both the neck and abdomen are prepped because a graft of internal jugular vein is often used to bridge the gap between the SMV and left portal vein. A wide transverse subcostal incision is preferred. The SMV is located where patent, as determined by preoperative angiography. Usually the SMV can be located near the confluence with the splenic vein behind the head of the pancreas. The left portal vein is located by following the ligamentum teres (umbilical vein remnant) into the umbilical fissure, Rex's recess. Once the length of the gap is known, a segment of internal jugular vein is harvested, usually from the left neck. Anastomosis to the SMV or confluence is then completed before connecting to the left branch of the portal vein. Both are performed with fine monofilament suture (6/0 polypropylene). Once flow is restored and hemostasis is assured, the wound is closed in layers, with no drains.

POSTOPERATIVE CARE

Children treated by endosclerosis for acute hemorrhage must be monitored closely for ongoing or recurrent bleeding. Once initial control has been achieved, follow-up endosclerosis may be performed on an ambulatory outpatient basis. Postoperatively, many patients experience low-grade fever and may have mild retrosternal discomfort. Severe chest pain and hyperpyrexia may indicate esophageal perforation (rare) and should initiate chest radiography and a contrast study. Ulceration at the injection site is common and is sometimes associated with mild self-limited hemorrhage a few days after the procedure. Because of the increased incidence of major complications and of bleeding, variceal banding has become the preferred technique. Postoperative fever, chest pain, and delayed bleeding are less common than with endosclerosis. The only perforation that has occurred in the author's experience was caused by the use of an over-tube, which tore the cervical esophagus; subsequently, the use of an over-tube in pediatric patients was abandoned.

For all portosystemic shunt procedures, nasogastric decompression should be maintained until normal bowel function has returned. Peptic ulcer prophylaxis is routine. Early ambulation and the use of elastic stockings may prevent lower extremity edema, which is seen occasionally after end-to-side mesocaval shunts. Vitamin K supplementation may be indicated based on prothrombin times. Dietary protein should initially be restricted and advanced only gradually to prevent hepatic encephalopathy. Ascites, which is common, may require salt restriction, diuretics, and the administration of exogenous albumin. Most surgeons use some type of anticoagulation postoperatively. We begin an infusion of dextran 40 immediately postoperatively and then administer aspirin (81 mg) daily for the antiplatelet effect.

OUTCOME

The author's results and those of others have shown good control of variceal hemorrhage using endosclerosis with reg-

ular surveillance sclerosis as needed to eradicate varices. Especially in children with portal vein obstruction, the tendency toward variceal hemorrhage decreases with time, as spontaneous natural retroperitoneal shunts develop. Death is rare, except when related to cirrhosis and liver failure. Some authors have reported alterations in esophageal motility, esophageal strictures, and even esophageal cancer after repeated endosclerosis. The control of hemorrhage and eradication of varices obtained using endoscopic variceal ligation have been equal to or better than those obtained using endoscopic sclerotherapy. The authors have not encountered the development of strictures or motility disturbances using endoscopic variceal ligation, and now use the technique almost exclusively for endoscopic treatment of variceal hemorrhage.

A properly performed portosystemic shunt is highly effective in controlling bleeding varices; however, because of the size of the vessels and therefore the propensity to thrombosis, re-bleeding rates of 10–25 percent have been reported. The Rex shunt restores normal portal flow to the liver, and therefore, with the distal splenorenal shunt, carries the theoretical advantage of decreased encephalopathy. The incidence of encephalopathy in children is difficult to estimate because different standards have been used to determine its presence and severity. In children with extrahepatic portal vein thrombosis, the risk of encephalopathy is reported to be low, but in the cirrhotic group encephalopathy is directly related to liver synthetic function. The long-term prognosis for children undergoing portosystemic shunts also largely depends on the severity of hepatic dysfunction. Probably those patients with advanced cirrhosis are best referred for liver transplantation and esophageal variceal bleeding managed by endoscopic means. For those patients with PVT, variceal bleeding can be successfully managed by endoscopic means, with low morbidity but with repeated visits and anesthetics. Shunts are associated with some surgical morbidity and mortality but provide good control and usually improve growth retardation and hypersplenism. The most important factor in the choice of management is the expertise and experience available. Thoughtful application of the various options can decrease the complications and mortality associated with variceal hemorrhage from portal hypertension.

FURTHER READING

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Laparoscopic cholecystectomy

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HISTORY

Cholelithiasis is a relatively uncommon condition in childhood except when it is associated with increased hemolysis such as congenital spherocytosis or sickle-cell anemia. Open cholecystectomy was the gold standard for the removal of a diseased gallbladder until 1987, when the French gynecologist Mouret described laparoscopic cholecystectomy in a human. The report was soon followed by similar reports of successful laser laparoscopic cholecystectomy by Reddick and Olsen in North America and Dubois and Perissat in France. These reports saw the explosion of advanced laparoscopic surgery, which has established laparoscopic cholecystectomy today as the gold standard and the operation of choice for the removal of a diseased gallbladder.

There are now two commonly recognized methods of performing a cholecystectomy, the French method (between the legs) and the America method, as described by Reddick and Olsen.

DIAGNOSIS AND ETIOLOGY

The diagnosis of cholelithiasis is usually made on abdominal ultrasound performed for the investigation of abdominal pain in childhood. Another common presentation is in children presenting with hemolytic problems such as spherocytosis or sickle-cell anemia, where the diagnosis is made as part of the investigative work-up prior to splenectomy.

PREOPERATIVE ASSESSMENT

A high-resolution ultrasound examination should be performed in all children with cholelithiasis. The presence of a dilated common bile duct should alert one to the possibility of stones in the common bile duct. Jaundice in a child with hemolytic disease does not necessarily indicate the presence of stones in the common bile duct, and may simply be a manifestation of significant ongoing hemolysis.

If a dilated common bile duct is seen on ultrasound examination, it is necessary to exclude the presence of a stone in the duct. The preferred method of investigation is to perform magnetic resonance cholangiopancreatography (MRCP). If a calculus is found, our preference is to proceed with endoscopic retrograde cholangiopancreatography (ERCP) and sphincterotomy in older children, prior to laparoscopic cholecystectomy.

Anesthesia

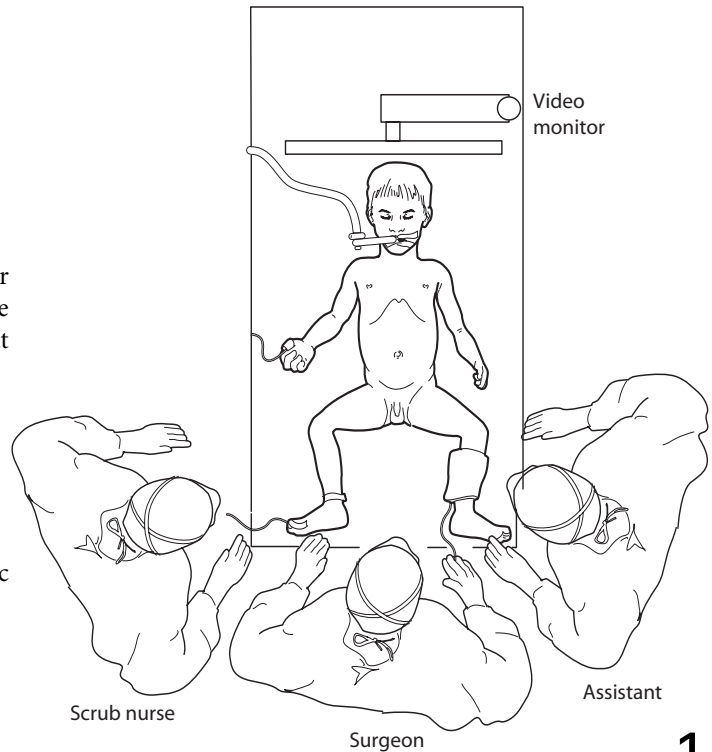
General anesthesia with full relaxation and endotracheal intubation is mandatory.

A 10-Fr nasogastric tube is inserted to decompress the stomach and ensure that it is kept empty.

OPERATION

Patient positioning

1 The patient is positioned at the foot of the bed as per fundoplication and the video monitor is placed at the head of the table. The patient should be tilted head up about 20°.



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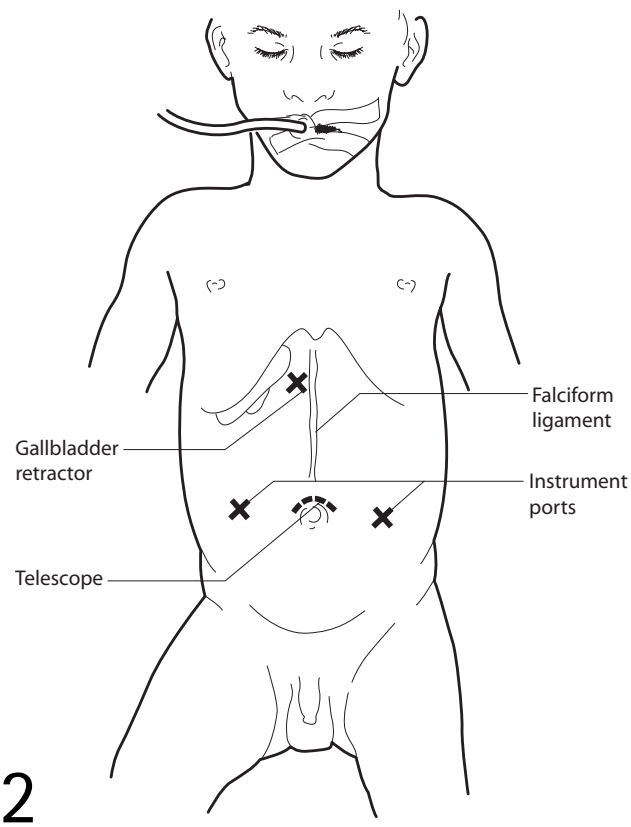
Laparoscopic instrumentation

The following are the instruments required for laparoscopic cholecystectomy.

- 0° 5 mm telescope.
- 11 mm Hasson trocar and cannula.
- Three 6 mm instrument ports.
- Liver retractor.
- Tissue graspers.
- Monopolar hook diathermy.
- Bipolar scissors.
- Bipolar diathermy forceps.
- Endoscopic clip applicators.



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Port placements

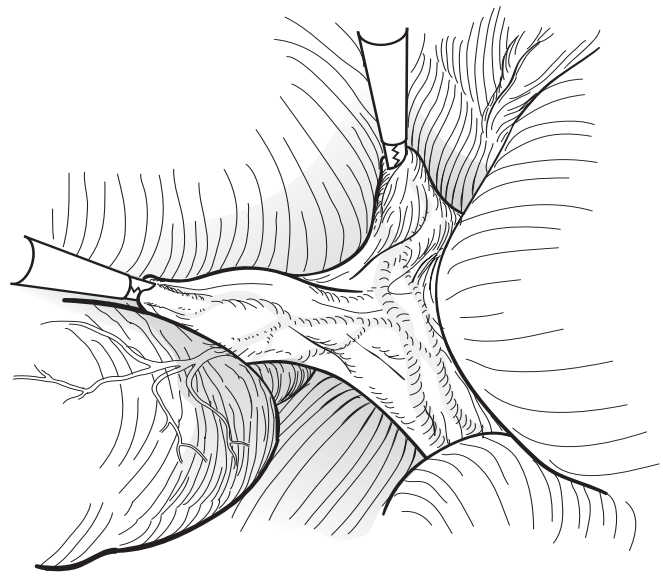
2 An open laparoscopy is performed to insert a Hasson cannula in the umbilicus. Two further instrument ports are inserted under direct endoscopic visualization, and a further port is inserted in the epigastrium. In small children, it may be preferable to place the instrument ports lower, parallel to the umbilical port. The liver is often bigger and overhangs the costal margin, and placing instrument ports in the conventional position in the upper quadrant will severely limit the ability to manipulate the instruments within the abdominal cavity.

Technical steps

The French method is described. Ensure that the stomach is completely empty, as a full stomach not only encroaches on the dissection field, but will also distend the duodenum, with the attendant risk of duodenal injury during the dissection of Calot's triangle.

GRASPING THE GALLBLADDER

3 The gallbladder should be easily visualized within its fossa. Hartmann's pouch should then be grasped with a toothed, ratcheted grasper and retracted towards the right hip. A retractor is then placed in the epigastric port to retract the liver away from Hartmann's pouch to display Calot's triangle, which should be splayed apart.



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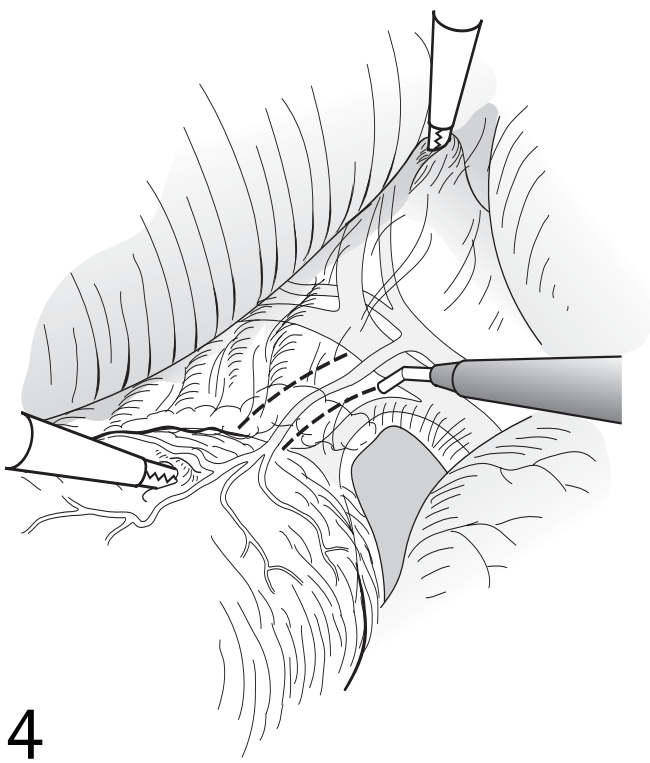

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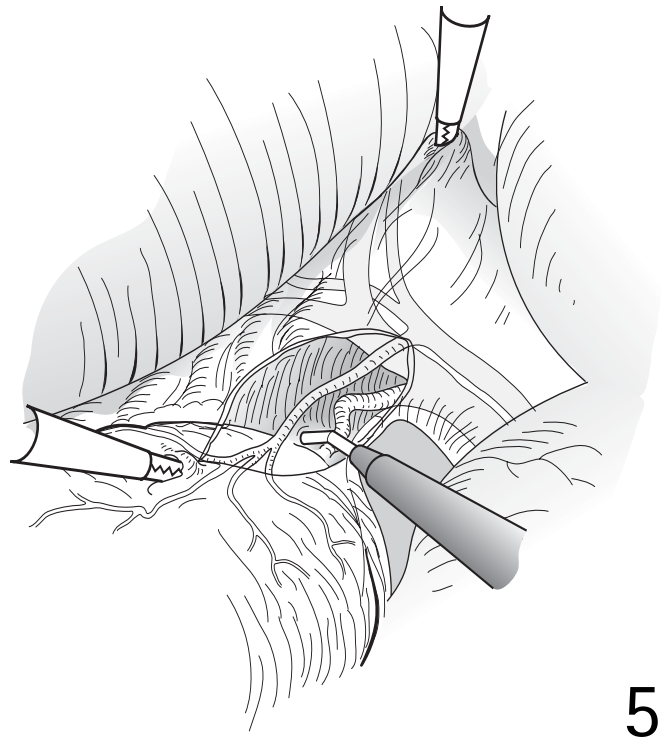
EXPOSING CALOT'S TRIANGLE

4 The peritoneum overlying the cystic duct should be opened with monopolar hook diathermy, taking care not to damage the duodenum or common bile duct. If employing hook diathermy, it is preferable to use the heel to push, or else to hook away from these structures to avoid inadvertent damage. The peritoneum covering the free edge of the cystic duct should be opened, as should the peritoneum covering the posterior part of Calot's triangle. This will facilitate the dissection of Calot's triangle. Constant countertraction must be maintained on Hartmann's pouch.

EXPOSING THE CYSTIC DUCT

5 The cystic duct and cystic artery should be exposed by dividing the respective overlying peritoneal lining. Hook diathermy can be used to incise the peritoneum between the upper margins of the cystic duct and the porta hepatic, and between the cystic duct and cystic artery to display Calot's triangle. A window is created between these two structures. Unlike conventional open cholecystectomy, it is *un*wise to expose the cystic duct at its junction with the common bile duct. Most reported cases of common bile duct injuries occur during this dissection.



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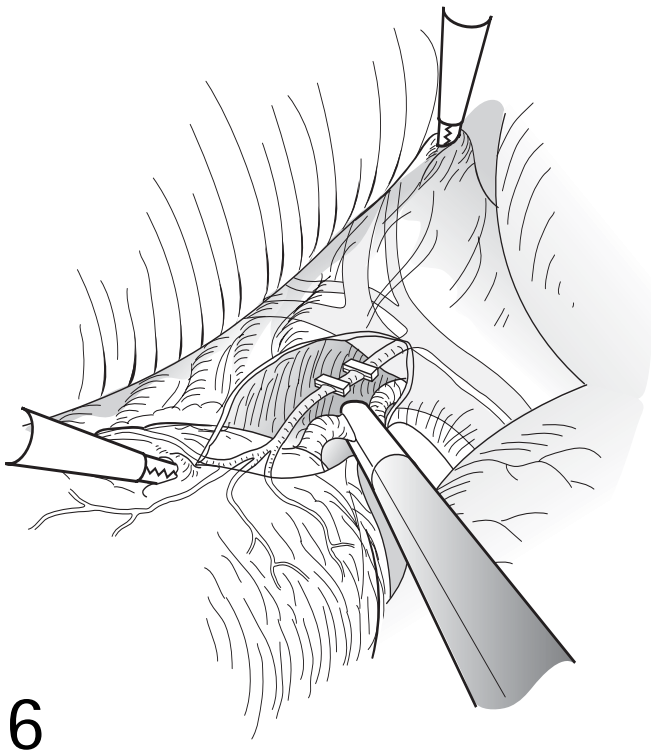
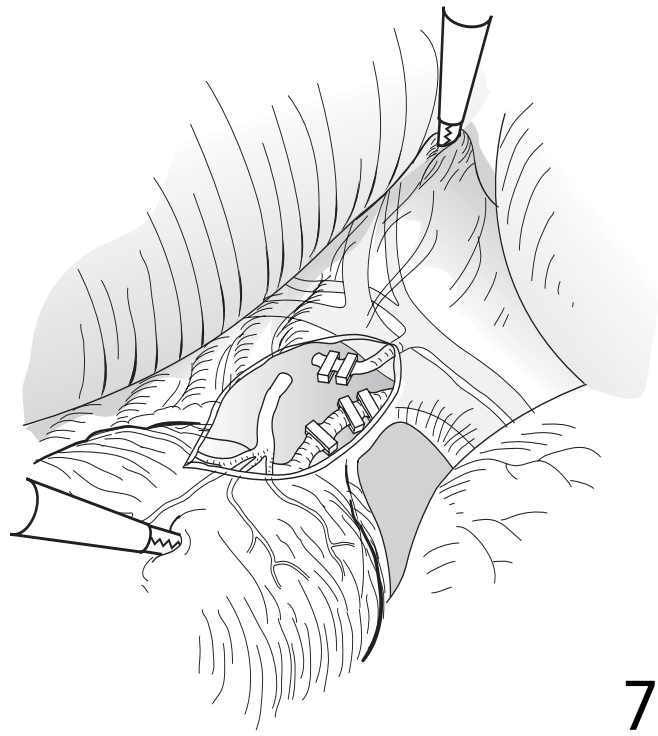


6

DIVIDING THE CYSTIC ARTERY AND CYSTIC DUCT

6 When an adequate length of cystic artery and duct has been exposed, the artery is clipped with two titanium clips on its proximal side. It is usually unnecessary to place a clip on the distal part of the cystic artery which is an end artery.

7 Three clips should be applied on the cystic duct, two on the 'stump', and one on the gallbladder side. The cystic artery should always be divided before the cystic duct, otherwise there is a risk of avulsing the cystic artery during manipulation of the cystic duct.



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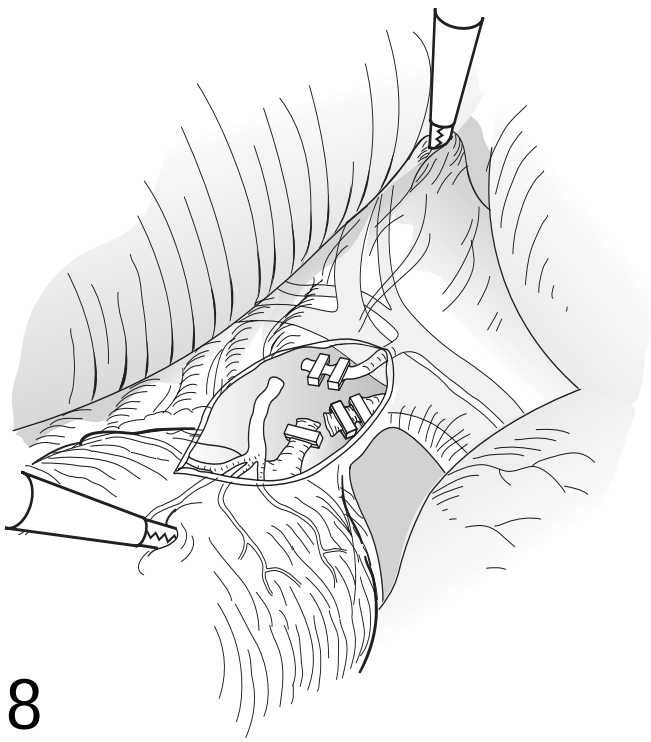
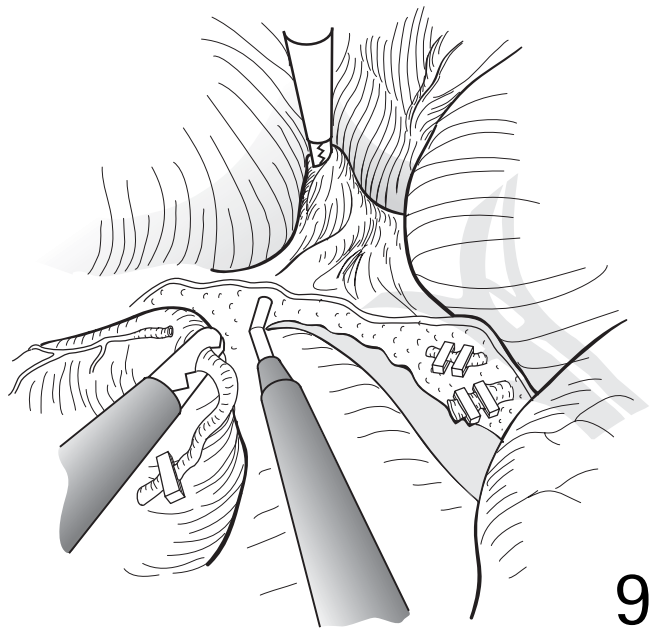


8

CHOLECYSTECTOMY

8 When the cystic artery and cystic ducts have been divided, the gallbladder can be removed from its fossa using monopolar hook diathermy.

9 It is important to maintain adequate countertraction on the free edge of the dissection, as this helps to develop the tissue plane between the gallbladder and its fossa.



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The gallbladder can then be parked on the diaphragmatic surface of the liver while checking the gallbladder fossa for bleeding. If there is bleeding, it may be wise to introduce a Redivac drain through the epigastric port site, after removing the trocar.

A small endoscopic bag can then be introduced through the 11 mm Hasson port, by transferring the 5 mm telescope in to the left-sided instrument port. This will allow you to insert the bag into the larger Hasson port to manipulate the gallbladder into the bag, which can then be removed through the umbilical port.

WOUND CLOSURE

The Hasson port site is closed with a purse-string suture, usually the one used to secure the Hasson port at open laparoscopy. The fascia of all instrument ports should be closed with a single absorbable suture and the skin can be closed with cyano-acrylic glue and left unprotected.

POSTOPERATIVE CARE

Most patients will be able to tolerate free fluids when fully recovered from the effects of the anesthesia. Strong oral analgesia is usually required for about 2 days, and patients can be discharged when ambulant and tolerating food.

ACKNOWLEDGEMENT

Dr A Tien, for assistance with the compilation of the further reading list.

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65

Surgery for biliary atresia

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HISTORY

John Thomson from Edinburgh described an infant dying from cirrhosis secondary to congenital biliary obstruction in 1891. Surgical exploration was advocated from the 1930s, and the terms 'correctable' and 'non-correctable' became prevalent to describe what could be done with a conventional surgical operation (e.g., hepaticojejunostomy). However, as most cases of biliary atresia were anatomically 'non-correctable', their outlook was poor and true survivors were exceptional.

During the 1950s and 1960s, Morio Kasai, a Japanese surgeon, developed a more radical approach to the dissection, exposing residual microscopic bile ductules within the apparently solid parts of the porta hepatis. This operation (portoenterostomy) resulted in a much larger proportion of children who, following the restoration of bile flow, lost their jaundice. Nevertheless, most still developed chronic liver disease. Liver transplantation became a practical option during the 1980s, initially for children with life-threatening complications of liver disease (e.g., portal hypertension) and then for infants of whatever age who had had no response to a Kasai operation.

PRINCIPLES AND JUSTIFICATION

Biliary atresia remains a rare disease, with a frequency of between 1 in 10 000 and 1 in 18 000 live births. It is more common in Japan and China than in Europe or the USA, although the reasons are not apparent. There is a slight female preponderance in most large series.

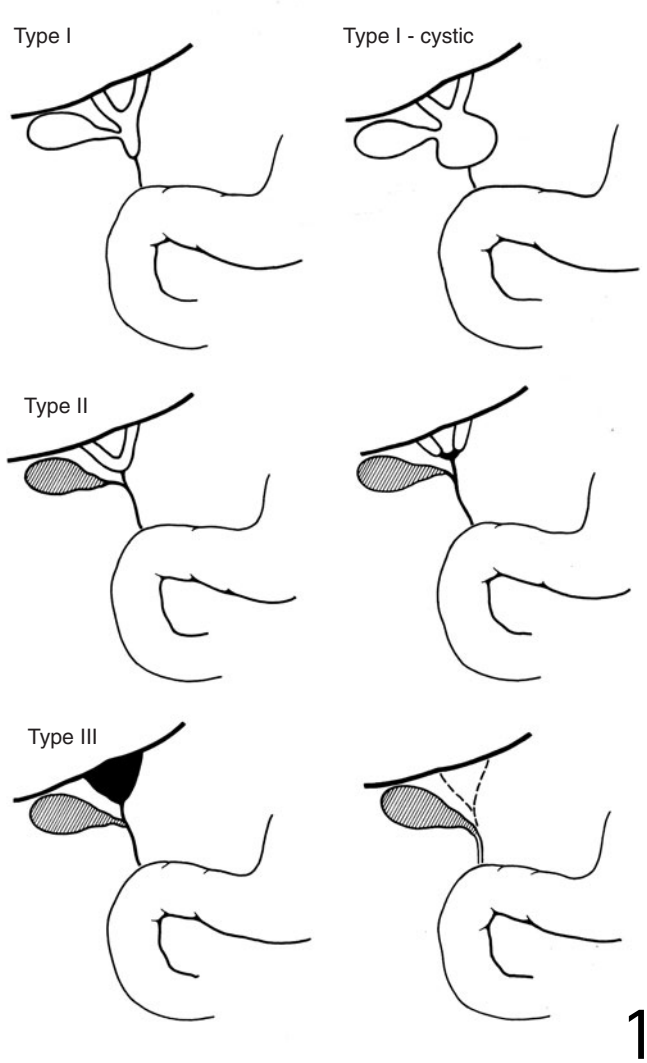
The etiology of biliary atresia is not known, although several hypotheses have been advanced. For instance, there is a consistent association (approximately 10 percent of all cases) with other non-biliary anomalies such as polysplenia, asplenia, cardiac malformation, situs inversus, absence of the inferior vena cava (IVC), preduodenal portal vein and malrotation, for which the term *biliary atresia splenic malformation* has been used. Such cases may result from some 'insult' within the first trimester of pregnancy that causes abnormal development of susceptible organ systems. Although such 'insults' are hard to define, maternal diabetes, maternal thyrotoxicosis, and drug abuse etc. have been reported. In most cases, only the biliary tree is abnormal, and no obvious prenatal cause is identified. This has led to speculation that a perinatal hepatotropic virus infection (e.g., Rotavirus, REOvirus, cytomegalovirus) has been able to damage an otherwise normally developed biliary tree.

Biliary atresia is a cholangiopathy of the extrahepatic and intrahepatic parts of the biliary tree, which, if untreated, leads to hepatic fibrosis and ultimately cirrhosis. The histological appearance of the liver is characterized by portal tract edema, bile duct plugging and proliferation, a small-cell infiltrate, and a variable amount of giant-cell formation. Bridging fibrosis is a late feature but in some cases can be seen at diagnosis.

1 The lumen of the extrahepatic duct is obliterated at a variable level and this forms the basis for the commonest classification in use. Thus, there are three main types of biliary atresia:

- *Type 1* (5 percent), where the level of obstruction is within the common bile duct (the gallbladder therefore contains bile);
- *Type 2* (3 percent), where the level is within the common hepatic duct;
- Type 3, the commonest (>90 percent), where there is no visible bile-containing proximal lumen and the obstruction is within the porta hepatis.

Cystic change can be seen in about 5 percent of cases, of whatever type. Some contain mucus, while some contain bile and invariably cause diagnostic confusion with a choledochal cyst. In such cases, the wall is thickened and communicates poorly with abnormal, non-dilated intrahepatic ducts, which are demonstrable on cholangiography.



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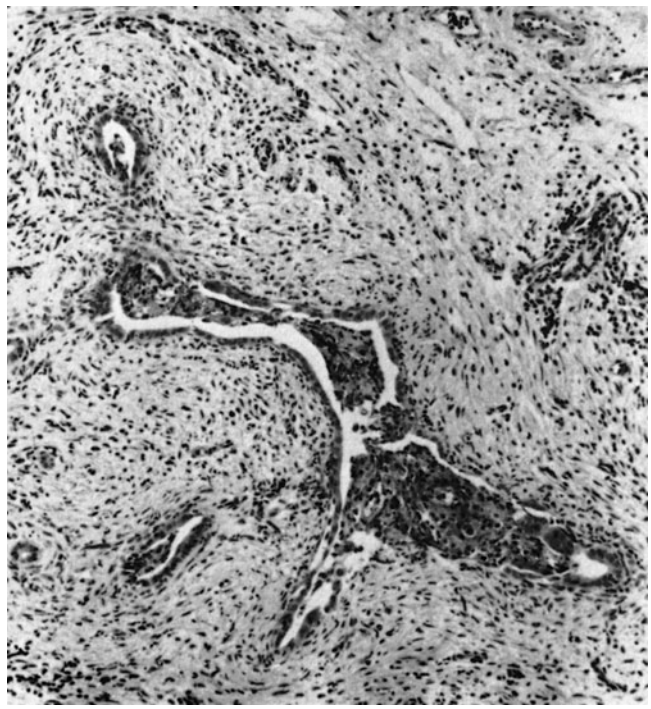
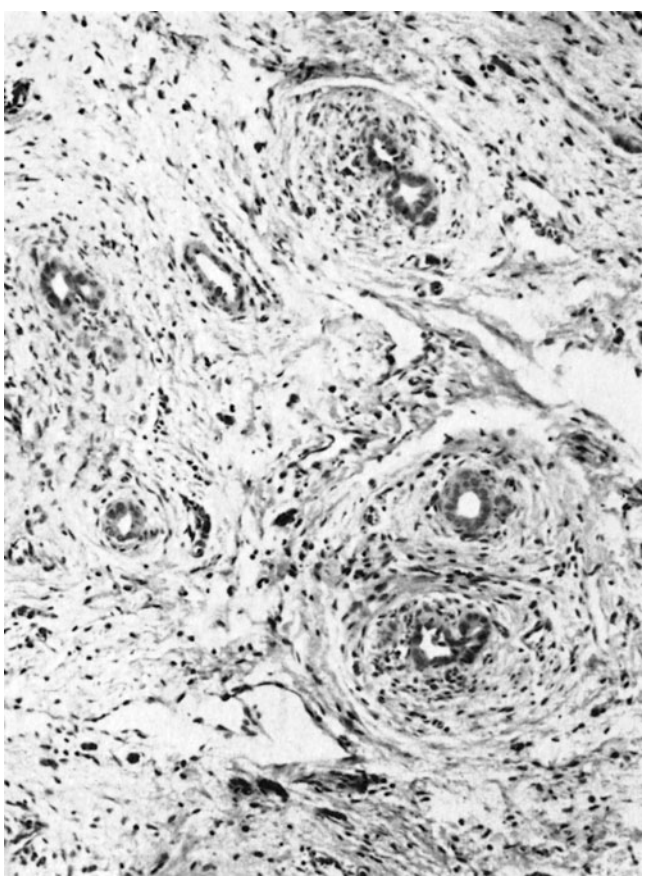


2 The porta hepatis of the commonest variant, Type 3, contains microscopic epithelial-lined bile ductules up to 300 μm in diameter. Partial destruction and desquamation of the epithelium has occurred, and the ductules are surrounded by fibrous tissue, containing inflammatory cells.



3 Large ductules are often absent, but serial sectioning has shown that even small channels may communicate with intrahepatic ducts.

PREOPERATIVE ASSESSMENT AND PREPARATION

All infants with biliary atresia will be clinically jaundiced, and have pale stools and dark urine when looked for. This is due to the inability to excrete conjugated (i.e., water-soluble) bilirubin into the gastrointestinal tract, which is then excreted into the urine, causing its color to darken. Such alternative pathways of bilirubin excretion are more developed (or perhaps better retained) in the neonate, and very high levels of bilirubin (>300 μmol/L) are not a feature (as might be seen in adults with complete biliary obstruction). Some infants will have had an abnormal antenatal maternal ultrasound scan (approximately 5 percent), because of cystic change in the biliary tree.

The differential diagnosis of conjugated jaundice in infants is long and can be complicated to work out. Surgical causes, other than biliary atresia, are uncommon, but include obstructed choledochal malformation, spontaneous perforation of the bile duct, and the inspissated bile syndrome (as seen particularly in preterm infants with other problems). The medical causes include biliary hypoplasia (as seen in Alagille's syndrome), neonatal hepatitis, α-1-antitrypsin deficiency, giant-cell hepatitis, and cystic fibrosis.

The diagnostic work-up should always include ultrasonography, biochemical exclusion of α-1-antitrypsin deficiency, and cystic fibrosis, and (in our institution) a percutaneous liver biopsy. Using this, about 80 percent will have a positive diagnosis prior to laparotomy. Other techniques that have been described include endoscopic retrograde cholangiopancreatography (ERCP), percutaneous cholangiography, duodenal intubation, and measurement of bile. These may be useful (or, more likely, more practical) in selected cases and other centers. Currently magnetic resonance cholangiopancreatography (MRCP) is not detailed enough to diagnose biliary atresia confidently, although it has shown promise for the more obvious structural problems such as a choledochal cyst. Radio-isotope hepatobiliary imaging (e.g., using iminodiacetic acid derivatives) was formerly a popular investigation in distinguishing biliary atresia from neonatal hepatitis, but it can be difficult to interpret and is not in current use as a diagnostic tool in our center.

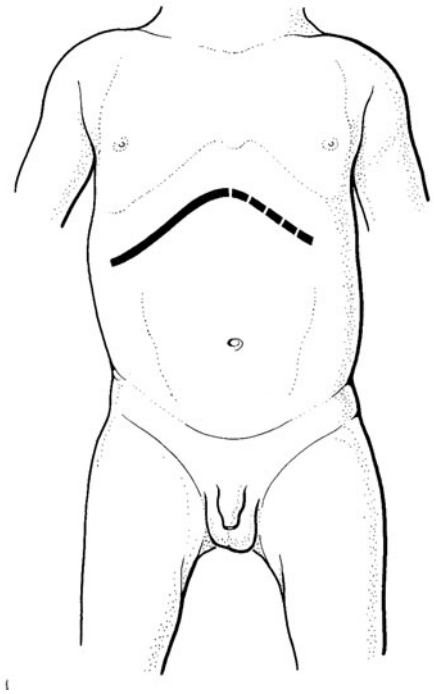
Most infants presenting within 80 days will not show clinical features of cirrhosis or irretrievable liver damage (e.g., gross ascites, nodularity on ultrasonography, or histological cirrhosis on liver biopsy) and should undergo laparotomy with the intention of performing a Kasai portoenterostomy. Others who do show these features, may be considered for liver transplantation as a primary procedure. Nevertheless, it should be realized that prognosis is difficult to predict with any certainty, and even a poorly functioning Kasai operation may delay the need for a donor organ.

The blood investigations should include coagulation tests (e.g., International Normalized Ratio, INR) to exclude a vitamin K-dependent coagulopathy, and all infants coming to surgery should have parenteral Vitamin K (phytomenadione 1.0 mg/day) supplementation. The choice of parenteral antibiotics depends on local policies, but should be broad spectrum with reasonable bile penetration (e.g., second-generation and third-generation cephalosporins). In our institution these are given intravenously for 5 days and then orally for a further 25 days.

OPERATION

The aim of the surgery is to excise all extrahepatic biliary remnants to allow a wide portoenterostomy reconstruction onto a portal plate denuded of all tissue. This should be the object not only in Type 3 biliary atresia, but also in those who do have a visible bile-containing proximal communication. It should be obvious, therefore, that frozen section, formerly used to confirm patent ductules, is not necessary because it should not be possible to resect any further biliary tissue.

4 A short right (or left if situs inversus) upper quadrant muscle-cutting incision should be performed initially to confirm the suspected diagnosis or, if not immediately obvious, to perform an on-table cholangiogram. If there is no bile, or only clear mucus, in a collapsed, atrophic gallbladder, it is invariably biliary atresia. In those circumstances where there is obvious bile in the gallbladder, an on-table cholangiogram is indicated.



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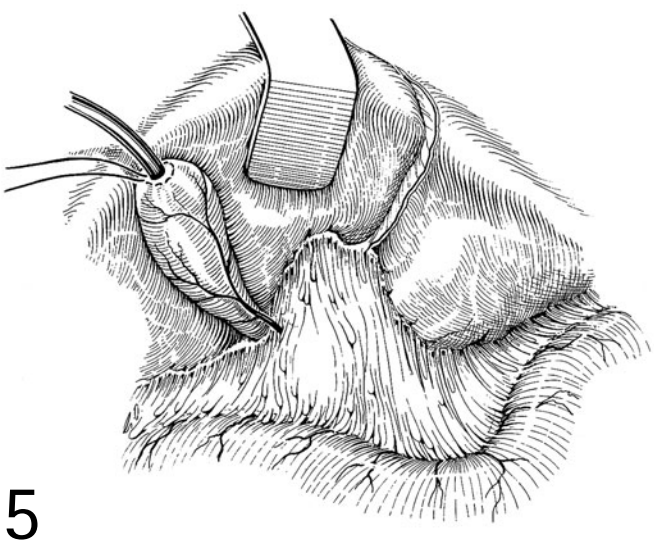

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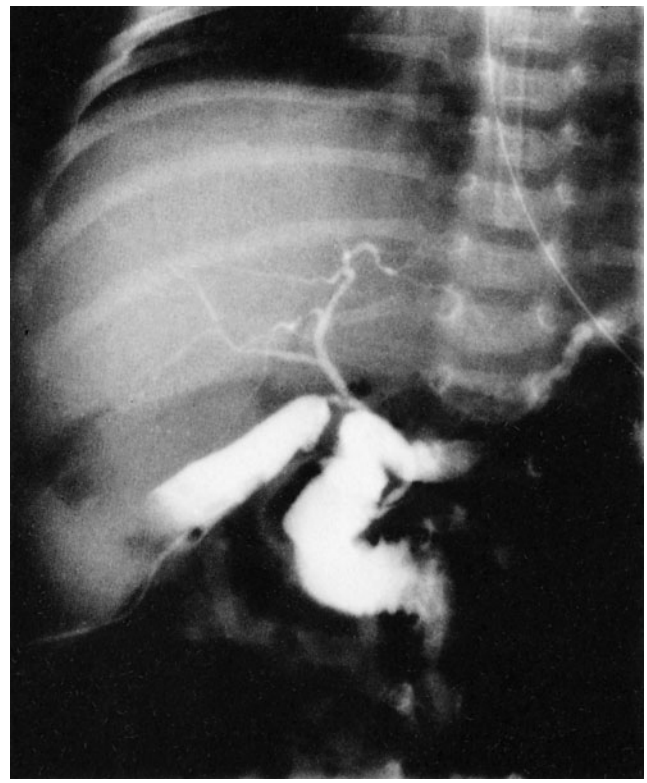
5 This is done by inserting a small feeding tube (4 Fr) into the gallbladder secured by a purse-string suture.

6 The demonstration of a patent common bile duct and intrahepatic ducts excludes a diagnosis of biliary atresia.



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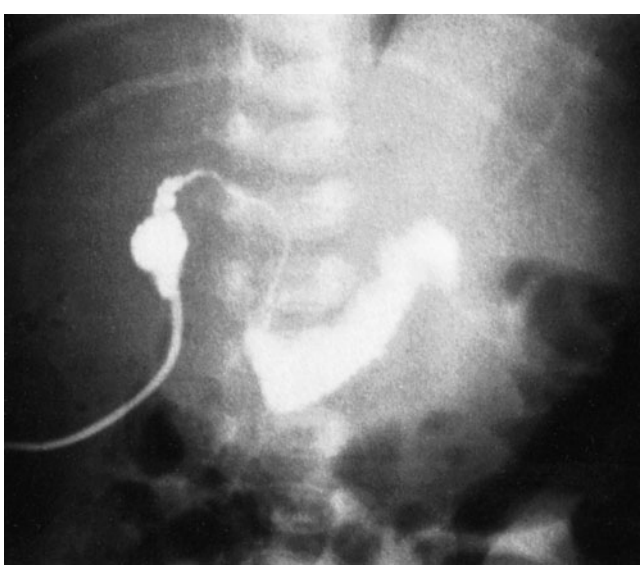

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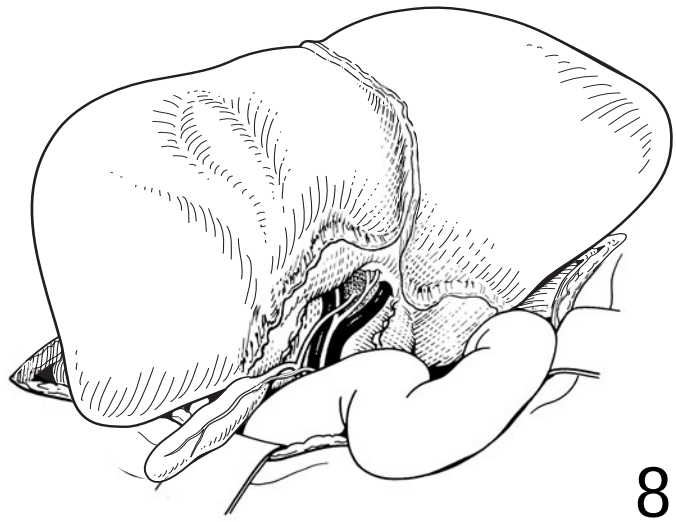
7 Sometimes proximal passage of contrast into intrahepatic ducts can be difficult to demonstrate, as typically it preferentially fills only the distal duct and duodenum. This can be prevented by a small vascular or 'bulldog' clamp on the common bile duct.

The incision can then be lengthened after confirmation of the diagnosis to cross the midline and ligate and divide the falciform ligament. The laparotomy should look carefully for other anomalies (e.g., polysplenia, preduodenal portal vein, absence of the inferior vena cava, and malrotation), which may alter the subsequent technique.

Mobilization of the liver

8 The liver should be fully mobilized by dividing the falciform ligament, coronary ligaments, and right and left triangular ligaments such that the organ can then be everted outside of the abdominal cavity. This is a crucial step, which allows full exposure of the portal hepatis and facilitates the subsequent detailed dissection. It is necessary to warn the anesthetist at this stage, as the maneuver impairs venous return to the heart by kinking the cava and will need an increase in intravenous volume support.



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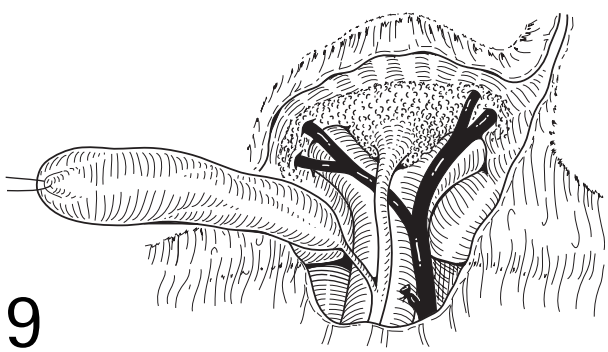


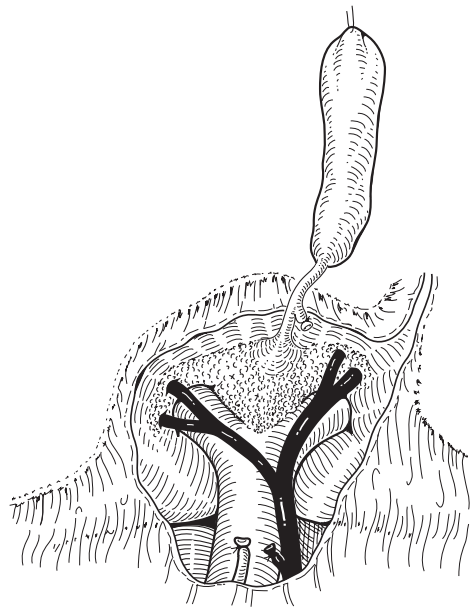
9

Mobilization of the gallbladder and bile ducts

9 A stay suture on the gallbladder allows its elevation, and it is then separated with bipolar diathermy off its bed. The peritoneum overlying the portal triad (the hilar plate) is then divided and the various vascular (hepatic artery and portal vein) and biliary structures positively identified. Parts of the biliary tract may be missing, but usually there is an intact biliary structure conforming to the usual arrangement of gallbladder, cystic duct, common hepatic and common bile ducts. The distal common bile duct is ligated and divided and the proximal part is elevated from underlying connective and lymphatic tissue. Following the divided end of the cystic artery should lead to identification of the right hepatic artery, which may be superficial or deep to the common duct and may need slinging.

Dissection and exposure of the porta hepatis

10 Gradual separation of the biliary remnant will lead proximally to the porta, and its relationship to the bifurcation of the portal vein will become clearer.



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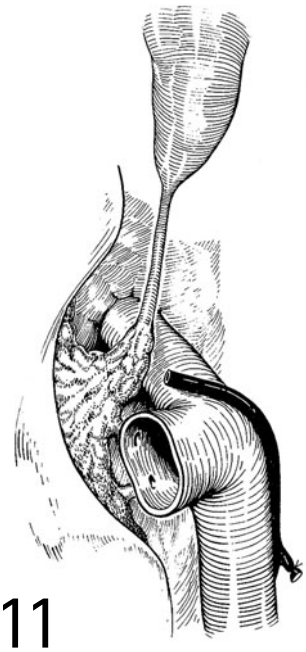


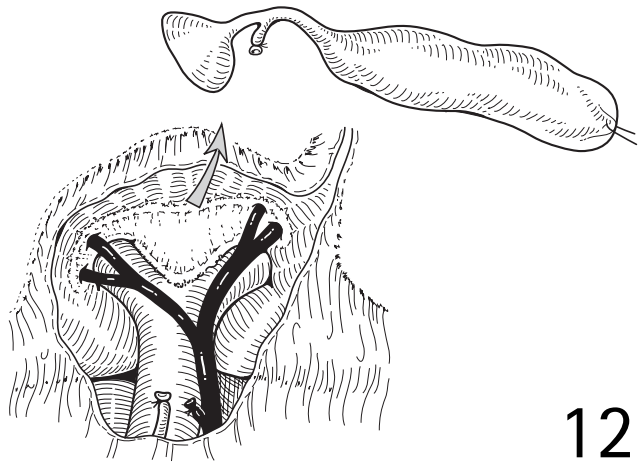
11

11 The portal vein bifurcation is gradually freed from the porta, typically by careful division of small veins crossing into the central portion.

In the posterior part of this dissection, the caudate lobe should become visible. The dissection is then extended to both right and left aspects. On the left, consideration should be given to exposing the recessus of Rex by dissecting the umbilical vein down to its origin from the left portal vein. An isthmus of liver parenchyma (from segment III to IV) may need division by coagulation diathermy to achieve this. This then becomes the proximal extent of the left-sided dissection. On the right side, remnant biliary tissue can be identified almost hemi-circumferentially around the right vascular pedicle, into the origin of the gallbladder fossa. Although some authors advocate slinging the right and left portal veins, this may compromise subsequent liver blood flow, and simple judicious retraction should give more than adequate exposure.

Excision of biliary remnants

12 The remnant can now be removed. Beginning at the right side and using a combination of scalpel and small, curved, sharp tenotomy scissors, a plane is developed between solid white biliary remnant and the underlying liver, flush with the capsule. The appearance at the transected portal plate should be almost translucent and denuded of tissue. Excising liver parenchyma itself does not improve bile drainage, presumably because any divided ductules simply become obliterated by subsequent scar tissue. Avoid untoward attempts at hemostasis of the porta by diathermy at this stage. The area should be packed and the liver returned to the abdominal cavity, which will relieve venous congestion.



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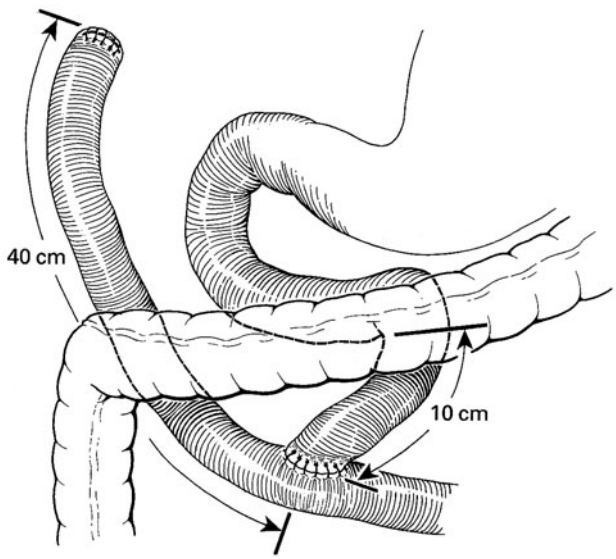
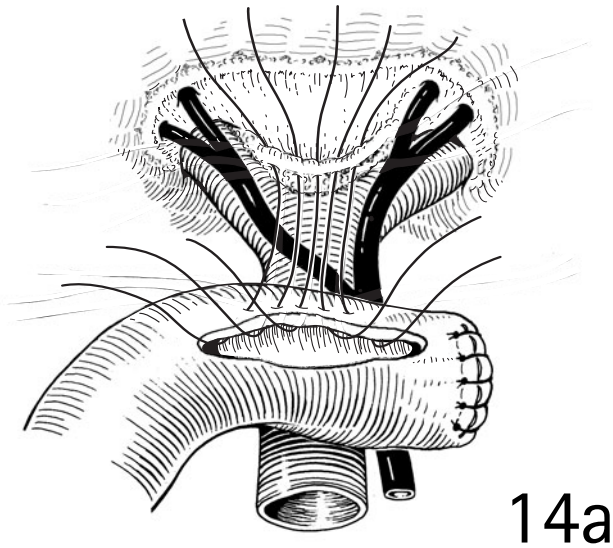


13

Formation of Roux loop

13 The duodenojejunal junction should be identified and a point about 10 cm from this freed to be the site of the jejunal anastomosis. The bowel is then divided using a stapled linear cutter, and a Roux loop, measured along the antimesenteric border at approximately 40 cm, constructed. This loop can then be brought through a right-sided window in the transverse mesocolon. An end-to-side jejunal anastomosis is usually constructed using whatever technique the operator is comfortable with.

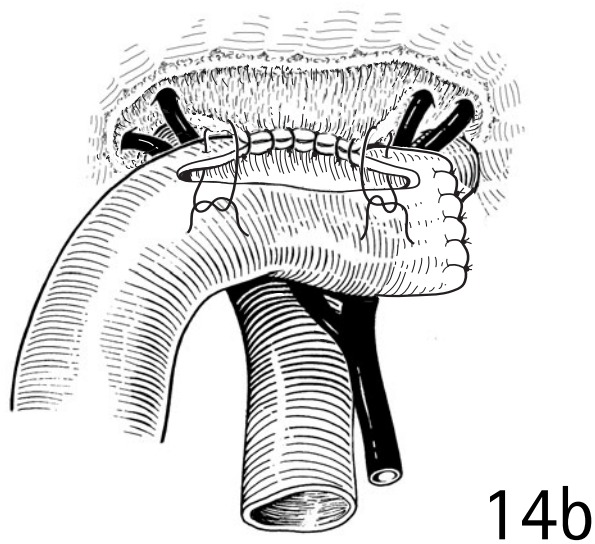
The end of the Roux loop should have enough length to reach the porta without undue tension.



14a

Portoenterostomy

14 The liver is again everted outside of the abdominal cavity. A non-crushing bowel clamp placed along the jejunal loop keeps this part steady, and an opening is made in the antimesenteric aspect about 1–2 cm from the stapled end.



14b

It is easier if the posterior row of sutures (e.g., 5/0 polydioxanone, PDS) is placed sequentially and held with mosquito clips until the row is complete. The jejunal loop can then be 'parachuted' down to the porta and the sutures tied. The anterior row is then completed. The anastomosis should be wide, measuring about 2 cm, if there has been an adequate dissection of right and left sides.

The mesocolic window around the Roux loop is closed, avoiding any kinking. A drain can be left, particularly if there has been more than a small quantity of ascites, but is not essential. The wound closure, however, needs to be secure and watertight.

Surgical alternatives

There have been many alternatives to the original description of the Kasai operation. The crucial part (viz. the portal dissection) has certainly become more extensive and widened on both right and left sides, although the *principle* of complete

excision has not been superseded. Most alternatives have included variations of the Roux loop, either involving the creation of stomas or interposed 'antireflux' valves. The aim of these has been to reduce postoperative cholangitis. None has stood the test of time, however, and a straightforward long (≥40 cm) Roux loop reconstruction is all that is required. Laparoscopic portoenterostomy has also been reported, but it is difficult to see any advantage, given that the only outcome that matters (restoration of bile flow, clearance of jaundice) is so crucially dependent on an appropriately radical dissection – difficult enough even when performed as an open operation.

POSTOPERATIVE CARE

Intravenous fluids and nasogastric aspiration are continued until the return of bowel function (approximately 3–4 days). Careful monitoring of blood glucose, electrolytes, and INR is important in the early phase. Liver biochemistry (including bilirubin) may well worsen in the first week whatever the eventual outcome, but by about the fourth week there should be a definite fall in bilirubin and consistently pigmented stools in those who will do well.

The role of phenobarbitone, corticosteroids, and ursodeoxycholic acid is unproven, although they are widely prescribed.

Strict attention to nutritional needs is important and all infants need regular vitamin supplementation. Medium-chain triglyceride (MCT) formula milk (e.g., Caprilon) is advocated to maximize calorie input and facilitate lipid absorption.

OUTCOME

There are many factors that will influence surgical outcome in biliary atresia. Some are unalterable (e.g., degree of cirrhosis at presentation, absence of, or paucity of, microscopic bile ductules at the level of section) and some are subject to change (e.g., surgical experience, untreated cholangitis). In large centers with experienced surgeons, about 50–60 percent of all infants will clear their jaundice and achieve a normal (<20 μmol/L) bilirubin level. These infants should do well and have a good-quality long-term survival with their native liver. In those with no effect from the Kasai operation (usually apparent within 2–3 months), active consideration should be given to early liver transplantation.

There is a relationship of surgical outcome to age at surgery (see Fig. 65.1). This seems to suggest that there is similar outcome up to 80–100 days of age, beyond which the expectation of native liver survival declines.

The two principal complications that can be life threatening are cholangitis and the development of esophageal varices.

Cholangitis occurs most commonly in the year following primary surgery in about 40–50 percent of children. Paradoxically, it only occurs in children with some degree of bile flow, not in those with early failure. Clinically, it is characterized by worsening jaundice, fever, and acholic stools. The diagnosis may be confirmed by blood culture or by percutaneous liver biopsy, but it is important to treat suspected cases early with broad-spectrum antibiotics effective against Gram-negative organisms.

Increased portal venous pressure has been shown in virtually all infants at the time of the Kasai operation; however,

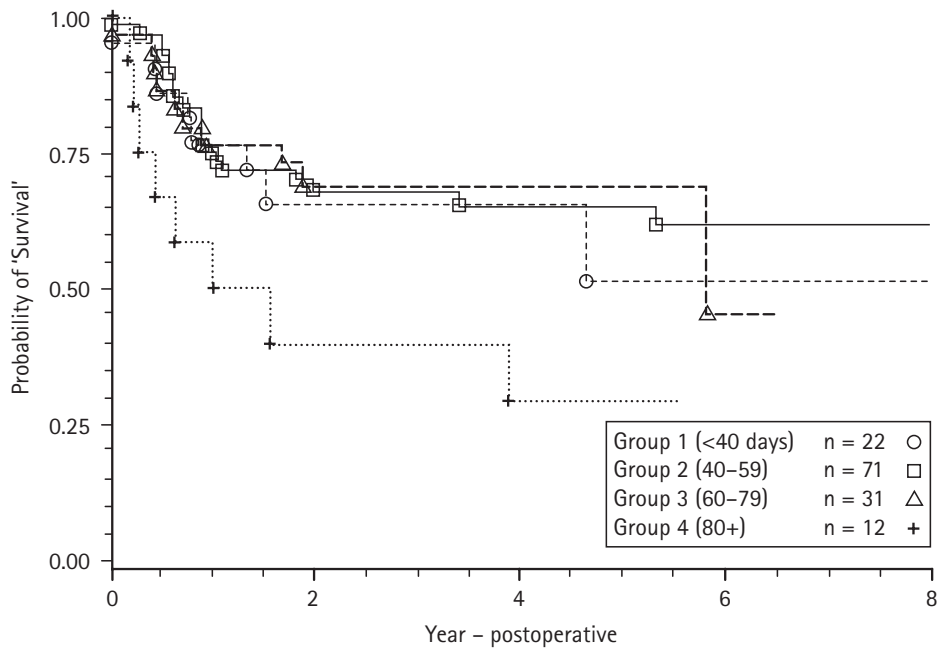


Fig. 65.1 Actuarial native liver survival curve in 137 infants presenting to King's College Hospital from 1992 to 1997, divided according to age at surgery. The end-points are death or transplantation. (From Davenport et al., 2003.)

subsequent portal hypertension depends on both the degree of established fibrosis and, most importantly, the response to surgery. There is a relationship with biochemical liver function and variceal development, and in those who fail and need early transplantation, about 30 percent will have had a significant variceal bleed. In those who respond well to initial Kasai but who have established fibrosis, variceal development can be delayed and presentation with bleeding may only occur after 2–3 years.

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66

Choledochal cysts

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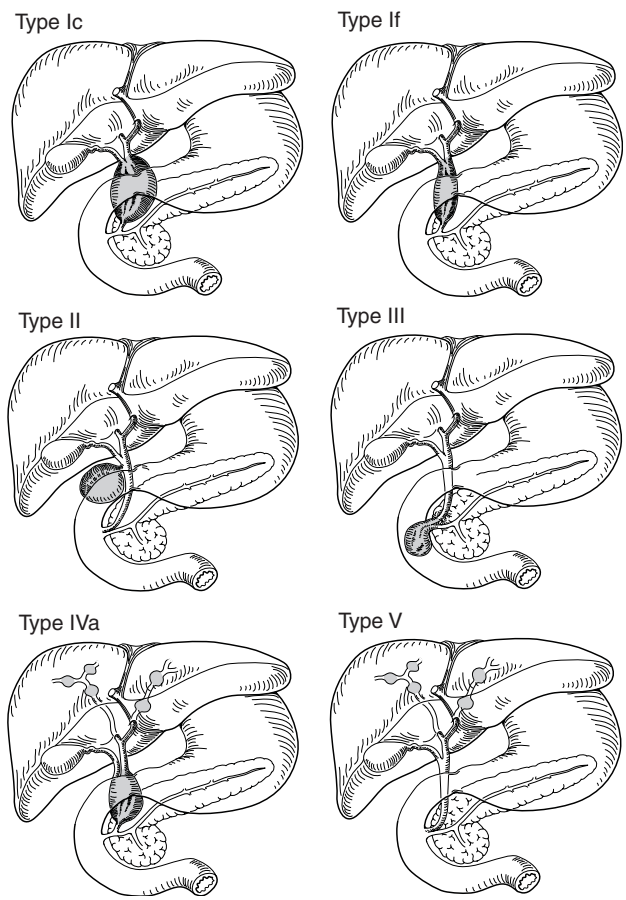
PRINCIPLES AND JUSTIFICATION

Congenital bile duct dilatation is a better description of this spectrum of congenital anomalies of the biliary tree, but 'choledochal cyst' is the traditional term.

1 The following classification is commonly used.

- Type I – cystic (Ic) or fusiform (If).
- Type II – diverticulum of the common bile duct.
- Type III – choledochocele (dilatation of the terminal common bile duct within the duodenal wall).
- Type IV – multiple cysts of extrahepatic and intrahepatic ducts (IVa) or multiple extrahepatic duct cysts (IVb).
- Type V – intrahepatic duct cyst (single or multiple [Caroli's disease]).



Type I choledochal cysts predominate. Together with type IVa cysts, they account for more than 90 percent of cases. Intrahepatic duct dilatation that resolves after successful treatment of a type I cyst should be distinguished from the irregularly dilated intrahepatic ducts found with type IVa cysts. Caroli's disease is characterized by segmental saccular dilatation of the intrahepatic bile ducts; it may affect the liver diffusely or be localized to one lobe.

Pathology

There are three components to the pathology of a choledochal cyst: the cyst, which may be inflamed and thick-walled, and any abnormal bile ducts; the associated liver histology, which varies from normal to fibrotic or cirrhotic; and the existence of pancreatobiliary malunion, which is present in most but not all cases (Fig. 66.1). In the last, the terminal common bile duct and pancreatic duct unite to form a common channel well outside the duodenal wall. Since this common channel is not surrounded by the normal sphincter mechanism, pancreatic juice refluxes into the biliary tree and high concentrations of pancreatic amylase and/or lipase are typically present in the bile. Occasionally, bile refluxes into the pancreatic duct causing pancreatitis. In most cases, the common channel represents a simple union of the two ducts, but in some patients the anatomy is complex. Rarely, pancreatobiliary malunion occurs without biliary dilatation.

Whenever possible, the preoperative and intraoperative assessment of a choledochal cyst should include an evaluation of all three pathologic features. Additional biliary tract abnormalities can occur, such as distal duodenal displacement of the papilla of Vater and, in type IVa cysts, hilar duct strictures. Associated anomalies outside the biliary tract are uncommon.

Clinical features

Girls are affected more often than boys. Typical presenting symptoms are abdominal pain and/or obstructive jaundice. The classic triad of abdominal pain, jaundice, and a mass is rare. Abdominal pain is often associated with hyperamylasemia due to pancreatitis or diffusion of pancreatic amylase from the cyst. A choledochal cyst should always be considered in the differential diagnosis of obstructive jaundice or pancreatitis in children. The bile duct dilatation of a fusiform type I cyst may be relatively subtle.

An increasing proportion of cystic type I choledochal dilatations are detected by routine prenatal ultrasonography. If these infants are jaundiced and a cystic variant of biliary atresia has not been excluded, surgery should be performed promptly. If the infant is otherwise well, surgery can be safely deferred until about 3 months of age. Delaying treatment further exposes the infant to the risks of developing liver fibrosis, cholangitis, or cyst perforation.

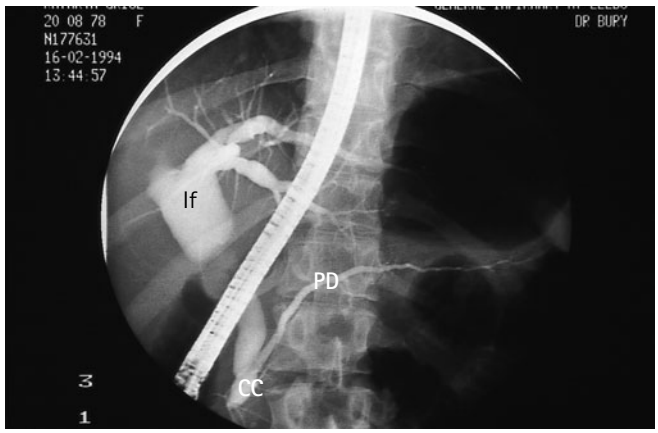


Fig. 66.1a Pancreatobiliary malunion with a fusiform type I choledochal cyst. If = type I fusiform choledochal cyst, PD = pancreatic duct, CC = common pancreatobiliary channel.

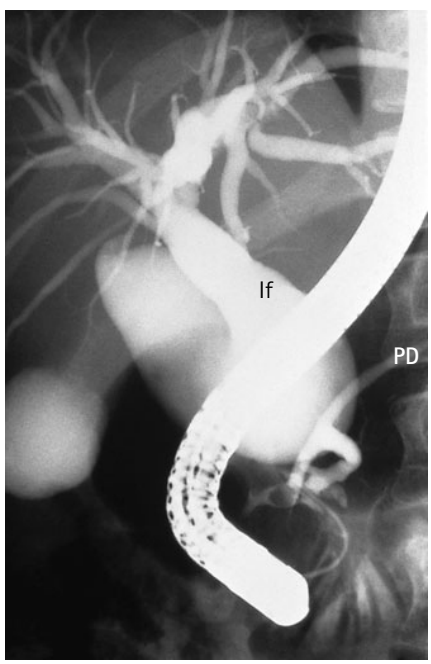


Fig. 66.1b Complex pancreatobiliary malunion. If = type I fusiform choledochal cyst, PD = pancreatic duct. (Reproduced from Stringer, 2002.)

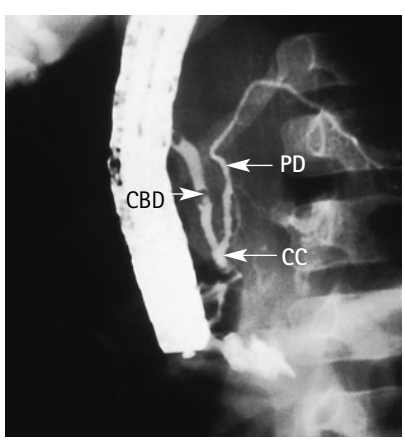
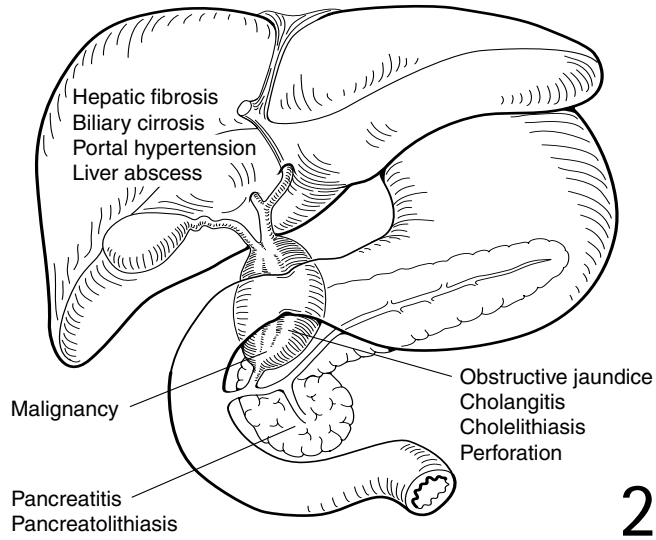


Fig. 66.1c Pancreatobiliary malunion without choledochal dilatation. Note filling defects in common bile duct. PD = pancreatic duct, CC = common pancreatobiliary channel, CBD = common bile duct.

Complications

2 Choledochal cysts are prone to complications. Perforation is rare and presents with an acute abdomen. Malignant change is a late complication, mostly described in adults, but has been recorded in teenagers. It is much more likely to occur after cystenterostomy than if the cyst has been excised. Prompt surgery consisting of radical cyst excision and biliary reconstruction eliminates or reduces the risk of complications.



PREOPERATIVE ASSESSMENT AND PREPARATION

Biochemical liver function tests may be normal or show evidence of biliary obstruction. The plasma amylase level may be elevated during episodes of abdominal pain. A prolonged prothrombin time secondary to cholestasis should be corrected with intravenous vitamin K. Routine preoperative blood tests should also include a full blood count and blood group and save.

Modern imaging methods provide an accurate diagnosis. Ultrasonography is the initial investigation of choice – the size, contour, and position of the cyst, the proximal ducts, vascular anatomy, and hepatic echotexture can all be evaluated (Fig. 66.2). Percutaneous transhepatic cholangiography and endoscopic retrograde cholangiopancreatography (ERCP) give excellent definition of the cyst and duct anatomy, including the pancreatobiliary junction. However, both investigations are invasive and have a small risk of inducing complications such as pancreatitis and biliary sepsis, and both require general anesthesia and antibiotic prophylaxis. Endoscopic retrograde cholangiopancreatography should be avoided during an episode of acute pancreatitis.

Magnetic resonance cholangiopancreatography (MRCP) is non-invasive and can be performed under sedation in small children without the use of contrast agents or irradiation (Fig. 66.3). However, definition of the pancreatic duct and common channel may be suboptimal in infants and small children.

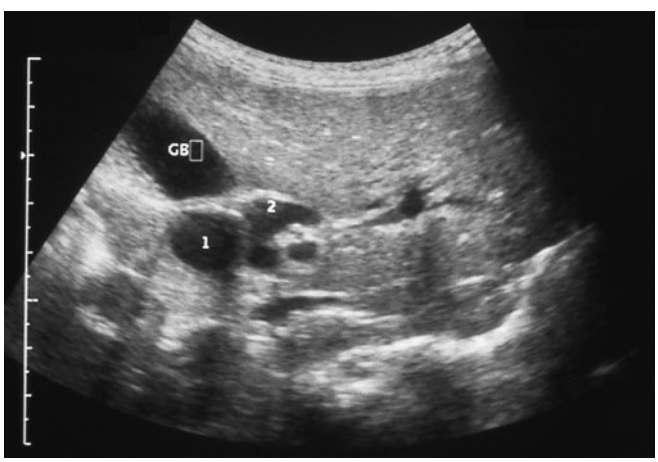


Fig. 66.2 Ultrasound scan of a type I choledochal cyst in a 3-year-old girl. GB = gallbladder. 1 and 2 are dilated extrahepatic bile ducts.

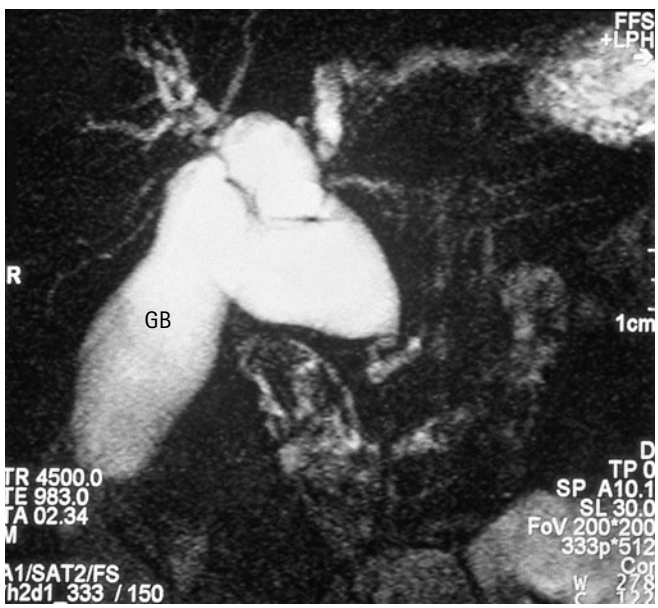


Fig. 66.3 Magnetic resonance cholangiopancreatography of the type I choledochal cyst shown in Fig. 66.2. Note the common pancreatobiliary channel (CC). GB = gallbladder.

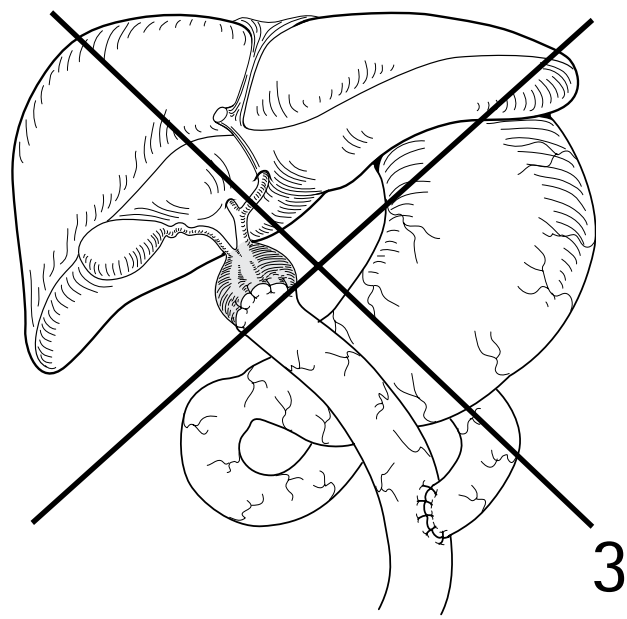
Hepatobiliary scintigraphy with technetium-^{99}m iminodiacetic acid is useful in selected cases by confirming biliary excretion into the cyst or when assessing biliary drainage after surgery. Contrast enhanced computed tomography may be indicated in some patients with pancreatitis or if an associated tumor is suspected.

In most patients, a detailed ultrasound scan supplemented by MRCP or intraoperative cholangiography provides sufficient anatomical information.

Consent

Radical cyst excision and reconstruction by hilar hepaticoenterostomy is the optimum treatment for the common types of choledochal cyst. This disconnects the pancreatic and biliary ducts. In experienced units, this operation can be performed safely at any age with a very low morbidity. Complications are rare, but early problems may include an anastomotic bile leak, bleeding, intra-abdominal sepsis, injury to adjacent structures, and wound complications. Potential late complications are a bilioenteric anastomotic stricture, stone formation (more common within ectatic intrahepatic ducts in type IVa cysts), pancreatitis from residual abnormalities in the common channel, and adhesive small bowel obstruction. Even after adequate cyst excision, malignancy may very rarely affect residual extrahepatic ducts (e.g., the terminal common bile duct) or abnormal intrahepatic ducts (type IVa cysts).

3 Internal drainage of the cyst by cystenterostomy has a prohibitively high long-term morbidity from cholangitis, stone formation, malignant degeneration, etc., and should be avoided. Definitive surgical excision may be unsafe in a critically ill child with perforation, uncontrolled biliary sepsis, or serious concomitant ill-health. In such cases, temporary external drainage of the choledochal cyst and delayed surgery once the patient has improved and the anatomy has been defined is safer.

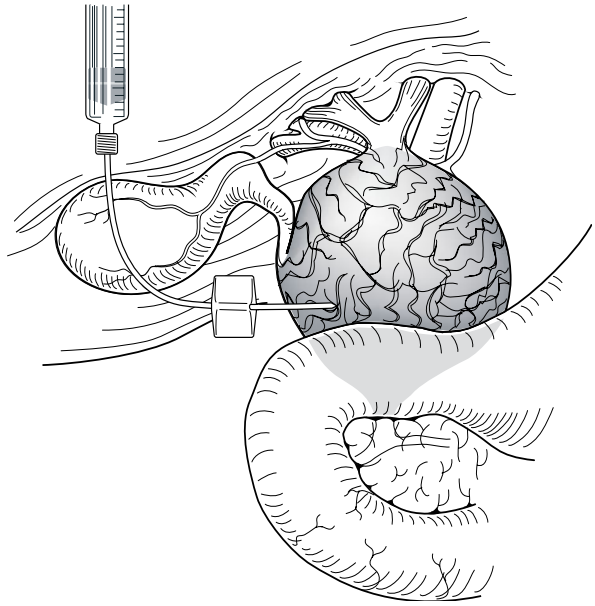


Anesthesia

Endotracheal intubation, muscle relaxation, and temporary nasogastric tube drainage are standard. Epidural analgesia can provide excellent perioperative pain relief. Broad-spectrum intravenous antibiotics are best given at induction of anesthesia and continued for 2–5 days postoperatively.

OPERATIVE TECHNIQUES

4 A high transverse or oblique right upper quadrant incision gives excellent exposure. The duodenum and head of pancreas may be displaced forward over the cyst. The appearance of the liver, spleen, and pancreas is noted. An intraoperative cholangiogram should be performed if the anatomy has not been clearly defined preoperatively. Bile is aspirated from the cyst and sent for culture and measurement of amylase/lipase. With large cysts, cholangiography is often best performed by the injection of contrast directly into the lower end of the common bile duct and into the common hepatic duct using a butterfly needle. Direct injection into a large cyst may fail to outline the intrahepatic ducts and obscure filling of the distal duct. It is important to try to identify the junction of the pancreatic and bile ducts.



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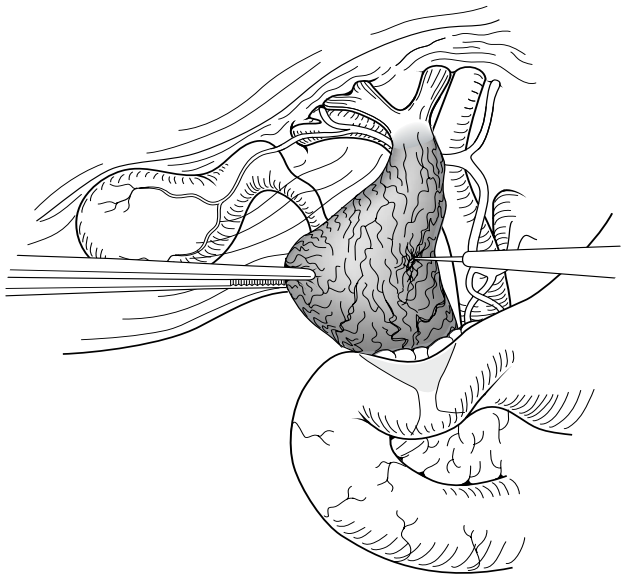

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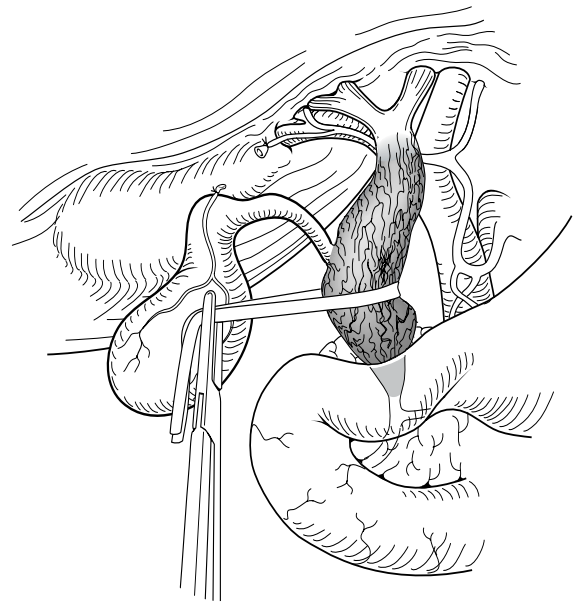
5 A plane is developed between the overlying peritoneum and the anterior wall of the cyst. The dissection extends inferiorly between the duodenum and the cyst and medially and laterally, keeping close to the cyst wall, using precise bipolar cautery to achieve safe and accurate hemostasis. Large cysts can be decompressed to facilitate dissection. The gallbladder and cystic duct are mobilized and the cystic artery ligated.

6 Where the bile duct begins to narrow down inferiorly, it is dissected circumferentially and encircled. In this region, small vessels arising from the pancreas need careful cautery. The distal common bile duct is dissected to just within the head of the pancreas and transected. The operative cholangiogram gives a useful guide to the distal level of bile duct transection. Protein plugs or calculi within a common channel should be removed using a combination of saline irrigation, balloon catheters and, when possible, intraoperative endoscopy using a pediatric cystoscope. The distal bile duct stump is then over-sewn with an absorbable monofilament suture (e.g., polydioxanone [PDS]).



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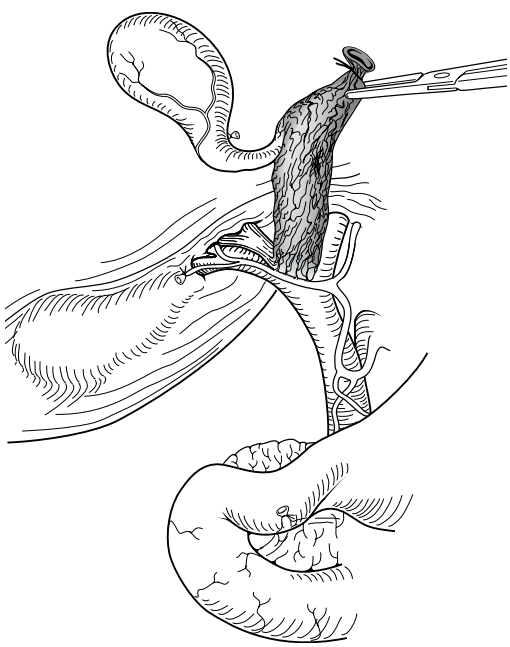

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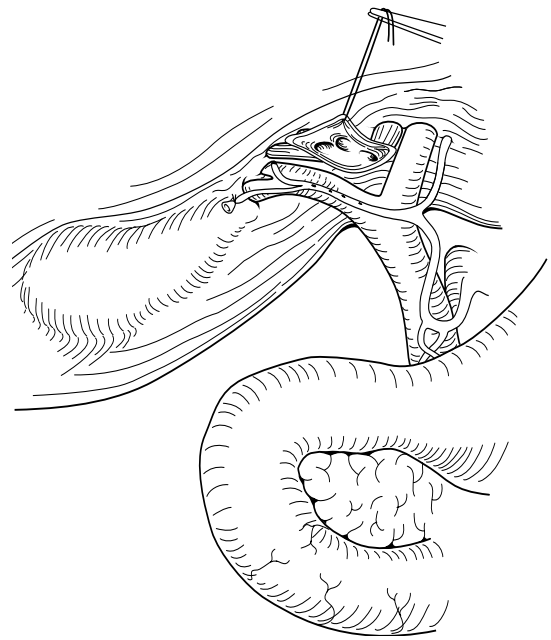
7 The cyst and gallbladder are lifted forward, exposing the portal vein behind. Sometimes the right hepatic artery crosses in front of the cyst and is adherent to its wall – it must be carefully freed and preserved. The common hepatic duct is divided at the level of the bifurcation, where it should appear healthy and well vascularized. Any dilated proximal intrahepatic ducts are cleared of debris by catheter irrigation with normal saline and, in larger ducts, with the aid of choledochoscopy.

8 The left hepatic duct is incised for a variable distance (5–10 mm) to allow a wide hilar hepaticoenterostomy. Anastomosis to a narrow common hepatic duct should be avoided because of the long-term risk of stricture.



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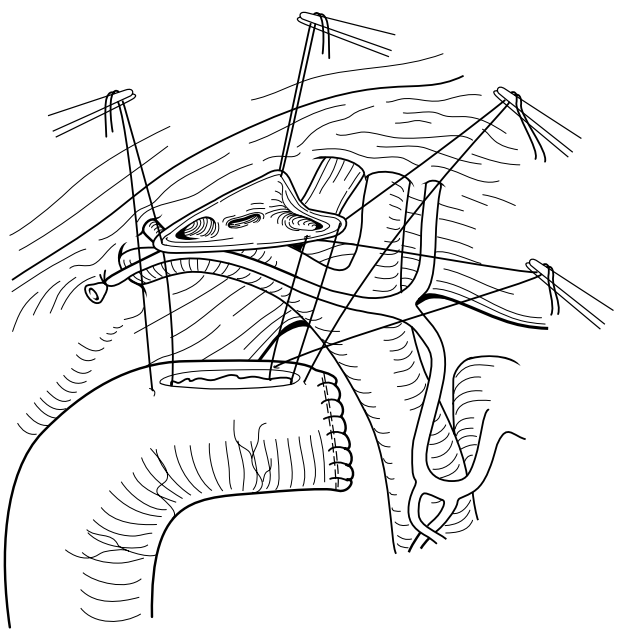

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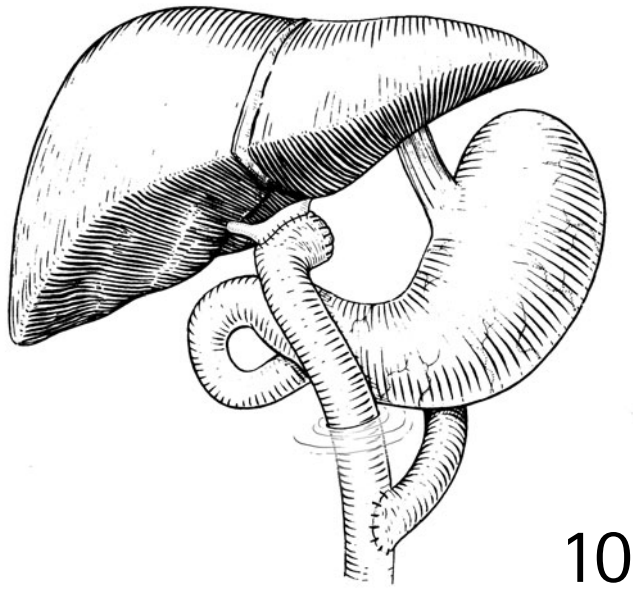
9 The duodenojejunal flexure is identified and the jejunum divided with a linear stapler approximately 15–20 cm downstream. At this point, there is a suitable vascular arcade to create a Roux loop that will reach the hilum of the liver without tension. The stapled end of the Roux loop is oversewn with an absorbable suture and passed through a window in the transverse mesocolon to the right of the middle colic vessels. The Roux loop of jejunum is widely anastomosed to the hepatic duct bifurcation at the hilum of the liver using fine, interrupted, absorbable monofilament sutures (6/0 or 7/0 PDS). Magnifying loupes help to ensure a precise anastomosis. The anastomosis is constructed a few millimeters from the end of the Roux to avoid the development of a blind pouch with future growth of the bowel.

10 A 40 cm Roux loop is adequate in most cases, but a shorter (30 cm) loop is adequate in infants. The proximal stump of jejunum is anastomosed in an oblique end-to-side fashion to the distal jejunum using a single layer of interrupted extramucosal PDS sutures. Mesenteric defects in the transverse mesocolon and small bowel mesentery are closed with sutures. A liver biopsy is performed at the end of the operation to document hepatic histology. The operative field is washed with warm saline and, in straightforward cases, the abdomen is closed without drainage. If a drain is left in place, it should be placed in Morison's pouch and not in direct contact with the anastomosis.



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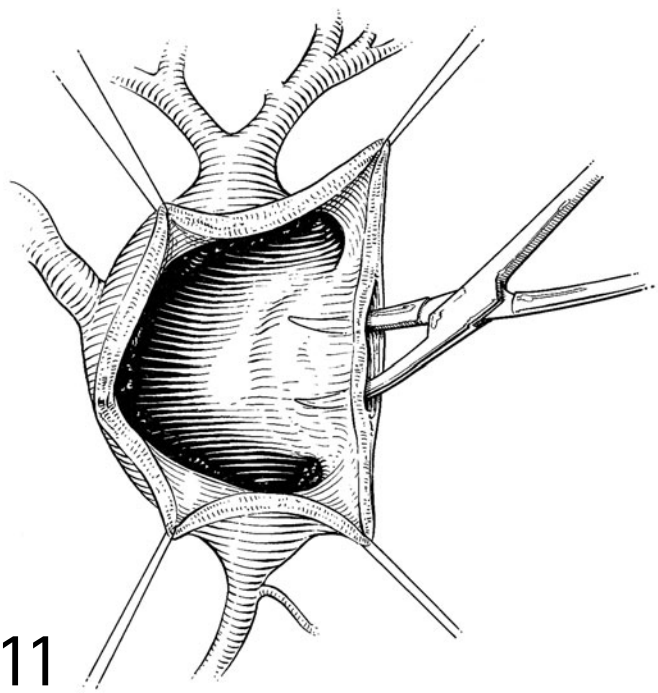
Alternative operative techniques

- Alternative biliary reconstructive techniques include an end-to-end hilar hepaticojejunostomy (in contrast to end-to-side) and hepaticoduodenostomy. Proponents of the latter argue that it is more physiological, associated with less risk of adhesion obstruction, and minimizes the loss of absorptive mucosa, but duodenogastric reflux of bile can be a problem and there are concerns about a long-term risk of anastomotic malignancy. Excellent results can be achieved with hepaticojejunostomy. The appendix should not be used as a conduit (hepatico-appendico-duodenostomy) because of a high incidence of subsequent biliary obstruction. An intussusception 'valve' offers no advantage in the Roux loop.
- Hilar ductal strictures may necessitate some form of ductoplasty or extended anastomosis.
- The addition of a transduodenal sphincteroplasty should be considered if the common channel is very dilated and contains debris.



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11 • Occasionally, portal hypertension or dense inflammation from previous infection or surgery makes radical excision hazardous. Intramural resection of the posterior wall of the cyst (excising only the mucosa and inner wall) can help to avoid damage to the portal vein. The cyst lining is completely removed, but a portion of the outer wall remains posteriorly.

- Endoscopic sphincterotomy or transduodenal sphincteroplasty alone cannot be recommended for fusiform choledochal cysts because the majority of these patients have a common pancreatobiliary channel, which results in an ongoing risk of recurrent pancreatitis and later biliary tract malignancy.
- Type II cysts: excision of the diverticulum and repair of the common bile duct is a satisfactory procedure for this rare variety of choledochal cyst.
- Type III cysts: large choledochoceles can be removed transduodenally. Smaller choledochoceles can be treated by sphincteroplasty or endoscopic sphincterotomy if there is no pancreatobiliary malunion.
- Type V cysts: if the cysts are multiple and confined to one side of the liver, hepatic lobectomy may be curative. If multiple cysts are distributed throughout the liver, recurrent cholangitis and stone formation are common. Antibiotics and drainage procedures are helpful, but liver transplantation should be considered in progressive cases.

Laparoscopic excision

After insertion of a nasogastric tube and urinary catheter and with the patient 30° head up, a 5–10 mm primary port (depending on the size of the child) is inserted at the umbilicus using an open technique; three secondary 5 mm ports are inserted in the right side of the abdomen, and in the right and left hypochondrium. The pneumoperitoneum is set at 8–12 mmHg. A combination of 3 mm and 5 mm instruments with cautery is used. The operative steps are as follows: needle puncture of the cyst and cholangiography; ligation and division of the cystic artery; cholecystectomy; dissection of the lower part of the choledochal cyst, opening it transversely; cautery dissection of the cyst keeping close to its wall; ligation and division of the distal common bile duct; proximal dissection of the cyst; and transection at the hilar bifurcation. If reconstruction is with a Roux-en-Y loop, this may need to be constructed manually after exteriorizing a segment of bowel through an extended incision at the umbilical trocar site. The jejunum is anastomosed to the hepatic duct bifurcation with interrupted sutures. Hepaticoduodenostomy is an alternative, as it avoids any extracorporeal procedure, but this is not ideal.

POSTOPERATIVE CARE

Anastomotic leakage should be rare and is likely to resolve with local drainage, intravenous antibiotics, and nasogastric decompression, but it may be followed by anastomotic stricturing (Fig. 66.4). Postoperatively, biochemical liver function tests (including gamma-glutamyl transpeptidase) should return to normal.

Cholangitis may signify an anastomotic stricture, an intrahepatic ductal stricture or stone, Roux loop obstruction, or calculi within a redundant blind pouch of an end-to-side Roux loop. Type IVa cysts are particularly at risk of developing intrahepatic calculi. Standard imaging methods supplemented by biliary scintigraphy and percutaneous transhepatic cholangiography (with antibiotic prophylaxis) help to define the problem. Interventional radiologic techniques may be able to clear stones and dilate strictures, but surgery is usually required to revise a bilioenteric anastomotic stricture.

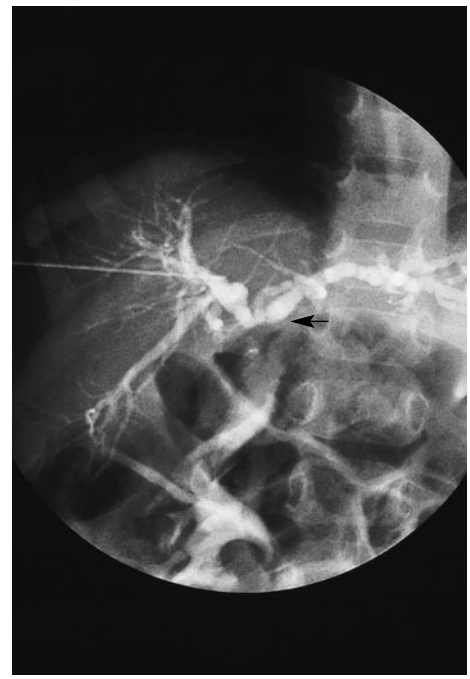

Fig. 66.4 Percutaneous transhepatic cholangiogram in a 2-year-old girl referred for biliary reconstruction after surgery for a type I choledochal cyst complicated by an anastomotic stricture (arrow). (Reproduced from Stringer, 2002.)

Pancreatitis may develop many years later in patients with a dilated or complex common channel containing protein plugs or calculi. This complication is usually preventable by appropriate primary surgery. Endoscopic retrograde cholangiopancreatography is a useful investigation in such patients, and endoscopic sphincterotomy may be curative.

OUTCOME

Radical cyst excision and hilar hepaticoenterostomy achieves consistently good results. In some older patients operated on late with established liver disease, the prognosis is more guarded. Late complications after radical cyst excision and bilioenteric anastomosis are uncommon. Nevertheless, biliary strictures causing cholangitis may develop up to 10 or more years postoperatively.

Todani (1998) reported a 10 percent re-operation rate after primary cyst excision in 103 children followed for a median of

14 years. Revisional surgery was necessary between 3 and 21 years after the initial operation because of cholangitis due to ductal or anastomotic strictures. These complications were more likely with type IVa cysts or after biliary anastomosis to the common hepatic duct. Re-operation was not required after hilar hepaticojejunostomy or hepaticoduodenostomy for type I choledochal cysts. In another large Japanese series of 200 children with type I or IVa cysts followed up for a mean of 11 years after cyst excision and hepaticoenterostomy (principally Roux-en-Y hepaticojejunostomy), there were 25 late complications in 18 (9 percent) children. These included cholangitis, intrahepatic and common channel calculi, anastomotic stricture, pancreatitis, and adhesive bowel obstruction. There were no instances of malignancy. No calculi or anastomotic strictures developed in children undergoing surgery before 5 years of age.

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67

Laparoscopic splenectomy

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HISTORICAL ASPECTS

Laparoscopic splenectomy has emerged as one of the most frequently performed laparoscopic solid organ procedures in children. The first laparoscopic splenectomy in an adult was reported in 1992 and in a child in 1993. Subsequently, many authors published their experience with pediatric laparoscopic splenectomy and a number of modifications of the original laparoscopic techniques have been described. These modifications include lateral positioning of the patient, the use of endovascular stapling devices, and the addition of novel energy devices such as the ultrasonic scalpel and the Ligasure®. Studies in children comparing laparoscopic splenectomy and open splenectomy have shown that the laparoscopic approach is safe and has several advantages, including reduced postoperative pain, a shorter stay in hospital, early return of normal activity, and improved cosmesis. Many reports have shown that operating time is longer with laparoscopic splenectomy compared to open techniques, but this may be due to the learning curve that applies to all laparoscopic procedures.

Indications

Splenectomy is often required in children with hematologic disorders such as sickle-cell disease, hereditary spherocytosis, thalassemia major, and idiopathic thrombocytopenic purpura. Rarely is laparoscopic splenectomy indicated for trauma.

OPERATIVE PROCEDURE

Preparation of the patient

The colon should be deflated to facilitate exposure. This may be achieved by giving an enema the evening before surgery if the child is constipated or if the colon appears distended. The stomach should be emptied with a nasogastric tube, and the urinary bladder should be emptied either by a Credé maneuver or by placing a Foley catheter.

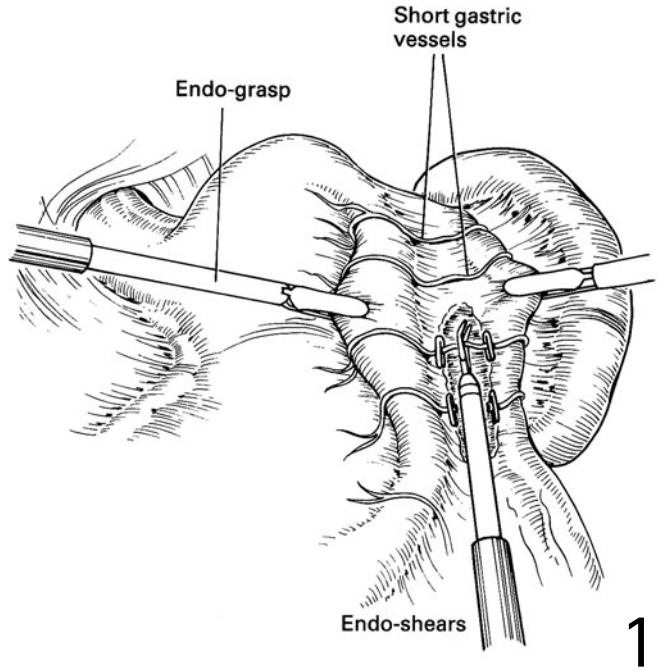
Trocar insertion sites

A 5 mm 30° laparoscope should be placed through the 10–12 mm umbilical cannula. The patient should be placed supine or in an exaggerated reversed Trendelenburg position and slightly rotated with the left side elevated. Alternatively, the table can be rotated as required. In either case, the patient should be appropriately secured so that further moving of the table to improve patient positioning will not put the patient at risk of slipping off it.

Two additional 5 mm cannulae are placed. Their exact location may vary depending on the size of the spleen and of the patient. In general, one 5 mm cannula is in the midclavicular line on either side. The cannula on the patient's right side is placed higher than the one on the left, which is usually placed below the tip of the spleen. Thus the three cannulae are usually placed in a diagonal line going from the patient's right midclavicular line, midway between the right costal margin and the level of the umbilicus, through the umbilicus, to the left midclavicular line between the level of the umbilicus and the top of the iliac crest.

Dividing short gastric vessels

1 The greater curvature of the stomach should be grasped gently with an atraumatic grasper or Babcock clamp passed through cannula on the right, and the gastrosplenic ligament should be retracted to expose the short gastric vessels. These vessels should then be divided with an energy device or between clips passed through the cannula on the left.



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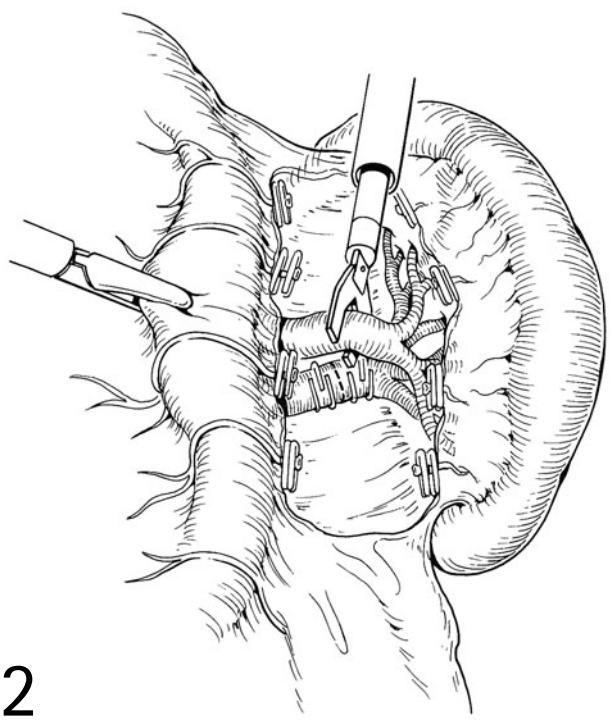
Freeing the spleen posteriorly

Once the short gastric vessels are divided, the spleen is retracted to the right and the posterolateral fascial attachments are divided to free the spleen so that its only remaining attachments are the splenic hilar vessels.



2

Dissection of the splenic vessels

2 As the dissection progresses, it may be helpful to rotate the patient toward the right and in a more reversed Trendelenburg position to allow the intestines to fall out of the way. The spleen should be elevated using the instruments passed through the lateral cannulae, and the splenic artery and vein at the splenic hilum just beyond the tail of the pancreas should be identified using gentle dissection with a curved or right-angled dissector. These vessels should be separately and carefully divided between ligatures or with a Ligasure® device in small children. Alternatively, we have isolated the splenic hilum and used the vascular load of an endoscopic stapler to divide both vessels at once, or they can be divided between clips, as illustrated. The endoscopic stapler is hemostatic in our experience and diminishes the operative time significantly. Care should be taken to avoid the tail of the pancreas during this dissection.

Dissection of ligamentous attachments

After the major splenic vessels have been divided, there are often some ligamentous attachments at the upper pole of the spleen that should be carefully divided using Metzenbaum-type scissors. At this point, the spleen should be completely mobilized and free in the peritoneal cavity.

Endo-Catch II for the isolated spleen

An Endo-Catch II (TYCO, Autosuture) should then be inserted directly through the port site of the 10–12 mm trocar with the trocar removed. This device acts like a net. When deployed in the abdomen, the freed spleen can be maneuvered into the sac and then the neck of the sac can be exteriorized through the trocar site.

Morcellating of the spleen

The neck of the sac should be opened and a tissue morcellator can be placed into the pouch. The organ can then be morcellated until it is sufficiently small to enable the entire pouch with any residual tissue to be withdrawn through the trocar site.

Because of the potential risk of the automatic morcellators breaking the bag, we break up the spleen with a finger, sponge holder, or ovary forceps. The trocar and cannula are then re-inserted, and a pneumoperitoneum should be re-established to check for hemostasis.

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SECTION V

Tumors

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68

Liver resections

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HISTORY

The anatomic approach to liver resection is a modern development, but its underpinnings stretch back many years. The remarkable regenerative capacity of the liver was prefigured in classical times by the story of Prometheus, whose liver was mythically able to regenerate daily after being devoured by an eagle. For many years, it was recognized that humans could survive with significant destruction of the liver due to amebic cysts, abscesses, and traumatic injury. However, the potential for significant hemorrhage from the liver was also recognized, and fear of such hemorrhage precluded safe resection. Thus, a surgical understanding of the structure and function of the liver is a development of modern times. One of the earliest reports of liver resection was by J.C. Massie in 1852. He described a 7-year-old boy who sustained a gunshot wound and subsequently herniated a portion of the right lobe of his liver through a defect in the abdominal wall. When this segment became gangrenous, the abdominal wall defect was enlarged, and a significant portion of the right lobe was removed. The patient ultimately recovered after this operation. The first elective liver resection was reported by Carl Langenbuch in 1888. He resected 370 g of the left lobe of the liver in a 30-year-old female with abdominal pain. Interestingly, this patient required early re-exploration for hemorrhage from the remaining liver, but subsequently recovered well. The modern understanding of the segmental anatomy of the liver was described by Couinaud in 1957 and serves as the basis for contemporary segmental liver resections. Advances in surgical techniques of liver resection were devised in the 1950s and 1960s by surgical pioneers such as Lortat-Jacob, Quattelbaum, Longmire, Fineberg, and McDermott. These improvements in technique – extrahepatic vascular control, parenchymal division with ligation of intrahepatic structures, and occlusion of the portal triad – allowed liver resections to be performed with an increased level of safety and ushered in the contemporary era of hepatic surgery.

PRINCIPLES AND JUSTIFICATION

Indications

In the pediatric population, liver resections are most frequently performed for malignant tumors. Hepatoblastoma is the most common hepatic malignancy in children and accounts for 40–50 percent of all primary liver tumors. The majority of hepatoblastomas are sporadic, although they can be associated with Beckwith–Wiedemann syndrome, hemihypertrophy, renal/adrenal agenesis, familial adenomatous polyposis, neurofibromatosis, and Prader–Willi syndrome. It is uncommon for hepatoblastoma to occur in the setting of underlying chronic liver disease. Children with hepatoblastoma typically present in the first 3 years of life with an abdominal mass, distension, loss of appetite, or pain. Males are affected three times more commonly than females. Because most of these tumors are asymptomatic, the majority of hepatoblastomas are unresectable at the time of presentation. Over 90 percent of children with hepatoblastoma will have an elevated serum alpha-fetoprotein (AFP), which can be helpful in making the diagnosis and assessing the response to therapy.

Hepatocellular carcinoma (HCC) is the second most common pediatric primary liver tumor, occurring in nearly a quarter of patients. It occurs most frequently in patients with underlying liver disease. Worldwide, the incidence of HCC is highest in regions of Asia, Africa, and South America, where

hepatitis B is endemic. In addition, HCC can occur on a background of metabolic liver disease (α-1-antitrypsin deficiency, glycogen storage disease, tyrosinemia, Wilson's disease), biliary atresia, neonatal hepatitis, and total parenteral nutrition-induced cholestasis. Because of its association with chronic liver disease, HCC tends to present in older children, typically aged 10–14 years. It usually presents with an abdominal mass, weight loss, and fever, although it can occasionally present with acute abdominal pain from tumor rupture. It is often present in more than one segment of the liver at the time of presentation, and it has extrahepatic spread or distant metastases at least half of the time. Serum AFP is elevated in two-thirds of cases, and transcobalamin is elevated in the fibrolamellar variant. These markers can also be used to follow response to treatment. Aside from hepatoblastoma and HCC, primary malignancies of the liver are extremely uncommon. Resection can play a role in the rare rhabdomyosarcoma or undifferentiated sarcoma of the liver.

Benign tumors account for almost one-third of primary liver tumors. Hemangiomas, including cavernous hemangiomas and hemangioendotheliomas, are the most common benign liver tumors. They usually present as an incidental finding on an imaging study, although they can present as an enlarging abdominal mass in infancy or childhood. Patients with hepatic hemangioendotheliomas may also have hemangiomas in other locations, such as the skin, the central nervous system, or the gastrointestinal tract. Very large hemangiomas may cause mass effect, high-output heart failure from left-to-right shunting, and thrombocytopenia from consumptive coagulopathy, known as Kasabach–Merrit syndrome. Small, asymptomatic hemangiomas may be followed expectantly, as many of these lesions regress spontaneously. Non-operative management of symptomatic hemangiomas includes corticosteroids, alpha-interferon, cyclophosphamide, and angioembolization. Operative management includes hepatic artery ligation, enucleation, or resection. Resection is the favored approach if the diagnosis of angiosarcoma cannot be reasonably ruled out. Other benign tumors of the liver include mesenchymal hamartomas, hepatic adenomas, focal nodular hyperplasia, and hepatic cysts. Resection is generally recommended for large lesions, symptomatic lesions, or lesions in which the diagnosis is uncertain.

Metastasis to the liver warranting resection is an uncommon occurrence in children. Patients with liver metastases from Wilms' tumor may benefit from liver resection as part of their multimodality therapy. Stage 4S neuroblastomas may have extensive involvement of the liver. However, these lesions are treatable without resection. Hematogenous metastases to the liver in stage 4 neuroblastoma have an extremely poor prognosis, and liver resection is generally contraindicated. The techniques of liver resection can be applied for control of locally advanced tumors of non-hepatic origin. Anatomic or non-anatomic liver resection may be used to obtain a negative margin in the resection of primary Wilms' tumor, adrenocortical carcinoma, pheochromocytoma, and germ-cell tumors.

Contraindications

Patient factors that may limit the feasibility of liver resection include medical suitability to undergo a major operation and the need for sufficient functional liver mass following resection. In light of the significant vascularity of the liver, coagulopathy, thrombocytopenia, and anemia should be corrected prior to operation. These factors may occur as a consequence of underlying liver disease or as a side-effect of therapy for the tumor. Platinum-based chemotherapy, used in protocols for both hepatoblastoma and HCC, may cause ototoxicity and nephrotoxictiy. Patients with a serum creatinine level higher than 1.3 who undergo extended liver resection have a significantly higher mortality than those who do not. Doxorubicin may also be used in preoperative chemotherapy regimens. The risk of cardiotoxicity is related to the cumulative dose and should be considered in patients with a history or symptoms of heart failure prior to operation. Patients with biliary obstruction and cholangitis have a higher risk of mortality with large liver resections. Measures to alleviate the cholangitis should be aggressively pursued prior to operation, including fluid resuscitation, intravenous antibiotics, and biliary decompression (endoscopic or percutaneous).

Children with liver tumors, particularly HCC, may also have significant underlying primary liver disease. Safe hepatic resection is determined by the size of the anticipated resection and the functionality of the remaining liver parenchyma. In a patient without concomitant liver disease, resection of 85 percent of the liver's total mass may be safely performed with minimal risk of postoperative hepatic dysfunction. Patients with Child's class A cirrhosis generally have good hepatic reserve and have a 1–2 percent risk of postoperative mortality related to their liver disease. This number approaches 10 percent in Child's B cirrhosis and 50 percent in Child's C cirrhosis. In equivocal cases, liver biopsy may be helpful to establish the extent of cirrhosis.

Anatomic features of the tumor may preclude safe liver resection. Liver tumors are considered unresectable if they involve all four sectors of the liver, or if multiple tumors involve all four sectors. Extension of a malignant tumor outside the confines of the liver is considered a contraindication to primary surgical resection. Involvement of the hepatic veins or inferior vena cava based on imaging studies should be anticipated prior to attempted resection. While this may not be an absolute contraindication to resection, it may require a significant modification in technique, including extracorporeal bypass to complete the operation successfully.

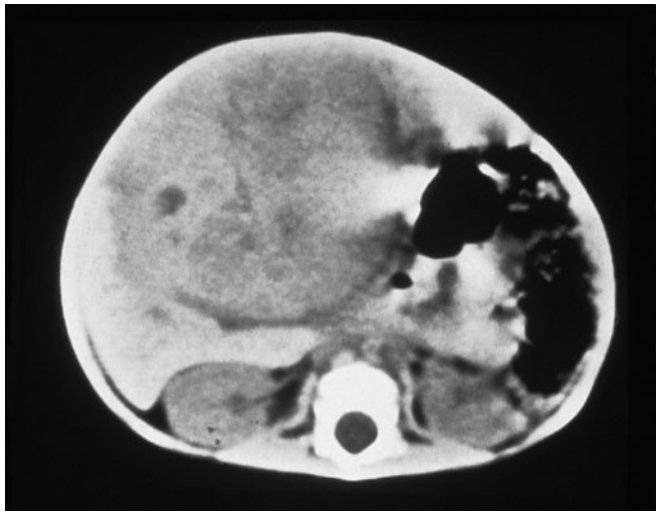
Neoadjuvant chemotherapy can benefit children with primary hepatic malignancies. Tumors treated preoperatively are generally smaller and better demarcated, favoring complete resection. Preoperative chemotherapy can promote lesser resections in patients whose tumors may be amenable to primary resection, thus promoting the preservation of hepatic parenchyma. Between 70 and 85 percent of patients who present with unresectable tumors will have sufficient

response to neoadjuvant chemotherapy to be able subsequently to undergo successful resection.

Liver transplantation should be considered as an alternative to liver resection in selected cases. Total hepatectomy serves as the definitive resection for extensive tumors. Hepatic function of the transplanted liver is superior to that of a small segment of residual liver, particularly in the face of underlying liver dysfunction. In addition, tumor recurrence or second primaries may be less likely in the transplanted liver. These potential benefits must be weighed against the scarcity of available organs, the possibility of disease progression while waiting for an organ, and the risks of lifetime immunosuppression in the case of a successful transplant.

PREOPERATIVE ASSESSMENT AND PREPARATION

1 Accurate imaging can prove invaluable in the planning of a successful liver resection. Computed tomographic (CT) scanning is the favored imaging study for the assessment of liver tumors in children. It is a high-resolution study that can be completed quickly, minimizing the need for sedation. The CT scans demonstrate the size and anatomic location of the liver tumor. The choice of a liver resection should be guided by the need to obtain a negative margin around the tumor and the need to preserve functional liver parenchyma. The administration of intravenous contrast can give additional information, as scanner timing can be coordinated to image the liver during hepatic arterial and portal venous phases of the contrast. Contrast-enhanced scans reveal the relationships of the tumor to the hepatic veins, inferior vena cava, and portal structures. These relationships are pivotal, as an anatomic liver resection is defined by the preservation or sacrifice of hepatic veins and portal structures. Computed tomographic scans may not reveal with certainty whether a tumor merely abuts or truly invades a vascular structure, but it does reveal the proximity of this relationship. Contrast CT scanning may reveal aberrant hepatic arterial anatomy, although it is not as accurate as conventional arteriography or intraoperative exploration. The presence of aberrant arteries, if noted, is helpful for operative planning. In addition, preoperative CT scans inform the surgeon about possible involvement of the tumor with extrahepatic structures (e.g., vena cava, diaphragm, retroperitoneum, kidney, stomach, and regional lymph nodes). A chest CT scan should be performed in children with potentially malignant liver tumors in order to evaluate for pulmonary metastases.



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Magnetic resonance imaging (MRI) can be useful for the evaluation of large, complex, or multifocal liver tumors. It may more accurately delineate hemangiomas than CT, as some hemangiomas have a radiodensity similar to that of normal liver parenchyma. When combined with magnetic resonance angiography (MRA), it reveals the relationship of the tumor to the major vascular structures of the liver and can reveal aberrant hepatic arterial anatomy. The resolution of hepatic MRA compared to conventional angiography has not been extensively studied in children. In addition, magnetic resonance cholangiopancreatography (MRCP) can be helpful in elaborating the patient's biliary anatomy in cases where a biliary reconstruction is anticipated, such as the patient with a previous Kasai procedure for biliary atresia.

Ultrasonography is often performed early in the work-up of a patient with abdominal or biliary symptoms. It can show the size and extent of the liver tumor, but the static images are less helpful for surgical planning of an anatomic resection than are the images from CT. Duplex ultrasonography can be helpful, however, if invasion or patency of the hepatic veins or vena cava is questioned.

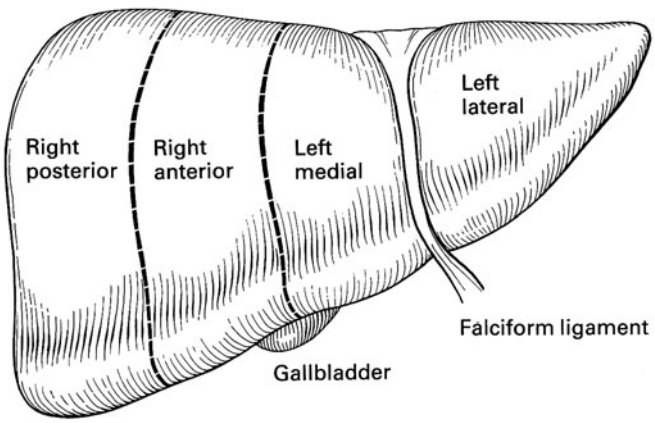
Conventional angiography is no longer routinely performed in patients with liver tumors. It is most useful in a patient with a vascular malformation for the performance of angioembolization as primary therapy. Conventional angiography may also be helpful to determine the hepatic arterial anatomy in cases where a hepatic artery infusion pump may be placed.

Preoperative laboratory investigation should be guided by the patient's clinical situation. Most children with liver tumors have normal liver function. A complete blood count and clot for cross-match are useful for all children. Anemia and thrombocytopenia should be identified and corrected prior to operation, and two units of cross-matched red blood cells should be available at the time of operation. Children who undergo neoadjuvant chemotherapy are more prone to anemia and thrombocytopenia as a result of bone marrow suppression. These children should also have a complete chemistry panel preoperatively, as platinum-based regimens can be nephrotoxic. Children with underlying parenchymal liver disease should have liver function tests as well as a prothrombin time and partial thromboplastin time prior to operation. Impairment of hepatic synthetic function may lead to a potentially correctable coagulopathy. Alpha-fetoprotein is a tumor marker that is expressed by most hepatoblastomas and HCCs. A baseline AFP level should be obtained for all children with these tumors, as it can be an indicator of tumor response to therapy, and it can be an early marker of tumor recurrence.

Prior to liver resection, a thorough and honest discussion should be held with the patient and family. As the liver is a highly vascular organ, the risk of intraoperative hemorrhage should be discussed. Although the risk of hemorrhage is related to the extent of resection, it is not unheard of to lose up to a blood volume of 80 mL/kg in a major liver resection. The likelihood and risks of transfusion should also be acknowledged. Despite the accuracy of current imaging, the most accurate assessment of tumor resectability is operative exploration, and the possibility of an unresectable tumor should be discussed preoperatively, particularly if imaging suggests possible hepatic vein or inferior vena cava involvement. Biliary injury, due either to direct mechanical injury or to indirect ischemic injury, should be mentioned. Postoperative risks include hemorrhage requiring transfusion or re-exploration, infectious complications (wound infection, subphrenic abscess, infected biloma), bile leak, pulmonary complications, wound complications, and long-term complications (hernia or adhesive small bowel obstruction). Postoperative hepatic insufficiency is rare, but it should be mentioned prior to liver resection in patients with cirrhosis. Overall mortality related to major liver resection should be less than 5 percent.

Anatomy

2 The ligamentum teres is the fibrotic remnant of the fetal umbilical vein, and it runs from the umbilicus within the falciform ligament, through the umbilical fissure, to the porta hepatis, where it joins the left portal vein. Similarly, the ligamentum venosum is the remnant of the fetal ductus venosus and travels from the left portal vein to the left hepatic vein. These two ligaments, along with the falciform ligament anteriorly separate the morphologic left and right lobes. The falciform ligament extends to the diaphragm, where it is continuous with the coronary, left triangular, and right triangular ligaments. These ligaments demarcate the bare area on the dome of the liver. The true anatomic left lobe and right lobe are separated by an imaginary line (Cantlie line) that runs from the gallbladder fossa to the suprahepatic vena cava. The middle hepatic vein lies in this plane. The anatomic left lobe is divided into a left lateral sector and a left medial sector by the umbilical fissure and falciform ligament externally and by the left hepatic vein internally. The right lobe is divided into a right anterior sector and a right posterior sector by the right hepatic vein.



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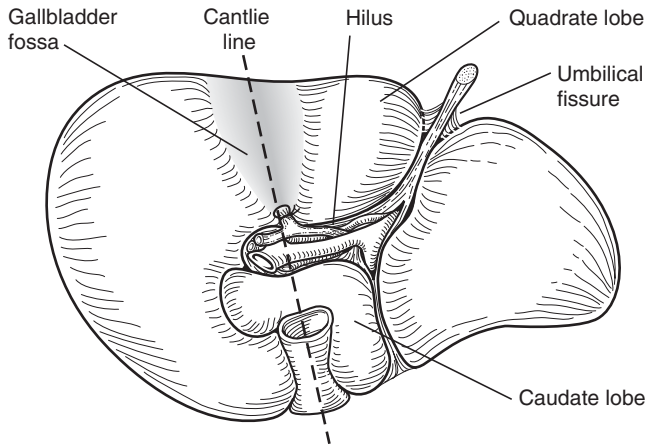
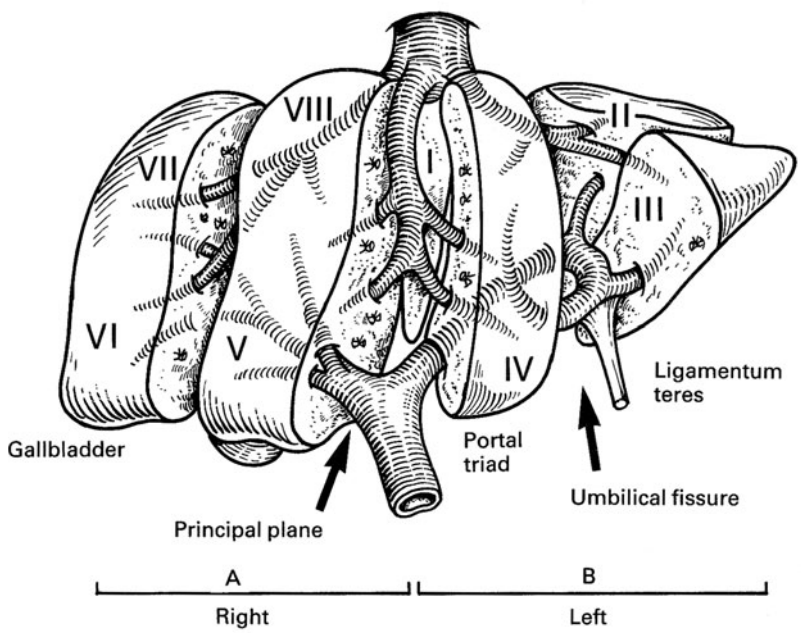


2b

3 The liver can be further divided into eight anatomic segments. Each segment is supplied by a portal pedicle, and each drains into a tributary of a hepatic vein. Segment I is synonymous with the caudate lobe. It is supplied by branches off the left and right portal triads, and its venous drainage is directly into the inferior vena cava. Segments II and III comprise the left lateral sector, and segment IV comprises the left medial sector. Segments V and VIII form the right anterior sector, and segments VI and VII form the right posterior sector. Appreciation of the segmental anatomy of the liver serves as the basis for liver resection, as it is preferable to obtain extrahepatic control of vascular structures prior to the division of liver parenchyma. Although lesser resections may be performed on the basis of this anatomy, the five most commonly performed liver resections are right hepatectomy (right lobectomy), left hepatectomy (left lobectomy), extended right hepatectomy (right trisegmentectomy), extended left hepatectomy (left trisegmentectomy), and left lateral lobectomy (left lateral segmentectomy).



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Anesthesia

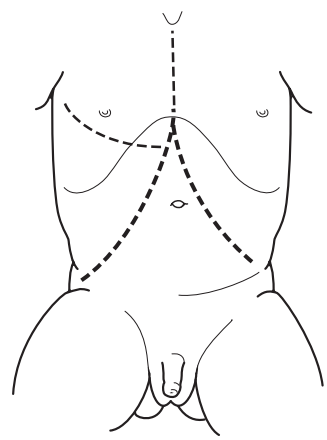
A general endotracheal anesthetic is administered. In appropriate patients, an epidural catheter is placed to facilitate analgesia in the intraoperative and postoperative periods. A Foley catheter is placed for intraoperative monitoring of urine output. A nasogastric tube is placed to decompress the patient's stomach.

In light of the possibility of significant hemorrhage, vascular access is secured in the form of two large-bore intravenous lines in the upper extremities or neck. It is important to secure vascular access above the nipples in case the inferior vena cava needs to be cross-clamped for total hepatic vascular exclusion. Central venous catheters in the internal jugular or subclavian veins may be helpful for this purpose. In addition, central venous pressure (CVP) monitoring can help the anesthesiologist with intraoperative fluid management. While we do not restrict the intraoperative CVP to a pressure of 5 mmHg or less, we do favor judicious infusion of intravenous fluids as long as the patient is well perfused. Avoidance of elevated CVP may help to reduce blood loss from tributaries to the hepatic veins and the inferior vena cava. A radial arterial line is placed for continuous blood pressure monitoring. This line is also useful in blood sampling for arterial blood gases, blood counts, and coagulation profiles during the operation.

Hypothermia can have deleterious effects on coagulation, particularly in the face of a large incision and significant blood loss. The patient's temperature requires close attention throughout the operation. Normothermia is maintained through ambient room temperature, warming blankets, and infusions of warmed fluids and blood products.

OPERATION

4 The patient is placed supine on the operating table with a small rolled towel serving as a bump under the upper lumbar spine. The operating table should be placed in a slight Trendelenburg position, to minimize the risk of air embolism. The table can be rotated with the patient's right side up to facilitate exposure. A single dose of a first-generation cephalosporin is given prior to incision. In the event that a median sternotomy or thoracoabdominal extension is needed to expose the tumor or obtain control of the inferior vena cava within the pericardium, the patient's abdomen and chest are both prepared and draped. A right subcostal incision is made one finger's breadth below the right costal margin. It is extended across the midline in a gently curved chevron below the left costal margin. The incision extends from the lateral border of the left rectus sheath to the right anterior axillary line. This incision allows for an extension in the superior midline if additional exposure is needed. It can be combined with a median sternotomy if it is difficult to control the inferior vena cava within the abdomen.



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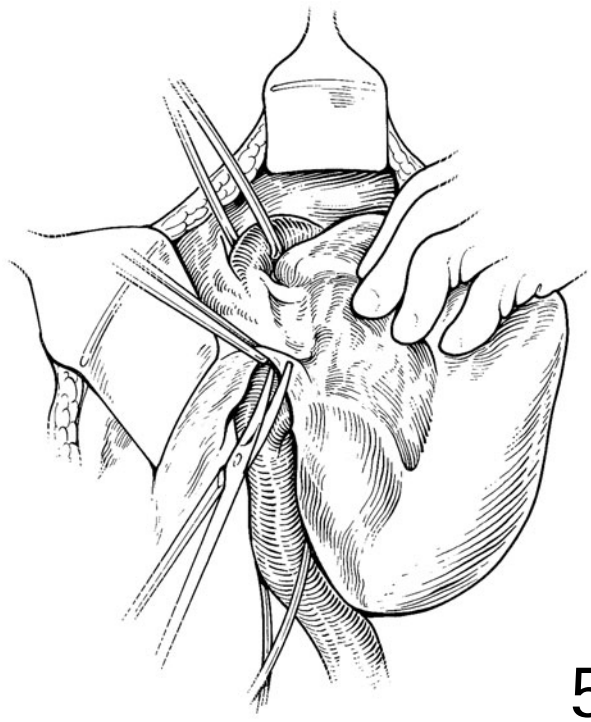
Dissection

5 Upon entry into the peritoneal space, the abdomen is explored by palpation and inspection. Particular attention is paid to the size and location of the primary tumor, adhesion to adjacent structures, enlarged lymph nodes, or additional tumor elsewhere in the liver. The ligmentum teres is ligated and divided. The liver is mobilized by sharply dividing the falciform ligament along its course up to the diaphragm. This dissection is continued along the coronary, left triangular, and right triangular ligaments in order to free the liver from the diaphragm. A self-retaining retractor is used to optimize exposure. The liver is manually retracted inferiorly to expose the suprahepatic inferior vena cava. The anterior and lateral surfaces of the inferior vena cava are dissected free of loose connective tissue. The liver is then rotated to the patient's left side to expose the posterolateral aspect of the inferior vena cava. A blunt instrument is used to dissect circumferentially the inferior vena cava and encircle it with an umbilical tape above the hepatic veins. Some dissection of the hepatic veins outside of the liver can be performed at this point, but, in general, circumferential dissection of the hepatic veins is postponed until the portal dissection is completed. Similar to the dissection above the liver, the inferior vena cava below the liver is circumferentially dissected and encircled with an umbilical tape above the level of the renal vein. This maneuver should be performed with care not to injure the lumbar veins as they enter the inferior vena cava, as bleeding from these veins can be difficult to control. A window is created into the lesser sac adjacent to the porta hepatis. The hepatogastric ligament is inspected and palpated for an aberrant left hepatic artery. The portal structures are encircled with a vascular tape. This allows for occlusion of the portal vein and hepatic artery (Pringle maneuver) if this is needed for the control of hemorrhage. Palpation along the posterior surface of the portal vein may reveal the pulse of an aberrant right hepatic artery.



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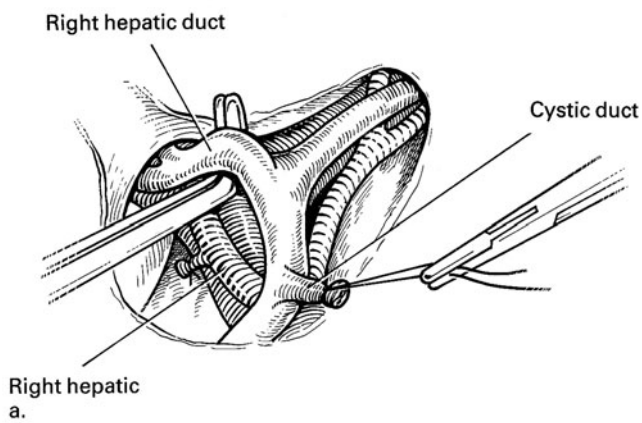
Right hepatectomy

6a–d Portal dissection is begun with a cholecystectomy. The cystic artery and cystic duct are divided between ligatures, and the cystic duct stump is followed to its confluence with the common hepatic duct. The common hepatic duct is followed into the hilum of the liver, where it bifurcates into right and left hepatic ducts. During this dissection, care is taken not to dissect the common hepatic duct extensively or circumferentially in order to minimize disruption of the blood supply of the duct, with the subsequent risk of ischemic stricture. The right hepatic duct is circumferentially dissected and divided between ligatures. The right hepatic artery runs just deep to the right hepatic duct. It is circumferentially dissected, ligated, and divided. The right portal vein is the most posterior structure of the portal triad. It is circumferentially dissected and occluded proximally and distally with vascular clamps. The vein is divided between the clamps, leaving a longer cuff on the side adjacent to the confluence. This end is suture ligated with a running 6/0 polypropylene suture. The hepatic side of the divided right portal vein is suture ligated with 4/0 silk. Once the right portal structures are divided, the right lobe of the liver will become ischemic, with a line of demarcation extending up the liver towards the vena cava, along the path of the middle hepatic vein.



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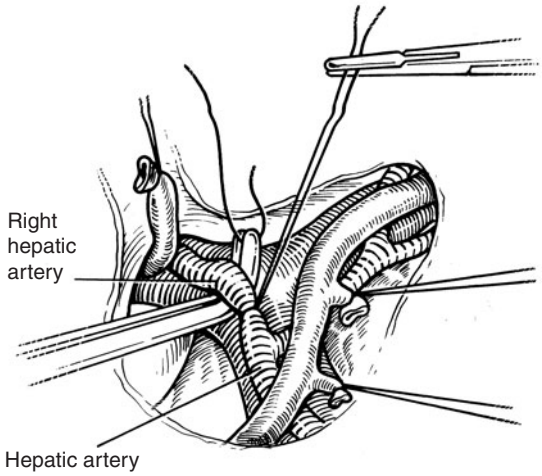

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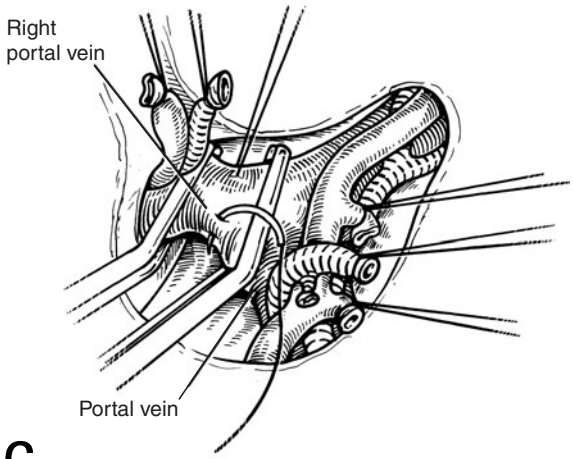

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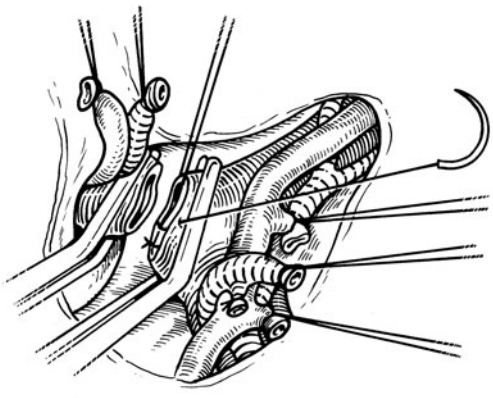
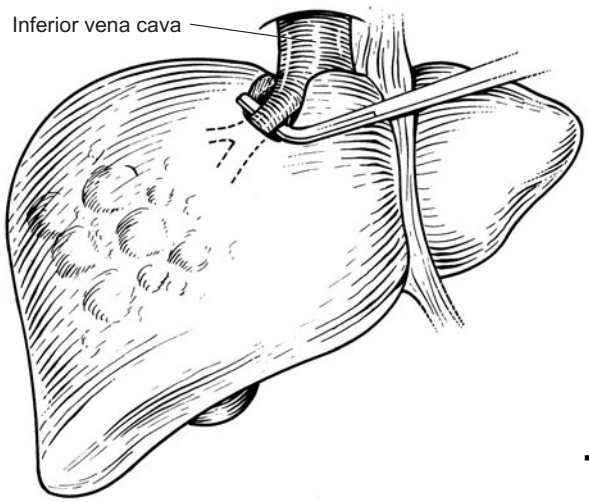


6d

7 Dissection and division of the right hepatic vein require great care, as an injury to the vein or adjacent vena cava can be accompanied by significant bleeding, which may be difficult to control. The course of the right hepatic vein may be very short as it exits the liver and enters the vena cava. If tumor is not immediately adjacent to the hepatic vein, additional length can be obtained by cautiously dividing a small amount of liver parenchyma that overlies it. The right hepatic vein is circumferentially dissected and divided between vascular clamps, and the cuff on the vena cava side is suture ligated with a running 4/0 polypropylene suture. The hepatic side is over-sewn or secured with a 2/0 silk.



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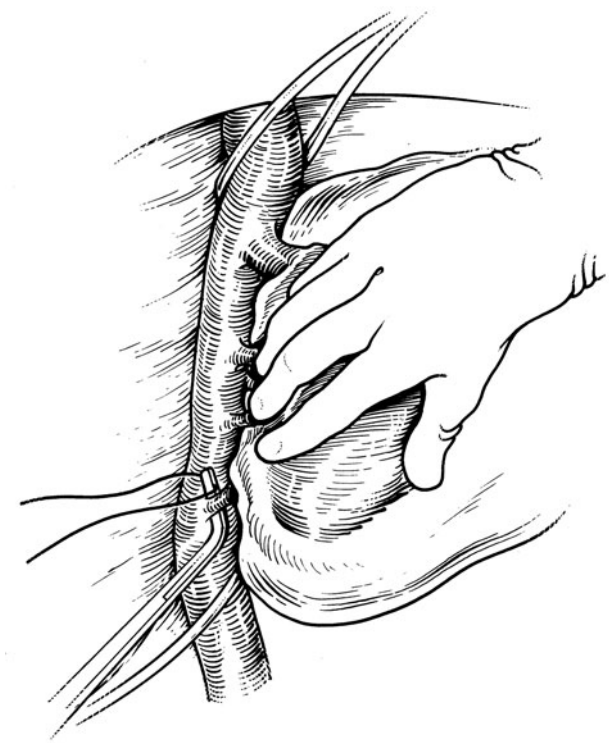

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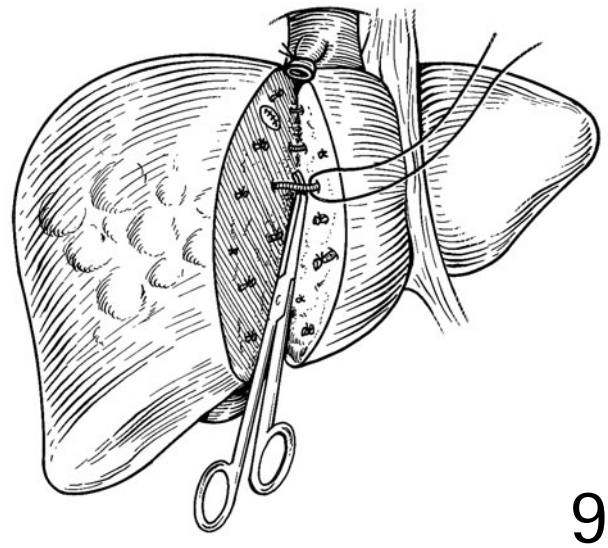
8 The right lobe of the liver is retracted to the patient's left side, to expose the length of the inferior vena cava. Numerous unnamed hepatic veins empty directly into the vena cava from the right lobe of the liver. These veins are divided between 4/0 silk ties, beginning with the most inferior veins and progressing superiorly toward the main hepatic veins. If the right hepatic vein cannot be safely controlled from an anterior approach, it can be approached from this posterolateral exposure. With the unnamed hepatic veins divided, the liver can be retracted anteriorly away from the inferior vena cava. A right-angle is used to dissect cautiously along the medial aspect of the right hepatic vein. With the vein circumferentially dissected, a vascular clamp is placed on the vena cava side of the right hepatic vein. A 2/0 silk ligature is placed along the hepatic side of the vein, and the vein is divided. If the cuff on the vena cava side is long enough, the vein is suture ligated with a running 4/0 polypropylene suture. If the cuff is not long enough, a second vascular clamp is placed beneath the first, flush with the inferior vena cava. The first clamp is removed, revealing additional venous cuff for suture ligation. If bleeding is encountered during these maneuvers, vascular clamps can be applied to the inferior vena cava, directed by the previously placed umbilical tapes. A Pringle maneuver may be performed to occlude inflow to the left lobe of the liver, and thereby decrease blood flow through the left and middle hepatic veins.

9 At this point the major vascular structures to the right lobe of the liver have been divided, and the parenchymal dissection is begun. Glisson's capsule is scored in the interlobar fissure using the electrocautery. The surgeon and first assistant compress the hepatic lobes adjacent to the line of transection during the division of the parenchyma. Although different methods may be used to divide the hepatic parenchyma, we favor the use of the ultrasonic scalpel, as it seals small vessels and ducts as it divides the parenchyma. Only modest amounts of liver parenchyma are divided with each application of the ultrasonic scalpel. This piecemeal transection exposes larger vessels and ducts. Vessels and ducts larger than 5 mm are ligated with 4/0 silk ties. Hepatic venous tributaries from segments V and VIII are encountered before they join the middle hepatic vein. These are divided between suture ligatures. Care is taken to avoid injury to the middle hepatic vein during the transection of the liver parenchyma. The parenchymal dissection tends to be accompanied by ongoing blood loss, and if this becomes excessive, a Pringle maneuver can be performed.



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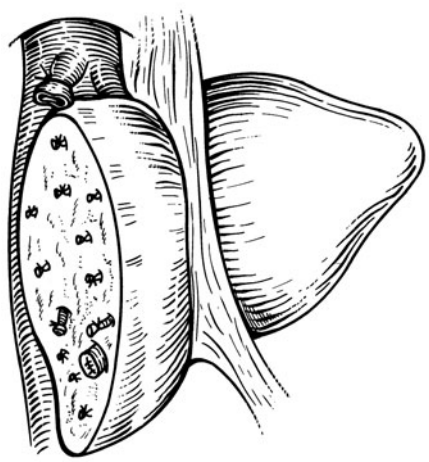

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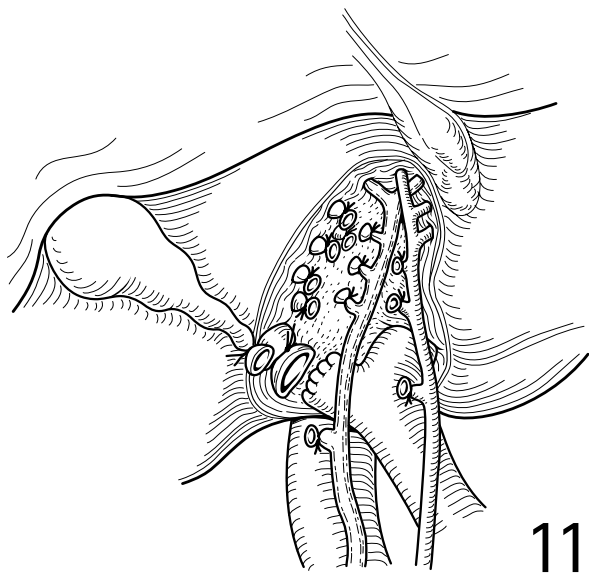
10 With the right lobe of the liver removed, attention is directed to the cut surface of the left lobe. Points of bleeding or bile leakage that were not controlled during the parenchymal dissection are closed with 4/0 silk suture ligatures. The argon beam coagulator is applied to the raw surface to coagulate points of bleeding not amenable to ligation. Horizontal mattress sutures to compress the liver edges are not routinely used. Topical hemostatic agents are not routinely used either, but these may be useful to reduce the diffuse ooze of blood from the cut surface while the coagulopathy is being corrected. A clean surgical sponge is packed against the cut surface during the remainder of the operation until just prior to closure of the abdomen. This sponge acts as a monitor for a small bile leak that may not be apparent on initial inspection. When the sponge is removed, it is inspected for small bile staining, which would direct the surgeon to a point on the cut surface for suture ligation. The left lobe of the liver is allowed to lie in its 'resting' position. As long as the remaining hepatic venous and portal structures are without kinks or twists, no hepatopexy to the diaphragm or retroperitoneum is performed.

Extended right hepatectomy

This operation is used for large right-sided tumors that extend across the interlobar fissure but do not involve segments II and III (the left lateral segment). The conduct of this operation is similar to the right hepatectomy, but segments I and IV are resected in addition to segments V through VIII.

11 As with the right hepatectomy, the portal dissection proceeds with division of the right hepatic duct, the right hepatic artery, and the right portal vein. Dissection proceeds along the anterior surface of the left hepatic duct towards the falciform ligament. This dissection is carried out within Glisson's sheath, which surrounds the portal structures as they enter the liver and bifurcate within it. The overlying hepatic parenchyma may need to be divided to facilitate this exposure. Branches of the portal structures come off the main trunks superiorly to enter segment IV. Branches to the caudate lobe come off posteriorly. These superior and posterior branches are ligated and divided from the main trunks up to the falciform ligament and the umbilical fissure. The left hepatic duct is particularly vulnerable, and it must be painstakingly preserved. No structures are divided to the left of the umbilical fissure and falciform ligament, in order to minimize the risk of damage to the left hepatic vein, which runs deep to these structures. The ligamentum teres serves as a posterior landmark that leads to the left portal vein. As these structures are divided, a zone of ischemia should develop, extending to but not beyond the umbilical fissure. The right hepatic vein is divided as previously described. Veins from the caudate lobe that empty directly into the inferior vena cava are divided as well. The middle hepatic vein typically joins the left hepatic vein within the liver before emptying into the inferior vena cava. Thus, extrahepatic ligation of the middle hepatic vein is seldom possible. It is secured during the parenchymal dissection. The liver parenchyma is divided to the right of the falciform ligament and umbilical fissure. This division proceeds superiorly towards the inferior vena cava. During the upper part of this dissection, the middle hepatic vein is exposed. At this point, it is divided and suture ligated. The parenchymal dissection is completed and the specimen is removed. Because of its small size, the remaining liver (segments II and III) may be more prone to torsion than a larger section of liver would be. This may be especially true if this section of liver is supplied by an aberrant left hepatic artery off the left gastric artery or celiac trunk. The falciform ligament and the ligamentum teres may be tacked to the diaphragm and retroperitoneum to maintain the liver in a neutral position.



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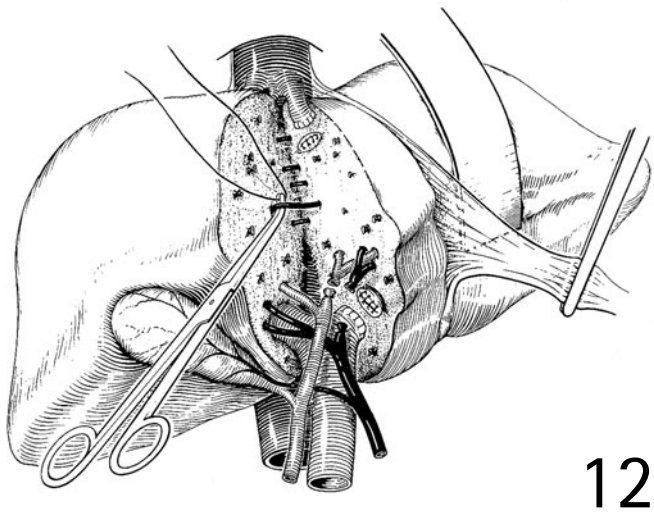
Left hepatectomy

12 Initial dissection proceeds in a similar fashion to the right hepatectomy. Cholecystectomy does not necessarily need to be performed, but it is customarily done. The portal dissection proceeds, and the left hepatic duct, the left hepatic artery, and the left portal vein are divided near their bifurcations. If the caudate lobe is to be preserved, the left portal structures are divided distal to the posterior branches to the caudate lobe. Division of the vascular inflow to the left lobe should produce a zone of ischemia to the left of the interlobar fissure. The gastrohepatic omentum is explored, and aberrant left hepatic artery is ligated if present. The remainder of the gastrohepatic ligament is divided to permit full mobilization of the left lobe. With the liver rotated anteriorly and to the patient's right side, dissection along the ligamentum venosum will lead to the left hepatic vein. The left hepatic vein is explored. If the left hepatic vein passes outside the liver before it becomes confluent with the middle hepatic vein, it can be divided and suture ligated prior to the parenchymal dissection. If the two veins join within the liver, the extrahepatic confluence can be encircled with a vascular tape by passing a right-angle to the right of the termination of the ligamentum venosum, at the top of the caudate lobe. This can be used to occlude the hepatic veins if significant hepatic venous bleeding is encountered during parenchymal dissection. Division of the hepatic parenchyma proceeds just to the left of the interlobar fissure. The middle hepatic vein is preserved, and tributaries from segment IV are suture ligated as they are encountered. As the parenchyma is divided superiorly, the left hepatic vein is identified before its confluence with the middle hepatic vein. The left hepatic vein is suture ligated and the middle hepatic vein is preserved. The parenchymal dissection is then completed and the specimen is removed.



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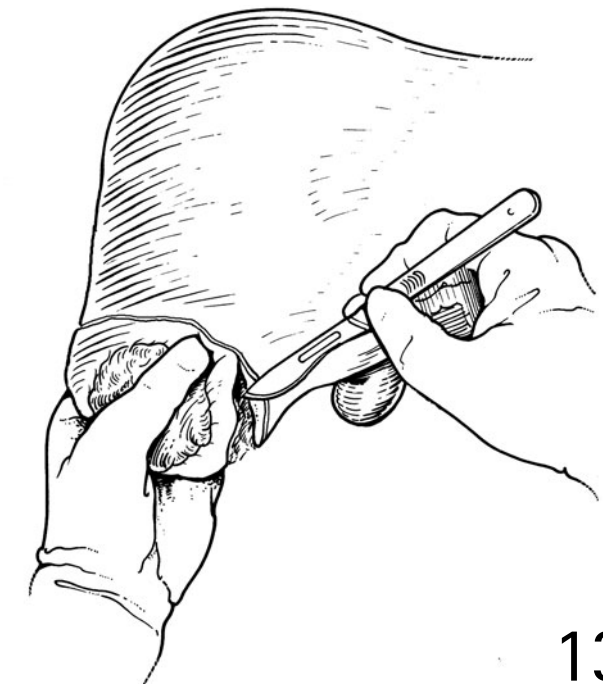
Non-anatomic liver resections

Wedge resections are generally performed for small, peripheral, or incidentally discovered liver tumors. They are usually done for diagnostic purposes. However, in certain circumstances, a wedge resection with an appropriate margin (generally 1 cm) may be used as a therapeutic operation. Large wedge resections and central wedge resections are discouraged, as bleeding may be excessive and difficult to control deep in the apex of the parenchymal defect. Inadvertent division of large intrahepatic vessels may unnecessarily jeopardize segments of the liver distant to the site of resection.

13 Because wedge resections are generally performed for smaller tumors, the liver is typically not mobilized extensively. The surgeon should not hesitate to enlarge an existing incision or divide ligaments of the liver if doing so would facilitate safe control of the liver during the resection. Dissections of the inferior vena cava and hilum of the liver are not performed in order to obtain segmental vascular control. In the case of a modest sized tumor (2–5 cm), the porta hepatis may be dissected in the hepatoduodenal ligament in order to perform a Pringle maneuver if needed. Wedge resections are usually performed without the use of instruments such as the argon beam coagulator or ultrasonic scalpel, which may be used in anatomic liver resections. The peripheral liver tumor is grasped and gently retracted. The liver capsule is divided in the path of intended resection using the scalpel or electocautery. The liver is manually compressed proximal to the margin of resection. Small lesions near the edge of the liver are sharply excised with the scalpel. The use of electocautery for excision of the specimen is discouraged, as thermal artifact may adversely affect subsequent histology. The parenchyma around larger lesions is fractured with a Kelly clamp. Vessels and ducts larger than 5 mm are ligated in continuity and divided. The cut surface of the liver is coagulated with the electrocautery. The current on the cautery can be increased to create an electrical arc to the liver surface. This facilitates coagulation of the surface without the cautery tip adhering to the coagulum from direct contact.



Wound closure

A closed suction drain is brought through a stab incision below the subcostal incision. The drain is placed adjacent to the cut surface of the liver. The purpose of this drain is to identify and potentially control a delayed bile leak. The field is irrigated with warm saline. The abdominal wall is closed in two layers with running, absorbable, monofilament sutures. The skin is closed with a running, subcuticular, absorbable, monofilament suture.

POSTOPERATIVE CARE

Following major liver resection, patients are admitted to the pediatric intensive care unit (ICU) for continuous hemodynamic monitoring. Intraoperative resuscitation is continued in the ICU to target a temperature > 36.5°C, urine output of 0.5–1 mL/kg per hour, hematocrit > 22 percent, platelets > 100 000/μL, and International Normalization Ratio (INR) < 1.5. Over-resuscitation is avoided, and a CVP < 8 mmHg is preferred, provided other indicators of perfusion are appropriate. The closed suction drain is monitored for volume and quality of output. The drain is kept in place to monitor for bile leak until the patient is tolerating a regular diet. The nasogastric tube is continued until abdominal distension resolves and appetite returns, at which time a clear liquid diet is started. Perioperative antibiotics are continued for 24 hours following operation. An epidural catheter is the preferred means of postoperative analgesia. It is continued until the patient is able to tolerate oral analgesics. The patient's Foley catheter is removed once the epidural is turned off.

Complications

Hemorrhage is the most potentially catastrophic postoperative complication. It should be suspected in a patient with poor perfusion, labile hemodynamics, inappropriate response to transfusion, or failure to improve with resuscitation in the ICU. The closed suction drain is not a reliable indicator of intra-abdominal hemorrhage. Significant bleeding can occur even when small volumes of serosanguinous fluid are noted in the drain. The surgeon should maintain a low threshold for operative re-exploration and accept the possibility of a negative exploration rather than delay the operative control of significant bleeding.

Metabolic derangements are unusual following liver resection in children with normal preoperative liver function. Hypoglycemia may occur for the first few days following a large liver resection, and this can be treated with 10 percent dextrose until hepatic glucose metabolism returns to normal. Patients with an elevated prothrombin time postoperatively

may benefit from the administration of exogenous vitamin K. Hepatic function may be transiently impaired and patients may have delayed clearance of medications such as analgesics and sedatives, which are metabolized by the liver. Hepatic function tests are abnormal in the early postoperative period and may not return to normal until several weeks following operation. Hepatic regeneration is remarkable, and most patients will have a normal liver volume within 3 months after liver resection.

Fever in the postoperative period can have a number of causes. Necrotic liver at the margin of the resection can cause fever. As long as the necrosis is not extensive or infected, the symptoms are self-limiting. An undrained bile leak can cause fever, whether or not it is infected. Once drained, almost all bile leaks stop over time without any further intervention. Prolonged or recurrent fever approximately 1 week after resection should raise the suspicion of a subphrenic abscess. Once identified, most subphrenic abscesses can be drained percutaneously, with ultrasound or CT guidance.

OUTCOME

Hepatic resections are complex operations that require vigilant care both inside and outside the operating room. Mortality from major liver resection is 5 percent or less in contemporary series. In patients with favorable risk factors, this mortality should be less than 3 percent. Overall outcome is related to the patient's underlying diagnosis. Successful liver resection is generally curative for benign disease. Resection forms the cornerstone of therapy for primary hepatic malignancies. With multimodal therapy, there is currently a 75 percent cure rate at 5 years for hepatoblastoma. This survival rate has been steadily increasing over the last two decades. The use of neoadjuvant chemotherapy has increased the resectability of these tumors and promoted tissue-preserving liver resections in patients whose tumors respond. The outcome for HCC is not as encouraging. Hepatocellular carcinoma is more prone to metastasis and multifocality than hepatoblastoma, and it typically occurs on a background of underlying liver disease. The 2-year survival rate for children with HCC is 41 percent. Survival may be somewhat better with liver transplantation, but limited organ availability prohibits its use as the primary surgical modality for most HCCs. Clearly, much work remains to be done to improve patient outcome with this disease.

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69

Surgical management of Wilms' tumor

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INTRODUCTION

In 1899, Max Wilms described a group of children with kidney tumors and since that time his name has been applied to nephroblastoma. The first nephrectomy for this malignancy was reported by Jessop in 1877 and the first radiation treatment was given in 1915. Farber, in 1956, introduced the chemotherapeutic agent actinomycin.

The dramatic improvement in survival rate is a result of the coordinated use of chemotherapy, surgery, and radiation (from 15 percent with surgery alone to 85 percent plus with combination treatment). Thus, the surgeon must be a member of an oncological team when managing a child with this condition.

Nephroblastoma is the most common solid childhood abdominal tumor and presents as a painless abdominal mass in an otherwise well child. The peak age of presentation ranges between 3 and 6 years and the tumor is bilateral in 6 percent of cases.

PRINCIPLES AND JUSTIFICATION

Indications for surgery

In continental Europe and the UK, all tumors are treated with vincristine and actinomycin before surgery. In the UK, all tumors are biopsied before chemotherapeutic treatment; this does not occur in the rest of Europe. In the USA, radical nephrectomy is still the primary treatment for unilateral operable Wilms' tumor. Secondary deposits, caval tumor, bilateral disease, or tumor in a solitary kidney are indications also for Tru-cut biopsy, chemotherapy, and delayed partial or total nephrectomy.

PREOPERATIVE

Blood

- Full blood picture: to monitor the effects of chemotherapy.
- Coagulation studies: to detect the bleeding disorder von Willebrand's disease – a typical abnormality in Wilms' tumor.
- Liver function tests: performed to monitor the effects of chemotherapy.
- Overall kidney function: measured by serum creatinine.

Urine

A sample is monitored to measure vanylmandelic acid (VMA), which, if present in excess, may denote an intrarenal neuroblastoma.

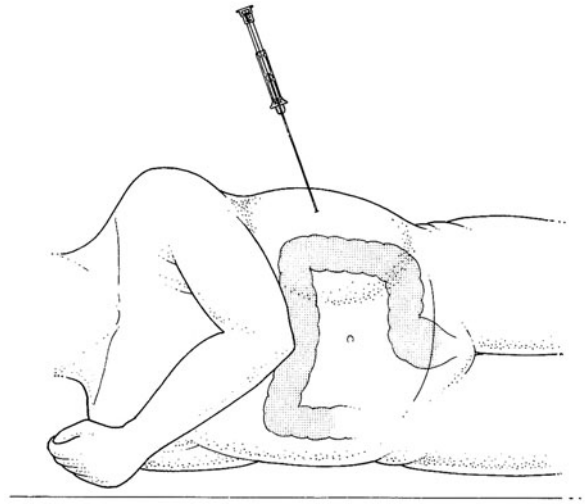
Radiology

An ultrasonographic scan will demonstrate a solid tumor arising from a kidney and possibly extension of the tumor through the renal vein into the inferior vena cava. It will also denote the presence or absence of liver or abdominal secondaries. Initial imaging must include the contralateral kidney to detect bilateral disease. Chest radiography (anterior and lateral) is still performed and is used in the European trials, but computed tomographic (CT) scanning is more sensitive in detecting abnormalities of the contralateral kidney, lymphadenopathy, lymph enlargement, and lung secondaries. Since CT scanning is so sensitive in detecting the presence of a contralateral renal tumor, it is now at the discretion of the surgeon as to whether the contralateral kidney is explored at operation for bilateral disease.

OPERATION

Biopsy of the tumor

1 With the patient in the lateral oblique position, Tru-cut biopsy may be performed with ultrasound guidance. Minimally invasive laparoscopy may have a role in the future in obtaining biopsy specimens under direct vision.



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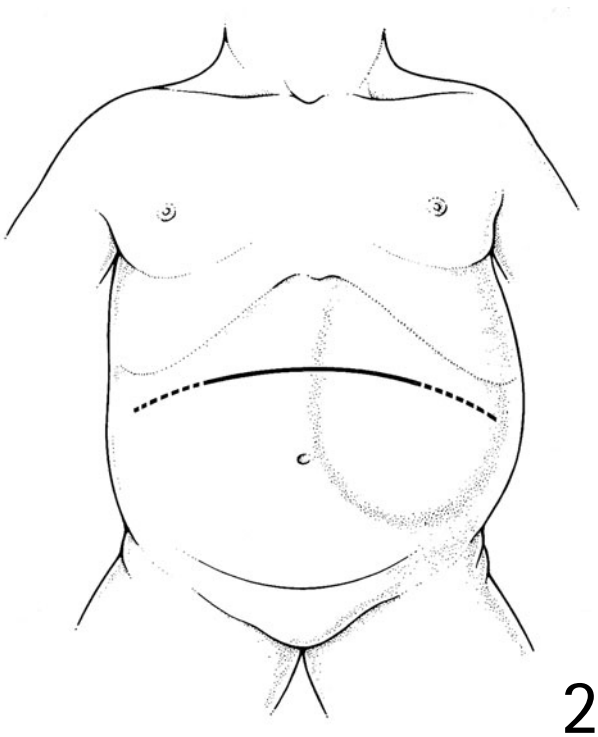

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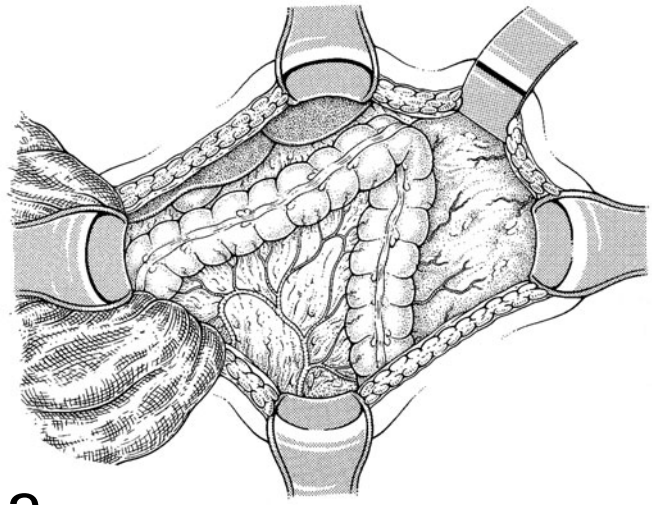
2 A generous transverse upper abdominal incision is required to facilitate complete exposure of the kidneys and easy mobilization of a large renal mass.

The rectus sheath is incised with diathermy. Both rectus muscles are completely divided. The falciform ligament is divided between ligatures. The peritoneum is entered with care to avoid breaching the anterior surface of the tumor, and the incision is extended laterally under direct vision.

3 The abdominal contents are carefully examined for liver and peritoneal secondaries.



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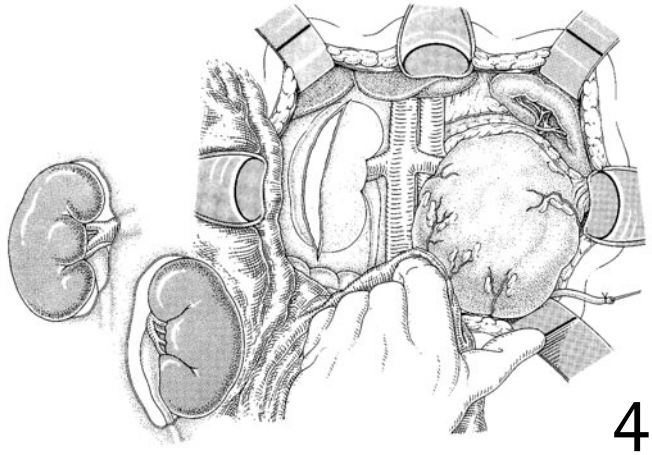
4 The contralateral kidney may be mobilized and examined in both anterior and posterior surfaces to detect possible bilateral disease. If a CT scan is performed, the visual and manual inspection of the contralateral kidney is at the discretion of the surgeon. If unsuspected bilateral disease is detected, the main tumor and contralateral lesion are biopsied with no further excisional surgery. The patient is subjected to chemotherapy with subsequent nephron sparing surgery.



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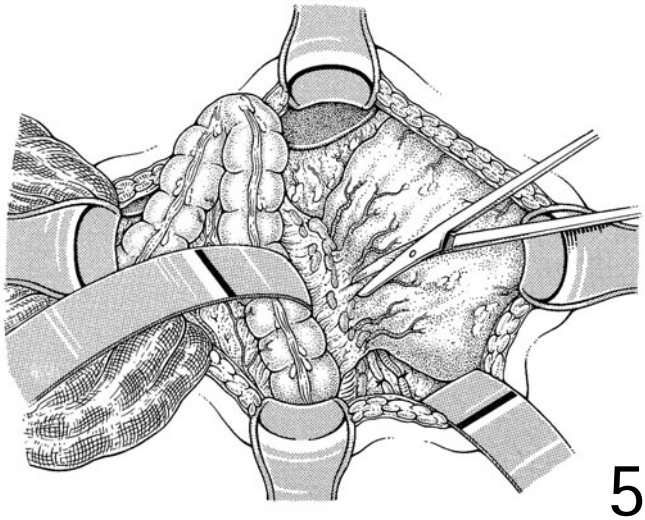

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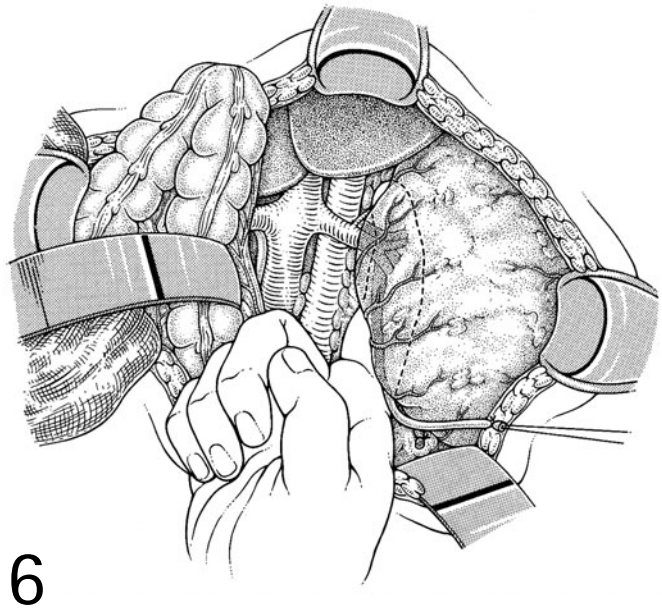
5 The colon with its mesentery is dissected from the anterior surface of the tumor.

6 The ureter and gonadal vessels are ligated and transected. A plane is developed between these structures and the great vessels by gently inserting a finger into the paravertebral space. If possible, the tumor is not mobilized at this stage.



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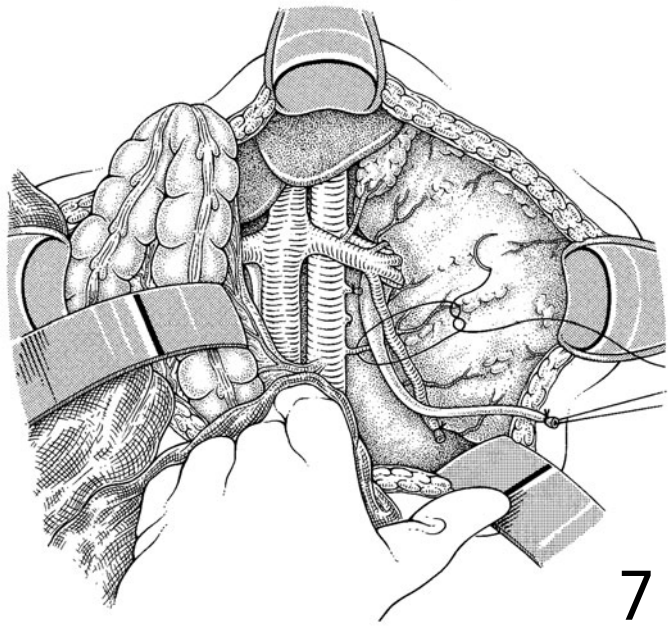
7 Dissection begins caudally, sweeping adventitial tissue and lymph nodes laterally off the great vessels. The renal vein and its branches are gently exposed. Careful palpation of the vein may detect venous extension of the tumor, although this should have been noted on preoperative investigations. Early mobilization of the inferior vena cava and renal vein may be required to prevent embolization of the tumor into the inferior vena cava, heart, and pulmonary artery. On the right side, the second part of the duodenum is encountered during this dissection.



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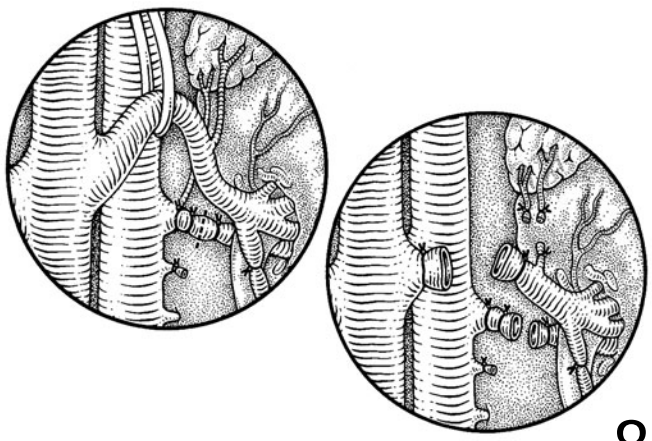

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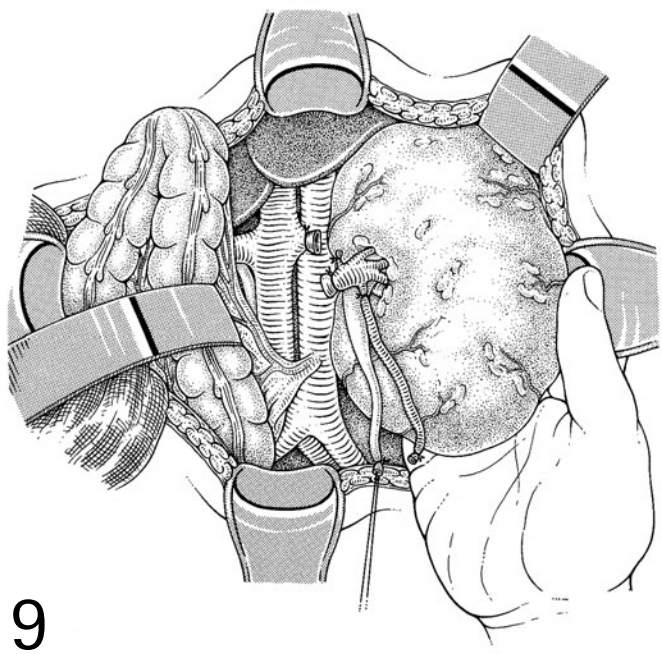
8 The renal vein is gently mobilized and elevated with a vascular sling to expose the renal artery, which is typically situated behind the upper border of the renal vein. It is preferable to ligate and divide the renal artery first so as to prevent swelling of the kidney with arterial blood. Due to the size of the tumor, this sequence of ligation is not always possible.

9 The posterior aspect of the kidney is partially mobilized by blunt dissection.



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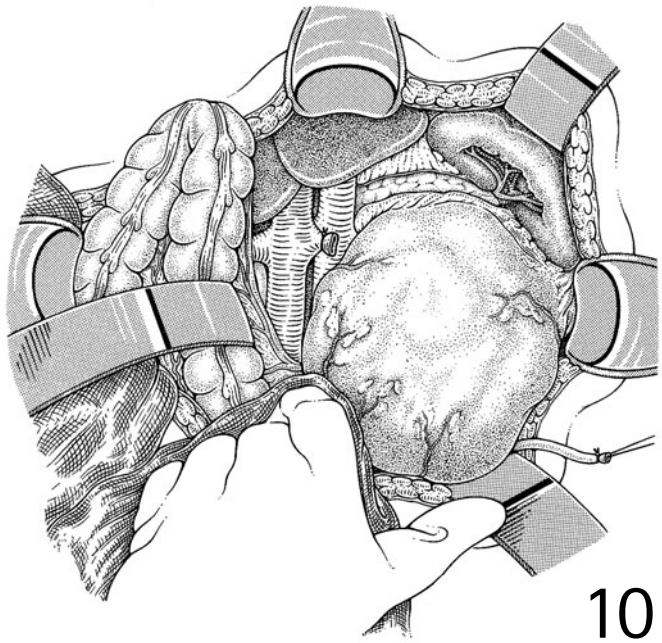
10 The superior dissection is hazardous on the left side, and damage to the spleen and tail of the pancreas must be avoided. The adrenal gland is removed only if the tumor is in the upper pole of the kidney.



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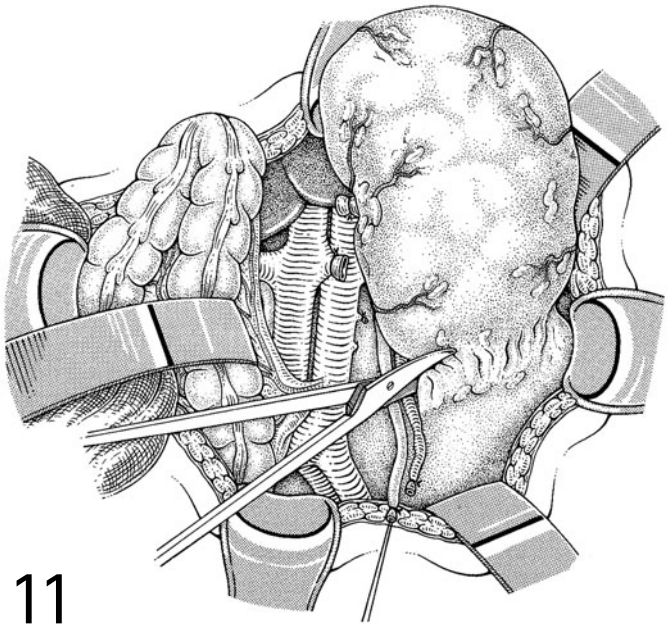

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11 The kidney within Gerota's fascia is lifted out of the abdomen and the posterior dissection is completed under direct vision. The renal bed is inspected following removal of the kidney. Any remaining lymph nodes on the great vessels are removed for histology, and this is essential for adequate staging.

POSTOPERATIVE CARE

Unilateral disease

Vincristine and actinomycin are administered for 4 weeks and surgery is performed on week 5–6. Postoperative chemotherapy is related to the histologic type of tumor and may not be required if there is low-risk histology or may include a combination of actinomycin and vincristine to which is added doxorubicin with or without abdominal radiotherapy for higher grade tumors.

Metastatic disease

Pulmonary metastases documented on X-ray or CT of the chest preoperatively will demand pretreatment with vincristine, actinomycin, and doxorubicin. This will be followed by the same triple chemotherapy after surgery, with or without radiotherapy, if there has been a poor response.

Bilateral disease

Biopsy is only performed in the presence of the bilateral disease, and combination chemotherapy is then administered. Subsequent surgical management may include a total or partial nephrectomy, bilateral partial nephrectomy, or bilateral nephrectomy with dialysis and delayed renal transplantation. Such decisions are complex and require a team approach.

CONCLUSION

The management of Wilms' tumor consists of chemotherapy combined with surgery and, less frequently, radiotherapy. Radiotherapy is mainly administered in the presence of high-risk tumors, local spillage, or secondaries in the lung that do not respond adequately to chemotherapy. The management of Wilms' tumor includes a team consisting of a surgeon, oncologist, and histopathologist.

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Neuroblastoma

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HISTORY

Neuroblastoma is the commonest abdominal malignancy of childhood, the annual incidence being about six cases per million children. These tumors arise in primitive precursors of the sympathetic nervous system and consequently are seen in the adrenal medulla and sites of sympathetic ganglia. Almost one-third are detected in the first 12 months of life and a further 50 percent between the ages of 1 and 4 years. Only 5 percent occur in children over the age of 10 years.

The majority of tumors are locally extensive or metastatic at the time of diagnosis. The first successful excision of a neuroblastoma was reported over 90 years ago by Dr Willard Bartlett of St Louis, Missouri, but, until recently, surgery has been helpful only in the management of those with localized disease. With the advent of more effective chemotherapy, primary tumors have been rendered smaller, more fibrotic, and more amenable to resection. Consequently, macroscopic clearance of disease is now a realistic expectation.

PRINCIPLES AND JUSTIFICATION

Clinical presentation

Less than 3 percent of such tumors are detected on antenatal ultrasound. Postnatally, the majority present with non-specific symptoms such as malaise, anorexia, weight loss, fever, and sweating. Bone and joint pain as a result of skeletal involvement may lead to an erroneous diagnosis of juvenile arthritis. Periorbital ecchymosis and proptosis may occur as a consequence of secondary disease in the skull. Less common presentations include massive hepatomegaly in stage 4S disease, or paraplegia as a consequence of spinal cord compression.

The diagnosis is usually suspected on clinical grounds alone. It is confirmed by the finding of elevated levels of catecholamine metabolites in the urine together with histologic examination of tumor biopsy specimens.

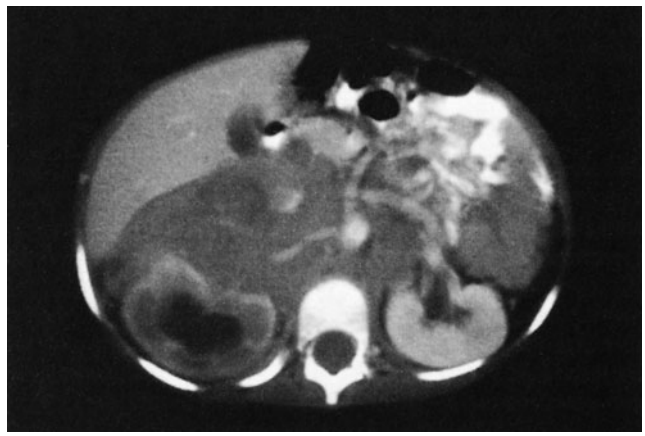
Cross-sectional imaging together with bone scanning and examination of bone marrow aspirates or trephines complete the initial work-up.

Ultrasound examination is almost always performed and is of limited use. Computed tomography (CT) or magnetic resonance imaging (MRI) is essential.

The typical appearance of an abdominal neuroblastoma on CT scan is of an irregular mass arising in the suprarenal or para-aortic region which enhances after the administration of intravenous contrast medium. Cross-sectional imaging will also help to evaluate the extent of vascular encasement, and serial scanning after therapy will document the response to treatment.

1a,b A representative scan is shown in Illustration a in a 4-year-old boy with stage 4 neuroblastoma. The extent of vascular displacement and envelopment by the tumor is readily apparent, with the aorta lifted off the vertebral bodies. In addition, there is bilateral hydronephrosis.

A further scan of the same child after chemotherapy shows the extent of tumor regression and resolution of hydronephrosis (Illustration b).



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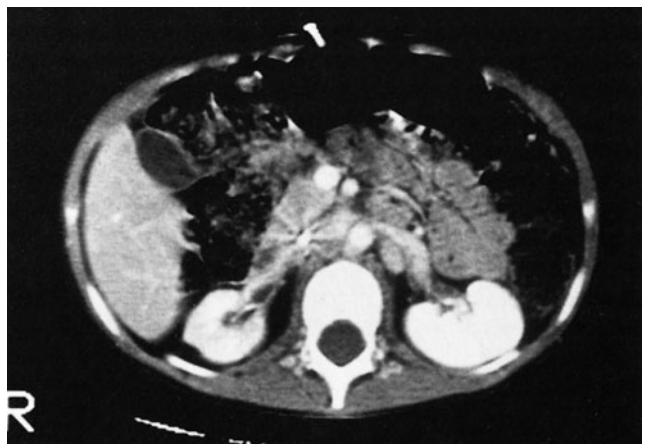

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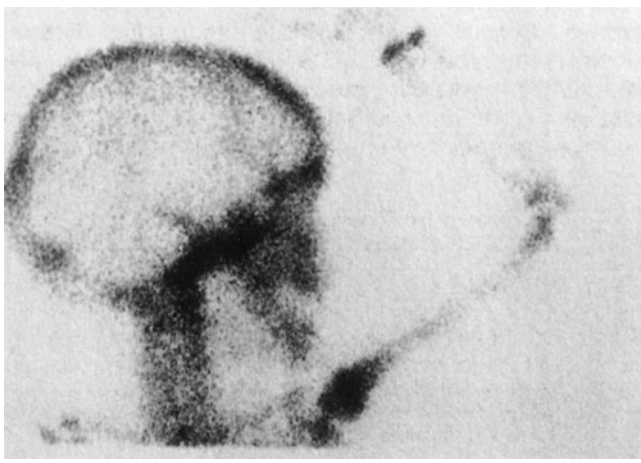
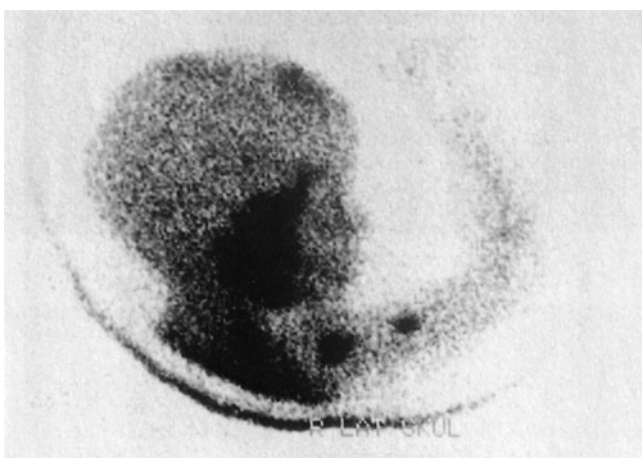


2a



2b

2a,b Radionuclide scanning is also considered essential in the evaluation of distal disease. Skeletal imaging with ^{99m}Tc methylene diphosphonate is more accurate than conventional radiology in demonstrating skeletal metastases. More recently, MIBG scanning (^{131}meta-iodobenzylguanidine) is used for the same purpose. Representative scans from the same child as shown on the CT images are demonstrated in Illustrations a and b. Limb and skull involvement is clearly seen on both images.

Staging

Staging systems are used to document disease extent and to stratify treatment accordingly. The commonest such system in use is the International Neuroblastoma Staging System (INSS, Table 70.1). About 25 percent of patients have stage 1 or 2 disease, 60–70 percent have stage 3 or 4 disease, and about 10 percent present with stage 4S disease.

At the present time, the biological profile of the tumor is not included in the staging system.

Pathology

The typical untreated tumor is rounded or lobulated and of varying consistency. More aggressive tumors are frequently friable and hemorrhagic.

After chemotherapy, tumors are substantially harder and less vascular. Calcification is generally present at the time of diagnosis and may be quite dense after chemotherapy.

Microscopic examination shows sheets of undifferentiated, small, blue nuclei. More differentiated tumors show varying numbers of ganglion cells.

TREATMENT

Surgery is the optimal treatment for those with stage 1 and 2 disease.

Patients with stage 3 and 4 disease are generally managed by initial chemotherapy followed by attempted surgical excision when metastases have been ablated.

Further therapy with bone marrow ablation and bone marrow transplant is used for those with high-risk disease, depending on local chemotherapy protocols.

Treatment protocols continue to evolve and are increasingly directed by adverse biological features.

Surgical management

Early surgery is appropriate for localized tumors where resection can be undertaken safely. For those with locally advanced or disseminated disease, the initial use of chemotherapy followed by surgery allows for a more complete resection.

The operation described here was developed to deal with the problem of vascular encasement. It also allows a planned and systematic approach to tumor excision and minimizes the risk of vascular accidents while achieving complete tumor clearance.

As neuroblastomas rarely invade the tunica media of major blood vessels, the dissection may be performed in the subadventitial plane with a scalpel. The success of the procedure depends entirely on the attitude and persistence of the surgeon.

The operation consists of three phases – vessel display, vessel clearance, and tumor removal. Vessel display is taken to mean the display of part of the circumference of each blood vessel that traverses the tumor, in continuity. If this can be accomplished, these vessels can generally be cleared of tumor. Once the vessels are free, the tumor can be removed piecemeal.

The order in which the vessels are displayed and cleared depends on the site and size of the tumor together with the surgeon's preference.

Table 70.1 *International Neuroblastoma Staging System criteria*

Stage	Definition
1	Localized tumor with complete gross excision, with or without microscopic residual disease; representative ipsilateral lymph nodes negative for tumor microscopically (nodes attached to and removed with the primary tumor may be positive)
2A	Localized tumor with incomplete gross excision; representative ipsilateral non-adherent lymph nodes negative for tumor microscopically
2B	Localized tumor with or without complete gross excision, with ipsilateral non-adherent lymph nodes positive for tumor; enlarged contralateral lymph nodes must be negative microscopically
3	Unresectable unilateral tumor infiltrating across the midline,[a] with or without regional lymph node involvement *or* Localized unilateral tumor with contralateral regional lymph node involvement *or* Midline tumor with bilateral extension by infiltration (unresectable) or by lymph node involvement
4	Any primary tumor with dissemination to distant lymph nodes, bone, bone marrow, liver, skin, or other organs (except as defined for stage 4S)
4S	Localized primary tumor (as defined for stage 1, 2A, or 2B), with dissemination limited to skin, liver, bone and marrow[b] (limited to infants younger than 1 year)

[a] The midline is defined as the vertebral column. Tumors originating on one side and crossing the midline must infiltrate to or beyond the opposite side of the vertebral column.

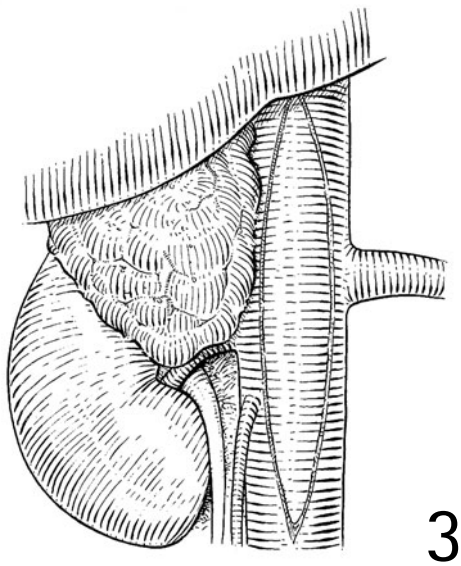
[b] Marrow involvement in stage 4S should be minimal (i.e., < 10% of total nucleated cells identified as malignant on bone marrow biopsy or on marrow aspirate). More extensive marrow involvement would be considered to be stage 4. The meta-iodobenzylguanidine scan (if performed) should be negative in the marrow.

Anesthesia

Full intubated general anesthesia is employed with intravascular pressure monitoring. Epidural anesthesia is often considered to reduce operative blood loss and provides very effective pain relief in the postoperative period.

Operation

3 The abdomen is opened through a transverse supraumbilical incision. For right-sided tumors, the ascending colon and duodenum are reflected medially. The inferior vena cava below the limit of the tumor is exposed by incision of the tunica adventitia longitudinally along the middle of the vessel. The dissection advances proximally, exposing the anterior wall in the 12 o'clock position. If tumor is encountered anterior to the vena cava, it is incised down to the tunica media. This dissection is advanced proximally until the liver is reached.



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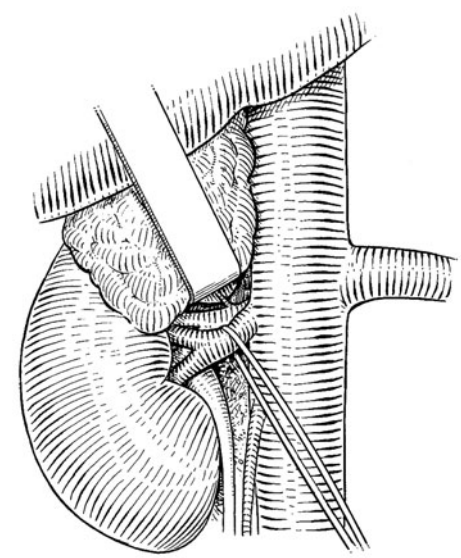
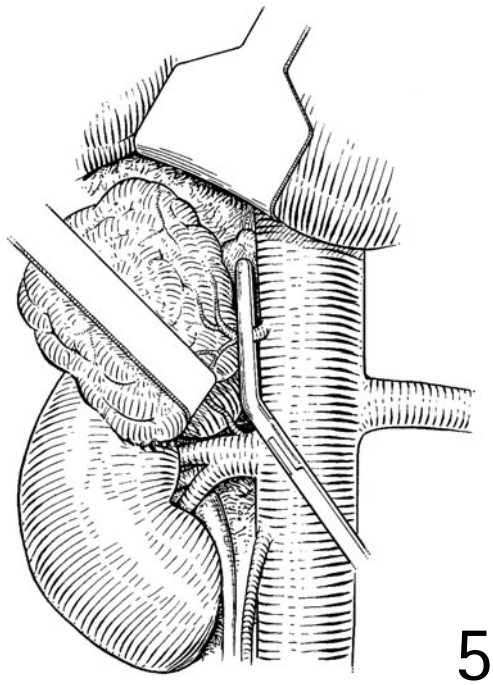


4

4 Subsequently, the right renal vein is displayed in similar fashion. The cava is then cleared of tumor and elevated to expose the right renal artery.

5 Once the right renal artery and vein are displayed, the cava above the renal vein may be cleared of tumor until the right adrenal vein is reached. It is as well to suture ligate this vein before the tumor is removed.



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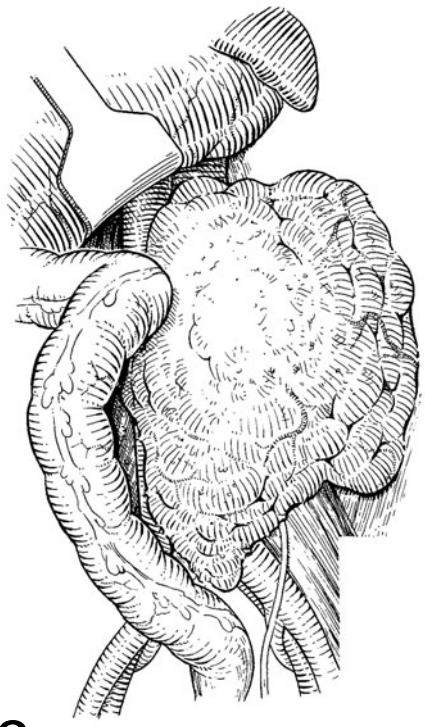
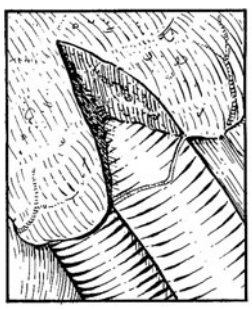


6a

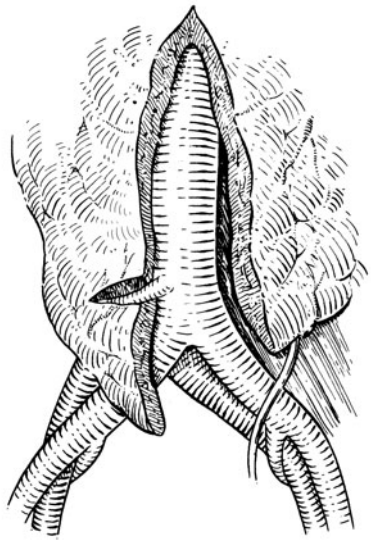
6a For left-sided tumors, the descending and sigmoid colon are mobilized, together with the spleen, pancreas, and stomach, and placed in an intestinal bag. The dissection commences just distal to the tumor edge along the middle of whichever artery is present.

6b The dissection then advances proximally, dividing tumor and adventitia down to the tunica media.



6b

7 Once the left renal vein is reached, a segment 4–5 cm in length is cleared of tumor in the same fashion until it can be retracted both superiorly and inferiorly. The dissection then continues along the aorta until the origin of the left renal artery is seen. The plane of dissection alters to the 2 o'clock position, the knife still coming on the wall of the aorta in perpendicular fashion. The dissection proceeds along the aorta until the upper limit of the tumor is traversed, frequently in the lower mediastinum.



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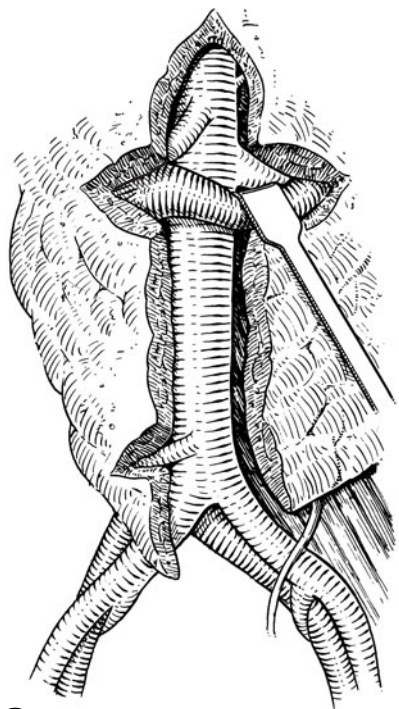
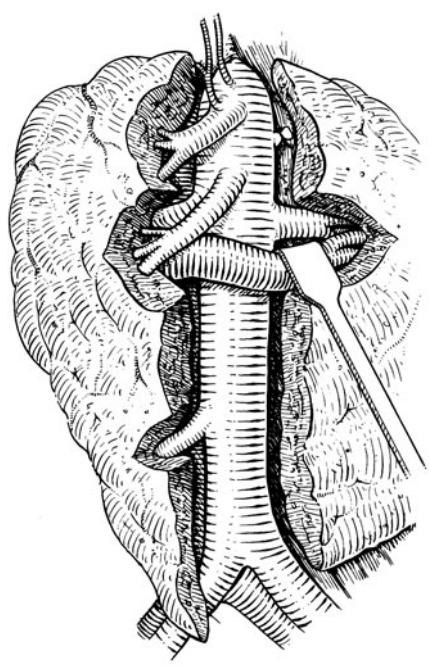


8

8 Each of the encased visceral arteries is dissected in turn – left renal, superior mesenteric, and celiac. The dissection advances distally on these vessels as far as is necessary. It is not unusual to have to dissect left gastric, hepatic, and splenic arteries for some distance.

9 Once all these vessels are safe and in view, they may be cleared prior to tumor removal.



9

POSTOPERATIVE CARE

Postoperative care follows the same principles as for any other major laparotomies. Intravascular monitoring is continued for 24–48 hours. Urine output is carefully measured, and intravascular volume adjusted accordingly.

Blood sugar levels are checked 4-hourly initially and subsequently less frequently.

Effective pain relief is delivered in the form of either epidural infusion or intravenous opiate infusion.

Enteral feeding usually resumes after about 3 days. Diarrhea is not uncommon when celiac and superior mesenteric arteries have been cleared of tumor.

OUTCOME

The surgical morbidity after this operation is similar to that encountered after other major procedures. Wound dehiscence or incisional hernia occurs in less than 1 percent of cases.

Chylous ascites is not unusual in the days following operation, but generally settles without the need for specific treatment. Five of the author's patients have required a peritoneovenous shunt for resistant ascites, having failed to respond to other measures.

The operative mortality is 1 percent.

The effects of age, tumor stage, and changes in chemotherapy protocols confuse attempts to analyze the influence of surgery on the long-term prognosis.

Of 255 patients operated on by the author, 6 had stage 1 disease and 42 had stage 2 disease. All those with stage 1 disease survived, and for those with stage 2 disease the survival was in excess of 95 percent.

For those with stage 3 disease, complete resection resulted in over 80 percent survival, as opposed to 40 percent with incomplete resection.

Of those with stage 4 disease, roughly one-third survived regardless of the extent of resection.

For many years, it has not been clear whether or not surgical excision contributes to survival in those with metastatic disease, who constitute the majority of children with neuroblastoma. However, as there has been no prospective evaluation of the role of surgery, it cannot be assumed that those who undergo successful surgery necessarily have the same type of disease as those in whom resection was unsuccessful. More recent results suggest a trend toward improved survival where surgery has been complete.

FURTHER READING

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71

Rhabdomyosarcoma

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HISTORY

The Intergroup Rhabdomyosarcoma Study (IRS), headed by Harold M. Maurer, was organized in 1970 by representatives of the existing three pediatric groups (CCSG, SWOG, and CALGB) and opened for patient entry in 1972. Five studies (IRS I, II, II, IV, and V) have been completed. The trend over these studies has been for less radical surgical intervention and neoadjuvant chemotherapy ± radiation. Since rhabdomyosarcoma is chemosensitive, some sites require biopsy only, followed by chemotherapy (i.e., head and neck). The goal is complete tumor excision in other sites either as primary therapy or after neoadjuvant chemotherapy (i.e., extremity). The surgeon plays a key role in diagnosis, staging, resection, and access for chemotherapy. Survival of patients with rhabdomyosarcoma continues to improve, with less radical or mutilating operations and a refinement in the chemotherapy ± radiation regimes.

Rhabdomyosarcoma is a primary malignancy in children and adolescents that arises from embryonic mesenchyme with the potential to differentiate into skeletal muscle. The first description of rhabdomyosarcoma in the English literature was by Raycoff in 1937. The tumor can arise anywhere in the body, and prognosis is affected by site and histology as well as by local control and metastatic speed.

PRINCIPLES AND JUSTIFICATION

Rhabdomyosarcoma is the most common soft-tissue sarcoma in pediatric patients and represents the third most common solid malignancy in this age group, behind neuroblastoma and Wilms' tumor. Rhabdomyosarcoma accounts for 4–8 percent of all malignant disease and 5–15 percent of all solid malignancies in childhood. The excellent response to chemotherapy has allowed less aggressive surgical intervention, and the concepts of delayed surgery and second-look surgery have been advanced. Although the sites of presentation of rhabdomyosarcoma are ubiquitous, only a few selected surgical sites are discussed in this chapter. Head and neck primaries are quite common, but usually only require biopsy initially, and further surgical management is not generally needed except in persistent or recurring cases. A clear understanding of the surgeon's role in the management of rhabdomyosarcoma is needed, as the surgeon plays a pivotal role in staging (preoperative clinical assessment) and in grouping (based on the extent of surgical resection and intraoperative findings).

The clinical manifestations of rhabdomyosarcoma vary with the site of origin of the primary tumor, the age of the patient, and the presence or absence of metastatic disease. The most common site of presentation is the head and neck region, accounting for 35 percent of all primary tumors. Genitourinary rhabdomyosarcomas account for approximately 25 percent of primary tumors and are divided into two distinct entities: bladder/prostate tumors (10 percent) and non-bladder/prostate tumors, seen in paratesticular sites, the perineum, vulva, vagina, and uterus. Other common sites include the extremities, trunk, and buttocks. The goal of diagnostic evaluation for rhabdomyosarcoma is to determine the histologic variant of the tumor, its primary site, and the extent of disease (both local and distant). Evaluation may include computed tomography (CT) and/or magnetic resonance imaging (MRI) of the primary area and chest, bone marrow evaluation, and, in selected sites, lymph node evaluation. The most common sites of metastatic spread include the lungs, bone marrow, lymph nodes, and the skeletal system. The most important part of the diagnostic process is obtaining adequate tissue for histologic and cytologic diagnosis and classification. This procedure is usually accomplished

by an open incisional biopsy under general anesthesia. Although percutaneous needle or core biopsies have been selectively utilized, they do not generally provide enough tissue for complete evaluation.

PREOPERATIVE STAGING

Staging for rhabdomyosarcoma is performed in order to determine the specific intensity of treatment as well as to compare outcomes. Pretreatment size is determined from the external measurement on MRI or CT, depending on the anatomical location. Computed tomography scanning is also used to evaluate nodal status, although there is a high risk of false negativity. The pretreatment staging evaluates the primary site, tumor invasiveness, tumor size, lymph node status, and metastases. The survival rate varies significantly with different primary tumor sites. Some tumor locations have a very favorable prognosis, e.g., orbit, vagina, vulva, and paratesticular. An intermediate prognosis is observed in cases of extremity, bladder, prostate, uterus, and non-parameningeal head and neck tumors. A relatively poor prognosis is observed with primary tumors of the parameningeal head and neck sites, retroperitoneum, buttocks, chest wall, trunk, and perineal and perianal regions. Poor prognosis is seen in patients with metastatic disease. The site of the tumor is, therefore, an important prognostic variable and has been taken into consideration in the pretreatment staging beginning with IRS IV (Table 71.1). Site groups and the primary anatomic sites are shown in Table 71.2. Chemotherapy regimens have been directed at these low-risk, intermediate-risk, and high-risk groups.

BIOLOGY AND PATHOLOGY

Rhabdomyosarcoma cells arise from undifferentiated mesodermal tissue and may appear in any part of the body, including tissues that do not ordinarily contain striated muscle. Histologically, rhabdomyosarcoma is classified within the category of small, round, blue cell tumors of childhood, a category that also includes neuroblastoma, Ewing's sarcoma, small-cell osteogenic sarcoma, and non-Hodgkin's lymphoma. Beginning in 1958, Horn and Enterline classified rhabdomyosarcoma into four different pathologic types: embryonal, botryoid, alveolar, and pleomorphic. Today the histologic variance of childhood rhabdomyosarcoma is grouped by a modification of the Horne and Enterline system

Table 71.1 *Tumor node metastasis (TNM) pretreatment staging classification*

Stage	Sites	T	Size	N	M
1	Orbit Head and neck (excluding parameningeal) Genitourinary (non-bladder/non-prostate)	T_1 or T_2	a or b	N_0 or N_1 or N_x	M_0
2	Bladder/prostate Extremity Cranial Parameningeal Other (includes trunk, retroperitoneum, etc.)	T_1 or T_2	a	N_0 or N_x	M_0
3	Bladder/prostate Extremity Cranial Parameningeal Other (includes trunk, retroperitoneum, etc.)	T_1 or T_2	a b	N_1 N_0 or N_1 or N_x	M_0 M_0
4	All	T_1 or T_2	a or b	N_0 or N_1	M_1

Definitions

Tumor

T (site)$_1$, confined to anatomic site of origin:
(a) ≤ 5 cm in diameter;
(b) > 5 cm in diameter.
T (site)$_2$, extension and/or fixation to surrounding tissue:
(a) ≤ 5 cm in diameter;
(b) > 5cm in diameter.

Regional nodes

N_0, regional nodes not clinically involved.
N_1, regional nodes clinically involved by neoplasm.
N_x, clinical status of regional nodes unknown (particularly sites that preclude lymph node evaluation).

Metastasis

M_0, no distant metastasis.
M_1, metastasis present.

Table 71.2 *Site groups and primary sites*

Orbit	**Genitourinary bladder/prostate**	*Perineum – anus*
Eye	Bladder	Anus
Orbit	Prostate	Perineum
Head and neck	**Extremity**	*Retroperitoneum*
Cheek	Arm	Pelvis, site indeterminate
Hypopharynx	Buttock	Retroperitoneum
Larynx	Elbow region	*Trunk*
Neck	Foot	Abdominal wall
Oral cavity	Forearm	Breast
Oropharynx	Hand	Chest wall
Parotid	Knee region	Paraspinal
Scalp	Leg	*Other*
Thyroid	Shoulder girdle	Adrenal glands
Other	Thigh	Bone
Parameningeal (PM)	**Other**	Brain, ventricles and central canal
Infratemporal fossa	*Gastrointestinal and hepatobiliary*	Brain, general
Middle ear	Esophagus	Cerebrospinal fluid
Nasal cavity/sinus	Gallbladder	Lymph nodes – distant
Nasopharynx	Intestine, colon/cecum/rectum	Lymph nodes – regional
Paranasal sinus	Intestine, small and duodenum	Marrow
Parapharyngeal area	Liver	Meninges
Pterygopalatine	Omentum	Multiple sites, excluding lungs
Cheek (with PM extension)	Pancreas	Muscle
Larynx (with PM extension)	Peritoneum	Peripheral nerves
Orbit (with PM extension)	Stomach	Pineal
Oropharynx (with PM extension)	*Intrathoracic*	Pituitary
Other head and neck (with PM extension)	Bronchi and bronchioles	Skin
Parotid (with PM extension)	Diaphragm	Spinal cord
Scalp (with PM extension)	Heart	Spleen
Genitourinary non–bladder/prostate	Hilum	Subcutaneous
Cervix	Lung and local sites	Unknown
Epididymis	Lung	Other
Kidney	Mediastinum	
Ovary	Pericardium	
Penis	Pleura	
Spermatic cord	Pleural effusion	
Testis – paratesticular	Thymus	
Urachus	Trachea	
Ureter		
Urethra		
Uterus		
Vagina		
Vulva		

into favorable-prognosis, intermediate-prognosis, and poor-prognosis groups. Favorable-prognosis tumors include the botryoid and spindle-cell variants. Botryoid rhabdomyosarcoma is best described as a 'cluster of grapes' in gross appearance. The botryoid variant appears primarily in young children in visceral cavities, such as the nasal/pharynx, vagina, and biliary tree, and is associated with the best prognosis of all types of rhabdomyosarcoma. Spindle-cell rhabdomyosarcoma has a predilection for paratesticular sites and has a favorable outcome. Intermediate-prognosis tumors are of the embryonal type. Embryonal rhabdomyosarcoma is composed of small, round, or spindle-shaped cells with variable cellularity and myogenous differentiation. Embryonal histology is the predominant type seen in infants and young children. Both the botryoid and spindle-cell variants are considered subvariants of embryonal rhabdomyosarcoma but are now classified in the favorable-prognosis category.

Unfavorable-prognosis tumors include alveolar and undifferentiated rhabdomyosarcomas. The alveolar variant is characterized by a prominent alveolar arrangement of stroma and dense, small, round tumor cells resembling those of lung tissue. Alveolar tumors frequently arise from the extremities, trunk, or perineum; they account for roughly 20 percent of rhabdomyosarcomas seen in children. Undifferentiated sarcoma is a poorly defined category of sarcomatous tumors whose cells show no evidence of myogenesis or other differ-

entiation. This sarcoma occurs most commonly in the extremities or head and neck sites and is associated with a very poor prognosis. Pleomorphic sarcoma is categorized by large pleomorphic cells with multinucleated giant cells and is often seen on the extremities or trunk; it is rare in children.

Embryonal rhabdomyosarcomas are known to demonstrate a loss of heterozygosity on the short arm of chromosome 11. Alveolar rhabdomyosarcomas have been demonstrated to have a characteristic translocation between the long arm of chromosome 2 and the long arm of chromosome 13.

A number of monoclonal antibodies have been shown to react with the elements of rhabdomyosarcomas and have been useful in their diagnosis. These include antibodies to desmin, muscle-specific actin, sarcomericactin, and myoglobin. Non-myogenous protein products that can be identified in these tumors include cytokeratin, neuron-specific enolase, S-100 protein, and Leu-7.

ANESTHESIA

Biopsies, resections, and staging are generally done under general endotracheal anesthesia. It is of value to perform bilateral bone marrow aspiration and placement of vascular access catheters at the same procedure.

OPERATIONS

Biopsy and general principles of operative resection

Surgical treatment of rhabdomyosarcoma is site specific. Two general principles are that (1) complete total excision of the primary tumor and surrounding uninvolved tissues should be performed while preserving cosmesis and function where possible, and (2) incomplete excision or tumor debulking as a primary procedure is generally not helpful and should suggest the need for neoadjuvant chemotherapy. Severely mutilating or debilitating excisions should not be performed as a primary procedure. Secondary excision after initial biopsy and neoadjuvant therapy has a better outcome than partial or incomplete resection and should be planned in cases in which primary excision is not possible. When initial excision is not possible, incisional biopsy only is employed, along with biopsy of clinically suspicious-looking lymph nodes as indicated. Certain sites such as the extremities and trunk may have a high incidence of regional node positivity (40–50 percent). Therefore, it is valuable to perform sentinel lymph node mapping in these patients. Positive sentinel lymph nodes will lead to regional radiotherapy.

Clinical grouping is determined by the extent of biopsy and/or resection as well as of metastatic disease. Biopsy may be by needle, incisional or excisional, depending on the site. Biopsy may be incisional in some sites such as the head and neck, or excisional in sites such as paratesticular. Most rhabdomyosarcomas are treated with biopsy followed by chemotherapy and then operative removal with or without radiation to the primary site and positive regional nodes. Biopsy of possible metastatic sites, such as the lungs, may also be warranted. Open or thorascopic techniques may be utilized. Solitary pulmonary nodules are usually not metastatic disease in rhabdomyosarcoma and should be biopsied to rule out an infectious focus.

The specific principles of surgical management vary with anatomic location, but, in general, the principle of wide local excision without destroying function is appropriate. The likelihood of gross or microscopic residual neoplasm when simple resection of an undiagnosed soft-tissue tumor has been carried out is so high that re-operation is recommended as the initial definitive approach to management. Re-excision has been shown by IRS studies to improve disease-free survival. Clinical grouping is shown in Table 71.3. Primary re-excision should be considered even if the margins are apparently negative when the initial resection was not a 'cancer operation' or when malignancy was not suspected preoperatively.

Second-look operations have been used for several pediatric tumors to evaluate therapeutic response and to remove any residual tumor after completing initial therapy. Second-look operations with re-excision appear to improve outcome. Second-look operations that demonstrate a complete response may also decrease the toxicity of further chemotherapy or radiation.

Site-specific surgery

HEAD AND NECK

The head and neck are the most common sites for primary rhabdomyosarcoma in children. Prior to the advent of effective chemotherapy and radiation therapy, radical excision of the tumor was the treatment of choice. Effective chemotherapy with an excellent prognosis is now available. Simple biopsy of the lesion is generally all that is required for head and neck lesions. The incidence of cervical node metastases, although not known for sure, appears to be low, and routine node biopsy is not warranted unless clinically suspicious nodes are evident.

GENITOURINARY TRACT

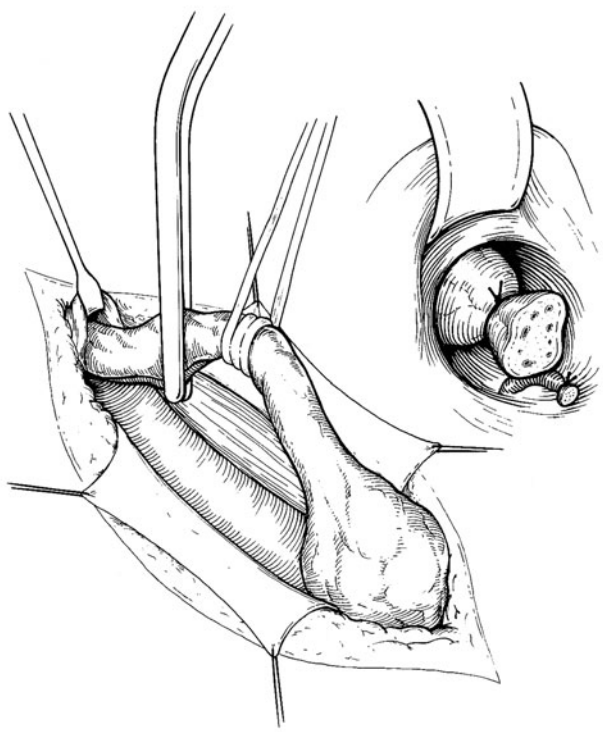
Rhabdomyosarcoma is the most common malignancy of the pelvis seen in children. Tumors in these locations are considered in two different categories because of their different prognoses. Bladder and prostate rhabdomyosarcomas may be difficult to distinguish from each other and have a worse prognosis than non-bladder/non-prostate tumors. This may be related to the size and location and the difficulty of tumor resection. Paratesticular, vulvovaginal, and perhaps uterus have a better prognosis and are highly chemosensitive.

Table 71.3 *Intergroup Rhabdomyosarcoma Study (IRS) clinical grouping classification*

Group 1	*Localized disease, completely resected* Regional nodes not involved – lymph node biopsy or dissection is required except for head and neck lesions. (a) Confined to muscle or organ of origin. (b) Contiguous involvement – infiltration outside the muscle or organs of origin, as through fascial planes. *Notation*: This includes both gross inspection and microscopic confirmation of complete resection. Any nodes that may be inadvertently taken with the specimen must be negative. If the latter should be involved microscopically, the patient is placed in group IIb or IIc (see below).
Group II	*Total gross resection with evidence of regional spread* (a) Grossly resected tumor with microscopic residual disease. (Surgeon believes that he/she has removed all of the tumor, but the pathologist finds tumor at the margin of resection and additional resection to achieve clean margin is not feasible.) No evidence of gross residual tumor. No evidence of regional node involvement. Once radiotherapy and/or chemotherapy have been started, re-exploration and removal of the area of microscopic residual does not change the patient's group. (b) Regional disease with involved nodes, completely resected with no microscopic residual disease. *Notation*: Complete resection with microscopic confirmation of no residual disease makes this different from groups IIa and IIc. Additionally, in contrast to group IIa, regional nodes (which are completely resected, however) are involved, but the most distal node is histologically negative. (c) Regional disease with involved nodes, grossly resected, but with evidence of microscopic residual histologic involvement of the most distal regional node (from the primary site) in the dissection. *Notation*: The presence of microscopic residual disease makes this group different from group IIb, and nodal involvement makes this group different from group IIa.
Group III	*Incomplete resection with gross/residual disease* (a) After biopsy only. (b) After gross or major resection of the primary (> 50%).
Group IV	*Distant metastatic disease present at onset* (Lung, liver, bones, bone marrow, brain, and distant muscle and nodes) *Notation*: The above excludes regional nodes and adjacent organ infiltration, which places the patient in a more favorable grouping (as noted above under group II). The presence of positive cytology in cerebrospinal fluid, pleural or abdominal fluids as well as implants on pleural or peritoneal surfaces are regarded as indications for placing the patient in group IV.

PARATESTICULAR

1 A paratesticular mass should be resected by inguinal orchiectomy, with complete resection of the spermatic cord to the level of the internal ring. Biopsy through the scrotum necessitates subsequent scrotal resection or radiation. If biopsy or resection is performed through the inguinal approach, the scrotal skin does not have to be resected unless there is fixation. If the margin is positive at the internal ring, higher resection is necessary. Once the diagnosis of paratesticular rhabdomyosarcoma is made, patients over the age of 10 years should undergo an ipsilateral retroperitoneal lymph node dissection. Children under the age of 10 years may be followed by thin-cut CT scans to evaluate nodal status. Positive nodes are radiated.



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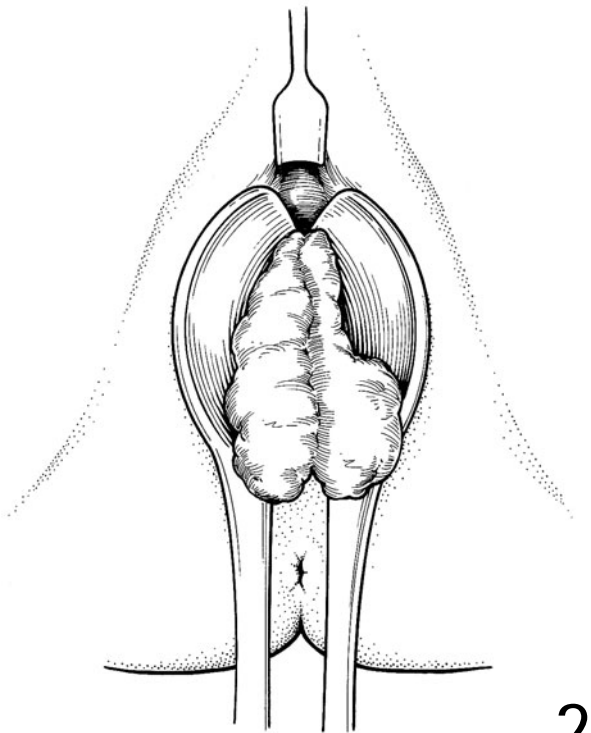
VAGINAL

2 Vaginal rhabdomyosarcoma presents with vaginal discharge, bleeding, or prolapse of a polypoid mass. The diagnosis is made by vaginoscopy and biopsy of the lesion. Occasionally, a tumor may present with a stalk, and primary resection is easy. Prior to the IRS studies, treatment included anterior pelvic exeneration, with good survival but significant sequelae. Most patients today undergo biopsy followed by chemotherapy, with little or any surgical intervention. On occasion, partial vaginectomy is performed or sleeve resection may be done. Rare patients with persistent or recurrent disease may require vaginectomy/hysterectomy. Bladder salvage is possible in almost all patients. Vaginal tumors most commonly arise on the anterior vaginal wall and may invade the vesicovaginal septum or bladder wall due to proximity. Initial biopsy and follow-up vaginoscopy and repeat biopsies generally demonstrate an excellent response to chemotherapy, and further resection is not indicated. Spread to the regional lymph nodes is extremely rare and lymph node evaluation is not done routinely.



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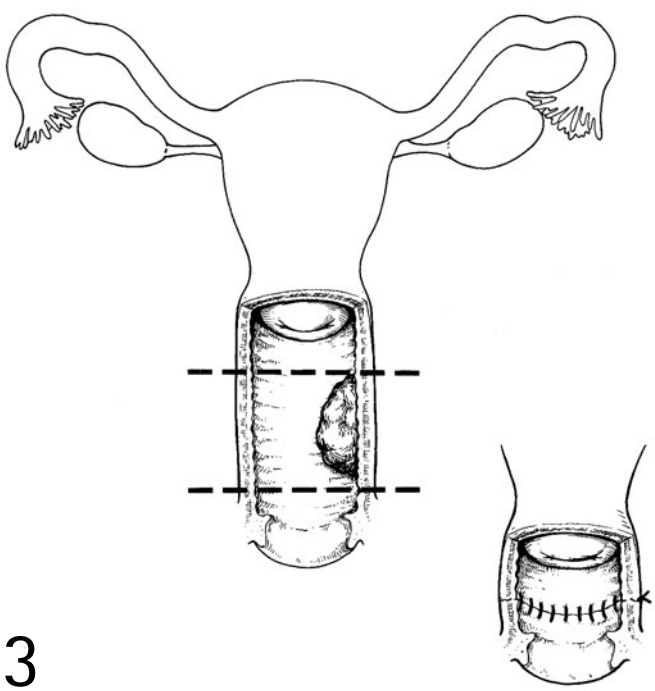

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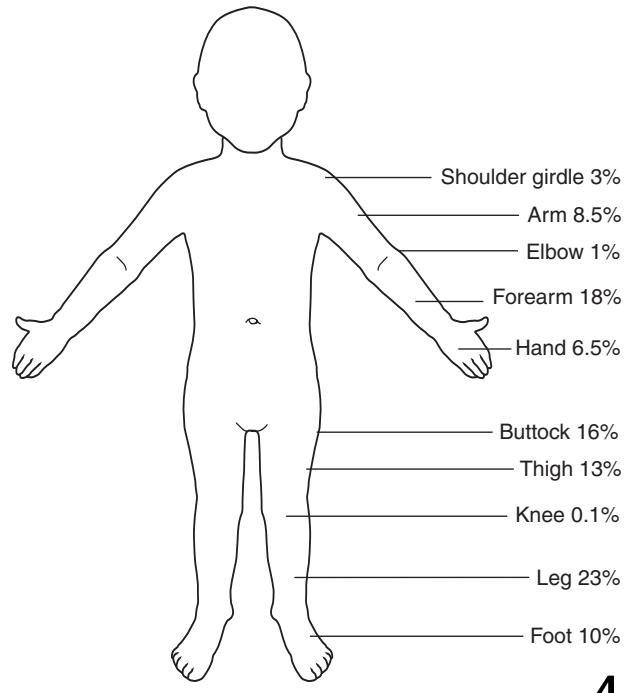
3 Tumors of the cervix and uterus are most commonly seen in somewhat older patients and may require hysterectomy. Persistent or recurrent tumors in the vagina can also be dealt with by local vaginal wall resection or sleeve resection.



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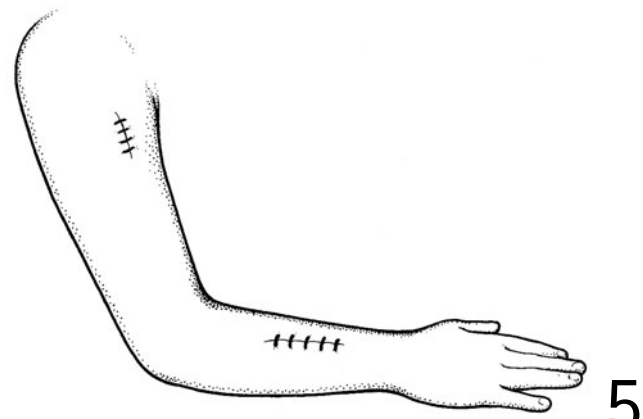
EXTREMITIES

4,5 Rhabdomyosarcoma arising in the extremities is seen in approximately 20 percent of patients. Alveolar histology and poor prognosis are more common in this site. Biopsy of extremity lesions should take into consideration the need for re-operation and wide excision. Therefore, longitudinal incisions for biopsy are preferable.



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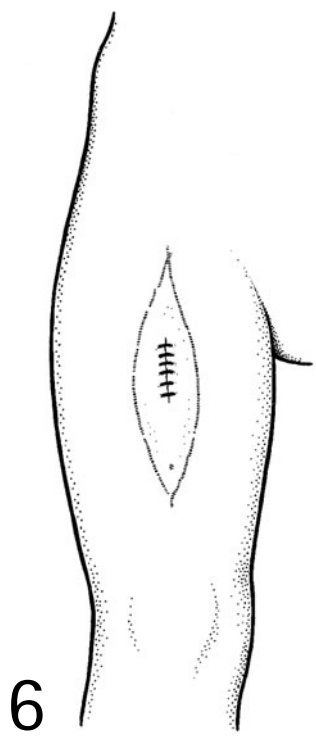

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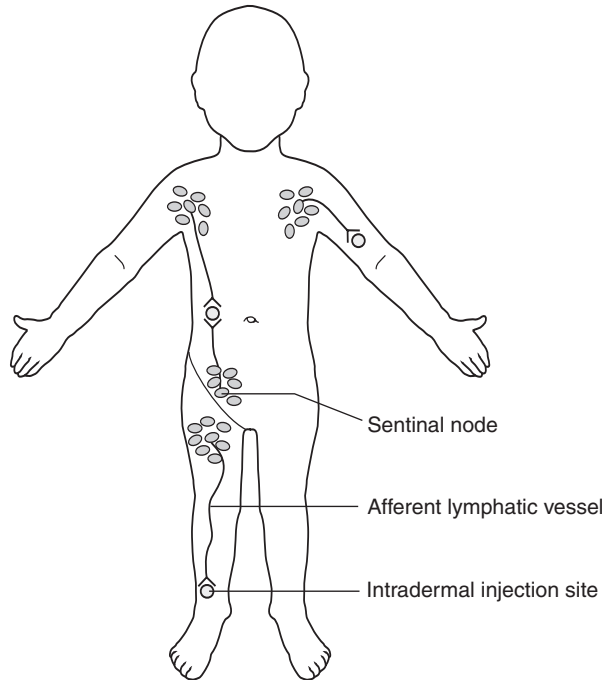
6 Small lesions may lend themselves to primary resection with sentinel lymph node evaluation. Primary re-excision appears to play a significant role in this site. Wide local excision with clear margins is preferable for definitive surgery whenever possible. Limb-sparing operation is the rule, and very few patients require amputation.

7 Regional nymph nodes should be evaluated by sentinel lymph node mapping, as physical examination and CT scanning are somewhat unreliable, with a high false-negative rate. Patients with extremity alveolar rhabdomyosarcoma are considered for post-chemotherapy radiation treatment. Wide local excision with clear margins provides the best prognosis, but the amount of margin necessary is unknown. It appears that, at least in rhabdomyosarcoma, a clear margin (no matter the extent) is more important than obtaining 2 cm of surrounding tissue. Larger tumors are best treated with neoadjuvant chemotherapy followed by second-look operation to limit the amount of mutilation. The sentinel lymph node is the first node in the regional basin that correlates with the nodal basin status. Positive sentinel lymph node mapping should lead to radiation to the regional basin. It appears that sentinel lymph node mapping is most helpful for extremity and trunk lesions. Delayed second-look operations to ensure a complete response or to confirm the need for further resection are indicated in selected patients.



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72

Sacrococcygeal teratoma

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INTRODUCTION

Sacrococcygeal teratomas were first described by Virchow in 1869 and, because of their complexity, were called 'teratomas' from the Greek *teratos onkoma*, meaning 'monstrous tumor.' They represent the most common germ-cell tumors of childhood (40 percent), with an incidence of approximately 1 in 35000–40000 live births, and a female preponderance of 3:1.

Pathology

Embryologically they arise from totipotent primordial endodermal germ cells, but may contain tissues originating from all three germ layers, including dermal elements (e.g., skin), muscle, glial tissue, intestinal mucosal, and pancreas. Ninety percent of lesions are benign at birth, but risk of malignancy increases with: age at diagnosis (up to 50 percent malignancy at 6 months and up to 75 percent at 1 year of age), gender, the anatomical type of the lesion (see American Academy of Pediatrics anatomical classification, p. 726; risk of malignancy is 8 percent in type I compared to 38 percent in type II), in recurrent lesions, and in those with incomplete resection. Histologically, teratomas are classified as mature (70 percent), immature with embryonic components (20 percent), or immature with malignant components (10 percent). Immaturity is defined by the mitotic activity and the extent of neuroepithelium present. Malignant sacrococcygeal teratomas almost exclusively arise from embryonal carcinomas or yolk-sac tumors.

Staging

Sacrococcygeal teratomas are currently staged using The Children's Cancer Study Group and Pediatric Oncology Group staging system for extragonadal germ-cell tumors (Table 72.1).

Table 72.1 *Staging system used for sacrococcygeal teratomas*

Stage	Extent of disease
I	Complete excision with coccygectomy Negative tumor margins Tumor markers positive but fall to normal if negative at diagnosis Lymphadenectomy must be negative for tumor
II	Microscopic residual tumor Lymph node negative Tumor markers positive or negative
III	Gross residual tumor Retroperitoneal nodes negative or positive Tumor markers positive or negative
IV	Distant metastases, including liver

Etiology

The precise etiology of the development of sacrococcygeal teratomas remains unclear. Several theories have been postulated, including the following:

- Presacral lesions occurring at sites of incomplete migration of endodermal cells along their normal pathway from near the origin of the allantois to the gonadal ridges.
- Postsacral lesions originating from remnants of Hensen's node (the midline primitive streak that comprises an aggregate of totipotential cells that are the primary organizers of embryonic development). These migrate caudally from the posterior embryo, finally resting anterior to the coccyx, and normally disappear by the end of the third week.

- Incomplete twinning theories. A familial link is found in 57 percent of cases, with an autosomal dominant mode of inheritance. Various genetic associations have also been reported, including amplification of a region (called 'i') on the short arm of chromosome 12 (12p) an association found with most germ-cell tumors.

DIAGNOSIS

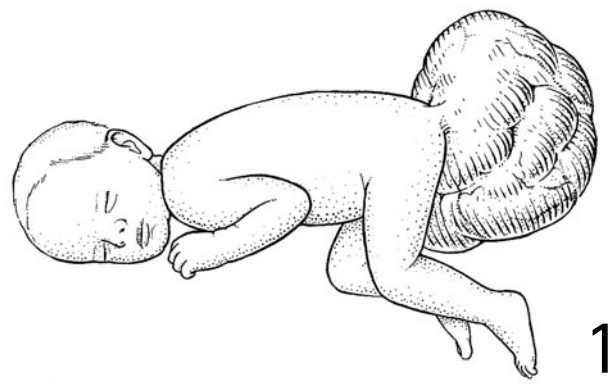
Prenatal diagnosis

The routine application of prenatal ultrasound has increased the prenatal diagnosis of sacrococcygeal teratomas and allowed for accurate identification of their site and details of any intrapelvic extension or urinary tract obstruction. Repeated ultrasound assessment of tumor size also helps determine the mode of delivery. Cesarean section delivery is advocated for tumors larger than 5 cm, as dystocia during vaginal delivery may cause tumor rupture, tumor avulsion, hemorrhage, and sometimes may result in the death of the infant. Finally, ultrasound may be used to prognosticate lesions. For instance, polyhydramnios (27 percent), placentomegaly, and hydrops fetalis are poor prognostic indicators. Recent-onset polyhydramnios is associated with premature labor. Twenty percent of tumors diagnosed prenatally develop hydrops from high-output cardiac failure secondary to vascular steal of blood flow through arteriovenous channels within the tumor, which is associated with a near-100 percent mortality.

Diagnosis in the neonate and children

CLINICAL

1 Most sacrococcygeal teratomas are visible externally and are therefore diagnosed clinically, either at birth or soon thereafter.



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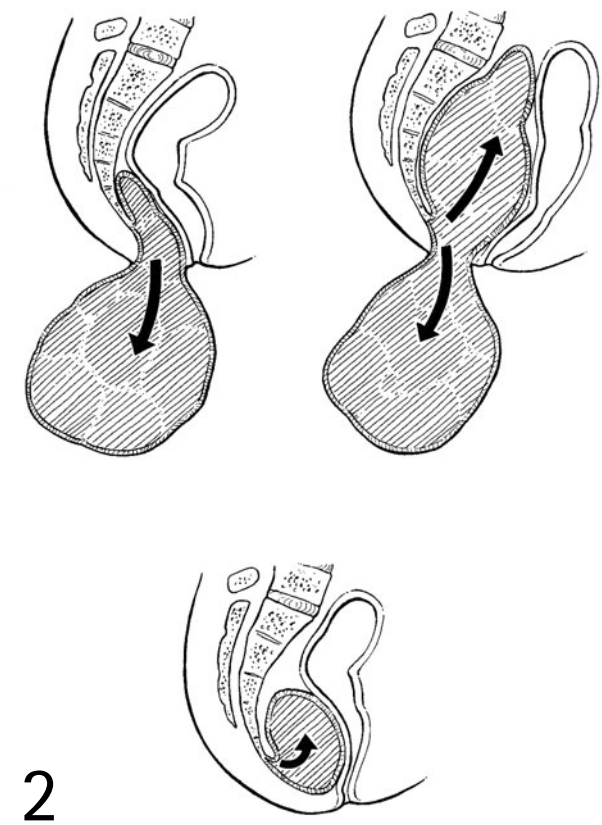

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2 The tumor is classified anatomically according to the criteria of the Surgical Section of the American Academy of Pediatrics.

- Type I (47 percent of lesions): a pedunculated tumor predominantly external with minimal extension into the presacral region.
- Type II (34 percent of lesions): a type I tumor with significant intrapelvic extension.
- Type III (9 percent of lesions): a predominantly pelvic tumor with abdominal extension, but minimal external component.
- Type IV (10 percent of lesions): a completely internal presacral tumor without external evidence of disease.

The tumor is always attached to the coccyx (therefore the necessity of coccygectomy during tumor excision), and may project to a varying degree into the presacral space between the sacrum and the rectum. Although most neonates with sacrococcygeal teratomas are asymptomatic, this upward extension into the pelvic space may compress and elevate the rectum, vagina, bladder, and uterus. Displacement of these pelvic organs may cause presenting symptoms of constipation, large bowel obstruction, urinary retention, an abdominal mass, or symptoms of malignancy, such as failure to thrive.

Most lesions are isolated; associated anomalies are identified in up to 18 percent of patients. The most common anomalies are those of the central nervous system (26 percent) and musculoskeletal system (24 percent); the most common being Currarino syndrome: a triad of anorectal malformation (either anal stenosis or agenesis), sacrococcygeal bony defect (hemisacrum with preservation of the first sacral vertebra), and a presacral mass (usually a presacral teratoma or anterior meningocele, although duplication cysts and dermoid cysts have also been described). Other associated anomalies include urogenital anomalies (hypospadias, vesicoureteric reflux), duplications of the vagina or uterus, orthopedic anomalies (congenital dislocation of hips in 7 percent, vertebral anomalies), central nervous system lesions (e.g., anencephaly, trigonocephaly, Dandy–Walker malformation, spina bifida, and myelomeningocele).

Differential diagnosis

The differential diagnosis of a sacrococcygeal teratoma is mainly meningocele or myelomeningocele. However, sacrococcygeal teratoma can be distinguished on the basis of its more completely cystic nature and its less abundant internal component. In addition, pressure on a myelomeningocele will often be noticeably transmitted to the anterior fontanelle. Less common differentials include lipomeningocele, hemangioma, lymphangioma, chordoma (50 percent occur in the sacrococcygeal region), pelvic neuroblastoma, sarcoma, hamartoma, cystic duplication of the rectum, neuroenteric cysts, dermoid cysts, meconium pseudocysts, and perirectal abscess.

Investigations

Aids to diagnosis include plain radiographs, which may demonstrate calcifications in the tumor in 60 percent of cases or identify spinal defects. Ultrasound will identify the lesion and any intra-abdominal or intrapelvic extensions. Detailed preoperative assessment of the lesion, any abdominal or pelvic extension, and its relationship to the adjacent structures, is made by computerized tomography (CT) and/or magnetic resonance imaging (MRI).

In addition, markers such as alfa-fetoprotein (AFP) and beta-human chorionic gonadotropin (β-hCG) may be useful for the assessment of disease progression. Serum AFP is normally elevated significantly at birth and remains high for up to 4 months, decreasing to adult levels only at 6–12 months. However, most yolk-sac tumors and some embryonal carcinomas also secrete AFP.

Since malignant elements of sacrococcygeal tumors almost exclusively arise from one of these two sources, AFP measurement is a useful marker for malignant degeneration of benign lesions, or for the presence of residual or recurrent malignant disease. It can be measured in the serum and demonstrated in the cells by immunohistochemistry. Persistently high levels may be an indication for further surgery or chemotherapy. β-human chorionic gonadotropin is another marker that may be elevated. It is produced by choriocarcinomas and, rarely, carcinoembryonic antigen, and may be measured in plasma.

PRENATAL MANAGEMENT

Following intrauterine diagnosis of the tumor, management is based on fetal lung maturity and the presence or absence of placentomegaly and hydrops fetalis (the latter conditions being associated with almost 100 percent mortality). Upon fetal lung maturity without placentomegaly and/or hydrops fetalis, early elective delivery by cesarean section is indicated.

Indications for surgery

The mainstay of the treatment of benign sacrococcygeal teratomas is early and en-bloc excision of the lesion within a few weeks of life, given that:

- the risk of malignant change in benign lesions increases with (a) age and (b) incompletely excised residual lesions; and
- the tumor's rich vascularity makes it vulnerable to spontaneous ulceration and hemorrhage if left unexcised.

PREOPERATIVE PREPARATION AND ANESTHESIA

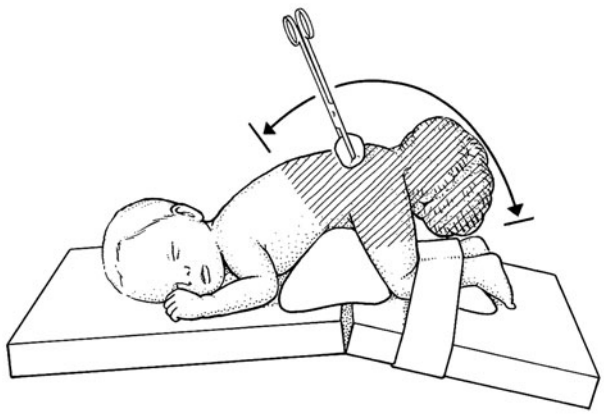
1. Appropriate imaging (ultrasonography/CT/MRI) to delineate the anatomy and extent of the lesion, as the surgical approach will be dictated by whether the lesion has intra-abdominal or intrapelvic extension.
2. Serum assays of tumor markers (AFP/β-hCG) for postoperative comparisons.
3. Adequate intravenous access and blood products should be secured before starting the operation, especially with large tumors, where there may be brisk intraoperative blood loss. Other vascular access, including an arterial line for blood pressure monitoring, and central venous line monitoring are beneficial.
4. General anesthesia is mandatory. High-output cardiac failure secondary to arteriovenous channels in the tumor may limit the use of inhalation agents, which have known cardiodepressant effects.
5. Broad-spectrum antibiotics should be given at the induction of anesthesia.
6. The stomach is emptied with a nasogastric tube, and an indwelling bladder catheter is inserted.

OPERATION

Position of patient and preparation

3 In the majority of cases, where the major component of the tumor is extrapelvic, the procedure is performed with the infant in the prone jack-knife position. This is achieved by supporting the pelvis and shoulders with rolled towels, thus also allowing free respiratory movements of the chest and abdomen during ventilation.

The rectum may be prepared for digital manipulation during the course of the dissection, using an enema (e.g., 1 percent solution of povidone-iodine). Alternatively, those who prefer to exclude the anus from the operative field pack the rectum with gauze impregnated with Vaseline, liquid paraffin, or povidone-iodine solution. Vaseline packing in the rectum facilitates its identification throughout the procedure.



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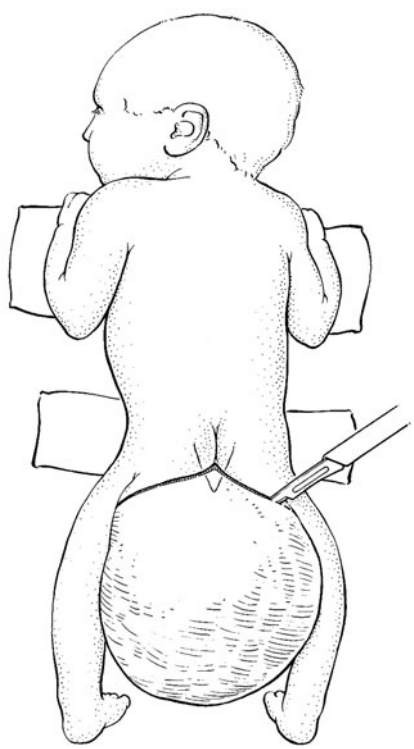

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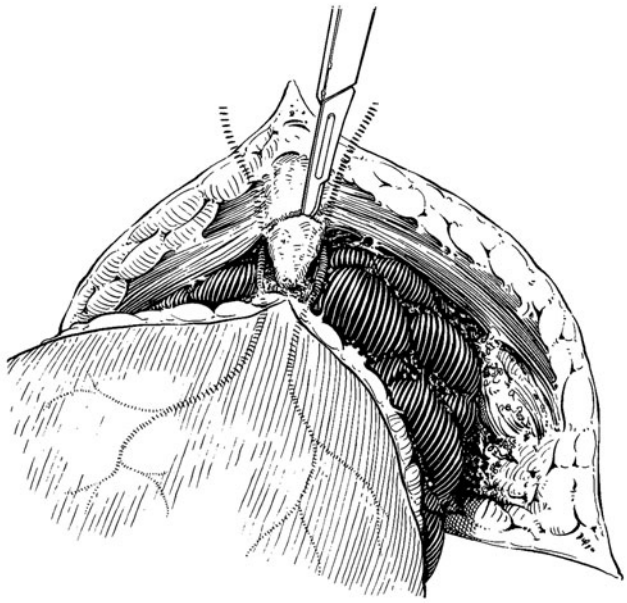
Incision

4 An inverted V-shaped chevron or curved incision is made with its apex over the coccyx and extending dorsolaterally along the base of the tumor capsule, which is well defined from the other tissues. Skin flaps are raised over the surface.

Removal of tumor and coccyx en bloc and ligation of sacral vessels

5 The tumor is dissected from the inferior and medial aspects of the gluteus maximus. The sacrum and coccyx are identified, and the coccyx is transected at the sacrococcygeal joint by cautery, taking the coccyx en bloc with the tumor. Failure to remove the coccyx is associated with a 30–40 percent incidence of recurrence, with more than 50 percent of cases being malignant.

The median and lateral sacral vessels are identified at this landmark, and also controlled and ligated in continuity with the tumor/coccyx. (With an extensive intra-abdominal tumor component, an initial transabdominal approach will be required to achieve vascular isolation of collateral vessels from the lateral sacral vessels.)



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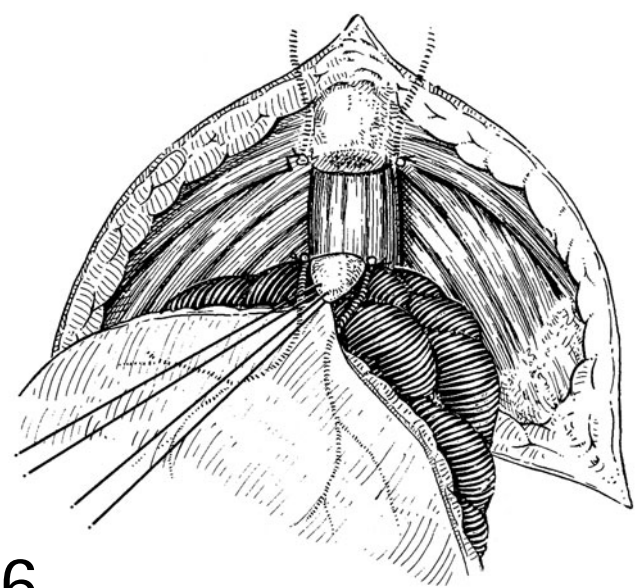

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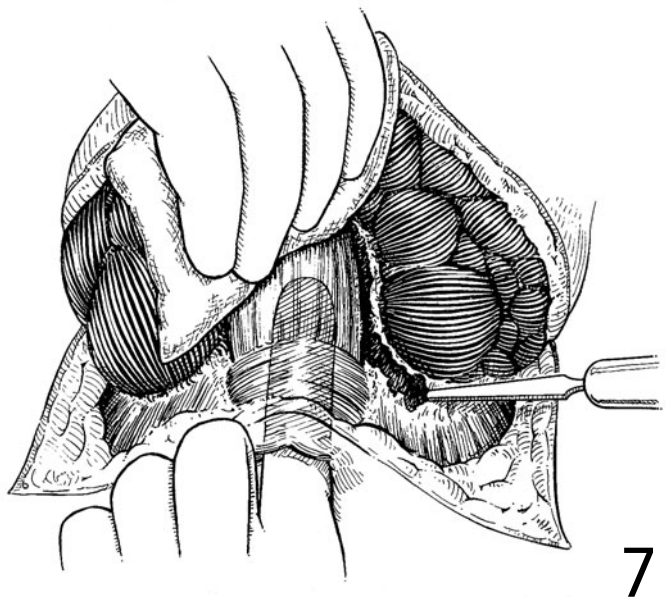
6 Using gentle traction, the coccyx and tumor are retracted and dissected free from the gluteus maximus and surrounding tissues, to the pelvic floor. The pelvic floor muscles are preserved and the proximal rectum identified. The tumor must be dissected and freed completely from the rectal wall and anorectal sphincter complex.

7 This dissection can be facilitated if necessary by placing a finger in the rectum. Rarely, the tumor completely surrounds the rectum, making total excision very difficult.



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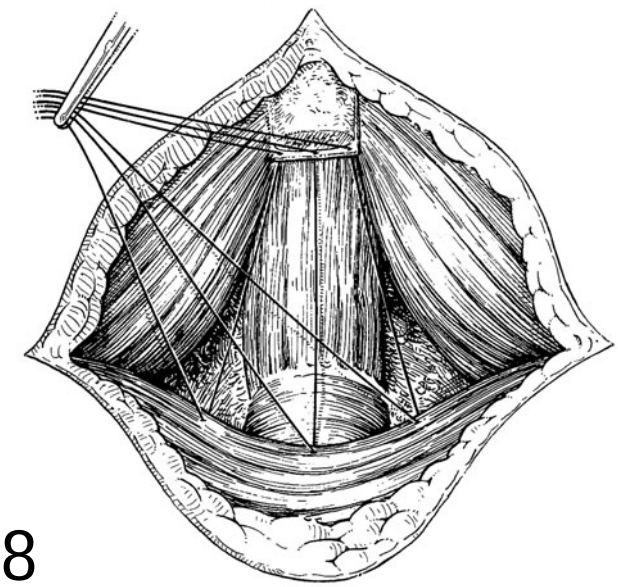
The excised tumor is sent for histological examination to identify the presence of any malignant components as well as to ensure tumor-free margins.



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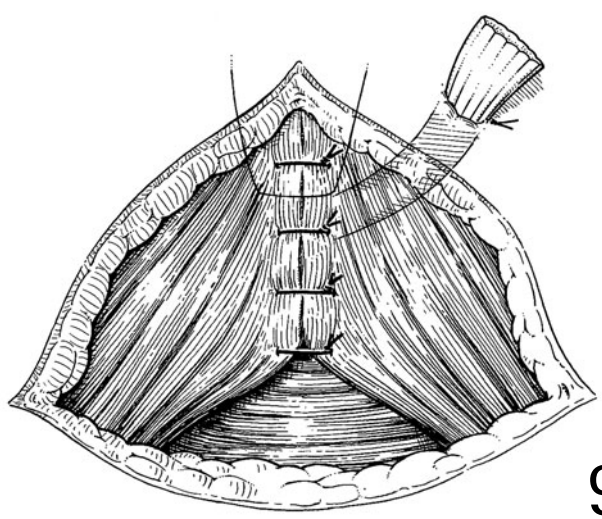
Pelvic floor reconstruction

8 The pelvic floor is reconstructed by suturing the superior and posterior portions of the levator muscles to the presacral fascia, behind the rectum. This allows the anus to assume a near-normal configuration and therefore the best possibility of achieving good fecal continence.

Closure

9 The gluteus maximus muscles are apposed in the midline using interrupted sutures. A drain may be left in place in the perirectal space prior to this closure, and brought out through a separate stab incision.



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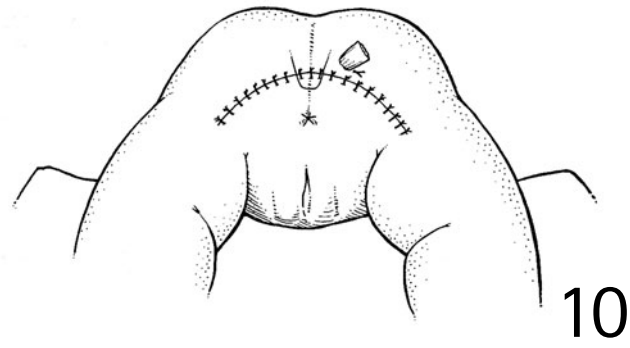
10 Excess skin may be trimmed to achieve best cosmesis, before closing with interrupted or continuous sutures.



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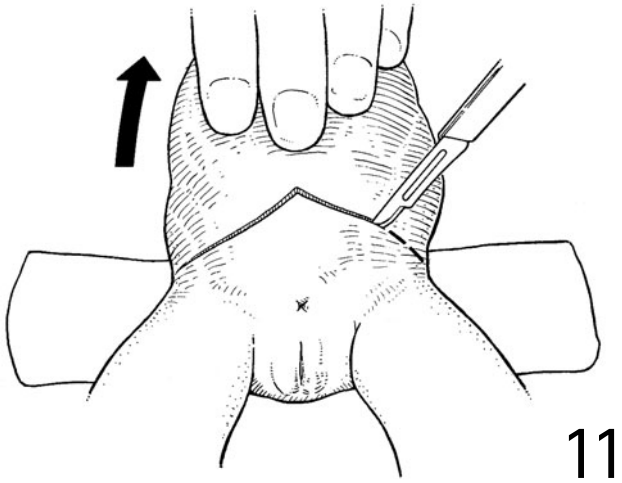

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Modifications for extensive or intra-abdominal tumors

11 In type III tumors where there is significant intra-abdominal involvement, the pelvic excision must be preceded by a laparotomy. In these cases, the operation commences with the patient in a supine position, with a rolled gauze pad under the buttocks, thus raising the pelvic floor. A lower midline or transverse lower abdominal incision approach is used, depending on the extent of the intra-abdominal component of the tumor. The tumor is dissected free from the lower abdominal and pelvic viscera, and the sacral vessels can be controlled through this approach. The abdominal wall is then closed, and the patient is turned prone to complete the operation as described above.

POSTOPERATIVE CARE

Immediate postoperative period

The child is kept in a prone position for 3 days to prevent soiling the wound with urine or feces. The urinary catheter is removed after 24 hours, and the drain after 48 hours. Oral feeding may be commenced when nasogastric aspirates are minimal, and the nasogastric tube may be removed thereafter. A pelvic neuropraxia may occur in the early postoperative period and may cause a poorly contracting, neurogenic bladder. This is generally temporary, but the patient will require intermittent catheterization until it resolves.

Chemotherapy

Malignant lesions respond poorly with surgery alone, with a 10 percent salvage rate. Adjuvant chemotherapy, particularly platinum-containing regimens, e.g., cisplatin, bleomycin, vinblastine and/or VP-16 (etoposide), addressing the specific malignant element, has improved survival with such lesions. This regimen may shrink the tumor, making it amenable to secondary resection. Resected tumors should also be examined for malignant elements, which may require subsequent chemotherapy.

Follow-up

Beyond the immediate postoperative care period, careful monitoring is required, as malignant germ-cell tumors can recur either from missed malignant elements in the original tumor or from malignant conversion of benign residual tissue. Follow-up, including rectal examinations, should be performed at monthly intervals for the first year, at 3-month intervals for at least 3 years (as most recurrences occur within the first 3 years), and then annually. Monitoring of serum AFP levels may also help detect the presence of malignant recurrence. Recurrent tumors should be excised, with preoperative chemotherapy for more extensive disease.

PROGNOSIS

The prognosis for patients with sacrococcygeal teratoma is dependent on the following:

- Antenatal/obstetric factors: hydrops or placentomegaly is associated with almost 100 percent mortality, whilst dystocia or tumor rupture during delivery may be associated with exsanguinating hemorrhage.
- Age at diagnosis: as the risk of malignant transformation in a benign lesion increases with age.
- Tumor histology and stage: up to 95 percent of benign tumors can be cured with excision surgery alone. Prognoses of malignant tumors are dependent on tumor type, stage, and location. Survival with malignant tumors can be achieved in up to 90 percent of cases using a combination of surgery and adjuvant chemotherapy, although the risk of late recurrences or second malignancies persists. Following surgical excision, mature teratomas are associated with an 11 percent risk of recurrence within 5 years.
- Associated anomalies.
- Surgical factors: recurrence is more likely with incompletely excised lesions or in the absence of coccygectomy. Failure to remove the coccyx is associated with a 30–40 percent incidence of recurrence, with more than 50 percent of cases being malignant.

In addition to tumor recurrence, long-term complications associated with sacrococcygeal teratomas are common: 40 percent will encounter mild bowel dysfunction (incontinence or constipation), whilst 10 percent will have urinary incontinence or neuropathic bladders, often associated with similar bowel symptoms.

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Surgery for hyperinsulinemic hypoglycemia

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HISTORY

Hyperinsulinemic hypoglycemia was previously known as nesidioblastosis – a term coined by Laidlaw in 1938 to describe diffuse proliferation of islet cells. Nesidioblasts were defined as 'cells that differentiate out of the duct epithelium to build islets'.

PRINCIPLES AND JUSTIFICATION

Hypoglycemia in the neonatal period is defined as a blood glucose concentration of less than 2.6 mmol/L. The hypoglycemia may be *transient*, as in the 'stressed neonate' or the infant of a diabetic mother or in Beckwith–Wiedermann syndrome, or *persistent*. In persistent hypoglycemia it is important to exclude leucin sensitivity and other endocrine disorders (e.g., hypopituitarism, cortisol deficiency) or inborn errors of metabolism (e.g., glycogen storage disease).

The commonest cause of persistent hyperinsulinemic hypoglycemia is loss of function of the beta cell adenosine triphosphate (ATP)-sensitive potassium channel (K^+_{ATP} channel). The K^+_{ATP} channel effectively couples the metabolic status of the pancreatic beta cell to membrane potential and hence insulin secretion. It consists of a hetero-octamer of two subunits – the sulphonyurea receptor SUR1 and a potassium channel pore protein Kir 6.2. Gene mutations of *ABCC8* and/or *KCNJ11* situated on the short arm of chromosome 11 are associated with hyperinsulinism.

Familial forms of hyperinsulinism normally exhibit autosomal recessive inheritance.

Diagnosis

The diagnosis of hyperinsulinism is based on the following criteria:

- inappropriately raised plasma insulin levels for blood glucose concentration;
- glucose infusion rate greater than 6–8 mg/kg per minute to maintain a blood glucose level above 2.6 mmol/L;
- low free fatty acids and blood ketone bodies during hypoglycemia;
- glycemic response to glucagon despite hypoglycemia.

DISTINCTION BETWEEN DIFFUSE AND FOCAL DISEASE

It is important to distinguish diffuse from focal hyperinsulinism. In the diffuse form, the histopathological features are of beta cells with enlarged nuclei and abundant cytoplasm throughout the pancreas. In focal hyperinsulinism, these changes are restricted to a small area, while the rest of the pancreas has 'resting beta cells' with small nuclei. Focal disease occurs in 30–50 percent of cases.

- *Pancreatic venous sampling* involves transhepatic catheterization of the pancreatic venous radicals and demonstrating high insulin levels throughout the pancreas (diffuse) or at a localized area (focal). The technique is technically demanding and carries a false-positive rate of 15 percent.
- *Intra-arterial calcium stimulation*, in which calcium is selectively injected into the gastroduodenal superior mesenteric and splenic arteries to stimulate insulin secretion, is better at identifying focal disease but inconclusive in diffuse disease.
- *Positron emission tomography (PET) scan*: PET scan has recently been shown to be capable of detecting focal areas of increased insulin secretion.
- *Laparoscopic biopsy of the pancreatic tail* and submitting the specimen for histopathologic diagnosis: if diffuse disease is diagnosed, near-total pancreatectomy is carried out; whereas if focal lesion is suspected, pancreatic venous sampling and calcium stimulation tests are performed to identify the precise location of the lesion.

Medical treatment consists of providing sufficient glucose to prevent hypoglycemia, which usually requires an intravenous infusion of 15 percent glucose solution. Diazoxide is the mainstay of medical management. It inhibits glucose-stimulated insulin secretion, and dosages of up to 25 mg/kg per day may be necessary. Its action is potentiated by the diuretic chlorothiazide. The somatostatin analog ostreotride infusion may be useful as a therapeutic adjunct in refractory cases.

Surgical treatment is indicated if the patient remains dependent on intravenous glucose despite full dosages of diazoxide and chlorothiazide.

Special investigations such as ultrasonography, computed tomography, nuclear magnetic imaging, and selective arteriography are of little value in the diagnosis of hyperinsulinism in infancy. In the older infant or child, they may be helpful in the localization of a focal adenoma.

Aim of surgery

The operative procedure consists of a near-total pancreatectomy for diffuse disease. The lines of resection showing the extent of pancreatic resection is shown in Illustration 1. For focal lesions, local resection should prove curative.

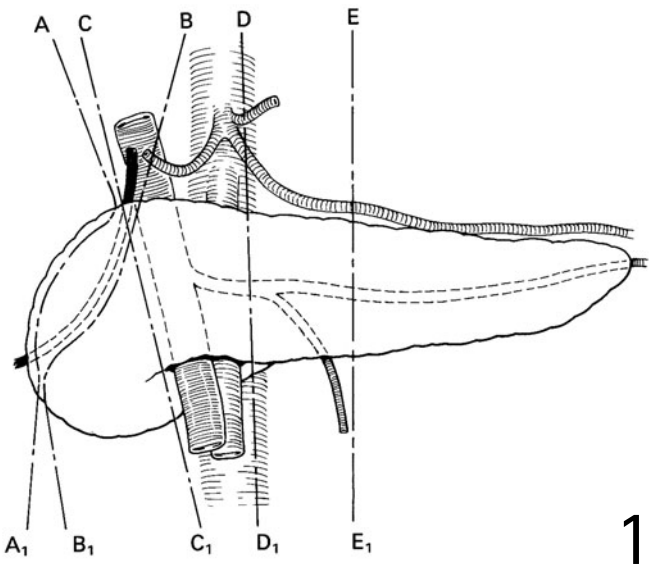
PREOPERATIVE PREPARATION

A central venous catheter is essential to monitor blood glucose levels at regular intervals, preoperatively, intraoperatively, and postoperatively. Prophylactic antibiotics (flucloxacillin and gentamicin) are advisable to prevent wound sepsis, as there is a high level of circulating blood glucose and an excessive amount of subcutaneous fat deposition.

OPERATION

1 The operative procedure consists of a 95 percent pancreatectomy. The line of resection showing the extent of pancreatic resection is shown.

A–A_1 = 99 percent resection.
B–B_1 = 95 percent resection.
C–C_1 = 80 percent resection.
D–D_1 = 50 percent resection.
E–E_1 = 30 percent resection.



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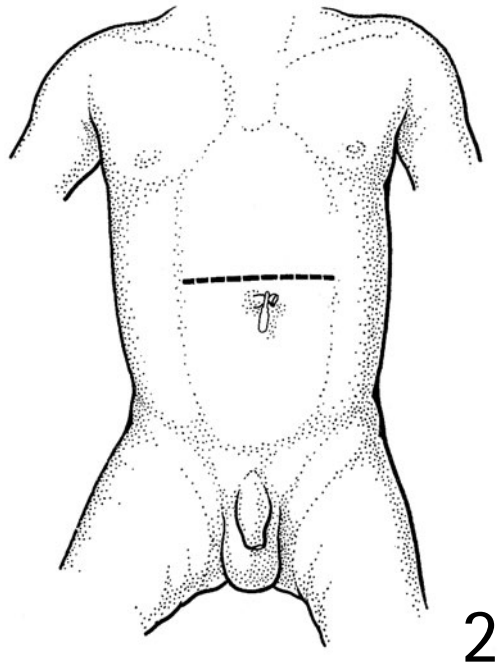
Incision

2 A laparotomy is performed via a generous supraumbilical transverse muscle-cutting incision, extending through both rectus abdominus muscles. A thorough search is made for sites of ectopic pancreatic tissue.



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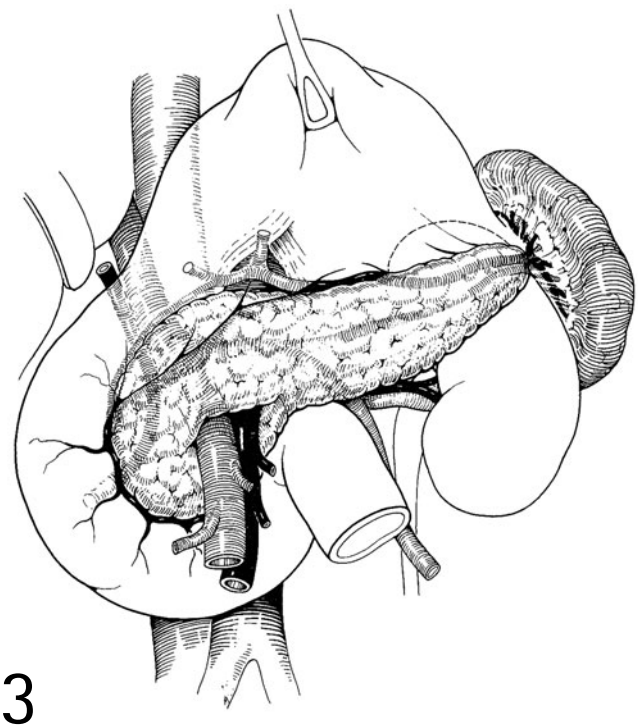

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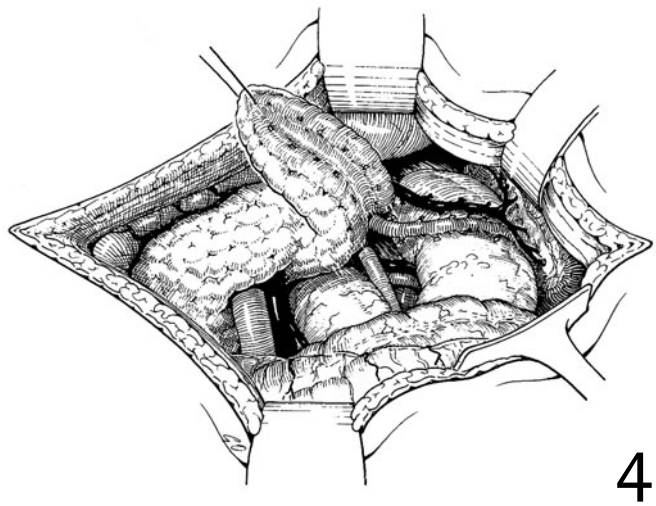
Exposure

3 The anterior surface of the pancreas is exposed by entering the lesser peritoneal sac via the gastrocolic omentum. Vessels in the greater omentum are ligated and divided or coagulated using bipolar diathermy, preserving the gastroepiploic and short gastric vessels.

The hepatic flexure of the colon is reflected medially and the duodenum Kocherized to expose the head of the pancreas. The entire pancreas is carefully inspected for the presence of an adenoma, which appears as a reddish-brown nodule on the surface of the greyish pancreas. Suspicious nodules should be excised and submitted for frozen-section histologic examination. The coexistence of a pancreatic adenoma and diffuse pancreatic disease is well recognized in early infancy. A generous biopsy of the tail of the pancreas to exclude diffuse disease should be performed before adenectomy alone is carried out.

Mobilization of the body and tail of the pancreas

4 The tail of the pancreas is carefully dissected out of the hilum of the spleen. The short pancreatic vessels arising from the splenic artery and vein are coagulated with bipolar diathermy and divided. The dissection of the pancreas proceeds medially from the tail toward the neck of the pancreas, which lies just to the right of the superior mesenteric vessels. It is essential for future immunological competence to preserve the spleen. This is accomplished by carefully exposing the short pancreatic vessels passing from the splenic vessels to the pancreas. These vessels, especially the veins, are extremely friable, but with meticulous dissection they can be individually coagulated and divided without traumatizing the main vessels. Should hemorrhage occur from damage to the splenic vein, direct repair of the vein should be attempted. In the event of failure to achieve hemostasis, the main splenic vein can be ligated in the expectation that splenic integrity will be preserved by collateral supply from the short gastric vessels. When the dissection has progressed to the right of the superior mesenteric vessels, attention is directed to the head of the pancreas and, in particular, the uncinate process.



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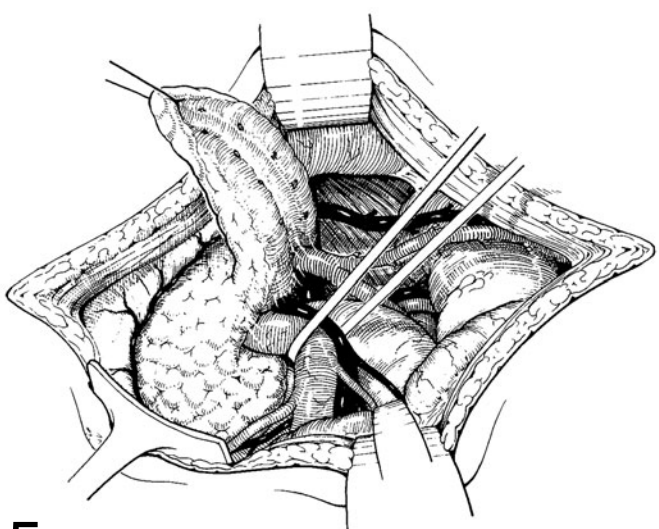

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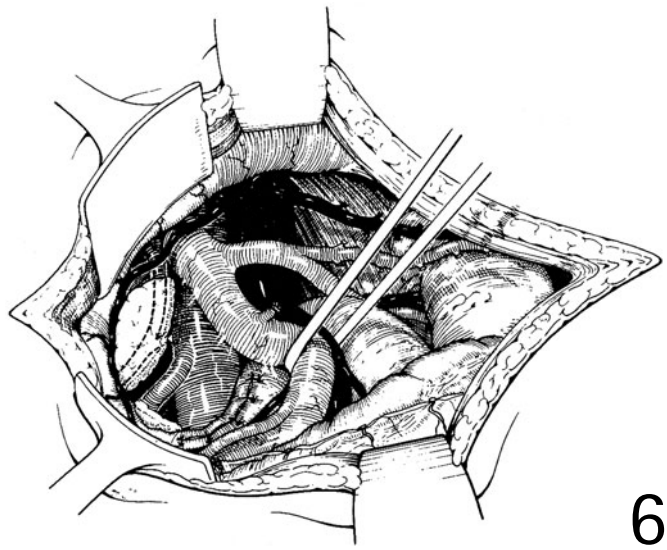
Mobilization of the uncinate process

5 The uncinate process, which can comprise up to 30 percent of the pancreatic weight, lies behind the superior mesenteric artery and vein. These vessels need to be retracted to the left, and the whole of the uncinate process should be carefully and meticulously mobilized, coagulating numerous short feeding vessels. Failure to resect the uncinate process exposes the patient to the risk of recurrent hypoglycemia.

Exposure of the head of the pancreas

6 It is essential accurately to define the course of the common bile duct, which may pass through the head of the pancreas or lie posteriorly, either on the posterior surface or in a groove between the pancreas and duodenum. This is best achieved by identifying the common bile duct above the first part of the duodenum and passing a sling around the duct at this point. A blunt forceps is now passed from the undersurface of the first part of the duodenum, within the concavity of the C-loop of the duodenum, behind the duodenum and the sling is grasped and passed into the operative field above the head of the pancreas. This will allow the course of the common bile duct to be kept in view during the dissection of the head of the pancreas.

The head of the pancreas can now be mobilized with safety without injuring the common bile duct. The superior and inferior pancreaticoduodenal vessels are ligated and divided to ensure hemostasis when completing the pancreatic resection.



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'95 percent' pancreatectomy

The head of the pancreas to the left of the common duct and in the concavity of the duodenal loop is excised, leaving a sliver of pancreatic tissue on the surface of the duodenum and on the left wall of the common duct. The pancreatic duct is identified and ligated with a non-absorbable ligature. Hemostasis is carefully and meticulously achieved. The remaining pancreatic tissue consists of that part of the gland between the duodenum and the common bile duct, and the sliver of tissue on the medial wall of the second part of the duodenum. This represents approximately 5 percent of the total volume of the pancreas. A suction drain introduced via a separate stab incision may be left in the pancreatic bed.

Closure

The wound is closed in layers or with an en-masse interrupted 3/0 polyglycolic acid suture. The subcutaneous fatty layer is closed separately with a running 4/0 absorbable suture. The skin edges are approximated with a 5/0 subcuticular suture.

POSTOPERATIVE CARE

Nasogastric decompression and intravenous fluids are continued during the period of postoperative ileus. Blood glucose levels are closely monitored postoperatively, and soluble insulin is administered as required on a sliding scale. Rebound transient hyperglycemia is common in the early postoperative period. Occasionally, more prolonged use of small amounts of insulin is required, but adaptation usually occurs within 3–6 months. In the long term, refractory diabetes mellitus may occur, the control of which may be extremely difficult.

Complications

- Trauma to the common bile duct (10 percent incidence). The defect may be amenable to direct repair. If the duct has been transected, end-to-side anastomosis to the first part of the duodenum should be performed. Late strictures can develop as a result of ischemia.
- Inadequate resection. This will become evident within 48–72 hours of surgery and it is advisable to carry out a further resection at this early stage rather than later, when fibrosis can render the procedure extremely difficult.
- Wound sepsis and adhesion intestinal obstruction.

N.B. Careful long-term follow-up is necessary to assess the adequacy of pancreatic exocrine function.

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SECTION VI

Urology

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Pelviureteric junction obstruction

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PRINCIPLES AND JUSTIFICATION

The pelviureteric junction (PUJ) is the most common site of obstruction in the pediatric upper urinary tract. The PUJ is formed during the fifth week of embryogenesis. Urine flows across it from the kidney and down the length of the ureter by the twelfth week. Flow occurs when renal pelvic pressure exceeds upper ureteral pressure. The pressure gradient is created partly by the hydrostatic force of the filtered urine, but principally by the peristaltic contractions originating in the region of the renal pelvis called the pacemaker and progressing across the PUJ and down the ureter. Hydronephrosis develops either when the PUJ is obstructed or when the normal peristaltic waves are impeded.

Obstruction more commonly results from an intrinsic defect in the smooth muscle layer of the PUJ. External compression and blockage by aberrant renal vessels and adhesive bands occur in one-third of cases. Light microscopy demonstrates that abnormal smooth muscle architecture and increased fibrosis are present in obstructed PUJ. Electron microscopy further shows disruption of the intercellular junctions needed to coordinate the transmission of peristaltic waves.

Incidence, signs, and symptoms

The incidence of obstruction of the PUJ is 1:1000 to 1:2000 live births and it is more common in boys than in girls. The left side is affected in 60 percent of cases, and 5–10 percent of cases are bilateral. In duplex systems, the lower pole moiety is more likely to be obstructed, although both systems can be involved.

Before the use of antenatal ultrasonography became widespread in the 1970s, the common signs and symptoms of PUJ obstruction were abdominal mass, gross hematuria, urinary tract infection, and pain. Antenatal ultrasonography now allows PUJ obstruction to be identified before becoming symptomatic and has increased the detection of asymptomatic obstruction fivefold, whereas the incidence of symptomatic obstruction has remained the same.

Older children and adolescents may have protean manifestations, including failure to thrive, vague sporadic flank and abdominal pain (especially with diuresis), nausea, vomiting, hypertension, and recurrent urinary tract infections. Hematuria or renal parenchymal injury may ensue from only minor trauma.

Particular attention should be paid to infants with other congenital anomalies. Conditions associated with PUJ obstruction include the VATER syndrome (vertebral defects, imperforate anus, tracheoesophageal fistula, radial dysplasia, and renal dysplasia), contralateral multicystic kidney, esophageal atresia, and vesicoureteral reflux.

PREOPERATIVE

Imaging studies

Clinically suspected PUJ obstruction should be confirmed by imaging studies. Ultrasonography is non-invasive, radiation free, and well tolerated. Characteristically, the PUJ produces a central lucency, which is the renal pelvis surrounded by communicating dilated calyces. A voiding cystourethrogram is advisable for all patients suspected of obstruction to rule out vesicoureteral reflux and other lower tract causes of hydronephrosis such as posterior urethral valves. Intravenous pyelography or urography provides excellent anatomic detail, and delayed films will best visualize the PUJ. The low glomerular filtration rate of neonates precludes the use of

intravenous pyelography. Advances in digital imaging algorithms allow computed tomography and magnetic resonance imaging to produce excellent images in the sagittal and coronal planes in addition to the traditional transverse images. These allow excellent visualization of renal anomalies such as horseshoe kidney, although their cost limits their widespread use.

Diuretic renal scintigraphy is particularly helpful in diagnosing and quantifying the severity of obstruction. The most common radionuclides are DTPA (^{99m}Tc-diethylenetriamine penta-acetic acid) and MAG-3 (^{99m}Tc-mercaptoacetyltriglycine). A bladder catheter should be inserted in all patients undergoing a renal scan when evaluating obstruction. A full bladder can exert back-pressure on the ureters and obscure the results. The scan produces two important results.

- It determines the percentage of total renal effort contributed by each kidney. A poor or non-functional kidney should be considered for removal rather than repair.
- The time it takes for half of the radionuclide to wash out of the kidneys after administration of diuretic (t½) is calculated. A t½ of more than 20 minutes is most commonly associated with obstructed systems. Recent studies in infants with antenatally detected hydronephrosis suggest that an elevated half-time alone may not always be associated with clinically significant PUJ obstruction.

Alternatives to pyeloplasty

The aims of surgery are to improve drainage of the PUJ, which should relieve symptoms, correct or improve the hydronephrosis, and prevent renal deterioration. For these reasons, obstructions are generally promptly repaired.

The management of PUJ obstructions detected by antenatal ultrasonography but remaining asymptomatic in the infant and young child has evolved over the past several decades. Many newborn kidneys continue to grow and develop normally despite imaging studies consistent with PUJ obstruction. A prospective 10-year follow-up study found that less than 25 percent of the observed infants ultimately progressed to surgery, and in nearly all cases it proved safe to watch the patients during their first 2 years of life.

These reports suggest that our understanding of PUJ obstruction and the methods used to identify it is still imperfect. In the presence of active symptoms and signs, however, particularly urinary tract infection, nausea, vomiting, stone formation, or declining renal function, surgical treatment is an obvious choice.

Endourological techniques used for the treatment of ureteral calculi have been adapted to the treatment of PUJ obstruction. The obstructed segment is incised and dilated through either a percutaneous or retrograde approach, and is left to heal over an indwelling stent. Reports suggest that this method may be useful in older children after previous failed pyeloplasty. Its utility in infants and children is limited by the smaller diameter of the ureter and the relatively larger size of the instruments currently available. Other constraints include large redundant pelves and crossing vessels.

Laparoscopic pyeloplasty is technically feasible in adults and has been applied to children and infants. The techniques are evolving and are discussed below.

OPEN OPERATION

The technique described is dismembered pyeloplasty. Unlike methods using flaps and advancements, it excises the pathologic segment and may be applied to a variety of anatomic configurations.

Anesthesia

General anesthesia with endotracheal intubation is required and epidural catheterization is helpful, reducing the total amount of general anesthesia used and providing pain relief postoperatively. The epidural catheter should run along the back away from the ipsilateral side. Postoperatively, care must be taken to check regularly on the site for signs of infection and to be sure that the patient is adequately turned and repositioned regularly, as decreased sensation may favor pressure effects on the skin.

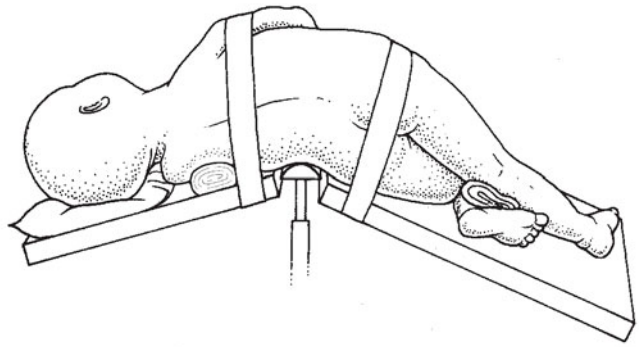
Instruments and equipment

In addition to a pediatric major surgery tray, fine forceps, magnifying loupes, 5 Fr and 8 Fr feeding tubes, hooked and pointed scalpel blades, needle point electrocautery, and 3/0 to 6/0 synthetic or biologic absorbable suture with atraumatic needles should be available. While the choice of suture varies greatly among surgeons, permanent synthetic or biologic sutures (e.g., silk or Ethibond™) should not be used because they can become nidi of infection and stone formation.

Cystoscopy and retrograde pyelography prior to turning the patient laterally ensure there are no other regions of ureteral narrowing. This step can be skipped if a previous imaging study has cleared the length of the ureter.

Position of the patient

1 After intubation, a Foley catheter is placed and the patient is turned to the lateral decubitus position. The surgeon should stand facing the patient's back, with the assistant opposite. Padding and support should be placed under all pressure points. A kidney roll and axillary roll are also placed. The kidney rest is elevated and the table flexed. In small children, a modified anterior approach can be employed, with a small roll under the ipsilateral shoulder and thoracic side.



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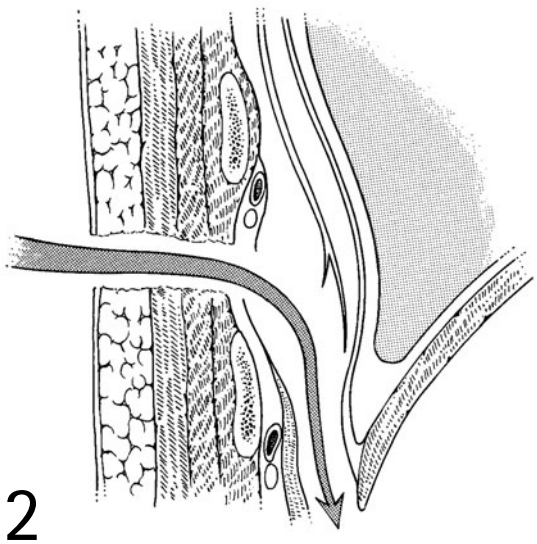

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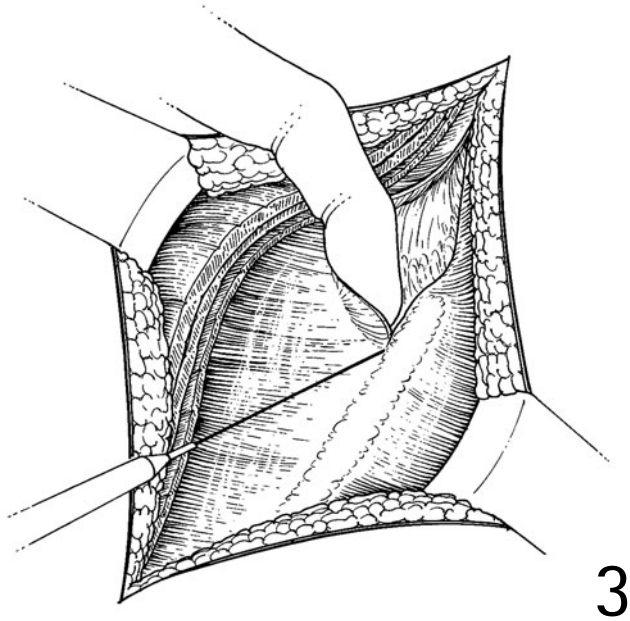
Incision

2 A flank incision is made using a supracostal extrapleural approach. The skin incision is made astride the twelfth rib and extended anteriorly following the natural skin creases towards the umbilicus. The external oblique and internal oblique, the latissimus dorsi, and the anterior edge of the serratus posterior muscles are divided until the periosteum of the rib is exposed. Other approaches include dorsal lumbotomy, anterolateral muscle splitting, and anterior transperitoneal. Lumbotomy allows direct access to the renal pelvis but creates a scar that crosses normal skin folds, and there is limited mobility of the kidney should further maneuvers be necessary. The muscle-splitting approach is suitable in infants and young children, with a modified anterior position. A supine transperitoneal approach is useful when access to the abdominal contents or the contralateral kidney is required. It is also useful when the PUJ obstruction is only one part of a larger urinary tract reconstruction (e.g., augmentation cystoplasty and PUJ obstruction repair).

Releasing the diaphragm

3 Entry into the retroperitoneum is made at the rib tip and extended posteriorly along the superior edge of the rib. The intercostal muscles should be carefully taken off the rib, keeping above the rib to avoid the nerves and vessels. The diaphragmatic fibers are freed from their attachment to the rib using sharp dissection. The translucent pleura falls away as the diaphragmatic fibers are released. The assistant on the ventral side of the patient often has a better view. Patience and gentle dissection allow the fibers to move away naturally. The peritoneum anteriorly is carefully peeled away from Gerota's fascia and retracted to allow for more working room.



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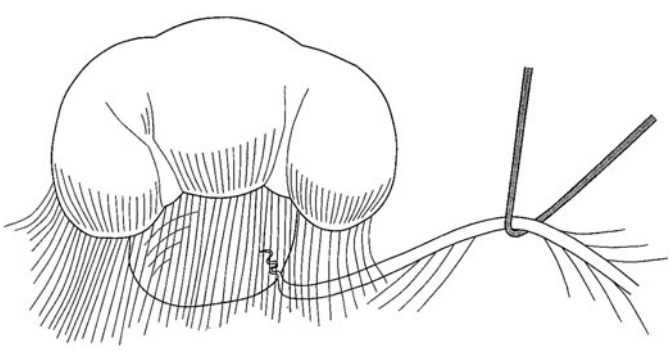
Kidney mobilization

A self-retaining retractor may aid exposure. Gerota's fascia is opened longitudinally and the overlying fat dissected away, taking care not to strip the renal capsule, which may be adherent because of previous pyelonephritis. Patients who may have had renal leak or placement of a decompressive nephrostomy tube also have an inflamed, scarred layer of fat. If possible, maintain the dissected fat as a single layer; it will be useful at the end of the case. Small blood vessels are coagulated. Branches and tributaries of the renal vessels around the anterior hilum should be sought. It is rarely necessary to skeletonize the vessels. The kidney should be mobilized sufficiently to expose the renal pelvis. (The kidney can be held either by a well-padded retractor or by an assistant's hand.) Displacing the kidney out of the depths of the incision may improve exposure, but exaggerated positions should be avoided because of the stretch imposed on the renal vessels and the risk of thrombosis.



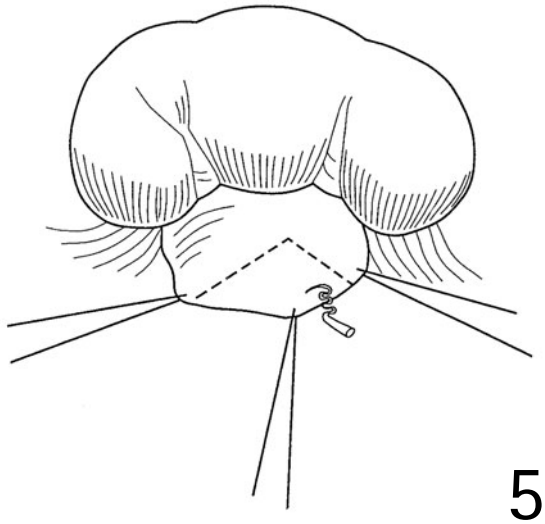
4

Identifying the ureter

4 The ureter is identified and traced toward the kidney. Care should be taken to ensure that the supporting adventitial vessels are not stripped away. Be careful on the left side not to confuse the gonadal vessels, which can have a similar course. Fine stay sutures and vessel loops are used to manipulate the ureter gently. Aberrant vessels, particularly those near the lower pole of the kidney, should be identified, teased away, and not divided.

Preparing the renal pelvis and dividing the PUJ

5 The PUJ is identified and the overlying fat and other attachments cleared away. The type of PUJ (for example high insertion) encountered, presence of crossing vessels, or other findings causally related to the obstruction should be noted. The ureter is divided just below the PUJ and managed on a stay suture. If cystoscopy and retrograde pyelography were not performed prior to surgery, the ureter should be intubated to ensure that there is no distal narrowing. The ureter should be gently moved to check that adequate length has been mobilized. If insufficient length is available, further mobilization is necessary. Stay sutures are next placed on the renal pelvis at points lateral, medial, inferior, and superior to the PUJ. These sutures help define the pyeloplasty.



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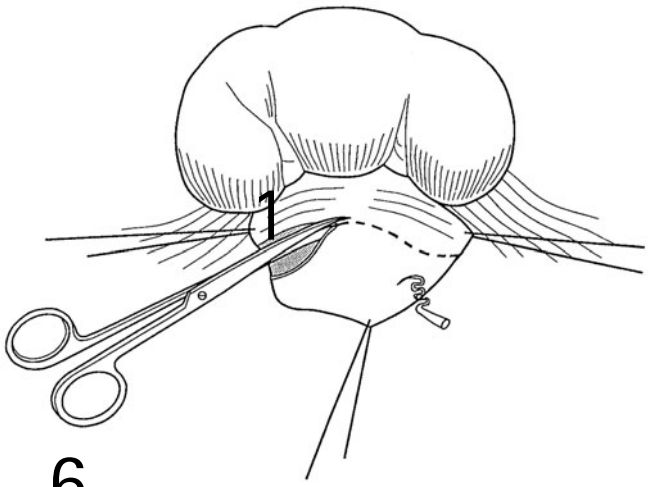
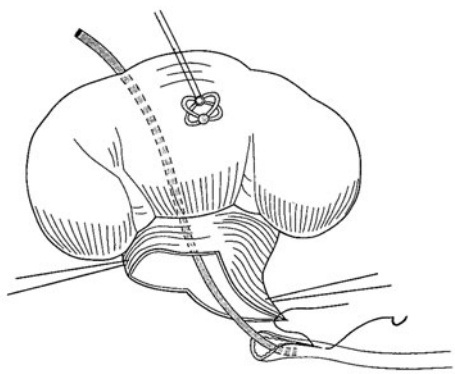


6

Excision of the PUJ and spatulation of the ureter

6 Traction is applied to the stay sutures to splay the renal pelvis flat. One smooth cut is made using sharp tissue scissors; repeated cuts tend to result in saw-tooth, jagged edges. A sharp hooked or pointed scalpel may be used instead of scissors. Irregular tags should be carefully trimmed. Be aware that if the patient has had a history of pyelonephritis or an indwelling nephrostomy catheter or stent, the wall of the pelvis can be thick. Additional pelvis may be excised if needed to reconfigure it easily into a funnel-shaped pyeloplasty. Debris or calculi are irrigated from the kidney. The course of the ureter is studied and noted. It is spatulated along the side that will be most likely to fit the lower extent of the pelvis without resulting in a twist or torsion of the ureter.

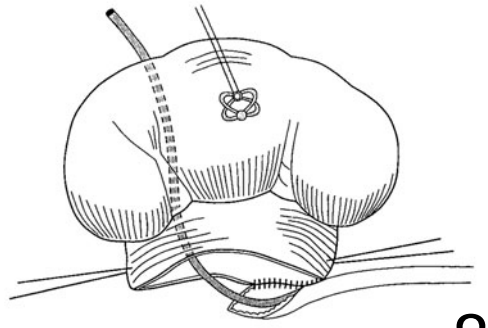
7 At this point we consider a nephrostomy tube and ureteral stent. Neither is necessary for uncomplicated pyeloplasty, but intubation is a useful reassurance in difficult cases, floppy kidneys, revisions, or very redundant pelves. When intubating, the authors prefer both stent and nephrostomy. These can be ready-made or adapted from feeding tubes and small-caliber (10–12 Fr) Malecot catheters.



7

Anastomosis

8 The anastomosis should begin at the caudal end, where the lower lip of the renal pelvis meets the spatulated apex of the ureter. The first suture (6/0 or 5/0 in older children and adolescents) is placed at the apex and the next two sutures are positioned on either side, approximately 2–3 mm apart. A running or interrupted technique completes the process. Sutures should advance up the posterior wall and then up the anterior wall, keeping each bite even so that the tissue is equally distributed and bunching does not occur. When the end of the ureter has been reached, suturing continues until the pelvis is closed.



8

Fat wrap

A tongue of fat freed from Gerota's fascia is carefully but loosely wrapped around the anastomosis and secured by stitching it to itself. A small drain should be placed in the region of the repair. Tubes are brought out through a separate stab incision. If no stents and nephrostomy tubes are used, a drain is mandatory.

Wound closure

The incision is closed in layers, with care taken to re-approximate the muscles. Local anesthetic can be infiltrated along the intercostal nerves for postoperative comfort. All tubes and drains should be securely sewn to the skin.

Postoperative care

The patient should take no solids overnight. Ice chips and sips of clear liquids may be possible if there are bowel sounds. The bladder catheter is removed the following morning and the diet advanced slowly. If the flank remains dry, the drain may be removed in the next day or so. If the repair is intubated, the tube should be left in place for 3 weeks, after which the stent may be removed and a nephrostogram performed. If no drainage is seen flowing down the ureter, the tube should be left to drainage for a further 2 weeks and the test repeated. If contrast flows across the repair, the tube may be removed. Follow-up renal scans and ultrasonography or intravenous pyelography are arranged 3–6 months after surgery.

COMPLICATIONS

The principal complication is stenosis of the anastomosis. Fortunately this is rare, and 90–95 percent of cases are successful. Although the repaired kidney may never look normal on ultrasound scan or intravenous pyelogram, it should function and drain normally on follow-up diuretic renal scan.

Special situations

DUPLEX SYSTEMS

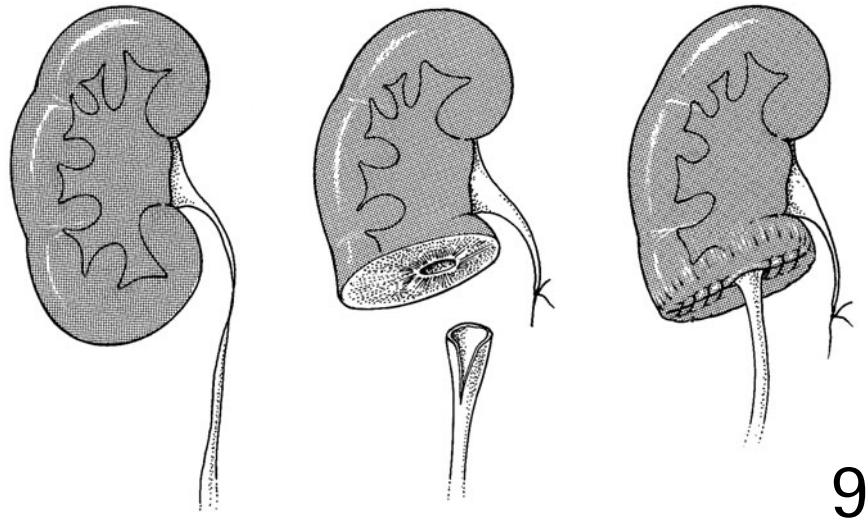
Obstruction is most commonly found in the lower pole moiety of duplex systems. The open pyeloplasty technique described above may be used, but the surgeon must be careful to identify and isolate the appropriate ureter. If a particularly long obstruction is found, ureteropyelostomy or a pyelopyelostomy may be advantageous.

Horseshoe kidneys

In horseshoe kidneys, the ureter often inserts high into the renal pelvis; a longer, more caudally placed anastomosis is needed. Division of the isthmus is usually not necessary. A Foley Y-V pyeloplasty may be useful in some anatomic configurations.

URETEROCALYCOSTOMY

9 If the renal pelvis is too small, intrarenal, or too fibrotic to achieve dependent drainage, a ureterocalycostomy may be necessary. The principal features of this rare procedure are the selection of the most dependent calyx and a thorough amputation of the lower pole of the kidney to prevent postoperative compression.



9

ILEAL URETER

In some situations, the gap is too long to bridge by mobilizing the kidney and distal ureter, usually in patients who have already undergone extensive surgery or trauma resulting in a long atretic ureter. In such patients, an interposition of a piece of prepared ileum is an option. Be sure to try to place it so that the normal peristaltic course of the ileum heads toward the bladder.

LAPAROSCOPIC PYELOPLASTY

Background

Laparoscopic pyeloplasty has been successfully carried out in adult patients, and although it has not yet achieved the same status as laparoscopic cholecystectomy, techniques are improving. Several methods are used: dismembered, flap, and Fenger plasty (a Heineke–Mikuliz-style repair). Initial reports noted longer operating times, and complication rates of up to 13 percent. Incidents of postoperative urinary ascites, blood transfusion, increased cost, and the difficulty of achieving a watertight anastomosis using intracorporeal sewing were among the difficulties. The early pediatric experience noted these same concerns and also the further difficulties of working in a smaller space and the relatively larger size of the instruments at that time. Most approaches to date have been transabdominal and have tried to find ways of improving the anastomosis by the use of specialized sewing aids (such as the Endostitch™). All of the common techniques also involve placement of an indwelling double-J stent either during or before the operation.

Other approaches being explored include the use of robotics and a retroperitoneal access. The DaVinci™ robotic laparoscopic system offers the ability to do true incorporeal sewing in a manner similar to that of open surgery coupled with three-dimensional viewing through a stereo-optic camera (Intuitive Surgical Inc., Mountainview, California, USA). It is, however, very expensive, requires larger ports (a 5 mm port system is being developed), and a specifically robot-trained operating room team. A retroperitoneal approach using a balloon dissector to expand a virtual space into a working one has been advocated by some as offering better exposure and less postoperative morbidity.

General comments

Laparoscopic pyeloplasty is an evolving procedure and no single method predominates. The equipment available is constantly being improved. Smaller diameter, more ergonomic instruments are being introduced. Improvements in vision, depth perception, and haptics are just some of the developments that are now being, or will soon be, implemented. For these reasons, the following description emphasizes those important common features that will probably survive any incremental changes in technology. Because it is still an evolving technique, one of the truly helpful and important aspects when doing this procedure is to have a thorough discussion of the surgical set-up and plan with members of the operating room team prior to the procedure and to review the major steps. Debriefing after a case is also worthwhile. A major surgical tray should be available, as well as drapes in case it is necessary to convert to an open procedure – a possibility that should be mentioned to the family when discussing the case with them. Sometimes an open procedure is best for a particular patient, because of body habitus, anatomical variation, and patient safety concerns, but this may not be determined until the operation has started. Conversion to an open procedure should not be regarded as a failure or complication.

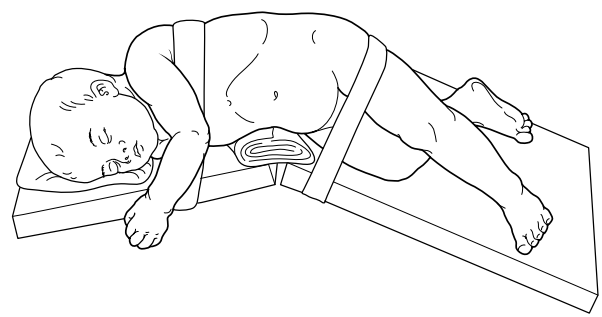
Instruments, equipment, and set-up

In addition to a standard operative laparoscopy set-up, a cystoscopy set-up, double-J stents, Penrose drain, 1/0 and 2/0 synthetic permanent suture on a short Keith needle or minimally curved needle, 5/0–6/0 biological absorbable sutures on atraumatic needles, and needle point and bipolar electrocautery should be available.

Preparation and position

The patient undergoes bowel preparation at home with magnesium citrate or polyethylene glycol the day before surgery, having been on a clear liquid diet the previous day. This will help prepare the bowel should injury occur. Intravenous antibiotics are administered, followed by cystoscopy and placement of a double-J stent. The stent helps when locating the ureter and allows a less traumatic way of controlling the ureter during the anastomosis. However, stents are not without their shortcomings. When in for a long time (more than 2 weeks), the ureter can develop inflammation and thickening of the adventitia and epithelium, thereby making the subsequent anastomosis more difficult. The stents should be about 2 cm longer than the length usually placed. The extra length is needed to reduce the risk of the stent pulling out or being displaced out of the bladder during the laparoscopic manipulation. The stent should also be slightly smaller than usual (3 Fr or 4.7 Fr instead of 5 Fr or 6 Fr) in order to have more spare room between it and the urothelium of the ureteral wall. General anesthesia is used and an oral or nasal gastric tube should be passed.

10 The patient is placed in a modified flank position, with a slight degree of flexion. The bottom leg is flexed at the knee and the top leg is straight. Be sure to check all pressure points and keep the patient well supported and padded. A Foley catheter should be placed. The patient should then be securely taped and strapped down to allow the table to be listed during the case.



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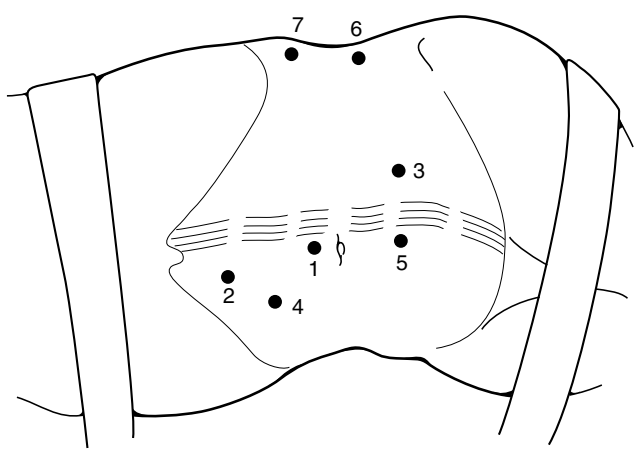


11

Port placement

11 The table is listed so that the patient's abdomen lies flat; typically, this requires the table to be tilted towards the ipsilateral side. Entry and pneumoperitoneum are obtained by a Veress needle or an open Hasson technique.

The port sites can be arranged as follows.

Site 1: just above or below the umbilicus, this site is typically the main camera port. For robotic systems, this is typically a 12 mm port, but with standard laparoscopy can be a 10 mm, 8 mm, or 5 mm port.

Sites 2 and 3: these are the main working ports. Site 2 is placed along the midline about halfway to two-thirds the distance above port 1 to the bottom of the sternum. Site 3 is along the line from the umbilicus to the ipsilateral anterior superior iliac spine. It should be placed beyond the edge of the ipsilateral rectus muscle and should be about halfway to two-thirds distance from the umbilicus to the anterior superior iliac spine. Care should be taken to see that the hip does not limit or hinder instrument movement. This is particularly true with procedures using robotics.

Sites 4 and 5: these are the port sites for accessory ports for use with such tools as irrigators and suction, suture-cutting scissors, and other accessory devices. Usually only one accessory port is used.

Sites 6 and 7: these are locations where sutures can be passed directly into the abdomen. Additional accessory ports can be placed here, and at the end of the operation a drain can be passed at these sites.

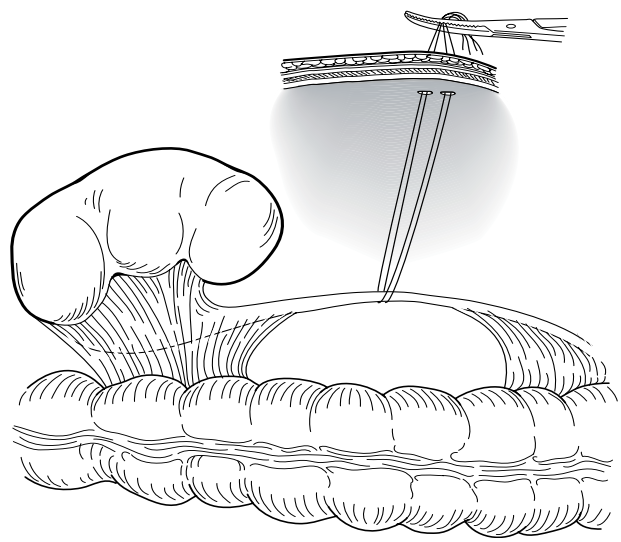
Depending on the size and shape of the patient, prior surgery, and the surgeon's experience, the location of the ports can be adjusted.

Exposure of retroperitoneum

The table is now leveled. Using atraumatic forceps in one working port and a hook dissector or Harmonic™ scissors in the other, the colonic attachments along the white line of Toldt are taken down and the colon is reflected medially. Attachments at the spleen or liver are taken down as needed. Usually these organs do not require retraction or distraction and do not interfere with the subsequent procedure. A rake or other atraumatic instrument can be passed through an accessory port to help distract the colon medially. The table can then be listed contralaterally to use gravity to hold the colon out of the working field.

Retroperitoneum dissection

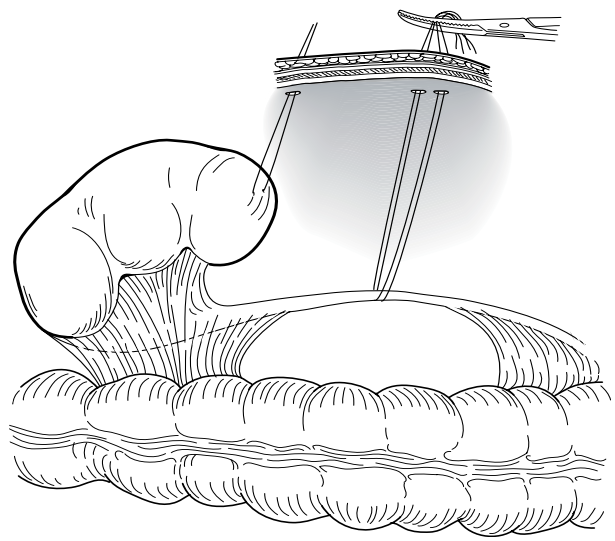
12 Starting at a point below the kidneys, roughly parallel to the camera port, begin working laterally to medially, gently spreading and lifting, always looking for the ureter. The ureter being stented should have a full, stiffer feeling to it. Be careful not to mistake the gonadal vessels for the ureter. If the gonadal vessels are found first, the ureter is usually a bit more posterior. A right-angle dissector and hook cautery are useful tools in this phase. After locating the ureter, pass a 1/0 or 2/0 synthetic suture on a short Keith or CT needle directly through the abdomen or an accessory port (see port site 6 in previous diagram), loop it around the ureter, and pass it back out of the abdomen. This gives control around the ureter. Loosely snap the suture with a mosquito clamp. Be careful not to put too much tension on this suture or it will act as a ligature and strangle the ureter. During the course of the procedure, periodically take tension off this suture to prevent ureteral ischemia. In some patients, the Keith needle may be too long and awkward to use easily inside the abdomen, and the standard swedged needles may be too curved to pass through the abdominal wall. In these cases, you may have to thread the suture onto a separate needle or gently straighten the bend of the needle.



12

Freeing up lower pole of kidney and pelvis

13 With the ureter identified, gently take down the inferior attachments. This action will give you more room and play on the ureter. Next, continue to dissect toward the kidney. As you encounter the lower pole of the kidney, begin to take down some of the attachments of the kidney to the surrounding muscle wall, but try to limit this dissection to the lower pole. It is advisable to avoid mobilizing the kidney too much, as it can flop down and make it harder to operate. Should this occur, pass another 1/0 or 2/0 synthetic stay suture through the abdomen or an accessory port (see port site 5 in previous diagram). Use it to sew gently and carefully to the renal capsule and lift up the renal pole. Next, repeat the maneuver and catch the renal pelvis and then pass it back out of the abdomen. This can be used to help stabilize the pelvis during surgery. As the lower pole is freed up, the pelvis should come into view. Carefully clear away the surrounding fat and attachments. At this point, be aware of any crossing vessels and make a careful assessment of the anatomy.



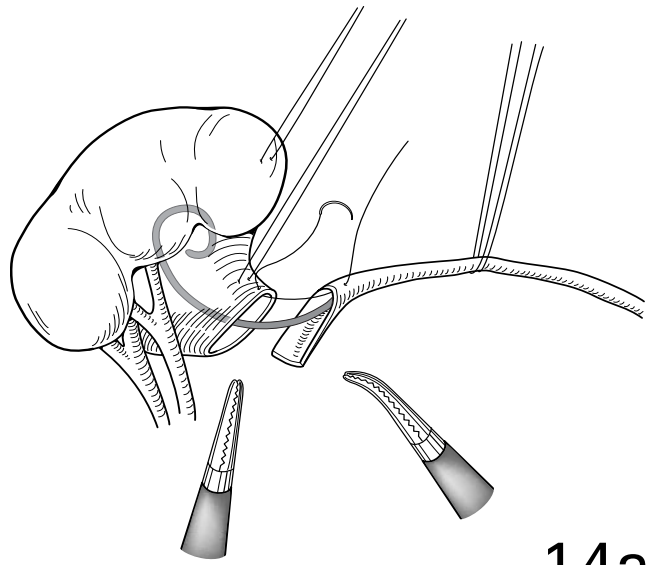
13

Cutting the PUJ and spatulating the ureter

Using sharp scissors, cut the ureter just below the PUJ. This can be tricky, and you may have to use whichever arm or port offers the best angle. Cut the ureter on a bias, slanting upwards. Spatulate the ureter on the side that would connect to the lowest point of the pelvis. Use the stent to help hold the ureter. Limit direct grasping of the ureter, particularly if you are using a robotic approach. Cut the PUJ away from the pelvis, again on a bias. The aim is to keep the front and back walls symmetric and avoid dog ears and irregular edges. If it has not been done previously, tag the pelvis with a stay suture. Cut the PUJ segment off the stent.

Anastomosis

14a–c Before sewing, be sure that the sutures have been cut to an appropriate length. Laparoscopic sewing does not require as long a suture as open surgery. Typical lengths are about 10–12 cm. For a robotic approach, because of the magnification, sutures as short as 6–8 cm can be used. Whether sewing using robotic or Endostitch™ assistance, first sew the lowest point of the pelvis to the spatulated corner of the pelvis. Sew the back wall first, which we usually do in an interrupted fashion. The corner stitch can be used to help maneuver the pelvis and ureter for better exposure, but great care should be taken not to tear. After the back wall is complete, the front wall is done. If there are any remaining gaps in the renal pelvis, these can then be sewn up.



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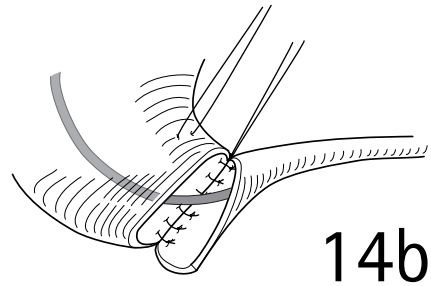

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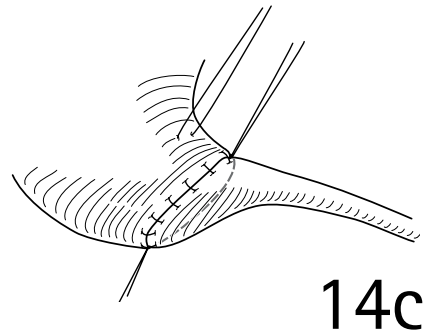

14c

Exiting

The pneumoperitoneum is then taken down to 8–10 mmHg and the area is checked for bleeding. After these are controlled, the pressure is returned to12–15 mmHg. Try to wrap some of the perirenal fat or Gerota's fascia fat around the anastomosis and sew it to itself. Take down the stay sutures. Pass a Penrose drain into the abdomen and lay it nearby. Secure the drain to the skin with a nylon suture. Remove the ports, and close the entry sites. Place small dressings over each site and a dressing over the Penrose site.

Postoperative care

Keep the patient on antibiotics for 1 week. Keep the Foley catheter in for 24 hours. Do not feed until there are bowel sounds. The Penrose drainage should be minimal and should subside by 48–72 hours. The patient can be discharged once able to take food and drink, get about, void, and manage with oral pain medications.

Complications

The complications are similar to those for an open surgical procedure. Some patients may complain about irritation and urgency from the stent. Anticholinergics will help limit these symptoms.

Follow-up

Follow-up should occur after about 2 weeks to check on the port sites. The double-J stent should be removed at 3–4 weeks, and prior to this, an intravenous pyelogram or a retrograde pyelogram can be performed at the time of the stent removal. A follow-up dynamic renal scan is done at 3–6 months.

FURTHER READING

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Peters CA, Scyhlussel RN, Retik AB. Pediatric laparoscopic dismembered pyeloplasty. *The Journal of Urology* 1995; **153**: 1962–5.

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Laparoscopic Anderson–Hynes dismembered pyeloplasty

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INTRODUCTION

Congenital obstructive uropathy involving the upper tract constitutes a significant proportion of the clinical caseload of a pediatric urologist. With many upper tract conditions being diagnosed antenatally, the challenge for pediatric urologists today is to identify the cases that should be treated early and those that can wait for planned surgery without detriment to the well-being of the patient. Whereas most cases of congenital obstructive uropathy can be watched safely, some cases of severe ureteropelvic junction obstruction may require early surgical intervention to preserve upper tract function.

Laparoscopic Anderson–Hynes dismembered pyeloplasty is one of the most technically challenging laparoscopic operations and requires meticulous attention to every detail, including patient positioning, floor plan, and especially port placements. With correct attention to detail, laparoscopic dismembered pyeloplasty can be completed within 90 minutes.

Patient selection

In general, there is very little indication for laparoscopic surgical intervention in the newborn. However, significantly impaired renal function with gross delay in excretion and evidence of increasing obstruction are some of the indications for pyeloplasty.

Indications and contraindications

Laparoscopic Anderson–Hynes dismembered pyeloplasty can be successfully performed with good functional results in any age group. However, this operation is not recommended in smaller babies unless you are very adept at laparoscopic suturing. Laparoscopic pyeloplasty can also be performed on horse-shoe kidneys. However, pelvic ectopic kidneys cannot be approached laparoscopically, as the ureteropelvic junction is usually beyond the reach of the laparoscopic instruments, the ureteropelvic junction being completely obscured from view, somewhat like the dark side of the moon.

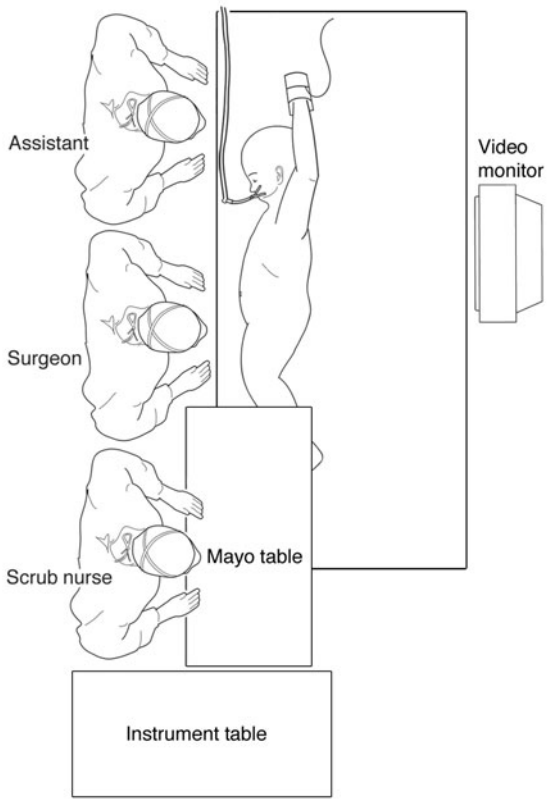
It has been the author's experience that right-sided pyeloplasties are more difficult because the liver prevents optimum placement of the suture port, and it is recommended that surgeons master the left-sided operation before attempting a right-sided repair.

Instrumentation and telescopes

It is not necessary to have an extensive range of laparoscopic instruments. Instruments that are only rarely used will add to the laparoscopic clutter. It is better to use a few, selected, good instruments. Our preference is a 5 mm telescope, which is not only more rigid but also provides adequate illumination and resolution to allow one to perform very delicate suturing. We prefer to use 3 mm 20 cm laparoscopic instruments for infants and young children. Finer needlescopic 2 mm instruments are widely available, but our experience with these is that they have limited tissue grip, provide insufficient leverage, and are easily bent. Avoid using long instruments in young children. The limited internal working space in a child results in these instruments protruding excessively, leading to exaggerated movements and constant instrument clash.

PREOPERATIVE PREPARATION AND GENERAL CONSIDERATIONS

Informed consent should be obtained for transperitoneal or conventional dismembered pyeloplasty. An enema should be administered, especially for left-sided ureteropelvic junction obstruction. While some authors advocate the insertion of a stent preoperatively, this author has not found this to be necessary, as it leads to complete decompression of the renal pelvis, making it more difficult to mobilize the pelvis and ureteropelvic junction. The stent may also become entangled with the suture, making an already challenging task more difficult. The author's preference it to insert an antegrade stent after completion of the posterior anastomosis.



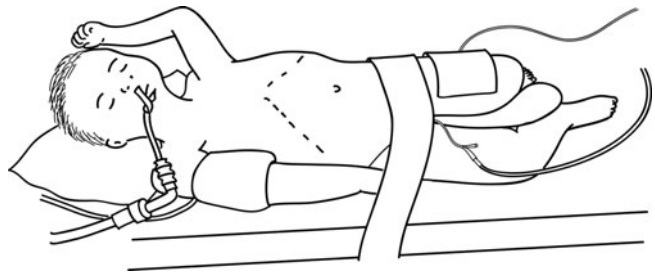
1

Floor plan

1 A transurethral Foley catheter should be inserted before the patient is positioned. The patient, surgeon, assistant, and scrub nurse should be positioned as per the floor plan. This gives the best ergonomics for this procedure.

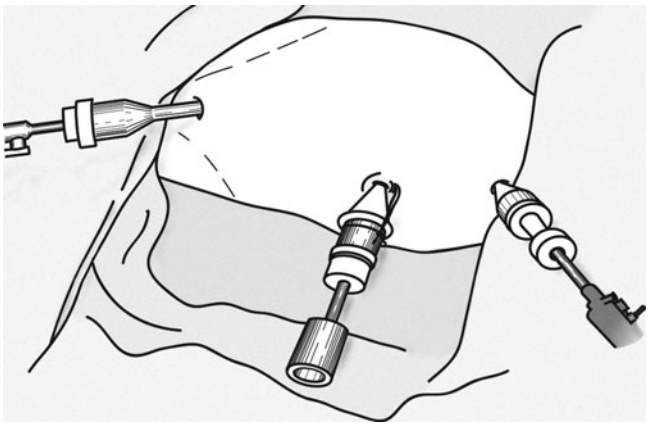
Patient positioning

2 It is important to ensure that the hips are not flexed, otherwise the legs will impede your ability to manipulate the instruments.



2

A superior circumumbilical incision is then made in the periumbilical skin crease down to linea alba, and the linea alba about 1 cm cephalic to the umbilical cicatrix is grasped between two hemostats, and opened. The peritoneum should be easily identified deep to this, and opened. A purse string is placed around the linea alba, and a 7 mm Hasson port is inserted and secured by tightening the purse string around it.



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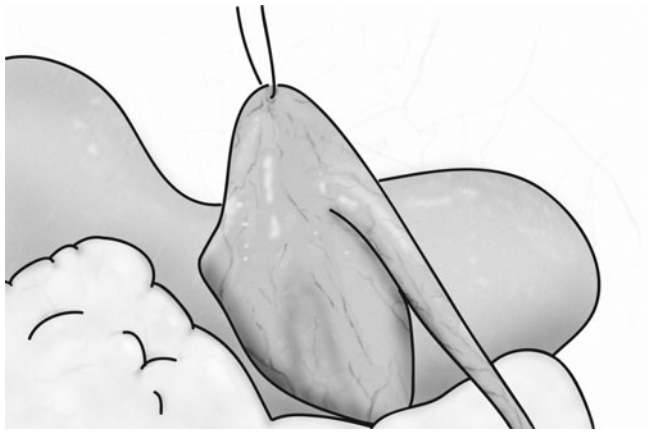
3 Port positions are critical for pyeloplasty. It is not possible to perform a satisfactory intracorporeal anastomosis if the needle driver is not in line with the intended line of anastomosis. This means that the suturing port should be placed as close to the midline as possible.

Only two instrument ports are used for the procedure, a 3.5 mm port and a 6 mm port, which is necessary to pass the laparoscopic suture into the abdomen. The 6 mm instrument trocar should be right handed, unless you are left handed, in which case port placement should be reversed.

The colon should then be detached from the kidney and Gerota's fascia opened. It is important to find the correct tissue plane at this point and to be absolutely sure you are right on the renal capsule. The renal capsule is covered by a thin film of loose adventitia, and it is important to be deep to this plane, keeping absolutely snug to the renal capsule. The plane is developed medially into the sinus, where the renal pelvis should then be easily identified. The renal vein may be exposed during this maneuver.

The ureteropelvic junction can usually be identified at this point, if you can identify the gonadal vessels which are usually located near the ureteropelvic junction. Once the ureteropelvic junction has been identified, the ureter should be mobilized gently by lifting it out of its bed to develop a window to hold it up. One should only mobilize just enough length of proximal ureter to create a window, to preserve the vascular supply. The renal pelvis may be mobilized more extensively, sweeping the renal vessels toward the upper pole. This is usually a bloodless procedure if you are in the correct plane. Clear about 2 cm of renal pelvis above the upper limits of your intended pyelotomy.

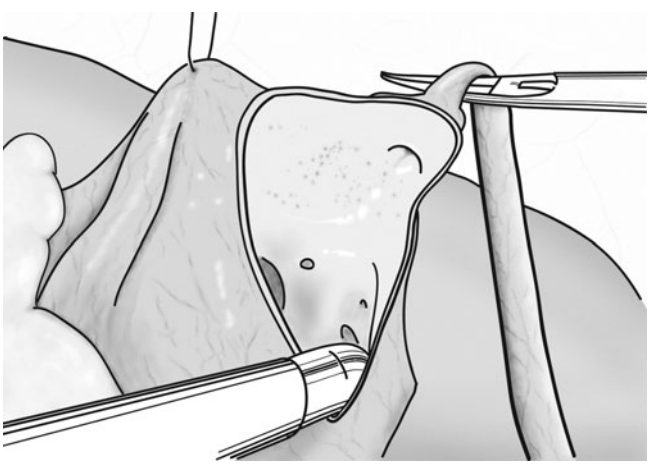
4 At this point, a 4/0 monofilament suture on a straight needle is then passed through the anterior abdominal wall at the mid-axillary or posterior axillary line just below the costal margin and grasped from the inside with a laparoscopic needle holder. The renal pelvis should be transfixed about 1–2 cm above the intended upper limits of the pyelotomy, and the needle is then passed through the abdominal wall near its insertion point. The renal pelvis can then be pulled out of its bed by this hitch stitch, displaying the ureteropelvic junction in full view. A pair of hemostats is applied to the hitch stitch on the outside just as it comes through the abdominal wall, and this will serve to stabilize the renal pelvis for the duration of the pyeloplasty.



4

It is important to leave a good length of the suture on the outside, as it may be necessary to loosen the hitch stitch if the anastomosis is under tension. Do *not* use a second hitch stitch on the ureter, as this makes for a more difficult anastomosis because of suture clash and will result in the ureter being detracted from the renal pelvis.

It is important at this stage to take a few minutes to clear the renal pelvis and proximal ureter of any loose connective tissue before dividing the ureteropelvic junction, otherwise the ureter will retract within the sleeve of connective tissue, resulting in a difficult anastomosis.



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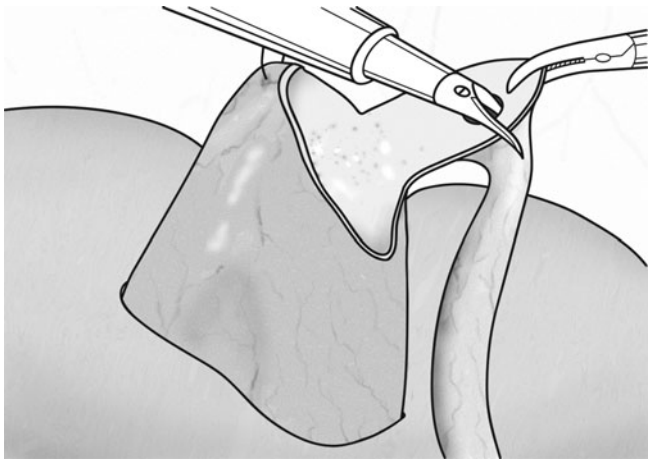
5 The most dependent part of the pelvis is identified and the renal pelvis opened at its most dependent position. The anterior wall of the renal pelvis is then opened along the entire length of the intended pyelotomy.

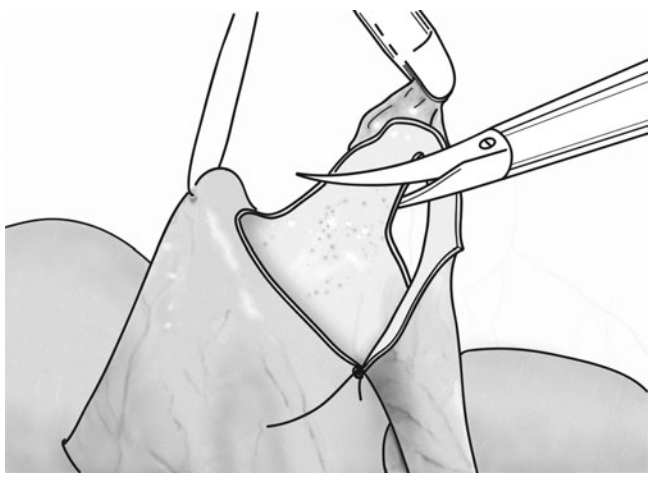


6

6 Do not completely dismember the ureteropelvic junction, as the partially attached posterior wall allows you to display the proximal ureter by grasping the portion of renal pelvis to splay the ureter open. The proximal ureter is then spatulated.

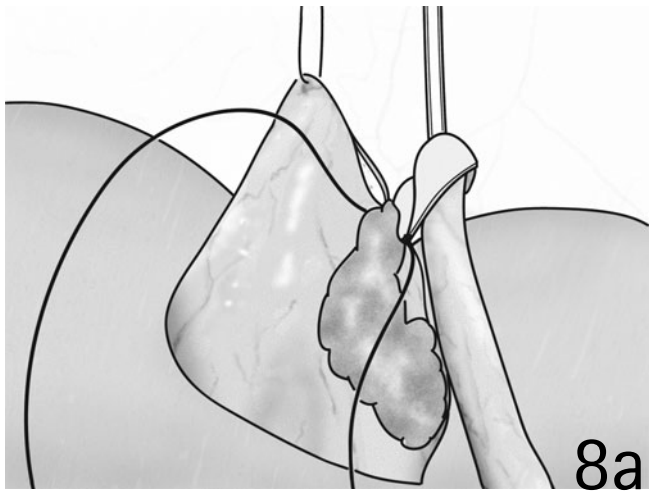
7 The first suture should be placed at the angle of the spatulated ureter to the most dependent part of the partially opened renal pelvis, before the pelvis is dismembered. This suture is best placed by inserting a fine, straight, non-toothed, atraumatic grasper into the proximal ureter to splay open the spatulated ureter to display its angle.

A 7 cm length of 6/0 PDS (5/0 in older children) with a preformed loop is held by its knot and introduced into the abdominal cavity. Leave about 2 cm of tail to enable you to use it to complete the anterior anastomosis. The needle is then mounted on the needle holder, in position for the first anastomosis. Once the angle has been re-anastomosed, the posterior pyelotomy can be completed.

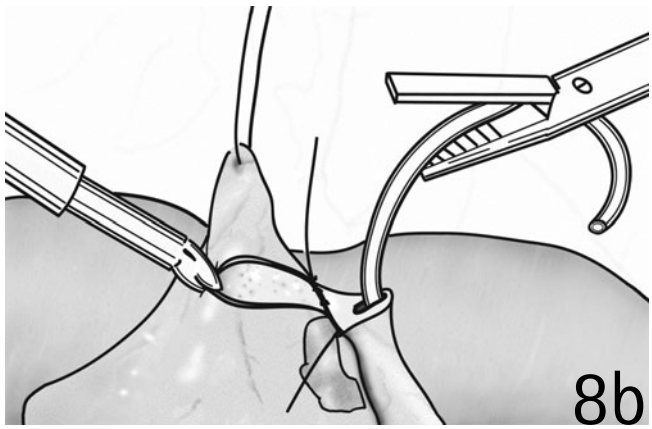


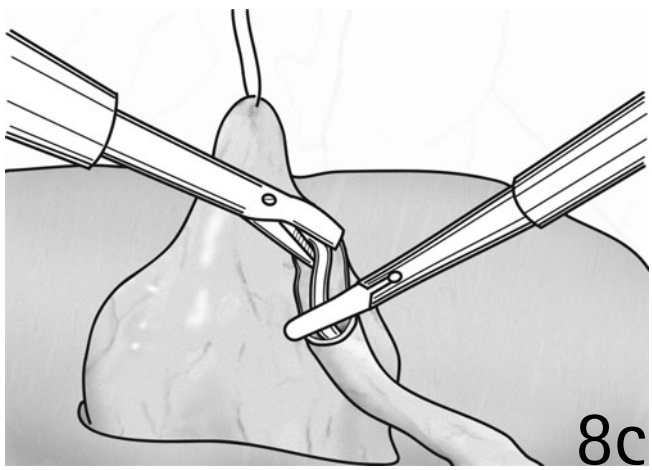
7

Do not excise redundant pelvis until the posterior anastomosis is completed. The redundant pelvis that is to be discarded can be grasped with impunity without risk of handling the urothelium. The posterior anastomosis is completed as a continuous suture. The anastomosis should then be inspected internally by grasping the redundant renal pelvis to open up the ureter.



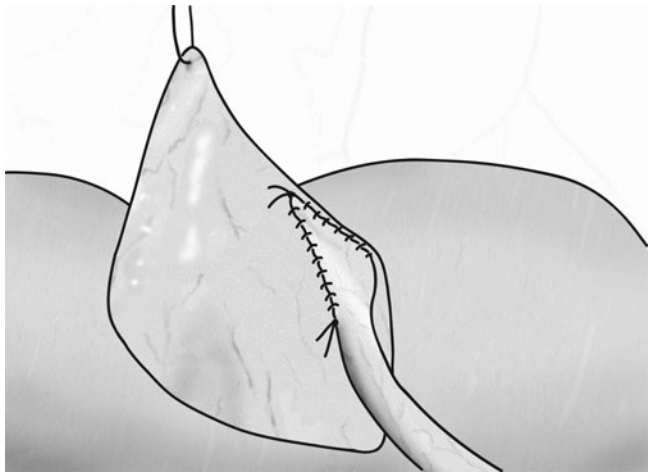
8a–c The antegrade stent should then be inserted at this point. A 19 Fr Teflon-sheathed needle is introduced just in front of the hitch stitch through the anterior abdominal wall; the needle is retracted into the Teflon sheath, and the Teflon sheath is guided into the proximal ureter. The needle is removed and a guidewire is introduced into the bladder. Some resistance is usually felt at the ureterovesical junction orifice. It is best to use a hydrophilic-coated 'slippery wire'. Once the guidewire is in the bladder, the Teflon sheath is removed, and a well-lubricated multi-length pig-tail catheter is passed over the guidewire and introduced into the bladder. It is best to pass the entire length into the bladder, leaving a single loop proximally, which can then be pulled out further if need be, to ensure that the distal end is in the bladder.





The previously inserted Foley catheter will prevent the stent from migrating into the urethra. After withdrawal of the guidewire, it should be possible to express one or two drops of urine by suprapubic compression to ensure that the stent has been correctly positioned. It is also convenient not to excise the redundant renal pelvis until after the insertion of the pig-tail stent, as it may be necessary to exert countertraction on the pig tail to pass it through the ureteropelvic junction, taking the tension off the completed posterior anastomosis. The proximal end of the pig-tail catheter is then introduced into the renal pelvis.

9 The redundant renal pelvis is then trimmed and discarded, and the anterior anastomosis completed with a continuous PDS suture, starting at the top of the pyelotomy down to the angle of the spatulated ureter. The loose end of the posterior anastomosis is then grasped and the anterior anastomosis completed by tying it to this loose end. A final inspection is then made of the anastomosis before the hitch stitch is removed and the pelvis and ureter returned to their bed.



9

Points of technique

A patient presenting with intermittent ureteropelvic junction obstruction with predominantly intrarenal pelvis presents a special challenge, as the intrarenal pelvis can severely restrict one's ability to manipulate the laparoscopic instruments. The patient must have a fluid load and frusemide to produce maximum diuresis. One should wait for maximal distension of the renal pelvis before placing the hitch stitch to transfix the renal pelvis out of the hilum. Under no circumstances should one divide the ureteropelvic junction before the renal pelvis is transfixed, as this will result in the pelvis retracting into the depths of the hilum, making suturing impossible. Crossing lower polar vessels are easily identified laparoscopically without the need of CT imaging.

If the pathology is an aberrant lower pole vessel, it is necessary to dismember the ureter from the pelvis completely in order to transpose the vessels, but spatulate the ureter before dismembering the ureteropelvic junction.

On completion of the pyeloplasty, tilt the patient head-up to allow the urine to drain into the bony pelvis, from where it can be aspirated. The patient is then tilted away from the surgeon, the bean bag supporting the back is removed, and the patient is placed as supine as possible. The colon is then re-manipulated into its original position. The peritoneal incision will be completely covered by the colon, leaving no raw surfaces. The author has not found it necessary to re-suture the colonic peritoneal attachment, as the colon becomes adherent quite quickly. There is no need for a perinephric drain.

With attention to detail, laparoscopic dismembered pyeloplasty can be completed within the same timeframe as conventional open pyeloplasty. While technically challenging, it is not beyond the realm of any laparoscopist who is competent at intracorporeal suturing. The 5/0 suturing technique can usually be mastered after several hours of practice, while 6/0 PDS suturing is considerably more difficult. Of all laparoscopic procedures performed by the author, none is more satisfying than a pyeloplasty.

ACKNOWLEDGEMENT

Mr Justin Kelly FRACS, for assistance with the illustrations.

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Partial nephrectomy

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HISTORY

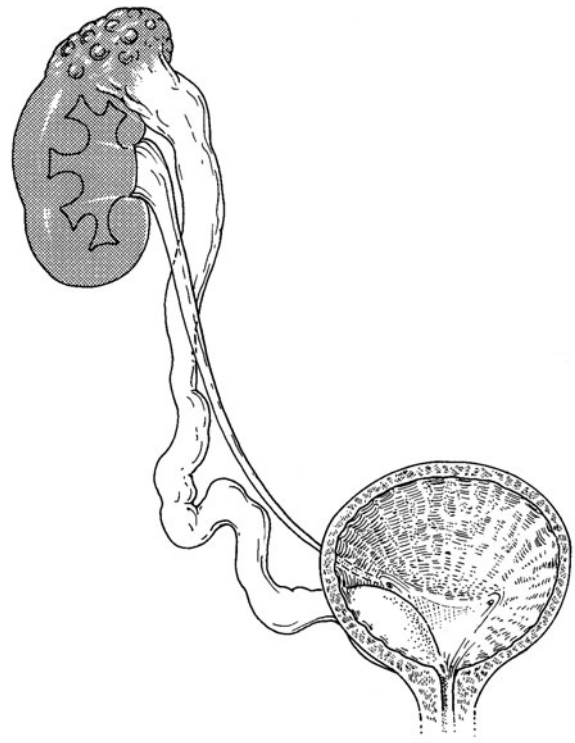
The first planned partial nephrectomy was by Kummell in 1889 for stone disease. Partial nephrectomy was initially performed predominantly for infectious problems and urolithiasis. Many patients in the early 1900s continued to have complete nephrectomy because of concerns about extensive bleeding following partial nephrectomy and the frequent occurrence of urinary fistula. Eisendrath reviewed the treatment of duplex kidneys in 1923 and found only 13 reports of heminephrectomy. With improvements in radiographic diagnosis and surgical techniques, this operation became more routine, even in younger children.

PRINCIPLES AND JUSTIFICATION

Indications

The indications for partial nephrectomy for benign disease have diminished. The treatment for urolithiasis has evolved from an open surgical approach to the use of endoscopic techniques and extracorporeal lithotripsy. Early recognition and treatment of vesicoureteric reflux and urinary tract infections have lessened the risk of severe segmental renal damage.

1 The most common indication for partial removal of the kidney in a child is complete ureteral duplication with a poorly functioning upper pole segment. The majority of these patients are diagnosed prenatally. The upper pole moiety becomes dilated because of obstruction secondary to ureterocele or ectopic insertion of the ureter.



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Evaluation of these patients consists of cystography, renal ultrasonography, and radionuclide scans. This will provide the correct diagnosis in most cases and enable the clinician to decide on the proper therapy. There are several treatment alternatives, including endoscopic incision of an obstructing ureterocele, ipsilateral ureteroureterostomy, pyeloureterostomy, or ureteral re-implantation. The merits of each approach continue to be debated. In the absence of concomitant vesicoureteral reflux, partial nephrectomy with segmental ureterectomy (open or laparoscopic) is the favored approach for a poorly functioning upper pole moiety.

Other less common indications for segmental resection of the kidney are trauma, renal cysts, and calyceal diverticula. The latter two problems are now usually managed with endoscopic or percutaneous techniques.

PREOPERATIVE ASSESSMENT AND PREPARATION

Advances in imaging techniques provide detailed assessment of the anatomy of duplication anomalies. Renal ultrasound is the preferred initial imaging study. Detailed images of the duplication anomalies in the fetus can be obtained early in gestation. Voiding cystogram is necessary in all children with duplication anomalies to exclude reflux. Assessment of the functional status of the upper pole segment is best done with either technetium dimercaptosuccinnic acid (DMSA) or ^{99m}technetium mercaptoacetyl triglycine (MAG-3) radionuclide imaging of the kidney. Intravenous urogram plays a limited role. In select cases, the anatomy of the kidney and collecting system may remain unclear. Magnetic resonance imaging (MRI) urogram has been shown to be very accurate in depicting the anatomy of duplication anomalies. When the child is brought to the operating theater, the lower urinary tract is examined endoscopically. Retrograde ureterography or puncture of the ureterocele with retrograde injection of contrast medium can be performed if it is necessary to define the anatomy further.

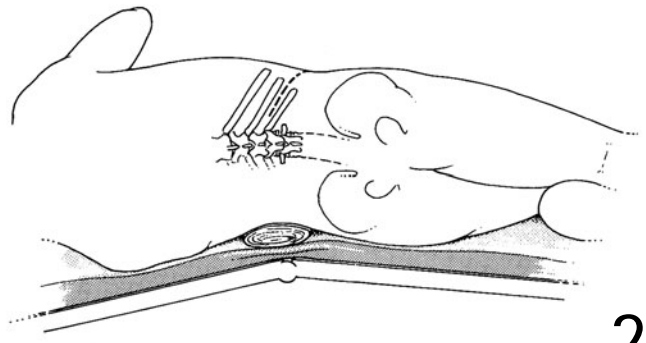
Anesthesia

General anesthesia is used in all cases. Caudal or epidural anesthesia is a useful adjunct for postoperative pain control. Intercostal or paravertebral nerve blocks are also effective for pain management.

OPERATIONS

Position of patient

2 The flank or lateral approach is preferred and the patient is placed in a full lateral position. The break in the table is utilized for older children, but a small towel or pad under the mid-section may suffice in the neonate. A bladder catheter is placed to monitor urine output. Proper padding of all pressure points is important. The patient is secured to the table with tape to prevent motion during the procedure.



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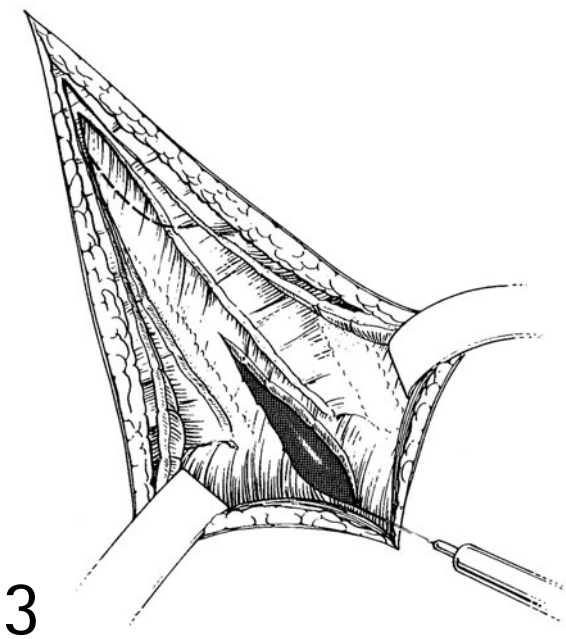

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Incision

3 The incision will vary with the indication for surgery. Trauma patients are likely to be undergoing exploratory laparotomy, and therefore a transabdominal approach is utilized. A subcostal incision or incision above the twelfth rib is used for most elective cases. The higher incision affords better exposure and decreases the need for traction on the kidney during upper pole nephrectomy.

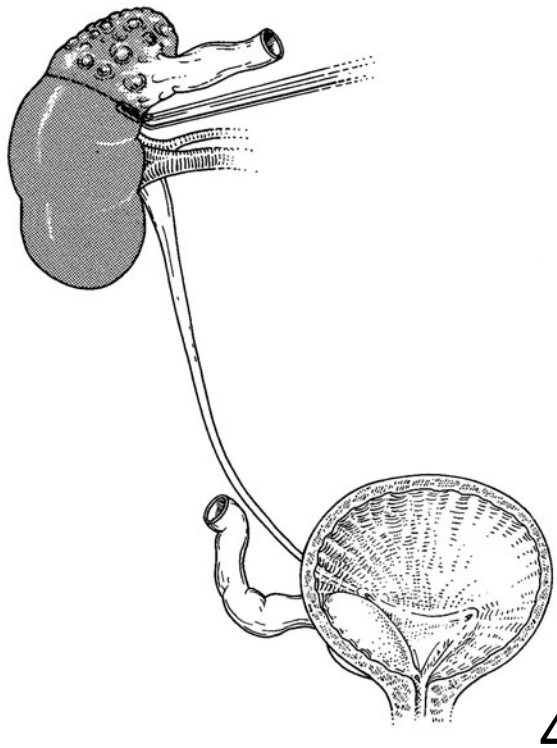
With a subcostal incision, the muscles can either be split in the direction of the fibers or divided with cautery. The neurovascular bundles are identified and preserved. The peritoneum is swept medially. With the approach above the twelfth rib, it is important carefully to free the diaphragmatic attachments to the lower rib and avoid tearing into the pleural space when the self-retaining retractor is placed.

Gerota's fascia is entered posteriorly after palpating the kidney. The perirenal fat is then dissected by both blunt dissection and cautery. At this point the ureter and renal vessels are identified. Mobilization and isolation of the renal vessels are generally not necessary, particularly in neonates, in whom the vessels are prone to spasm. Infiltration of the hilar area with lidocaine (lignocaine) (0.25 percent solution) and papaverine 15 mg has been recommended to prevent this problem when performing renal surgery in infants.

The renal segment to be removed is generally readily identifiable, either due to atrophy and thinning of the renal parenchyma or because of a dysplastic or cystic appearance of the involved region. The upper pole moiety is generally small, representing less than 15 percent of the total parenchymal volume of the kidney. Occasionally, a centrally located cyst or calyceal diverticulum may not be apparent on initial inspection.

Partial nephrectomy for duplication anomaly

4 In most cases, the dilated upper pole ureter is readily identified, but care must be taken as the lower pole ureter may also be abnormal. Division of the ureter is not undertaken until the origin from the upper pole is assured. After transection of the ureter, the collecting system is decompressed. This allows the junction between the upper and lower renal segments to be readily identified. In most patients, the ureters can be easily separated at the renal level. With careful dissection, the upper pole ureter is freed from its mate and transposed above the renal hilar vessels. Traction on the upper pole ureter should be minimized to avoid injury to the renal vessels. Maintaining the dissection plane close to the wall of the upper ureter allows access into the renal sinus between the two collecting systems. This helps define the line of resection through the renal parenchyma.



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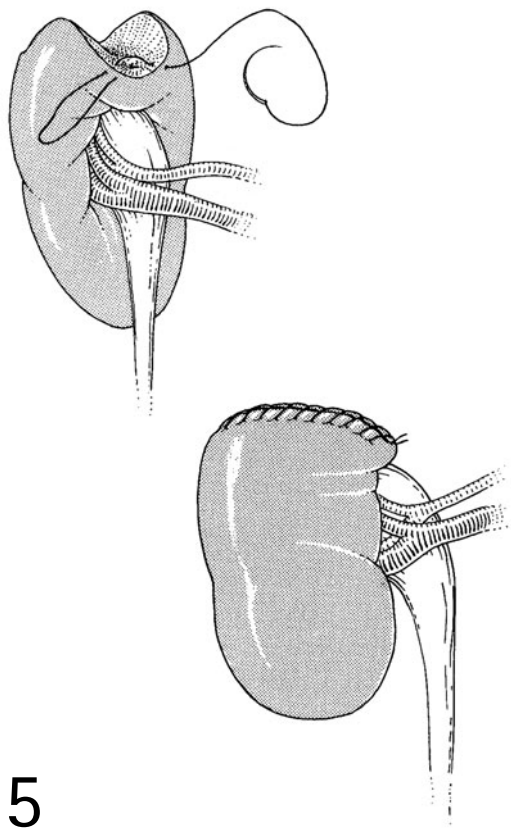

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5 The author prefers to incise the renal parenchyma with the cautery in those cases where it is dysplastic and/or atrophied. On the periphery, the parenchyma of the lower pole moiety often extends cephalad to the upper pole segment (see illustration). If a branch of the renal artery is clearly seen to enter the parenchymal segment being removed, it can be ligated and divided. If there is any doubt about the region supplied by an upper pole arterial branch, a vascular clamp can be placed and the line of demarcation observed. Removal of the cystic non-functional upper pole segment can generally be accomplished with minimal blood loss. After excision of the upper pole, control of bleeding is obtained with a combination of cautery and suture ligation. Clamping of the main renal vessels is not generally needed. Gentle manual compression of the parenchyma will control bleeding. Mattress sutures can be placed to approximate the cut edges of the renal parenchyma. Pledgets of perirenal fat or Surgicel® (oxidized, regenerated cellulose) will help prevent the suture cutting through the parenchyma.

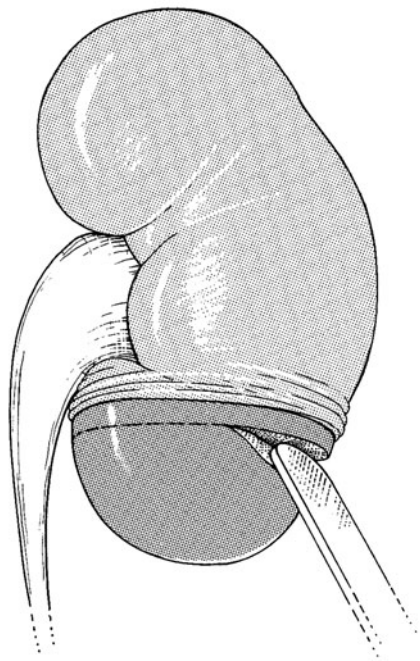
A potential future adjunct for obtaining hemostasis is the use of tissue sealants. Fibrin sealant is a biologic tissue adhesive consisting of fibrinogen and thrombin. There have been reports of the effective use of fibrin sealant for hemostasis after partial nephrectomy. These products have not been approved by the US Food and Drug Administration for this indication.

When the upper pole resection is complete, the ureter is traced distally. Care should be taken to avoid devascularization of the lower ureter. The plane of dissection should be very close to the upper pole ureter. If the ureters are closely adherent, a strip of the upper pole ureter can be left attached to the lower pole ureter, but this is rarely needed. The ureter is removed as low as possible via the flank incision. This will be below the level of the iliac vessels in most infants and small children. The ureter is then transected and a catheter is passed into the distal ureteral stump. This is irrigated with antibiotic solution and the catheter is aspirated. The ureter is left open, except in rare cases of reflux into the upper pole moiety; complete excision of the distal ureter or very low ligation is more appropriate in these patients. Drains are placed in both the renal bed and pelvis (near the transected region of the ureter).

Partial nephrectomy for other conditions

6 Once the segment to be removed has been identified, the capsule can be incised with a scalpel and peeled back (this may be adherent if the renal segment is diseased) or cautery may be used to incise the renal parenchyma. Classic teaching is that blunt dissection with the back of a knife handle can be used to separate the parenchyma without disrupting the major blood vessels, which can then be ligated.

As mentioned, if the renal parenchyma is atrophied, it is divided with cautery. If there is considerable renal parenchyma to be divided, the Cavitron Ultrasonic Surgical Aspirator (CUSA) is very effective. The parenchyma is separated without dividing the vessels, which minimizes blood loss.



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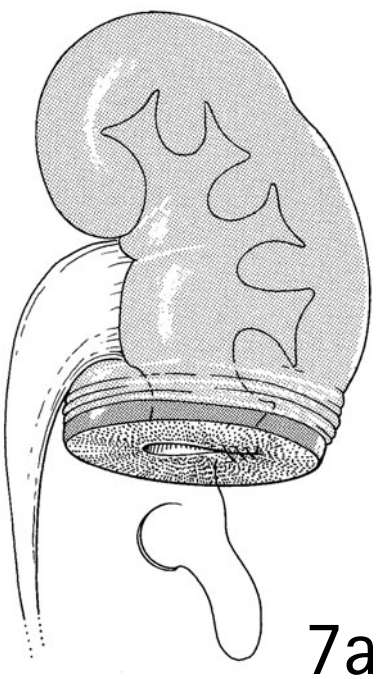

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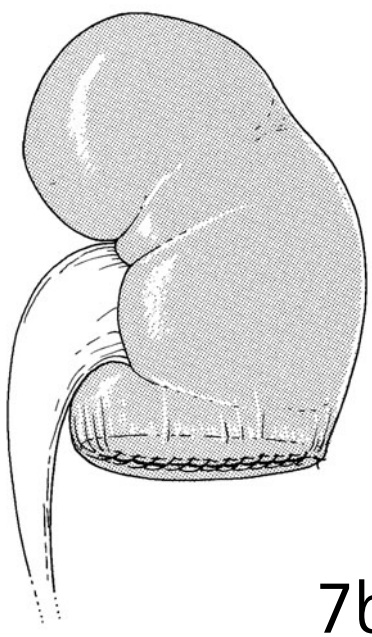

7b

7a,b After partial resection of a kidney with a single collecting system, closure of the transected calyx or infundibulum is required. A watertight closure with fine absorbable suture (5/0 or 6/0 polyglycolic acid) is recommended. The renal capsule is closed over the cut surface of the kidney. If the capsule is not available, a patch of peritoneum can be used.

POSTOPERATIVE CARE

The majority of children require only a short hospital stay after an uncomplicated partial nephrectomy for benign disease. Many infants are ready for discharge the day after surgery. The use of epidural catheters after operation can eliminate much of the postoperative discomfort. Most complications will be apparent before discharge from the hospital.

Complications

PNEUMOTHORAX

Before the wound is closed, the flank is filled with saline and the anesthetist holds the patient in deep inspiration to check for air leakage. If the parietal pleura has been opened but the visceral pleura is intact, a small catheter is placed into the pleural space and the air is evacuated. Closure of the pleural space and diaphragm is then performed and wound closure is accomplished. After closure of all the muscle layers, the catheter is removed. A chest radiograph is obtained in the recovery room to ensure complete expansion of the lung.

URINE LEAK

The potential for urine leak exists in all patients, particularly if closure of the collecting system has been required. A flank drain is left in place for 4–5 days after operation to assess urine leakage. Persistent urine drainage can be managed by placement of a stent or percutaneous nephrostomy tube into the remaining collecting system.

ISCHEMIA OF THE REMNANT KIDNEY

This complication is most commonly due to either vasospasm or a traction injury of the renal vessels. Prevention is the most important factor, as recognition of this problem after surgery is difficult. Imaging studies to assess renal function are not routinely done in the early postoperative period. Unless the operation was performed on a solitary kidney, the loss of function of the remaining renal segment will usually go unrecognized before discharge.

OUTCOME

Most children who undergo partial nephrectomy do very well. Children who undergo surgery for duplication anomalies need careful follow-up, as approximately 20 percent require secondary procedures on the lower ureter. Most of these operations are performed to correct vesicoureteric reflux into the ipsilateral lower pole ureter. Other indications for further surgery include stasis of urine in the ureteric stump with recurrent infections and persistence of a ureterocele. Renal function is generally well maintained, but patients with solitary kidneys should be followed throughout life, as there are reports of an increased risk of hypertension and renal insufficiency.

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Laparoscopic nephrectomy and heminephrectomy

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HISTORY

Laparoscopic procedures for the treatment of benign renal conditions in children are gradually replacing open procedures. Nephrectomy, heminephrectomy, and nephroureterectomy have now become standard procedures by laparoscopy in centers where expertise is available. The transperitoneal approach was initially popular due to the familiarity of surgeons with laparoscopic gastrointestinal surgery. After the landmark publication of Gaur describing the retroperitoneoscopic approach, it has found favor with many surgeons. Regardless of the approach utilized, the benefits to the child in terms of a faster postoperative recovery and improved cosmesis are without question.

Laparoscopy has also paved the way for novel techniques for managing children with end-stage renal disease who require bilateral native nephrectomy. These children can undergo retroperitoneoscopic nephrectomy, which maintains the integrity of the peritoneum and thereby allows for immediate postoperative peritoneal dialysis. Hemodialysis can therefore be avoided.

PRINCIPLES AND JUSTIFICATION

Approach

Every pediatric urologist should be familiar with the possible approaches for laparoscopic renal surgery in children. The choice is essentially between a transperitoneal and a retroperitoneoscopic approach, with each having its own merits. Although the transperitoneal approach was believed to be better for beginners, it has largely been replaced by the retroperitoneoscopic approach. The retroperitoneoscopic approach avoids colonic mobilization, the risk of injury to hollow viscera, and the potential risk of adhesion formation. However, it is believed to be more difficult to master, due to the reversed orientation of the kidney and hilum with the patient in a prone or semi-prone position. Another possible advantage of the retroperitoneoscopic approach is reduced postoperative pain due to the absence of peritoneal irritation by blood and/or urine.

Some surgeons still prefer the transperitoneal approach for operations such as pyeloplasty, as it provides for a larger working space, which facilitates intracorporeal suturing. It may also be preferable in children who have previously undergone surgery to the affected kidney, in whom fibrosis and scarring may prevent the creation of a retroperitoneal working space. Inevitably, the choice of approach will also be influenced by the surgeon's experience and training.

In this chapter, only the retroperitoneoscopic approach will be discussed, as this is the current technique of choice for laparoscopic nephrectomy and heminephrectomy.

Indications

NEPHRECTOMY

A laparoscopic nephrectomy or nephroureterectomy is indicated in the following cases:

- Congenital non-functioning or poorly functioning dysplastic kidney.
- Pelviureteric junction obstruction with loss of function.
- Multicystic dysplastic kidney that has failed to involute or is associated with systemic hypertension.
- Reflux-associated nephropathy.
- Congenital nephrotic syndrome causing intractable protein loss.

- Pre-transplant in children with focal segmental glomerulosclerosis.

HEMINEPHRECTOMY

A laparoscopic heminephrectomy is indicated in children with complicated renal duplication anomalies. An upper pole heminephrectomy is performed most commonly, typically in the setting of hydroureteronephrosis of the upper moiety with reduced or poor function. In some girls this surgery is performed for urinary incontinence when the upper moiety ureter drains ectopically into the vagina or into the urethra below the external urinary sphincter.

A lower pole heminephrectomy is performed when there is reflux-associated nephropathy of the lower moiety or in those rare cases of lower moiety pelviureteric junction obstruction with loss of function.

PREOPERATIVE

Nephrectomy

A recent renal ultrasound scan and MAG-3/DMSA scan must be available in the operating theater on the day of surgery. In children with a history of vesicoureteric reflux, the micturating cystogram images must also be reviewed. The renal ultrasound will provide information about renal size and the degree of hydronephrosis, when present. In children with a multicystic dysplastic kidney, the size and number of cysts must be noted, as this will determine whether specimen removal will be facilitated by cyst aspiration.

In general, the child will require routine preoperative blood tests, which should include serum creatinine, hemoglobin level, and a group and save of serum. Clotting parameters do not need to be checked routinely, unless there is a history of bleeding disorders.

All children should be administered a single dose of intravenous antibiotic, either prior to leaving the ward or at the induction of anesthesia.

Heminephrectomy

All relevant imaging must be reviewed and present in theater for a laparoscopic heminephrectomy. A detailed knowledge of the degree of hydronephrosis and ureteric dilatation, presence of collecting system debris, presence of ureterocele, reflux status, and variation in function within the affected kidney is essential. If the child has undergone previous surgery, e.g., ureterocele puncture, this information must be available, as it may influence whether the affected ureter is ligated or left open.

Routine preoperative blood tests should be performed, including serum creatinine, hemoglobin level, and a blood cross-match. Clotting parameters do not need to be checked routinely, unless there is a history of bleeding disorders.

All children should be administered a single dose of intravenous antibiotic, either prior to leaving the ward or at the induction of anesthesia.

Anesthesia

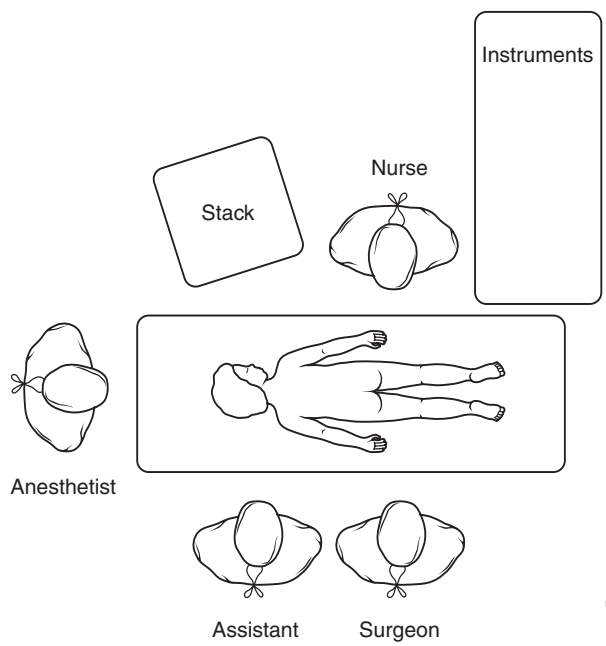
Endotracheal intubation is required in all cases using either a cuffed or reinforced endotracheal tube, securely fastened. This is to prevent tube dislodgement when the child is positioned prone for the surgery. Perioperative and postoperative analgesia is provided by pre-emptive local infiltration of the planned incisions with 0.25 percent bupivacaine.

OPERATION

Retroperitoneoscopic nephrectomy

ROOM SET-UP

1 As the patient is positioned prone for the operation, the laparoscopic stack system should be placed on the side opposite to the affected kidney, toward the head of the table, with the screen pointing toward the pelvis. The scrub nurse should be positioned adjacent to the laparoscopic stack, with both the operating surgeon and assistant on the side of the affected kidney.



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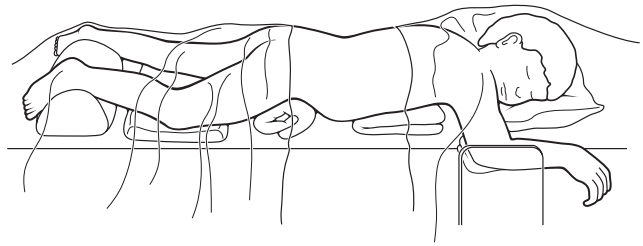
SURGICAL ACCESS

2 The patient is positioned fully prone under general anesthesia. The exposed dorsal and lateral aspects of the trunk are prepared and draped in a sterile manner. Topographic landmarks and anticipated port sites are marked as shown.



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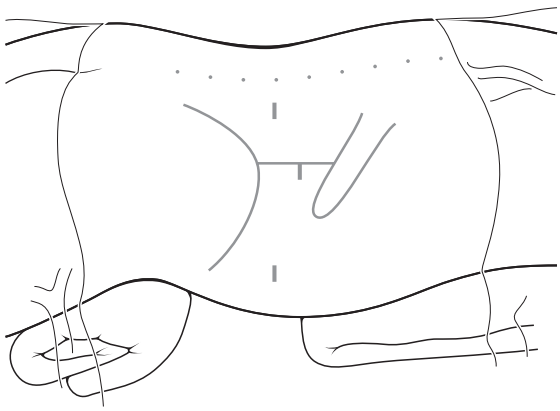

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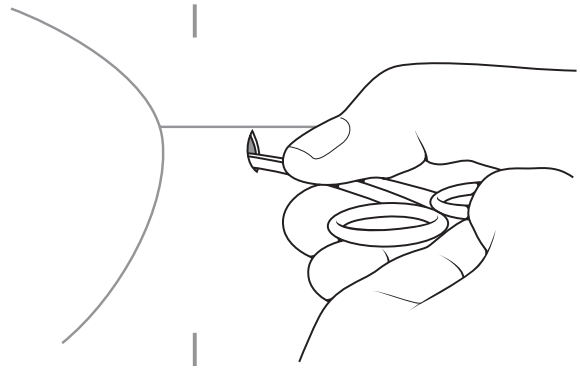
3 A 5 mm transverse incision is made midway between the iliac crest and the tip of the twelfth rib, just lateral to the outer border of the sacrospinalis muscle.

4 Through this incision, a small area of the retroperitoneum is dissected bluntly with artery forceps to allow insertion of the retroperitoneal dissecting device.



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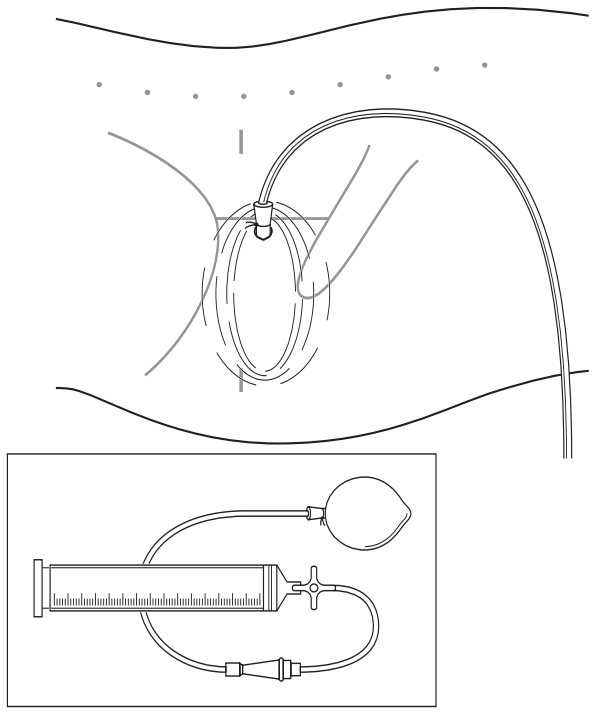

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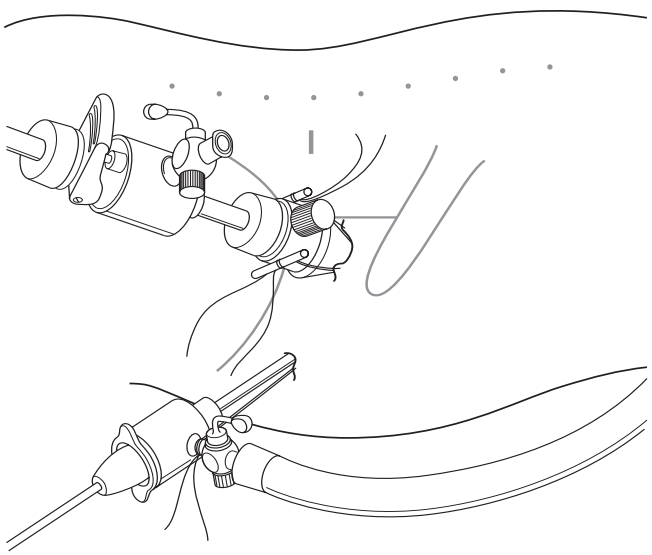
5 The retroperitoneal space is developed outside Gerota's fascia by a technique described by Gill. A dissecting balloon is made by securing the finger of a sterile surgical glove to the end of a 12 Fr Jacques catheter with a silk tie. The catheter is connected to a three-way tap and a 50 mL luer-lock syringe. Depending on the size of the patient, 100–250 mL of air is injected slowly to develop the retroperitoneal space. The balloon is left inflated for 2 minutes to promote hemostasis, and then deflated and withdrawn.

6 A 6 mm Hasson cannula is inserted into the port site, followed by insufflation of the retroperitoneum with carbon dioxide to a pressure of 10–12 mmHg. The Hasson port is secured by a suture to the skin. A 5 mm instrument port is placed under direct vision below the tip of the eleventh rib and above the iliac crest. A 30° 5 mm laparoscope is recommended for all retroperitoneoscopic surgery, as it can be rotated along its longitudinal axis to alter and maximize the viewpoint for the surgeon.



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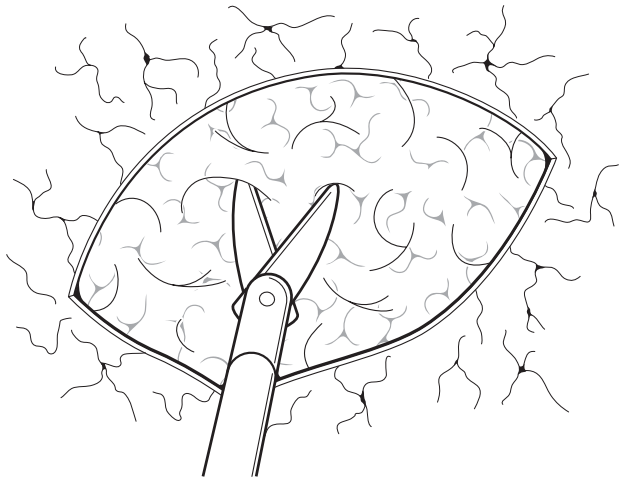
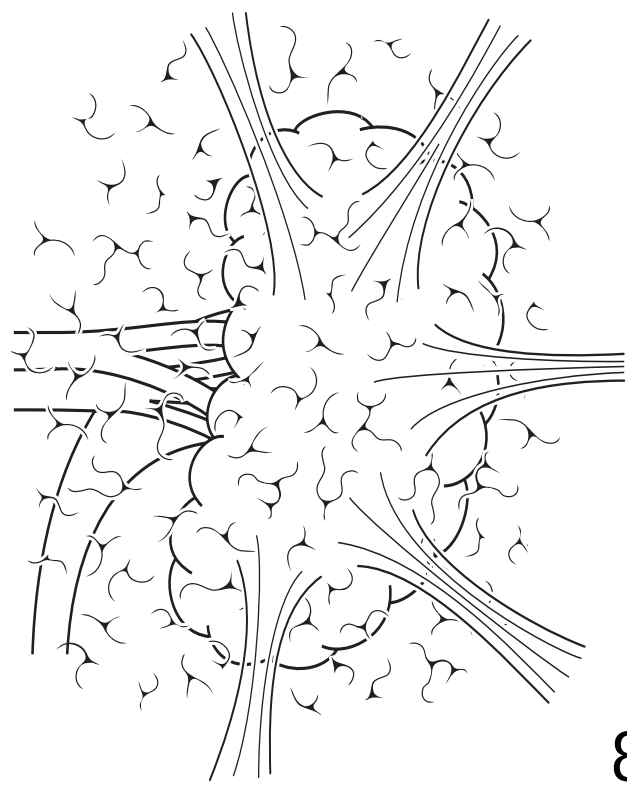


7

OPERATIVE TECHNIQUE

7 Gerota's fascia is incised longitudinally adjacent to the posterior abdominal wall using scissors. Bleeding points are meticulously controlled with monopolar diathermy. The loose areolar adventitial tissue can then be visualized through the window in Gerota's fascia. This tissue is dissected in a blunt manner to create a large perinephric working space, thereby exposing the posterolateral surface of the kidney.

8 In the author's experience, the entire operation can be performed safely through a single instrument port. However, for surgeons with limited experience, it is recommended that a second instrument should be placed. The second instrument will provide counter-traction for dissection and the application of hemoclips. It is the author's preference to use the Step Trocar system for the second instrument port, as these trocars can be inserted more easily through the paravertebral muscles.



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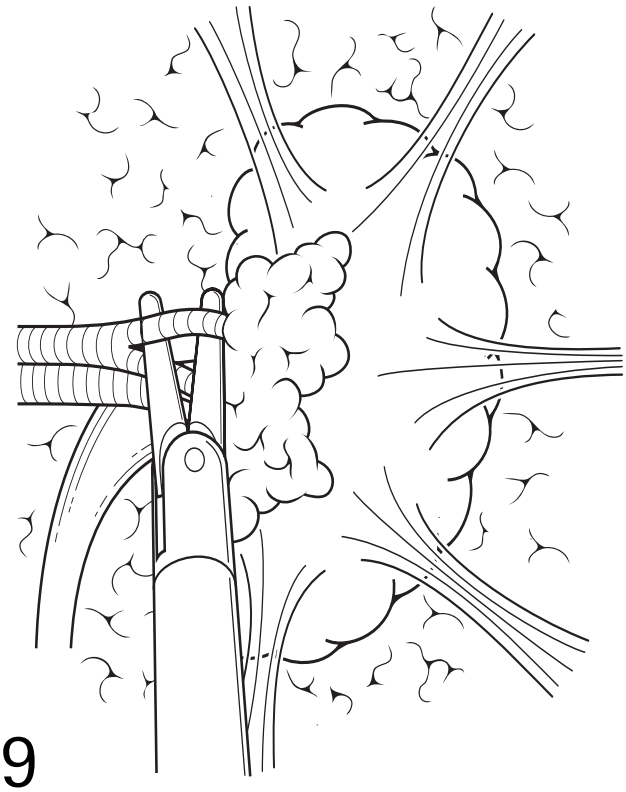

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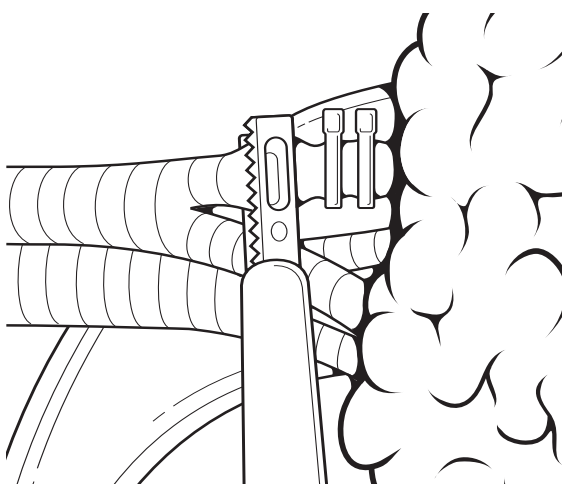
9 Dissection is started at the apex and continued along the medial aspect of the kidney, pushing it laterally and anteriorly to expose further the posteromedial surface, in particular, the renal hilum. The lateral and inferior attachments are maintained intact to facilitate exposure of the renal pedicle by gravity pull of the kidney on the vessels.

10 After adequate mobilization of the renal pedicle, the vessels are divided between hemoclips or with a Harmonic scalpel when the vessels are less than 3 mm in diameter. A minimum of three clips should be applied on all vessels, with at least two clips remaining behind on the proximal stump of the divided vessel. Care must be taken to identify and divide every possible vessel, particularly in the case of multicystic dysplastic kidneys, in which anomalous vessels are frequently found.



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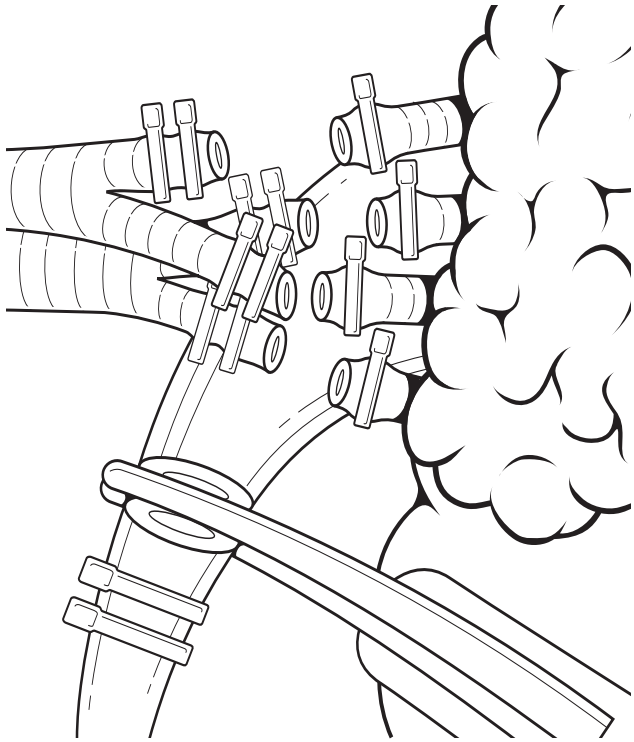

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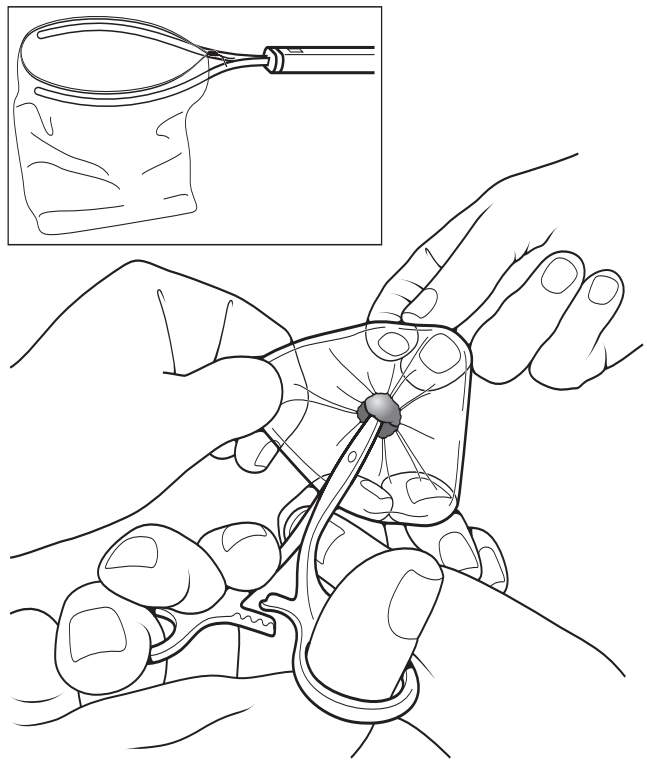
11 The ureter is dissected distally as far as required and divided/ligated. If a near-complete ureterectomy is intended, such as in cases with reflux-associated nephropathy, the retroperitoneoscopic approach will allow access into the pelvis to just below the level of the pelvic brim. In all cases of documented ipsilateral reflux, it is the author's practice to ligate the ureter with a 3/0 Vicryl endoloop suture. If this were not possible, an alternative would be to leave the ureteric stump open and to drain the bladder with an in-dwelling urethral catheter for 48 hours. In the absence of ipsilateral vesicoureteric reflux, the ureter can be safely left open without postoperative bladder drainage.

12 The remaining attachments of the kidney are divided using a combination of blunt dissection, monopolar diathermy, and/or the Harmonic scalpel. In the case of a large multicystic dysplastic kidney, complete intracorporeal mobilization can be technically difficult, time-consuming, and risks creating a tear in the closely attached peritoneum. In such cases, after all vessels have been divided and the cysts decompressed, the kidney can be withdrawn via the camera port incision and the remainder of the dissection completed in an extracorporeal manner.

12

13 The specimen can be extracted directly through the camera port incision in the case of a multicystic dysplastic kidney, grossly hydronephrotic kidney, or a small dysplastic kidney. Larger specimens are extracted with the use of a 10 mm Endopouch specimen retrieval device. The specimen is entrapped within the endobag and removed piecemeal with the use of sponge-holding forceps. The wound is closed in layers, without the use of a drain.



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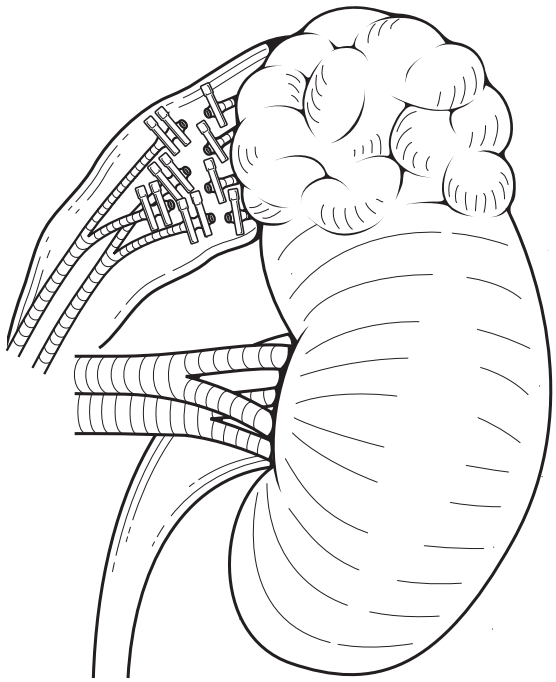
14

Retroperitoneoscopic heminephrectomy

The room set-up, patient positioning, and steps for surgical access are the same for a retroperitoneoscopic heminephrectomy as they are for a retroperitoneoscopic nephrectomy. In particular, the position of the patient and the port sites are identical. This applies whether an upper or lower pole heminephrectomy is to be performed.

14 Once the retroperitoneoscopic space has been created and Gerota's fascia has been incised, the dissection proceeds to expose the posterolateral surface of the kidney and display renal hilum as for a nephrectomy. It is mandatory to visualize clearly both upper and lower moiety ureters from the outset, as this will confirm the anatomy and provide a guide to the vascular supply to both renal moieties.



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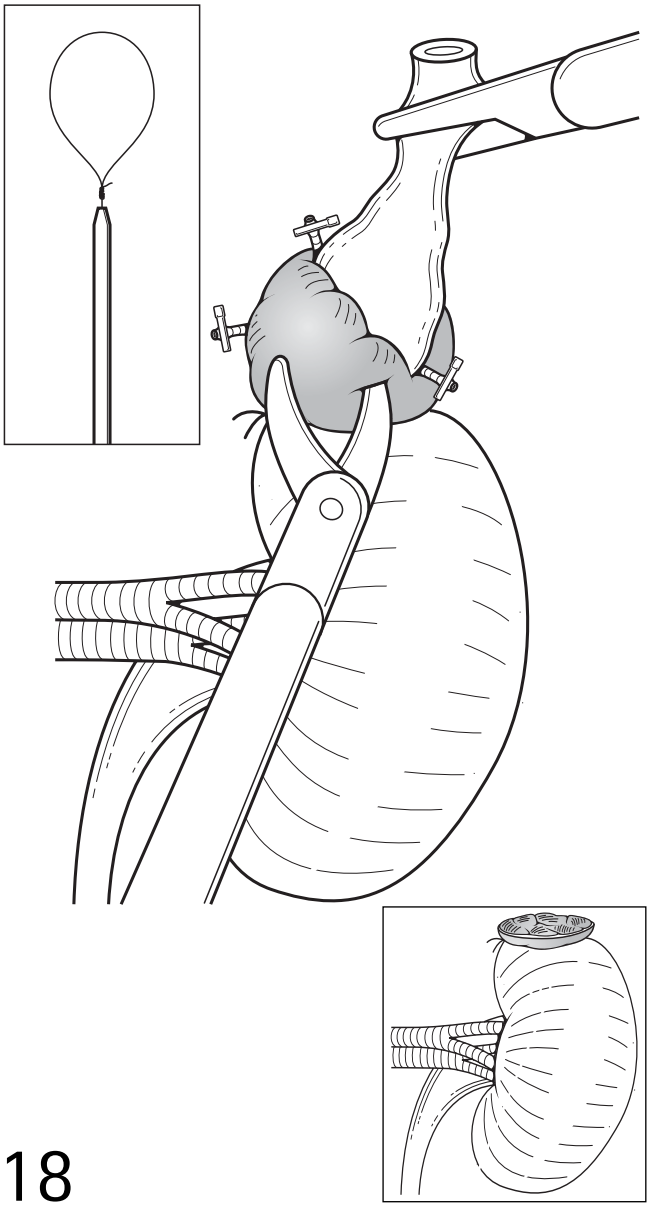
15 The vessels supplying the affected moiety are selectively identified and divided between clips or with a Harmonic scalpel. In some cases, the polar vessels will be clearly evident, whereas in other cases there will be short segmental vessels originating from the main vessels close to the renal hilum. The latter scenario is seen more frequently when the affected renal moiety is small and dysplastic.

16 The ureter from the affected moiety is separated from the non-diseased ureter and divided just distal to the pelviureteric junction. The proximal stump of this ureter is used to lift the kidney, rotating it laterally to expose the anteromedial surface. There may be additional vessels supplying the affected moiety, which only become evident with this maneuver. These should be secured and divided.

16

17 Once all the vessels to the affected moiety have been secured, there will be blanching of the parenchyma, indicating hypoperfusion. This will provide a guide to the portion of renal parenchyma to be resected. The renal capsule is scored with monopolar diathermy at the junction between the two moieties.



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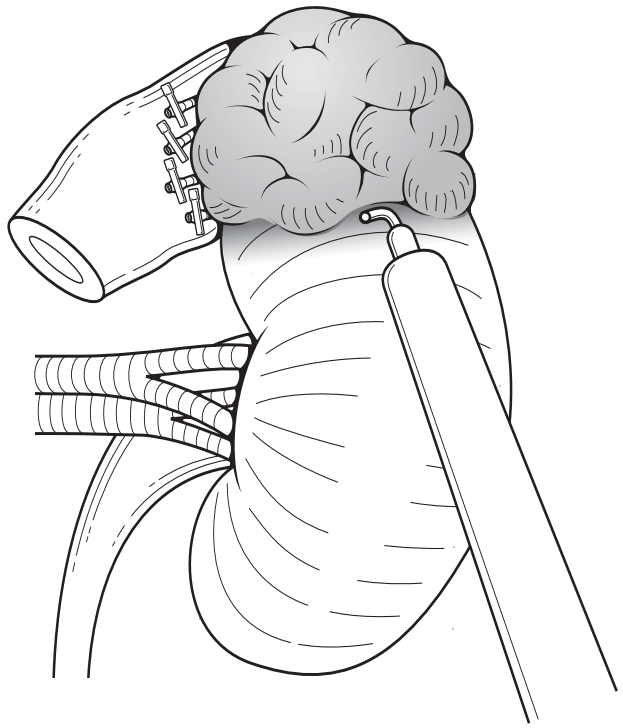

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18 A 3/0 Vicryl endoloop suture is placed over the affected moiety, using the proximal end of the divided ureter as counter-traction to facilitate this sometimes difficult step. The ligature is firmly tightened at the junction between the renal moieties, providing secure hemostasis and minimizing the risk of urine leak. The parenchyma is transected with hook scissors 5–10 mm distal to the ligature. The cut surface is carefully inspected to detect any remaining arterial bleeding points, which are rare with the endoloop technique, but would need to be secured with diathermy or a further endoloop suture.

The distal ureteric stump should be removed as far distally as possible. Care must be taken to visualize clearly the ureter from the unaffected moiety, especially in the pelvis, where both ureters lie within a common sheath. The ureter should be ligated in all cases with documented vesicoureteric reflux into the affected ureter.

The specimen can be extracted directly through the camera port incision in the majority of cases. Larger specimens are extracted with the use of a 10 mm endopouch specimen retrieval device. The wound is closed in layers, without the use of a drain.

POSTOPERATIVE CARE

In the immediate postoperative period, particular attention needs to be paid to the possibility of hemorrhage. The patient should be kept well-hydrated and the blood pressure and pulse monitored closely. Oral fluids and milk/diet can be commenced on return from theater. Children undergoing heminephrectomy are at risk of developing postoperative pyrexia, and intravenous antibiotics may need to be given if this occurs.

Most patients can be mobilized the day after surgery and allowed home provided they are stable and pain is well controlled.

Complications

PERITONEAL TEAR

The posterior prone approach minimizes the risk of a peritoneal tear as compared to other approaches for retroperitoneoscopic surgery. Peritoneal tear can occur when the dissecting balloon is inflated too rapidly, when the balloon is too small for the size of the patient, in adolescent children, and in children on peritoneal dialysis.

BALLOON RUPTURE

Rupture of the dissecting balloon can occur when the balloon is inflated too rapidly, with over-inflation of the balloon, or when excessive external pressure is applied over the balloon. When it occurs, the ruptured balloon must be carefully examined for lost fragments, which should be sought and removed from the retroperitoneal space.

INTRAOPERATIVE BLEEDING

Intraoperative bleeding is most likely to be due to slipping of hemoclips from a renal vein or to inadvertent damage to a renal vein or vena cava by a laparoscopic instrument. In most cases, hemorrhage can be controlled by the prompt application of hemoclips to the affected vessel or side wall of the vena cava. Uncontrollable hemorrhage will require conversion to an open approach to ligate or over-sew the bleeding vessel.

URINE LEAK

A retroperitoneal urinoma can occur from the reflux of urine from the distal ureteric stump or from the cut surface of the kidney following heminephrectomy. The risk can be kept to a minimum by the use of an endoloop suture on the renal parenchyma and by endoloop ligation of refluxing ureters as opposed to the use of hemoclips or the Harmonic scalpel to seal the ureter. Most urinomas will resolve with the placement of a urethral catheter for at least 48–72 hours. A persistent urine leak or an infected urinoma may require the placement of a percutaneous wound drain.

FURTHER READING

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Vesicoureteral reflux: endoscopic correction

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HISTORY

Primary vesicoureteral reflux (VUR) is the most common urological anomaly in children and has been reported in 30–50 percent of those who present with urinary tract infection (UTI). The association of VUR, UTI, and renal parenchymal damage is well known. Reflux nephropathy is recognized as a major cause of end-stage renal failure in children and young adults. Primary VUR is caused by congenital absence or deficiency of the longitudinal muscle of the submucosal ureters. This results in upward and lateral displacement of the ureteric orifice during micturition, thereby reducing the length and obliquity of the submucosal ureter. There has been no consensus regarding when medical or surgical therapy should be used. A number of prospective studies have shown low probability of spontaneous resolution of high grade of reflux during conservative follow-up. Furthermore, all of these studies revealed that observation therapy does carry an ongoing risk of renal scarring.

Since its introduction, endoscopic correction of VUR has become an established alternative to long-term antibiotic prophylaxis and open surgical treatment. We published our data regarding the long-term effectiveness of endoscopic STING (subureteral [Teflon] injection). Recently, a number of other tissue-augmenting substances have been used endoscopically for subureteral injection. At the present time, the most commonly used tissue-augmenting substance for the endoscopic treatment of VUR is dextranomer/hyaluronic acid copolymer (Deflux®). It has been reported that Deflux® is biodegradable, has no immunogenic properties, and has no potential for malignant transformation.

PRINCIPLES AND JUSTIFICATION

Indications

The indications for endoscopic correction of VUR are the same as for open antireflux operations. It is generally agreed that lesser grades of reflux (grade I or II international classification) can be managed conservatively. Grade II reflux is conservatively managed unless there are 'breakthrough infections' while on antimicrobial therapy, or poor compliance on medical management. Children with grade III to V VUR are generally considered candidates for surgery. In addition to primary VUR, the endoscopic procedure has been used successfully to treat VUR in duplex system, VUR secondary to neuropathic bladder and posterior urethral valves, VUR in failed re-implanted bladders, and VUR into ureteral stumps.

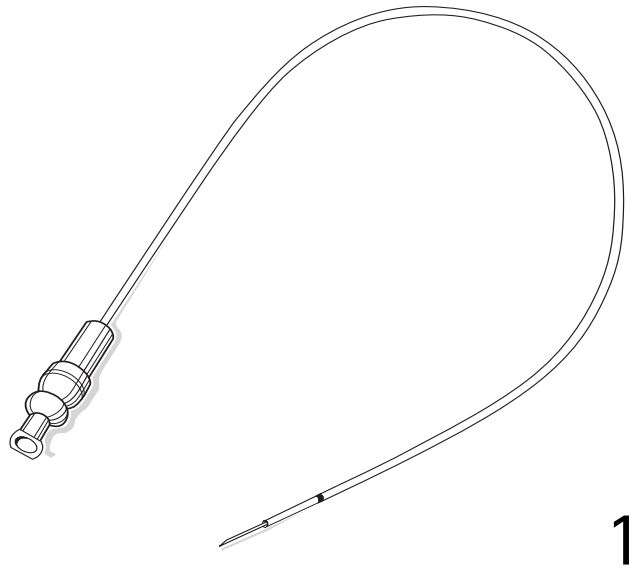
PREOPERATIVE

Material

The tissue-augmenting substance commonly used for subureteral injection is dextranomer/hyaluronic acid copolymer (Deflux®), which consists of microspheres of dextranomer in 1 percent high-molecular-weight sodium hyaluronan solution. Each milliliter of Deflux® contains 0.5 mL of sodium hyaluronan and 0.5 mL of dextranomer.

Instruments

1 The disposable Puri catheter for injection (Storz) is a 4 Fr nylon catheter onto which is swaged a 21-gauge needle with 1 cm of the needle protruding from the catheter. Alternatively, a rigid needle can be used.



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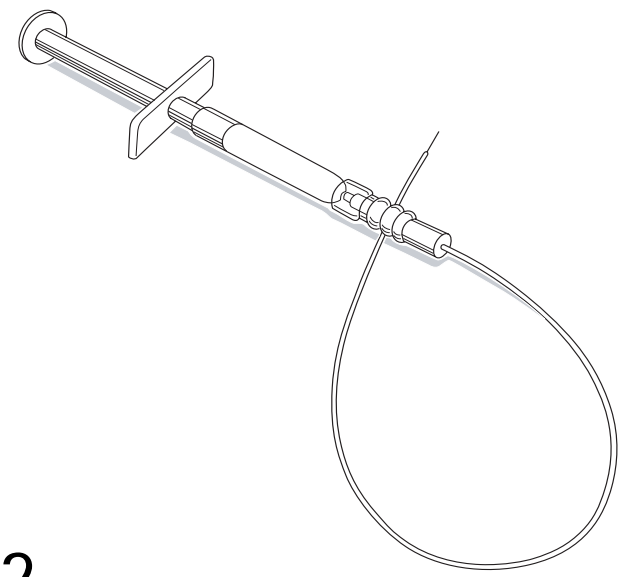

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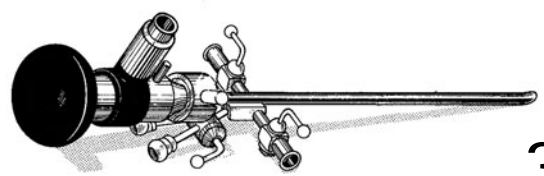
2 A 1 mL syringe pre-filled with Deflux® is attached to the injection catheter.

3 All cystoscopes available for infants and children can be used for this procedure. The injection catheter can be introduced through a 9.5 Fr, 11 Fr, or 14 Fr Storz cystoscope (Illustration a) or a 9.5 Wolf cystoscope (Illustration b). A 9.5 Fr or 11.5 Fr angled cystoscope through which a rigid needle or injection catheter can be used for injection (Illustration c).



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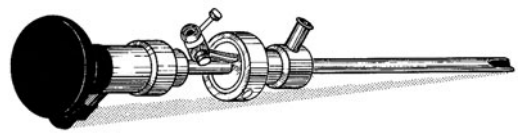

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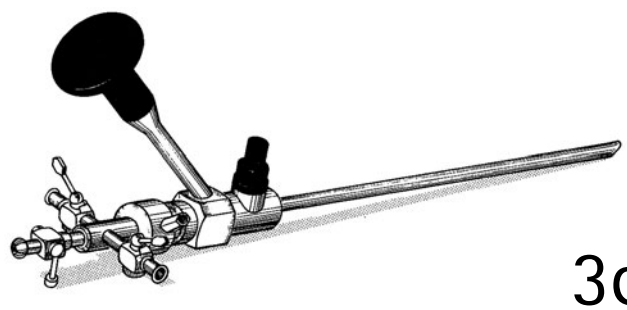

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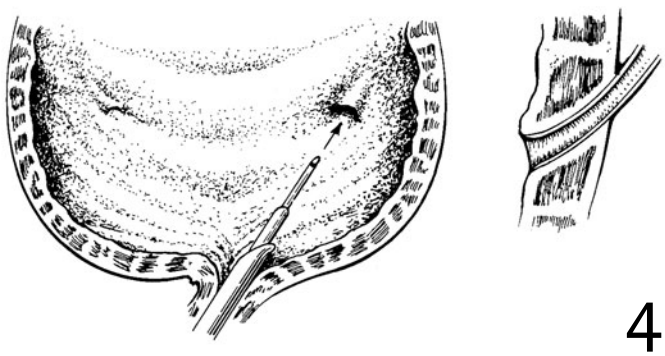
OPERATION

Subureteric injection technique

The patient should be placed in the lithotomy position. The cystoscope is passed and the bladder wall, trigone, bladder-neck, and both ureteric orifices are inspected. The bladder should be almost empty before proceeding with injection, since this helps to keep the ureteric orifice flat rather than away in a lateral part of the field.

4 The injection of Deflux® should not begin until the operator has a clear view all around the ureteric orifice. Under direct vision through the cystoscope, the needle is introduced under the bladder mucosa 2–3 mm below the affected ureteric orifice at the 6 o'clock position. In children with grade IV and V reflux with wide ureteric orifices, the needle should be inserted not below but directly into the affected ureteral orifice. It is important to introduce the needle with pinpoint accuracy. Perforation of the mucosa or the ureters may allow the paste to escape and may result in failure.



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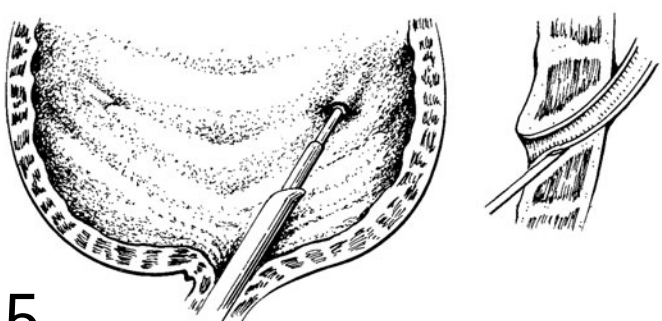

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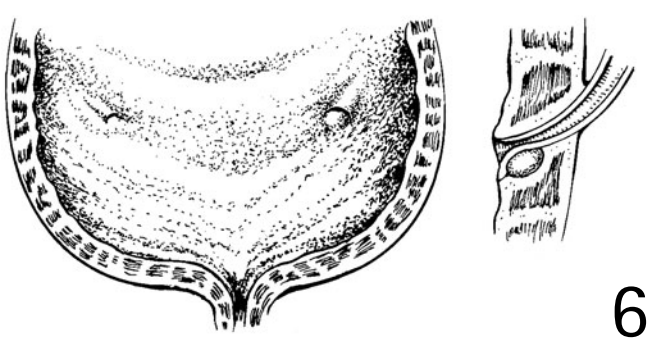
5 The needle is advanced about 4–5 mm into the lamina propria in the submucosal portion of the ureters, and the injection is started slowly. As the paste is injected, a bulge appears in the floor of the submucosal ureters. During injection, the needle is slowly withdrawn until a 'volcanic' bulge of paste is seen. The needle should be kept in position for 30–60 seconds after injection to avoid extrusion. Most refluxing ureters require 0.3–0.6 mL Deflux® to correct reflux.

6 A correctly placed injection creates the appearance of a nipple on the top of which is a slit-like or inverted crescentic orifice. If the bulge appears in an incorrect place, e.g., at the side of the ureter or proximal to it, the needle should not be withdrawn, but should be moved so that the point is in a more favorable position. The non-injected ureteric roof retains its compliance while preventing reflux.



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POSTOPERATIVE CARE

Postoperative urethral catheterization is not necessary. The majority of patients are treated as day cases. Co-trimoxazole is prescribed in prophylactic doses for 3 months after the procedure. Micturating cystography and renal ultrasonography are performed 3 months after discharge. Follow-up renal and bladder ultrasonographic scans are obtained 12 months after endoscopic correction of reflux.

Complications

Procedure-related complications are rare. The only significant complication with this procedure has been failure. This may be initial failure, i.e., the reflux is not abolished by the injection, or recurrence, where initial correction is not maintained. About 15–20 percent of refluxing ureters require more than one endoscopic injection of paste to correct the condition. Apart from failure to correct reflux, vesicoureteric junction obstruction is the only other reported complication following STING. A multicenter survey of STING procedures in 12251 ureters in 8332 patients revealed vesicoureteric junction obstruction in 41 ureters (0.33 percent) requiring reimplantation of ureters.

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79

Surgical treatment of vesicoureteric reflux

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HISTORY

Pozzi first described the concept of vesicoureteric reflux (VUR) at the end of the nineteenth century. The antireflux mechanism of the vesicoureteric junction was reported by Sampson in 1903, and in 1923 Graves and Davidoff stated that reflux does not normally exist.

Vesicoureteric reflux and its renal consequences (chronic atrophic pyelonephritis) remained poorly understood until 1960. Children with recurrent urinary tract infections tended to be treated by surgical procedures to the bladderneck and urethra. Nephrectomy for recurrent unilateral symptomatic pyelonephritis was not uncommon. In 1960, Hodson and Edwards clarified the relationship between VUR and renal damage and stressed the importance of pyelotubular backflow of urine into certain papillae. In 1973, Bailey introduced the term 'reflux nephropathy,' and later Ransley and Risdon published an essential study of papillary morphology.

The concept of early damage of renal parenchyma ('big-bang') at the first urinary tract infection in infants with VUR and the potential danger of intrarenal reflux of infected urine was proposed by Ransley and Risdon and Smellie et al. in 1975.

Surgical correction of VUR was widely practiced until it was documented that in many cases the reflux resolved spontaneously with growth of the child, and the only treatment required was prophylaxis against urinary tract infections. Antenatal detection of urinary dilatation and reflux now provides an opportunity to commence prophylaxis soon after birth.

PRINCIPLES AND JUSTIFICATION

Definition

Vesicoureteric reflux may be defined as a permanent or intermittent intrusion of bladder urine into the upper urinary tract due to a defective ureterovesical junction. The defect in the ureterovesical junction may be a primary disorder or may arise secondary to bladder dysfunction (neuropathic bladder; unstable bladder) or bladder outlet obstruction (posterior urethral valve).

The refluxing urine can fill the upper excretory system (ureters and renal pelvis) between and/or during micturition, and can sometimes penetrate into the renal substance (intrarenal reflux). The volume of refluxing urine can vary in the same patient at different times. Attempts to classify the degree of reflux are therefore of limited practical interest, as reflux may change in intensity in individual patients.

Pathophysiology and presentations of vesicoureteric reflux

The pathophysiology of VUR remains unclear, but there is a general consensus that intrarenal reflux of infected urine can cause renal damage (reflux nephropathy). It is also likely that other disorders such as immune reactions, associated renal dysplasia, change of urine biochemistry, etc. can also be responsible for renal deterioration. The difficulty is that there is no investigation able to distinguish renal lesions caused by

reflux from primary renal dysplasia, which can be associated with reflux.

There are essentially two different types of clinical presentation of VUR.

- Type 1: major VUR associated with dilated upper urinary tracts on prenatal and postnatal ultrasound scans, abnormal dimercaptosuccinate (DMSA) in more than 50 percent of cases, and a low spontaneous regression rate. This type occurs mainly in boys (90 percent) and is due to a congenital malposition of the emergence of the ureteric bud resulting in a morphological abnormality at the level of the vesicoureteric function.
- Type 2: vesicouretic reflux secondary to lower urinary tract dysfunction and abnormal bladder urodynamics. The reflux does not cause upper tract dilatation, is not detectable prenatally, is not associated with an abnormal DMSA scan, and usually occurs in girls with poor bladder and bowel function. The resolution rate is around 80 percent with bladder training. Radical treatment is seldom required.

Indications

As a consequence of the failure to reach a consensus on the precise indications for surgery, it is difficult to select the ideal treatment for each patient. The current policy is to prescribe prophylactic antibiotic treatment for prolonged periods (12–36 months) in expectation of spontaneous maturation of the ureterovesical junction (and a resolution of the VUR). This policy involves repeated investigations to check if the VUR is still present. The long-term side effects of prophylactic treatment are unknown, and there are reservations regarding its effectiveness in preventing infection. If the patient suffers from recurrent breakthrough infections or if there are signs of continued deterioration of the renal substance, more radical treatment should be offered. Surgery consists of reimplanting the ureters into the bladder; endoscopic treatment consists of injecting a substance behind the intramural ureter to create an effective posterior backing, which helps to restore the antireflux mechanisms.

The aim of these modalities of treatment is to stop the reflux. This does not necessarily mean that the progression of renal damage is halted or that further urinary tract infections are prevented. In cases where the reflux has caused severe damage to the kidney (relative function less than 10 percent), a nephroureterectomy may be the best option.

The choice between surgical and endoscopic correction of reflux is an individual matter, each technique having its own protagonists. The best result with endoscopic treatment is with low-grade refluxes (Table 79.1), where spontaneous resolution is high and where underlying bladder dysfunction is common. The place of endoscopic treatment is therefore unclear, and we reserve it mainly for reflux associated with neuropathic bladders where re-implantation is more challenging.

Table 79.1 *Reflux symptom (bladder dysfunction) and reflux disease*

	Reflux – symptom	Reflux – disease
Presentation	Recurrent urinary tract infections	Prenatal diagnosis
Ultrasound	Normal	Dilatation
Cystography	Grades I–III	Grades III–V
DMSA scan	Normal	Abnormal
Resolution	> 80%	< 50%
Etiology	Bladder dysfunction	Abnormal vesicoureteric junction + renal dysplasia
Treatment	Training	Surgery

DMSA, dimercaptosuccinate.

PREOPERATIVE

Assessment

Before planning operative correction of the VUR, complete assessment of the urinary system is mandatory: The investigation should include the following:

- Micturating cystography.
  - Direct contrast micturating cystography. This investigation is carried out during the first month of life when dilatation of the upper urinary tract has been diagnosed by antenatal scanning. It is also recommended following diagnosis of the first urinary tract infection in boys or girls at any age.
  - Indirect radioisotope cystography is preferred in older girls, as it is a less traumatic investigation.
- Direct isotopic micturating cystography. This has the advantage of a considerably reduced radiation dose, but is not available at all centers.
- DMSA or ^{99m}Tc- labeled mercaptoacetyl triglycine (MAG-3) renal scanning. This assesses the presence of renal scars and provides a measure of the relative function of each individual kidney.
- Urodynamic study. This is important for detecting an underlying bladder dysfunction.
- Ultrasonographic scanning of the urinary tract. Although a poor investigation to detect reflux, this can be useful for assessing the size, shape, and echogenicity of each kidney, the degree of dilatation of the ureter, and bladder wall thickness.
- Intravenous urography. This is rarely performed nowadays for this indication.
- Cystoscopy. Evaluation of the shape of the ureteric orifice or the length of the submucosal portion of the ureter is often subjective and will rarely alter the therapeutic decision. It is, however, recommended if the VUR is associated with a contralateral or ipsilateral ureterocele or if the reflux has occurred secondary to bladder dysfunction or bladder outlet obstruction.

Anesthesia

General endotracheal anesthesia is complemented by caudal anesthesia.

OPERATIONS

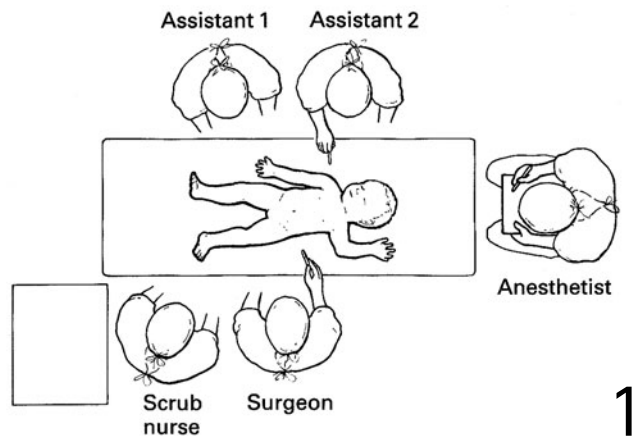
Transhiatal re-implantation of the ureter (Cohen's procedure; Glenn–Anderson's procedure) and suprahiatal re-implantation of the ureter are the two main surgical options. Their aim is to mobilize the distal segment of the ureter(s) (transmural ureter) and place it under a tunnel of bladder mucosa in order to restore the flap valve mechanism that is designed to prevent VUR.

Transhiatal re-implantation of the ureter

This is mainly represented by Cohen's procedure.

POSITION OF PATIENT

1 The patient lies supine on the operating table. A small sheet placed under the sacrum is useful to flatten the abdomen. A right-handed surgeon should stand on the left side of the patient, with the scrub nurse on his or her left. The two assistants should stand on the right side of the patient. A frame should be put between the surgeons and the anesthetist.



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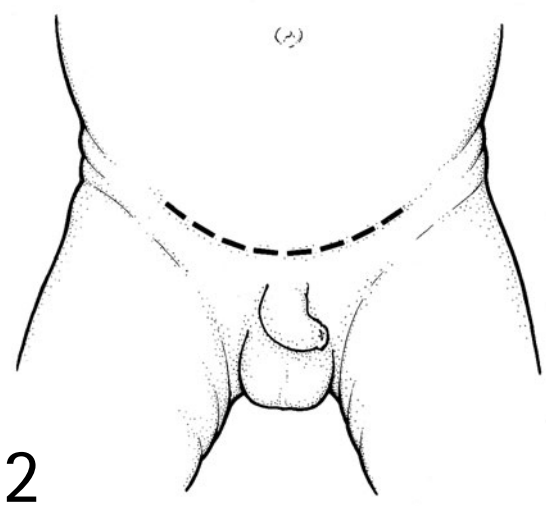
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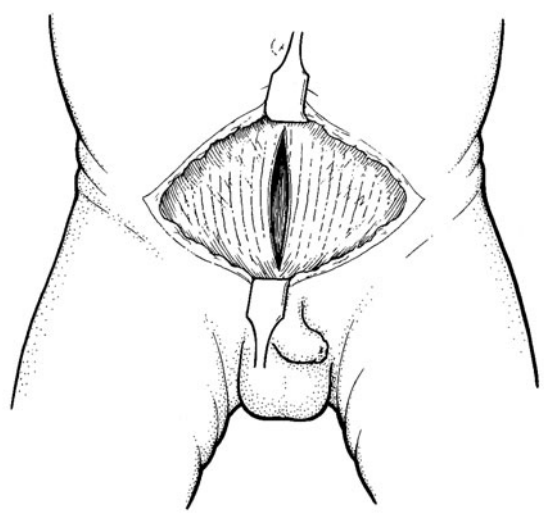
INCISION

2 A transverse suprapubic incision is made, 2 cm above the pubic symphysis, in the low abdominal crease.

3 The subcutaneous tissues are incised, exposing the rectus sheath, which is opened vertically in the midline. Both recti are separated and the peritoneum is gently pushed upwards, superiorly, providing good exposure of the bladder.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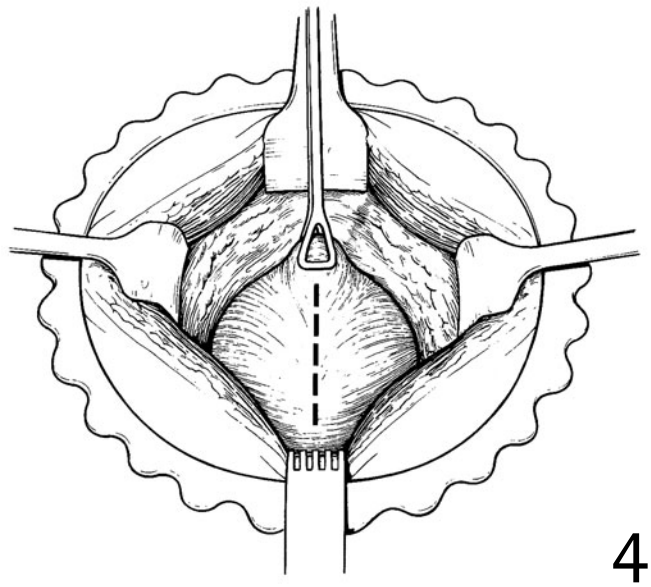
4 A Denis Browne retractor is inserted. The lateral blades retract the recti and the upper and lower blades retract the skin and subcutaneous tissues.



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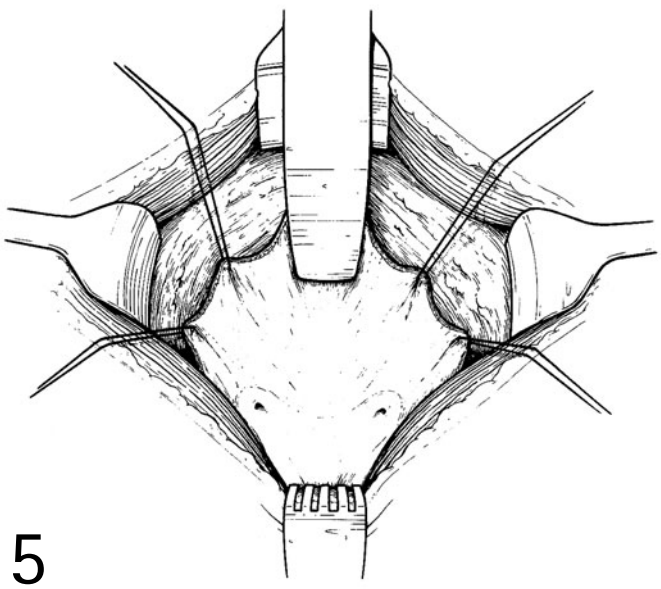

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EXPOSURE OF THE TRIGONE

5 The anterior wall of the bladder is incised vertically and two or three stay sutures suspend each edge of the vesicotomy. One or several swabs are put inside the bladder and retracted upwards with a Deaver retractor held by the second assistant, in order to expose the trigone. A 3/0 or 4/0 absorbable suture is placed at the lowest point of the vesicotomy to prevent splitting of the incision downwards into the urethra.

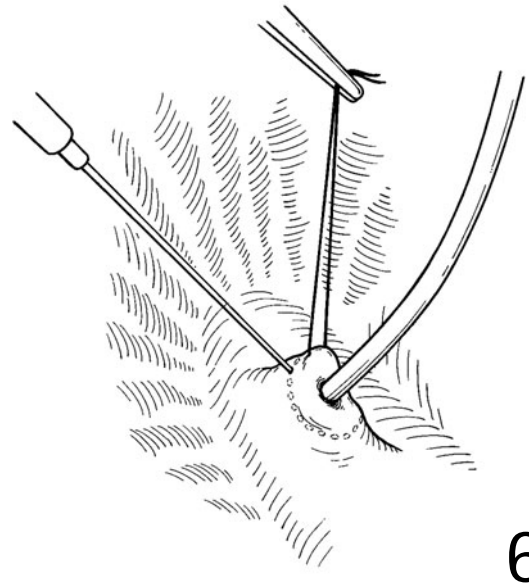
The blades of the Denis Browne retractor should not be placed within the bladder for three reasons: (1) retraction is vigorous and may damage the bladder; (2) access to the laterovesical spaces may be difficult; and (3) the bladder wall loses its natural mobility, rendering the procedure more difficult.

It is essential to avoid rigid retraction and to maintain the natural suppleness of the tissues.

The trigone is now well exposed, and an infant feeding tube (3, 4, 6, or 8 Fr) is inserted into each ureter. A stay suture (5/0) is placed around each ureteric orifice and tied over the feeding tube. The first assistant holds this stay suture with mild traction.

TRANSHIATAL DISSECTION OF THE URETER

6 The ureteric orifice is circumcised with diathermy (cutting and coagulation should be very low) and mobilization of the distal 2 cm of ureter can be performed with diathermy alone (these 2 cm will be excised later).



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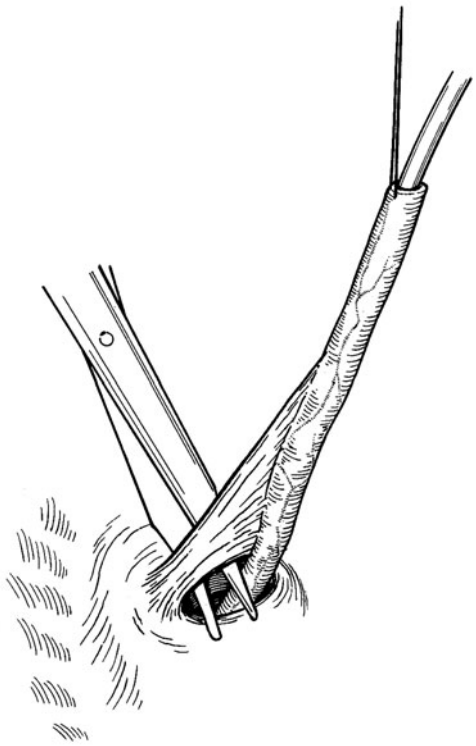

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7 It is essential to enter the correct plane between the bladder and the transparietal ureter, commencing below the orifice. Sharp scissors should be avoided, and Reynolds scissors make this procedure much easier. The tip of the Reynolds scissors elevates the muscle fibers that attach the ureter to the bladder musculature. These fibers are grasped with fine De Bakey forceps, coagulated, and divided. The dissection continues progressively, circumferentially until the ureter is completely free. Coagulation of the fibers should be carried out some distance from the ureter to avoid damaging its blood supply.

The peritoneum is visible at the end of this dissection and should be teased away from the ureter. In boys, the vas deferens may lie close to the ureter at this point and care must be taken to avoid damaging it.

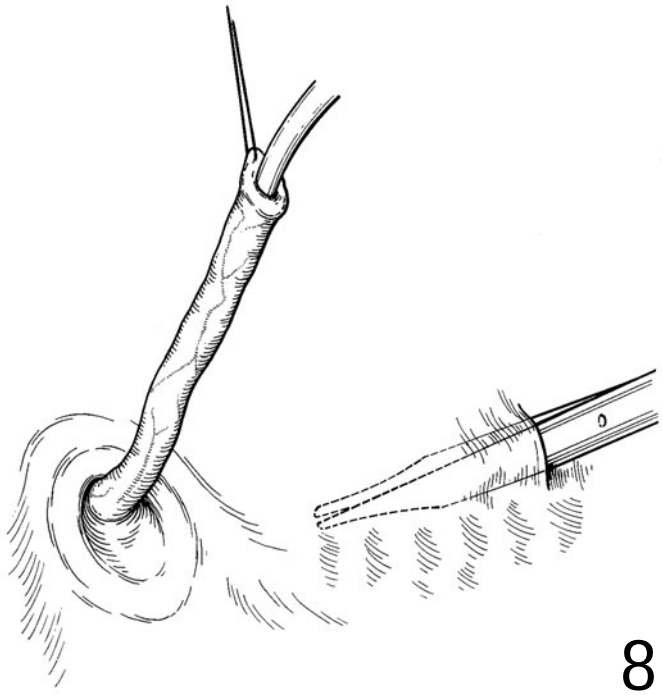
A similar procedure can be used for the opposite ureter. In cases of ureteric duplication, both ureters are dissected together and should not be separated, thus avoiding damage to their blood supply.

COHEN OR TRIGONAL RE-IMPLANTATION OF THE URETER

In some cases, the ureteric hiatus is wide and should be narrowed by one or two absorbable sutures. This is done to prevent the formation of a diverticulum. These sutures should narrow the hiatus, but still allow the free movement of the ureter and not restrict or constrict it.

8 The submucosal tunnel is then constructed. It is usually a horizontal tunnel, crossing the midline of the posterior surface of the bladder, just above the trigone. Its length should represent at least five times the ureteric diameter (Paquin's rule) and if this condition cannot be fulfilled, trimming or remodeling of the ureter should be considered (see below).

The site of the new ureteric orifice is selected and the bladder mucosa is lifted from the underlying bladder muscles with a pair of Reynolds scissors, starting either from the hiatus or from the new ureteric orifice. Again, sharp scissors should be avoided (especially Potts scissors) and Reynolds scissors are ideal. The tunnel should be wide enough to allow easy insertion of the ureter without constriction.



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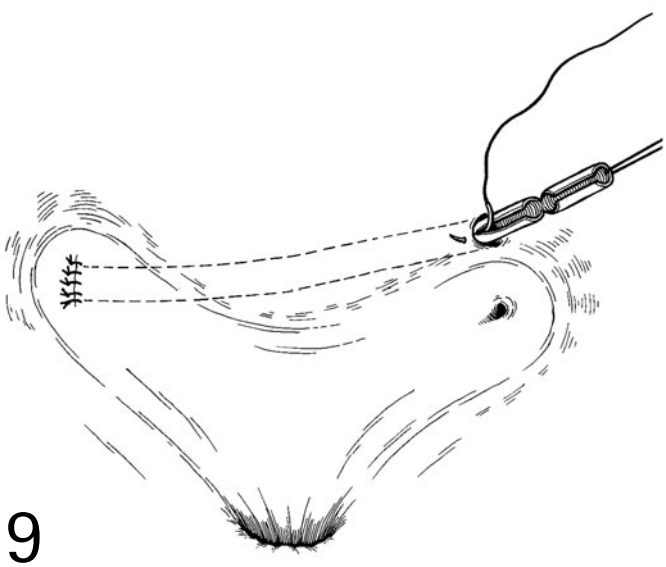

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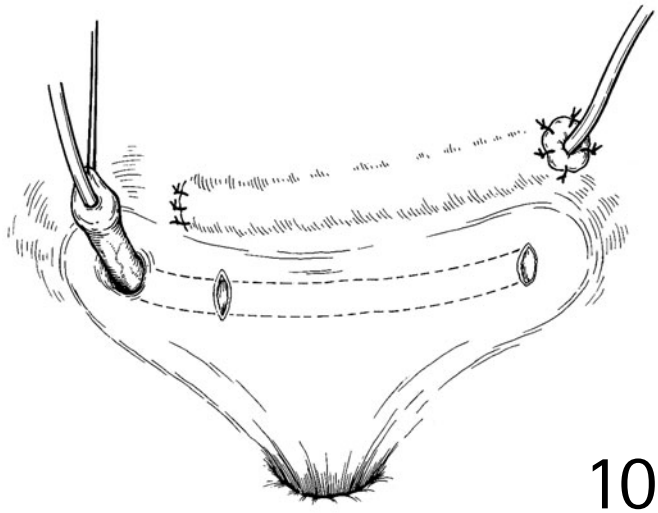
9 A similar procedure can be carried out for the opposite ureter in cases of bilateral re-implantation. The construction of the lowest tunnel that crosses the trigone can cause bleeding and the lifting of the mucosa is slightly less easy.

A pair of artery forceps or right-angled forceps is inserted through the tunnel, the stay suture is grasped and gently pulled to draw the ureter into place, taking care not to twist or kink it in the process.

10 The last 2 cm of ureter are excised and the ureteric opening is spatulated with a pair of angulated Potts scissors. The 5/0 absorbable suture anchors the ureter to the bladder muscles and the ureterovesicostomy is completed with interrupted 6/0 absorbable sutures.



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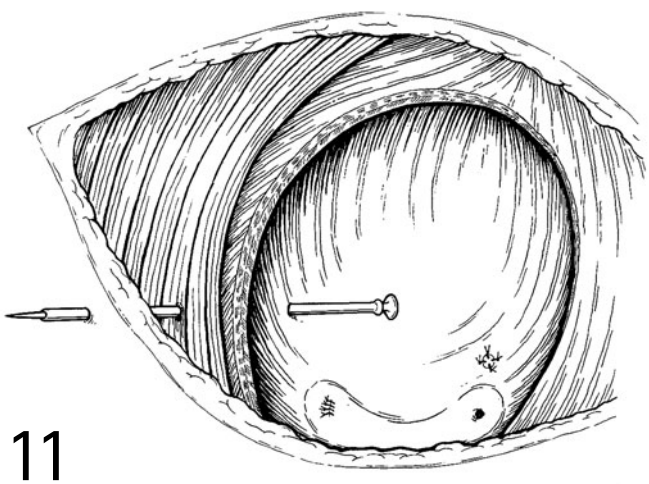


11

CLOSURE AND DRAINAGE

11 An infant feeding tube is inserted into the re-implanted ureter and exteriorized through the bladder wall, the rectus muscle, and the skin, using the punch of a suprapubic catheter. The feeding tube is left in position for 2 days, or for 10 days if the ureter has been remodeled. Alternatively, a retrievable JJ stent can be used.

There is no consensus on the efficacy of drainage of the re-implanted ureter and some authors do not leave any drain. The bladder is drained either by a transurethral catheter, which is left in situ for 5 days, or by a suprapubic catheter.

The bladder is closed with a 3/0 or 4/0 suture (interrupted or continuous). The prevesical and subcutaneous spaces are drained by a suction drain. The abdominal wall, the subcutaneous tissues, and the skin are then closed.

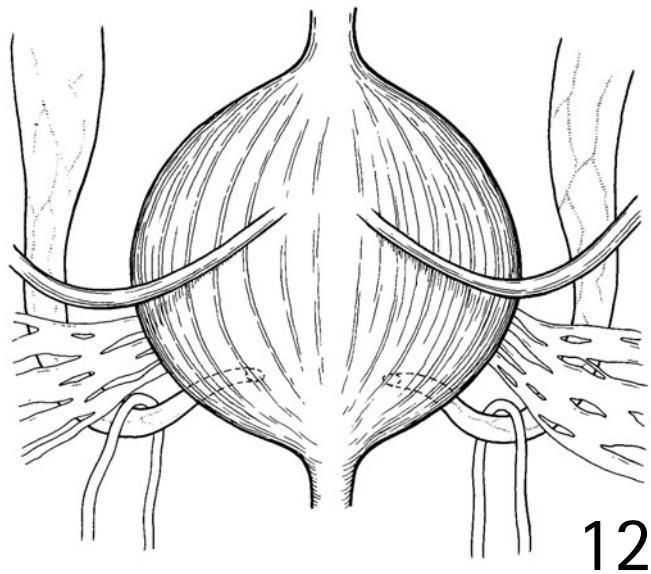
Suprahiatal re-implantation of the ureter

Megaureters are the principal indication for this technique, which should be performed by an experienced pediatric urologist. It is a difficult procedure, which carries a significant complication rate.

The approach to the bladder, the retraction with the Denis Browne retractor, and the exposure of the bladder mucosa are as in the transhiatal procedure.

The extravesical approach to the ureter is the main step in this procedure.

12 The peritoneum covering the dome and the lateral face of the bladder should be pushed upwards, which involves ligation of the obliterated hypogastric ligament. It is then easy to mobilize the peritoneum upwards and to expose the full length of the iliac vessels. The vas deferens and its pedicle are easily located and should also be freed before the ureteric re-implantation.



12

IDENTIFICATION OF THE URETER

The ureter is now identified passing over the iliac vessels close to their division into the external iliac and hypogastric arteries. The ureter is progressively mobilized from this point down to the bladder, preserving its blood supply and the blood supply of the bladder. There is, in fact, a plethora of small vessels and nerves arising from the pelvic pedicles to the bladder, which cross the distal part of the extravesical ureter and should be preserved.

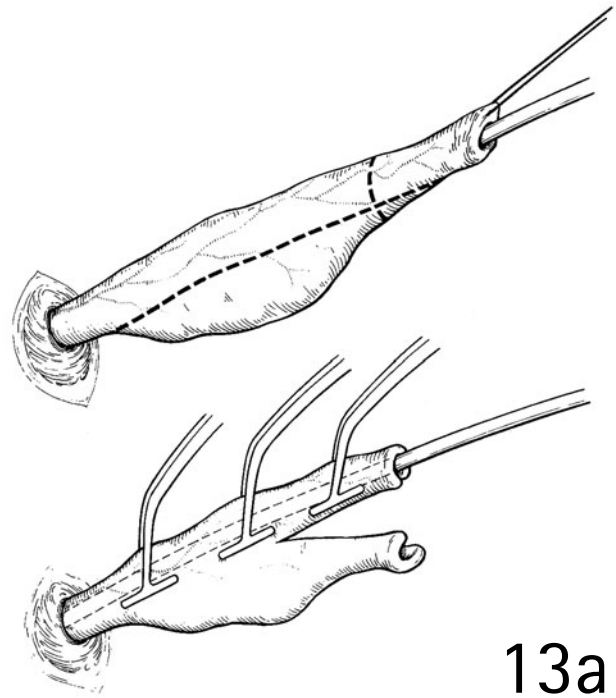
In severe megaureters the ureter is grossly dilated and kinked, and its dissection should be very meticulous to straighten it and maintain enough tissue around it to preserve its blood supply and innervation. The ureter is divided at its entrance into the bladder and a stay suture facilitates its mobilization. The ureter, which normally passes under the vas deferens, should be redirected over it to straighten it out.

EXCISION OF THE DISTAL SEGMENT OF URETER AND REMODELING

The distal segment of the ureter is usually narrowed and its excision allows urine to flow out freely. The ureteric diameter rapidly contracts and it is then possible to decide whether ureteric re-implantation can be achieved with or without remodeling or trimming. This decision is dictated by Paquin's rule: the length of the submucosal tunnel should represent at least five times the ureteric diameter.

13a,b If the ureter remains too large after excision of its distal end, its caliber should be reduced, either by excising a strip of ureter (Hendren's technique) or by infolding the ureter (Kalicinski's technique).

Excision of a strip of ureter may threaten the ureteric blood supply, whereas ureter infolding can create a certain degree of obstruction. Whichever technique is chosen, the length of the remodeled or trimmed segment of ureter should not exceed the length of the submucosal tunnel.



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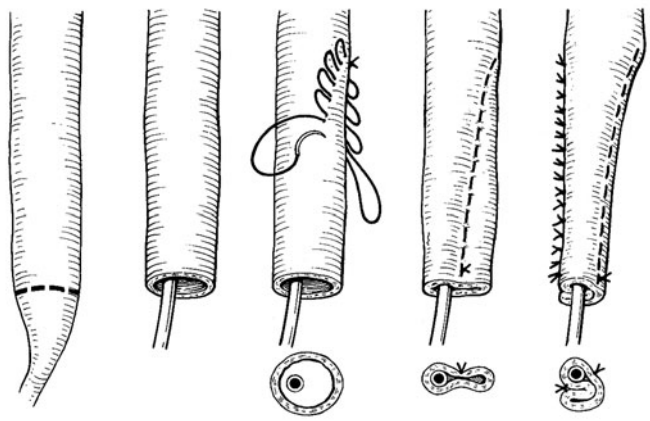
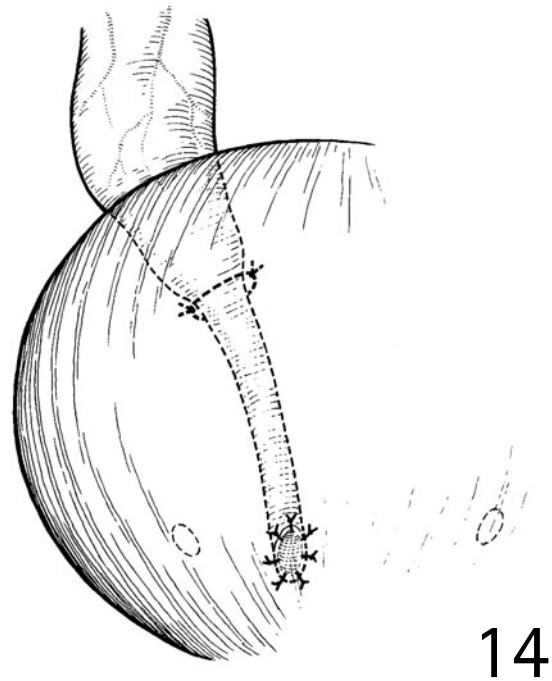


13b

RE-IMPLANTATION OF THE URETER CREATING A NEW HIATUS OF ENTRANCE

14 The hiatus of entrance of the ureter into the bladder should be medial and high at the top of the posterior surface of the bladder. The ureter should not be constricted at this level, and it is necessary to excise a disk of bladder to allow free passage of the ureter. The submucosal tunnel is fashioned as described above and should be vertical. Its distal end should open on the trigone. The passage of the freed ureter through the tunnel is the most difficult step of this procedure. The ureter should not be twisted or kinked, especially at the entrance into the bladder, and its pelvic course should be smooth. A few absorbable sutures are placed at its entrance into the bladder and sometimes the bladder itself is tacked down on the psoas muscle to maintain the smooth course of the ureter. The ureterovesicostomy is as described above.

A bilateral procedure may be performed, although bilateral extravesical approach of the ureters may affect bladder innervation and cause transient bladder dysfunction. Some authors prefer to perform a transureteroureterostomy to avoid bilateral suprahiatal re-implantation.



14

CLOSURE

Closure and drainage are as described above except that the ureteric stent is maintained for at least 10 days if the ureter has been remodeled.

POSTOPERATIVE CARE

The child is hospitalized for 5 days. The ureteric stent is removed after 2 days (or 10 days if the ureter has been remodeled). The bladder catheter is removed on the fifth postoperative day. Both suction drains are usually removed on the second day. Bladder spasms are common, and administration of oxybutinin can be useful to reduce the discomfort. Antibiotic prophylaxis with gentamicin (48 hours) and amoxicillin (10 days) is one possible option, followed by 3 months of trimethoprim-sulphamethoxazole (co-trimoxazole). Pain is controlled with diclofenac suppositories (12.5 mg).

The child should stay away from school for 2 weeks and avoid sport for 1 month.

A MAG-3 scan is performed after 6 months in order to detect indirect signs of reflux. Ideally, a repeat micturating cystogram should be performed, but this is usually poorly accepted by the patient. Repeated isotopic renal scans are useful to detect the possible progression of existing or new scars.

OUTCOME

It is essential to reiterate that these procedures only aim at stopping VUR. Their effects on the renal damage and on the recurrence of urinary tract infections are questionable.

Transhiatal procedures resolve VUR in more than 95 percent of cases. Suprahiatal procedures are more difficult and have a significant incidence of complications (around 10 percent), including persistent reflux, secondary dilatation of the upper tract, and stenosis.

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80

Ureteric duplication

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HISTORY

Ureteric duplication occurs with duplication of the kidney. Post-mortem studies show upper tract duplication in 0.8 percent of the population; 40 percent are bilateral. It can be inherited as an autosomal dominant trait, but with variable penetrance; affected families have an 8 percent incidence.

An understanding of the embryology is crucial to an appreciation of the anatomy of these conditions. At the beginning of the fifth week of gestation, epithelial out-pouchings from the mesonephric duct grow laterally (normally one ureteric bud on each side) into the metanephric blastema. A process of reciprocal induction between the metanephros and the ureteric bud causes nephrogenesis to occur. The metanephros gives rise to the glomerulus, the convoluted tubules and loop of Henle, whereas the ureteric bud undergoes several generations of bifurcation, and is the origin of the collecting ducts, calyces, and renal pelvis. Partial duplication occurs when a ureteric bud has started to bifurcate early. Complete duplication occurs when an accessory ureteric bud arises from the mesonephric duct. Ureteric buds arising proximally or distally to the normal site of origin on the mesonephric duct induce nephrogenesis in a less competent part of the metanephros, producing a dysplastic pole of the kidney. The part of the mesonephric duct distal to the ureteric bud(s) (also called the excretory duct, common mesonephric duct) becomes incorporated into the posterior wall of the developing bladder (which develops from the cranial part of the urogenital sinus) in a cranial to caudal direction – forming the trigone. Stephen's hypothesis describes how distal ureteric buds drain the lower poles of duplex kidneys and become incorporated into the bladder earlier, and more laterally, with a shorter intramural tunnel, lack of a flap valve, and consequent vesicoureteric reflux (VUR), whereas proximal ureteric buds drain the upper poles and become incorporated into the trigone later and more distally (which explains the Meyer–Weigert rule (see below). In extremely proximal cases, the part of the mesonephric duct to which the upper pole ureter is attached never becomes incorporated into the trigone or urethra, giving rise to a connection to a mesonephric duct derivative. (In the male, these derivatives include the vas, seminal vesicles, epididymis, and prostate, which are all suprasphincteric; in the female, they include epoophoron, oophoron , or Gartner's duct – urine drainage into these could lead to rupture into an adjacent structure, which may be above the sphincter mechanism (suprasphincteric) or below it (infrasphincteric to the introitus or vagina.)

The terminology describing the spectrum of abnormalities is reviewed here.

- **Duplex kidney** has two, separate pelvicalyceal systems, and consequently an upper and a lower pole. Duplication may be partial or complete. In incomplete duplication, the ureters may join at any point: a *bifid system* joins at the pelviureteric junction (PUJ); *bifid ureters* join at some point proximal to the bladder. Complete duplication occurs with *double ureters*, which drain their respective poles and empty separately into the genitourinary tract.
- **Ectopic ureter** drains into the urethra below the bladder-neck, or outside the urinary tract.
- **Weigert–Meyer rule** states that the lower pole ureter enters the bladder lateral and cephalad to the upper pole ureter. The lower pole ureter therefore has a deficient antireflux mechanism compared to the upper pole ureter, so the lower pole ureter refluxes, whereas the upper pole does not.
- **Ureterocele** is a cystic dilatation of the terminal part of the ureter (upper pole ureter if associated with a duplex system). A ureterocele may be *intravesical* (also called orthotopic, or simple), entirely contained within the bladder, or

ectopic, if any part of it passes through the bladderneck into the urethra. The orifice can be described as stenotic, sphincteric (orifice distal to the bladderneck), sphincterostenotic (tight and distal to the bladderneck), or cecoureterocele (intravesical orifice, but with an extension submucosally into the urethra, most severe type, and occurs in girls).

PRINCIPLES AND JUSTIFICATION

The vast majority of duplications are partial, and not clinically significant. Less then 0.1 percent of the population have complete duplications (approximately 10 percent of all duplications). These present during childhood, and more than 50 percent are now detected antenatally because of dilatation related to reflux, obstruction, or dysmorphism.

Duplex system ureteroceles

Duplex system ureteroceles are related to the upper pole ureters, and are present in 0.02 percent of the population (80 percent female). These may have other associations and consequences: obstruction of the upper pole ureter itself; VUR to the ipsilateral lower pole (50 percent); bladder outflow obstruction by ectopic ureterocele; compromise of contralateral kidney by bladder outflow obstruction; incidental contralateral reflux (25 percent); and, urethral prolapse of ureterocele in girls. Sixty percent are antenatally detected, otherwise they present in infancy with urinary tract infection (UTI) or urosepsis. Occasionally, they present with urinary retention. Baby girls may present with prolapse of the ureterocele. Surgical intervention for duplex ureteroceles should be considered: (1) if symptomatic; (2) for bladder outflow obstruction; and (3) for ureterocele prolapse.

Ectopic ureters

Ectopic ureters are detected antenatally if the upper pole ureter or upper pole of the kidney is dilated. Postnatally, supraphincteric ectopic ureters present with UTI (epididymo-orchitis in boys). Infraphincteric ectopic ureters occur in girls and present with a history of continuous dribbling of urine, but with a normal pattern of micturition.

PREOPERATIVE ASSESSMENT AND PREPARATION

Every case of duplicated ureter should be assessed and a specific management plan should be made that is tailored to the individual. The overall aims of treatment are preservation of renal function, urinary continence, reduction of infection risk by removal of obstruction, and treatment of VUR. All patients with antenatally diagnosed ureteroceles should receive prophylactic antibiotics postnatally. The following investigations may be useful:

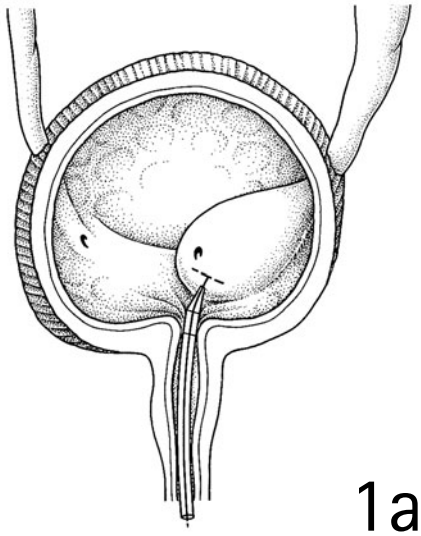
- Ultrasound will detect lower pole dilatation (may be VUR or PUJ obstruction) and upper pole dilatation (ureterocele or an ectopic ureter). Occasionally, girls may have a 'cryptic duplication' that is small and dysplastic, associated with an undilated ectopic ureter.
- Dimercaptosuccinnic acid (DMSA) scintigraphy will allow assessment of function of the renal units, and may reveal a 'cryptic' duplex kidney.
- Micturating cystourethrography (MCUG) will allow detection of lower pole VUR. Reflux, if present in both moieties, suggests partial duplication.
- Intravenous urography (IVU) is rarely required, but may be useful in identifying a poorly functioning, undilated 'cryptic' upper pole. The signs include missing calyces, lateral and downward displacement of the lower moiety ('drooping lily' sign), and a scalloped, tortuous lower moiety ureter, laterally displaced lower moiety ureter.
- Cystoscopy is useful to allow determination of the site of the orifice of a ureterocele, and the number and sites of any other ureteric orifices. Endoscopic puncture can be performed at the same time.

OPERATIONS FOR DUPLEX URETEROCELE

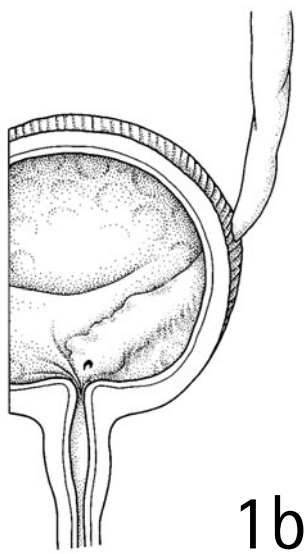
Endoscopic puncture of ureterocele

1a,b The ureterocele should be punctured at its base, proximal to the bladderneck, at the most medial end to allow for a long tunnel. Puncture can be done using a Bugbee electrode, resecting cold knife, or a resecting diathermy electrode. Large incisions of ureteroceles run the risk of allowing reflux to occur.

Following endoscopic puncture in intravesical ureteroceles, more than 90 percent will decompress and more than 95 percent will have maintained upper pole function; the risk of creating reflux occurs in 18 percent and 7 percent will require further surgery. For ectopic ureteroceles, the outcome is not as good: 75 percent result in upper tract decompression, upper pole function is preserved in less than 50 percent, reflux occurs in nearly 50 percent, and second procedures are needed in 50 percent.



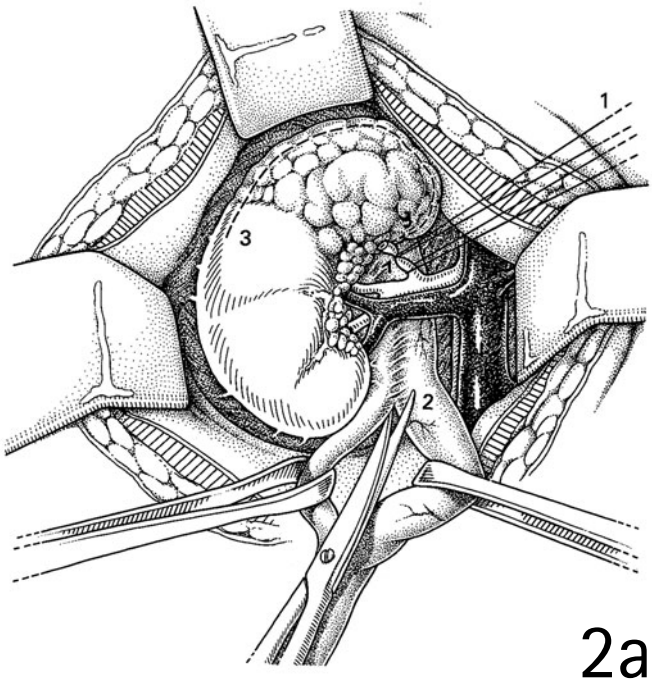
1a



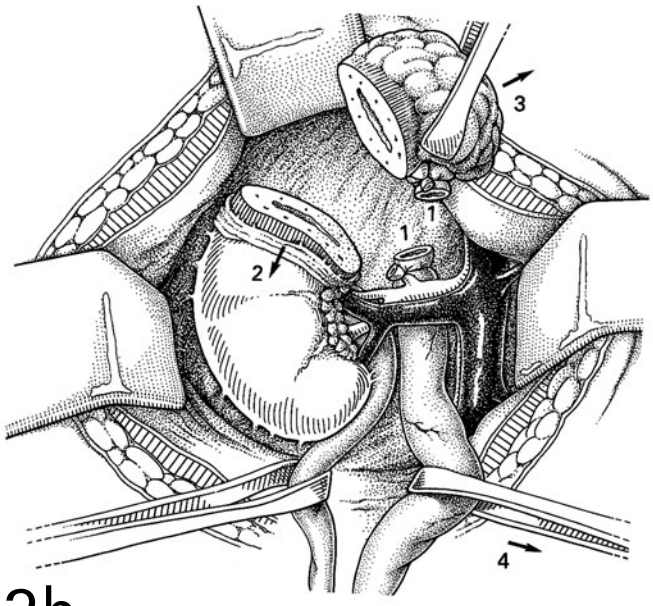
1b

Upper pole heminephrectomy

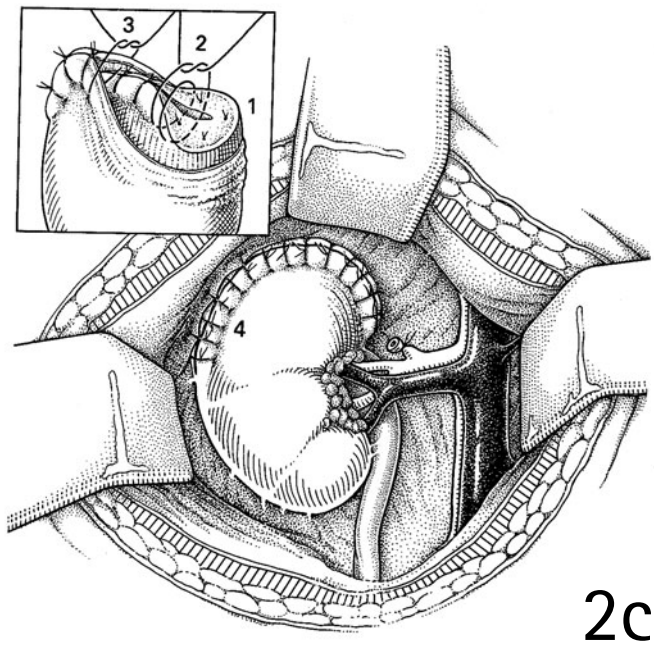
2a–c The indication for this procedure is a poorly functioning upper pole. The procedure can be performed by an open approach, through an anterior muscle-splitting incision or flank incision, or by a laparoscopic approach (either transperitoneal or retroperitoneal). The procedure involves transection of the upper pole ureter, allowing traction. Ligation of the upper pole vessels allows demarcation to occur, enabling preservation of the normal renal parenchyma. The upper pole capsule can be stripped off in continuity and used to cover the cut edge at the end of the procedure. Ureterectomy is performed with care not to damage the blood supply to the other ureter. The ureterocele should be aspirated through the upper pole ureter at the end of the procedure. Eight percent of patients will require further surgery if only a high approach is used.The risk of the need for further intervention is increased by ectopic ureterocele (re-operation risk 65 percent) and high-grade reflux into another moiety (50–70 percent re-operation rate).



2a



2b



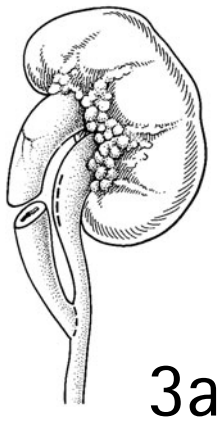
2c

Upper pole heminephrectomy, ureterectomy, and excision of ureterocele

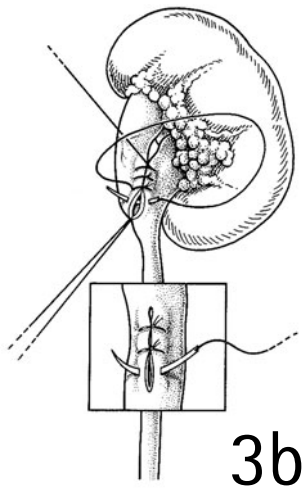
This is indicated for prolapsing ureterocele, ectopic ureterocele, high-grade reflux (≥ grade 3) to lower pole. This approach requires two incisions, the upper one as described and the lower one, a Pfannenstiel. The first part of the procedure is described above, the second below. There is a ≤ 15 percent risk of requiring re-operation, but also a risk of damage to the sphincter mechanism when an ectopic ureterocele is enucleated.

Ureteropyeloplasty

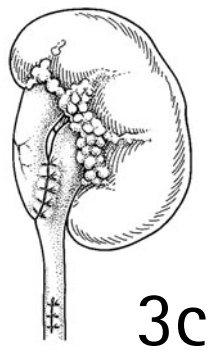
3a–c This allows preservation of a functioning upper pole in association with obstructed duplex ureterocele (rare). An open approach, through an anterior muscle-splitting incision or flank incision, or a laparoscopic approach, either transperitoneal or retroperitoneal, is made. Ureteropyeloplasty is performed. The ureterocele must be aspirated through the upper pole ureter, following which a ureteric stent is left in place for 5 days. There is a re-operation rate for continuing reflux into lower pole ureter, or into the upper pole stump. The risk of further surgery is increased, as described above.



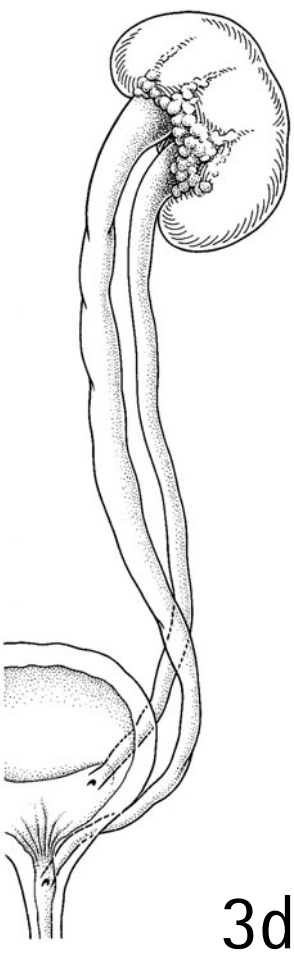
3a



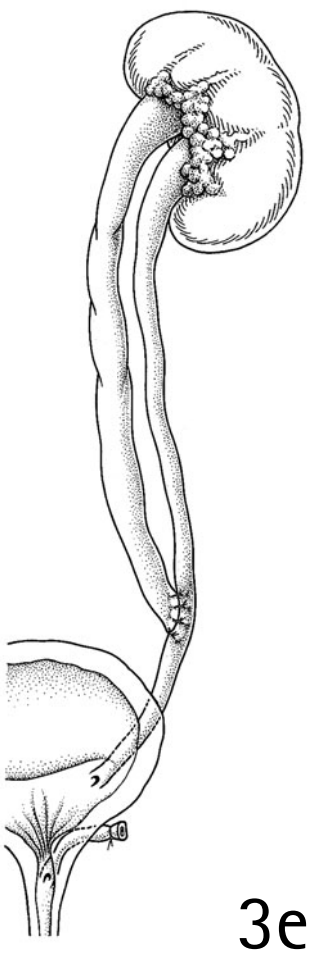
3b



3c



3d

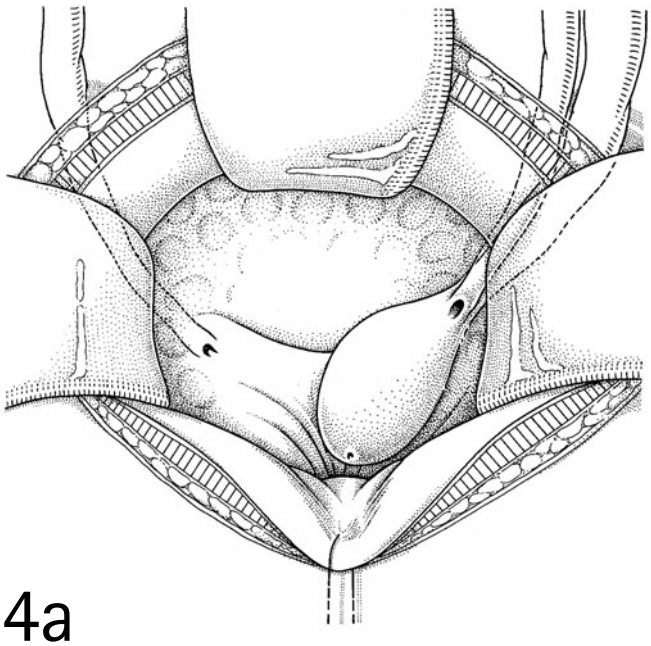


3e

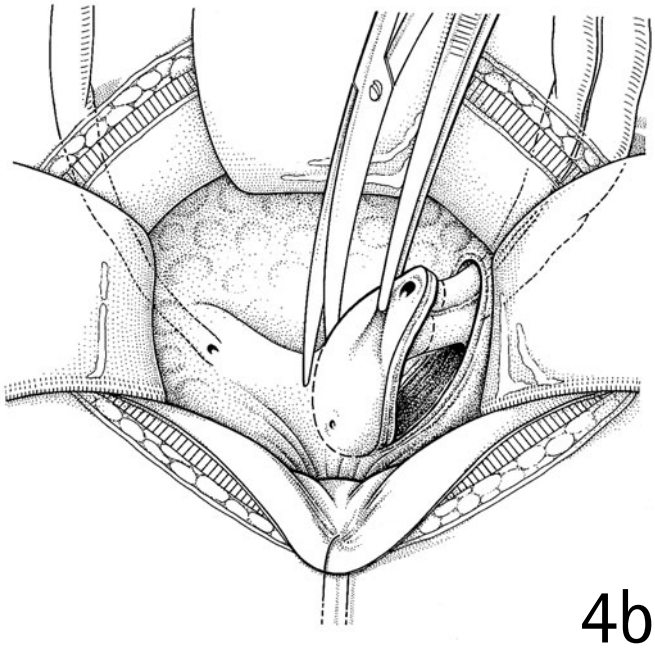
3d,e Increasingly, this operation is being replaced by a ureteroureteroplasty using an extraperitoneal groin incision.

REMOVAL OF INTRAVESICAL URETEROCELE AND RE-IMPLANTATION OF THE URETER

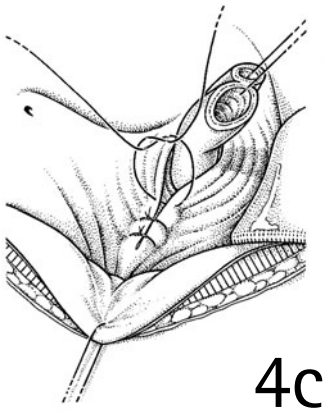
4a–d This is indicated by continuing reflux, infection, or obstruction of the upper pole ureter. A Pfannenstiel approach is used. The ureterocele is excised carefully, because the common sheath of double ureters means that the blood supply is common to both.



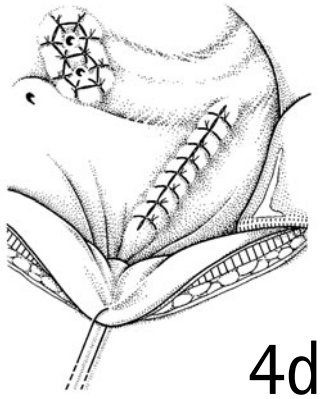
4a



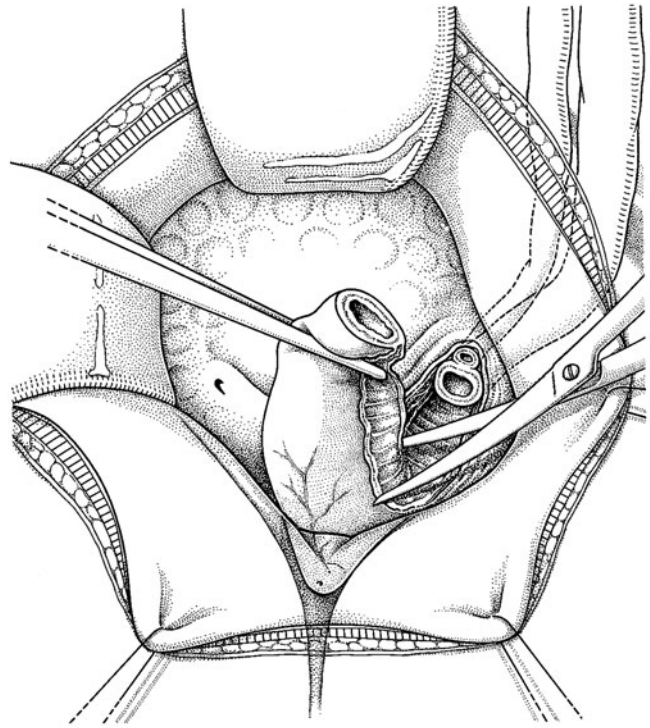
4b



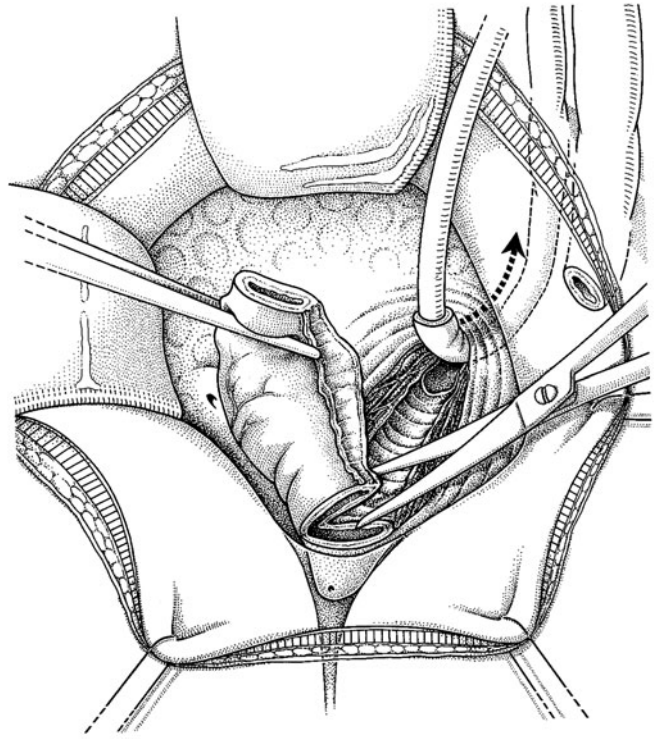
4c



4d



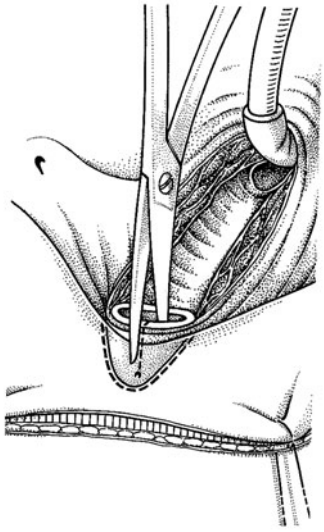
5a



5b

REMOVAL OF EXTRAVESICAL URETEROCELE AND RE-IMPLANTATION

5a–f The approach and procedure are as described above, but plication of the posterior bladder wall may be needed if the ureterocele is prolapsed and the posterior wall is deficient. Care must be taken with extravesical extension, as this can lead to damage of the sphincter mechanism.



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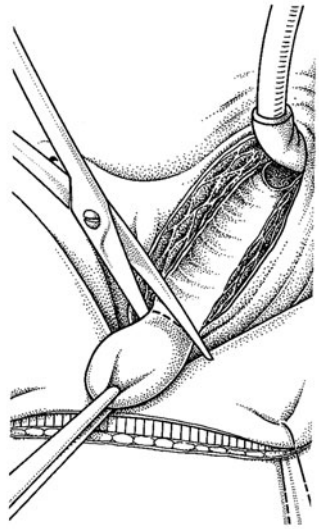
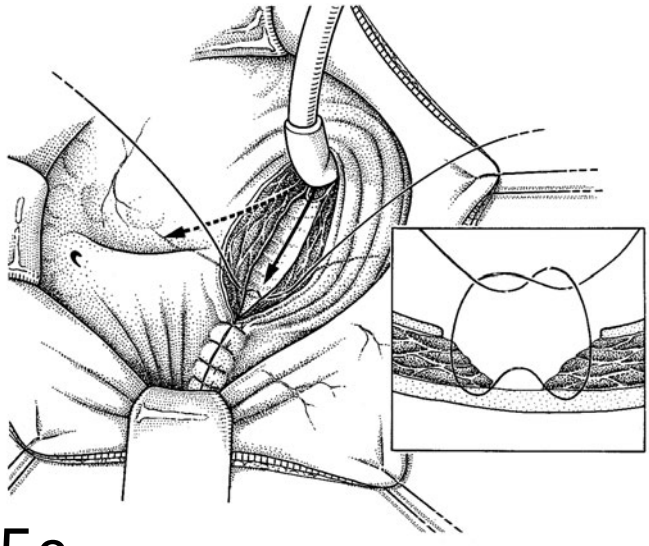


5d



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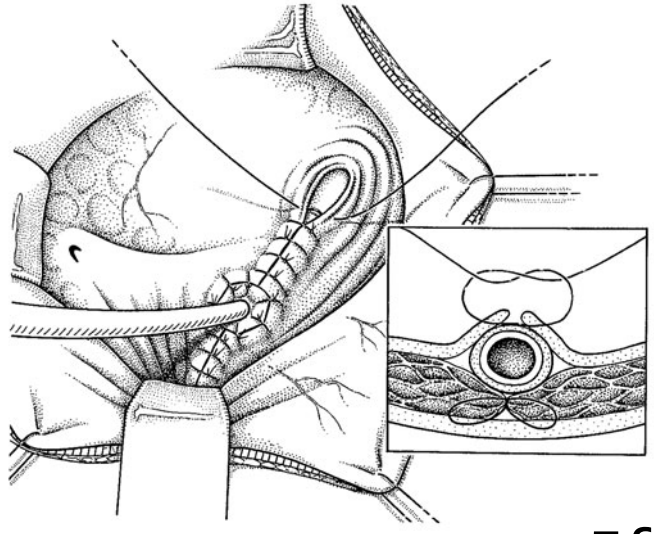


5f

Nephrectomy

This is described in Chapters 76 and 77.

OPERATIONS FOR ECTOPIC URETER

Suprasphincteric ectopic ureter

The indications depend on the symptoms from recurrent UTIs.

- Upper pole heminephrectomy (see Illustration 2). In most cases, this is indicated by lack of function.
- Ureteric re-implantation (see Illustration 6). This is only possible where there is a well-functioning upper pole. An en-bloc re-implantation is performed because both upper and lower pole ureters share a common sheath and blood supply.

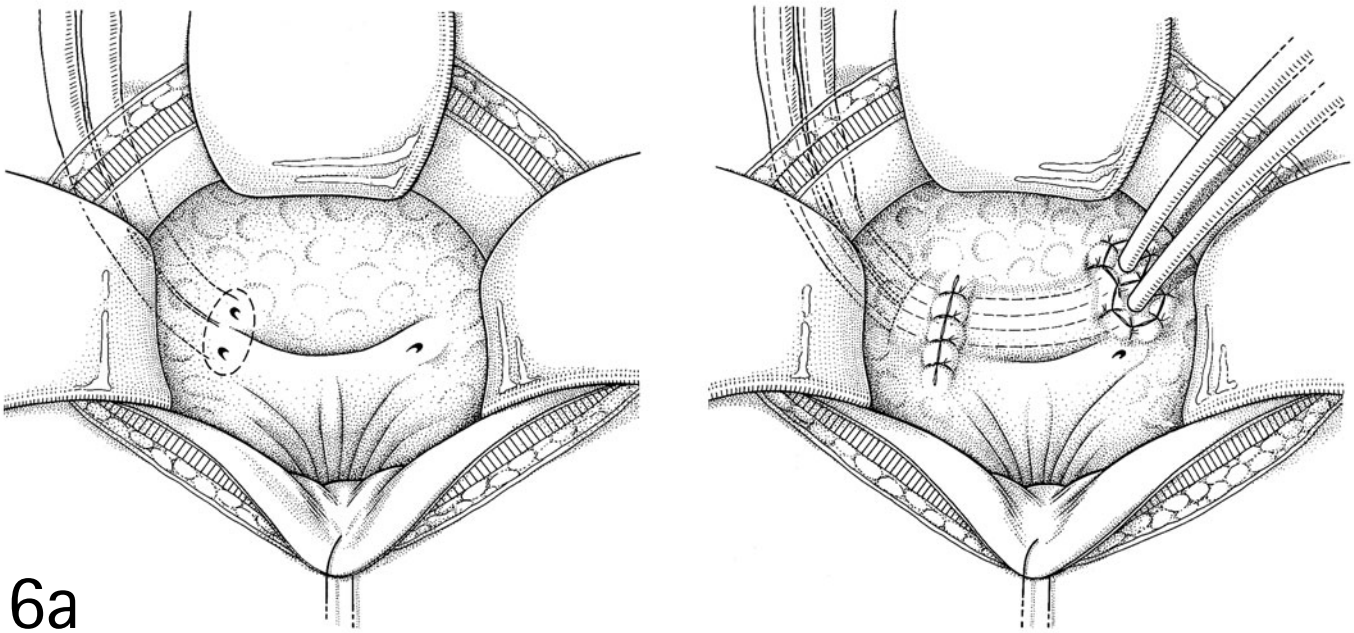
Infrasphincteric ectopic ureter

The main problem is diagnostic: identifying 'cryptic' upper pole, 10 percent of cases of which will have a coexistent and detectable contralateral duplex kidney. For poorly functioning moieties (the majority), nephrectomy of the upper pole will be curative (see Illustration 2). Good function is very rare; if re-implantation is needed, both ureters on the affected side will need to be re-implanted because of the presence of the common ureteric sheath distally (see Illustration 6 below).

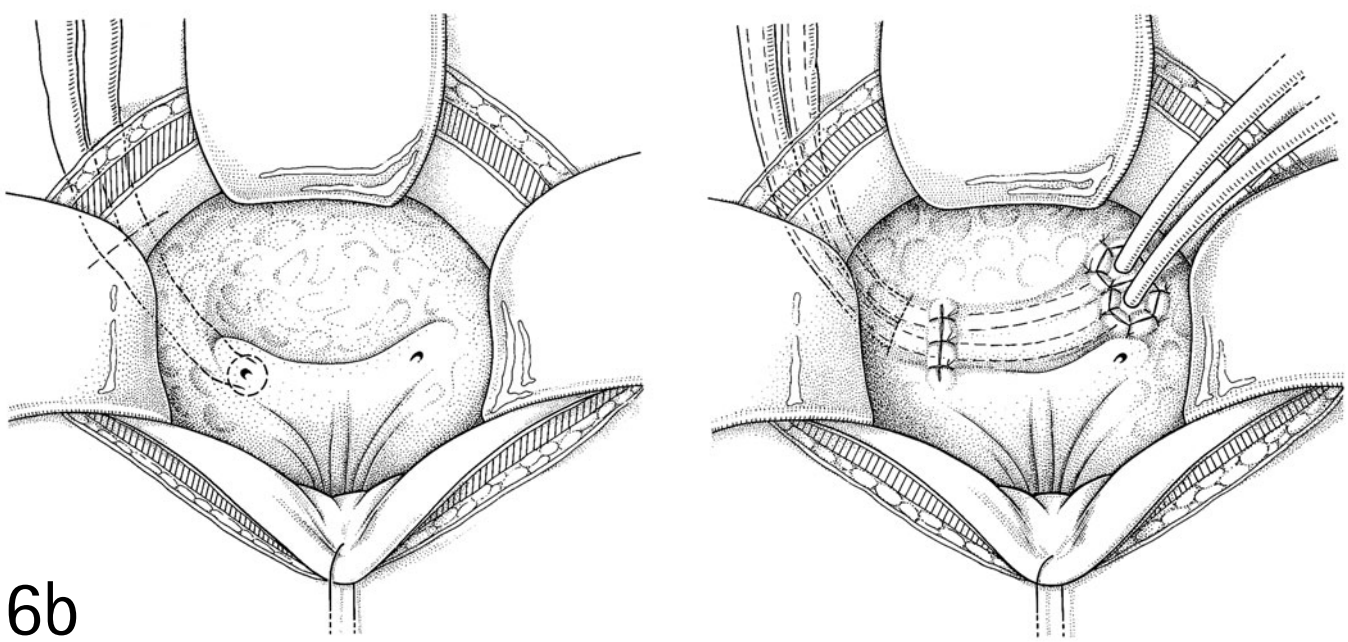
OPERATIONS FOR VESICOURETERIC REFLUX ASSOCIATED WITH DUPLEX

Ureteric re-implantation

6a,b This is indicated by persisting VUR with breakthrough infections. A Pfannenstiel approach is used for a Cohen cross-trigonal re-implantation. Care must be taken to re-implant double ureters en bloc, otherwise the shared blood supply associated with the common sheath will be impaired.



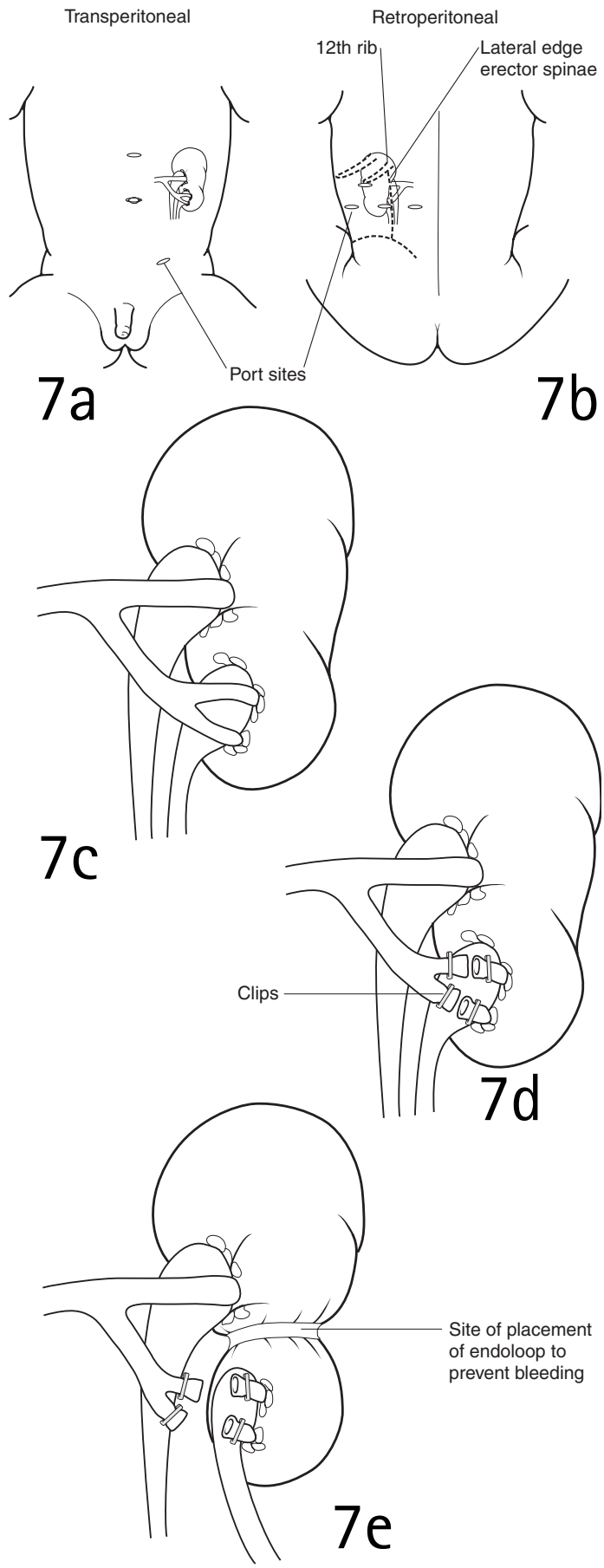
6a



6b

Lower pole heminephrectomy

7 The indication for this is a poor/non-functional lower pole, with recurrent UTIs. The procedure may be done with an open approach or laparoscopically. If performed laparoscopically, the kidney may be approached transperitoneally or retroperitoneally. The kidney is visualized and the blood vessels are ligated with clips and divided. The ureter is divided. An endoloop is placed at the point where demarcation occurs and tightened. The lower pole is then excised and removed.



POSTOPERATIVE CARE

Intravenous fluids and adequate analgesia (which may include a morphine infusion) are continued until patients are drinking adequately. No stents or drains are needed for nephrectomies or partial nephrectomies, whereas bladder drainage (suprapubic catheter) for 3–5 days is appropriate if cystotomy has been performed to allow excision of a ureterocele or ureteric re-implantation.

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81

Urinary diversion in children

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PRINCIPLES AND JUSTIFICATION

Urinary diversion, although uncommon in children, is a useful tool in the armamentarium of the pediatric urologist. The main aim of urinary diversion is to ensure adequate drainage of the urinary tract and thereby preserve renal function. Urinary diversions may be incontinent or continent. Incontinent diversions entail cutaneous drainage of urine into a pad, nappy, or stoma bag. A continent diversion aims to create a low-pressure, compliant, and capacious reservoir and a catheterizable conduit for drainage of the reservoir where the urethra is found to be unsuitable.

The majority of urinary diversions are performed for congenital anomalies of the urinary tract, e.g., neuropathic bladder due to spinal dysraphism, the exstrophy epispadias complex, consequences of posterior urethral valves, functional bladder disorders such as the Hinman bladder, or bladder malignancy. The choice of diversion depends on the child's diagnosis, body habitus, manual dexterity, compliance, and social circumstances. For example, a child with a neuropathic bladder, severe developmental delay, difficult social circumstances, and deteriorating upper tracts may be better off with a vesicostomy rather than a bladder augmentation and a Mitrofanoff conduit.

PREOPERATIVE ASSESSMENT AND PREPARATION

The assessment and preparation of children for surgery consist of three components:

- overall assessment,
- renal assessment,
- assessment and preparation for the specific procedure.

Overall assessment

This entails assessment of the overall fitness of the child for the surgery involved. For example, a child with severe kyphoscoliosis may have decreased respiratory reserve and may require a chest radiograph. Attention should also be directed toward the nutritional status, cardiovascular status, and neurological status of the child. Assessment by the anesthetist well in advance of surgery may be advisable in complicated cases.

Renal assessment

All children should have baseline biochemistry and hematology. Those children with chronic renal failure require input from the nephrologists preoperatively and postoperatively. All children for diversion should also have a baseline ultrasound scan and functional imaging – either a mercaptoacetyl triglycine (MAG-3) or dimercaptosuccinic acid (DMSA) scan. Additional investigations depend on individual patients, e.g., videourodynamics, micturating cystourethrogram, etc.

Assessment and preparation for the specific procedure

This forms a very important component of the assessment process. Before a decision can be made regarding the appropriate procedure for a particular child, the capabilities, compliance, and circumstances of the child and carers must be taken into consideration. In the authors' institute, this is conducted in part by the clinicians in outpatient consultations, but mainly by a dedicated team of clinical urology nurse specialists. Once a decision regarding the type of procedure has been made, the child and carers are prepared for the surgery

by being given leaflets with information about the procedure as well as other visual aids such as videos, and the child is allowed to interact with other children who have had a similar procedure. Surgery is carried out once the child and carers are deemed 'ready' for the procedure. Attention must be paid to the need for bowel preparation, e.g., in colocystoplasty. In general, for most urological procedures we use a broad-spectrum urinary antiseptic such as amikacin alone or a combination of amikacin, benzylpenicillin, and metronidazole at induction and for up to 48 hours postoperatively. Further need for antibiotics depends on the clinical situation.

Anesthesia

All procedures are performed under general anesthesia with endotracheal intubation and full muscle relaxation. Concomitant regional analgesia such as an epidural catheter or caudal block may also be used. Children deemed unsuitable for these regional techniques may receive postoperative nurse-controlled analgesia or patient-controlled analgesia.

OPERATIONS

The following operations are described:

- Incontinent diversions:
  - vesicostomy,
  - ureterostomy and pyelostomy,
  - conduit diversion – ileal conduit, colonic conduit.
- Continent diversions.
  - Surgery of the reservoir: ileocystoplasty, colocystoplasty.
  - The continent catheterizable conduit – the Mitrofanoff principle and its variants: appendicovesicostomy, ileovesicostomy (Yang–Monti), continent tube vesicostomy.
  - Stoma: the VQZ plasty.

INCONTINENT URINARY DIVERSION

Vesicostomy

A vesicostomy may be performed as a temporary measure for urinary diversion in selected cases such as posterior urethral valves, gross vesicoureteric reflux, or neuropathic bladder. In cases of posterior urethral valves, although primary fulguration is the ideal method of treatment, we have found there to be no difference in the outcome for boys with posterior urethral valves who had an initial vesicostomy and delayed ablation as compared to primary fulguration (unpublished data). Vesicostomy has a high incidence of stomal stenosis and prolapse in the long term to be considered as a permanent option.

1a–d

TECHNIQUE

POSITION

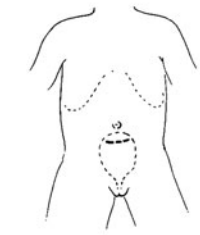
The patient is placed in the supine position.

INCISION

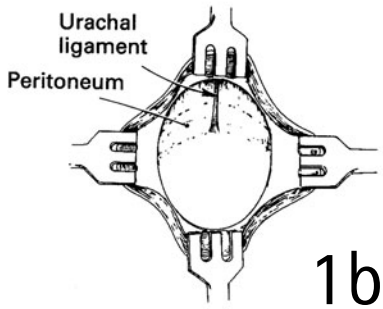
A short transverse incision a few centimeters in length is made above the pubic symphysis in an area that will drain into the pad/nappy.

PROCEDURE

The bladder is mobilized extraperitoneally and the dome is identified by the urachal remnant, which is ligated with an absorbable suture. The dome is sutured circumferentially with interrupted 4/0 absorbable sutures to the fascia. The dome is then opened and the mucocutaneous anastomosis is performed with circumferential 4/0 absorbable, interrupted sutures.



1a



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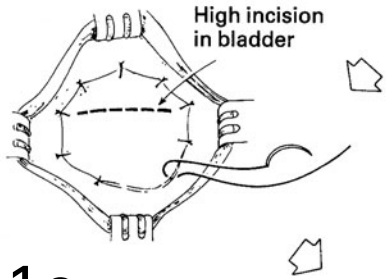


1c

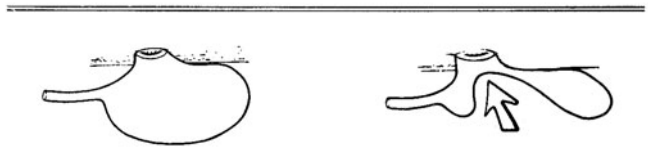
1d

POSTOPERATIVE CARE

A 12 Fr catheter is left in situ for 24 hours and then removed. Subsequently the vesicostomy is allowed to drain into the nappy.

COMPLICATIONS

1e If the vesicostomy is made too low, prolapse of the posterior wall may occur. Stomal stenosis may occur in the long term.



1e

Ureterostomy/pyelostomy

A cutaneous ureterostomy is used as a temporary measure in carefully selected cases for decompression of the upper tracts. Debate still exists as to its role in posterior urethral valves with deteriorating upper tracts either as a primary procedure or even after valve fulguration. We have used this method in selected children under 1 year of age with solitary kidneys and a ureterovesical junction obstruction with deteriorating renal function. A ureterostomy may also be performed where malignancy necessitates large pelvic resections. In selected cases of posterior urethral valves with gross unilateral vesicoureteric reflux, a 'refluxing' ureterostomy may be performed on the side of reflux concomitant to valve resection to act as a pop-off mechanism and thereby protect the upper tracts. The ureterostomy may be created distally or proximally. The description below is of a distal ureterostomy.

2a–g

TECHNIQUE

POSITION

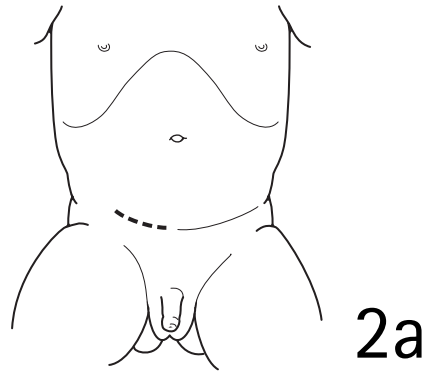
The patient is placed in a supine position; a more oblique position is required for a higher diversion.

INCISION

A short inguinal groin crease incision is made on the affected side (a). The site of the incision is modified depending on the site of the intended diversion; however, the approach to the ureter remains extraperitoneal.

PROCEDURE

A flap of skin is raised superiorly to gain access to the extraperitoneal space via a muscle split. The peritoneum is reflected medially. The ureter is identified by dividing the obliterated umbilical artery (b). In cases of end ureterostomy, the ureter is detached close to the bladder and the bladder end is over-sewn with 4/0 absorbable suture – either polyglactic acid (Vicryl) or polydioxanone (PDS). The ureter is brought out of the groin crease incision and the mucocutaneous anastomosis creating a small everted nipple is performed with interrupted 6/0 absorbable sutures (c). If a loop ureterostomy is performed, the ureter is opened longitudinally and mucocutaneous anastomosis of both limbs is performed with interrupted 6/0 absorbable sutures (d and e). A pyelostomy is performed using a similar technique (f and g).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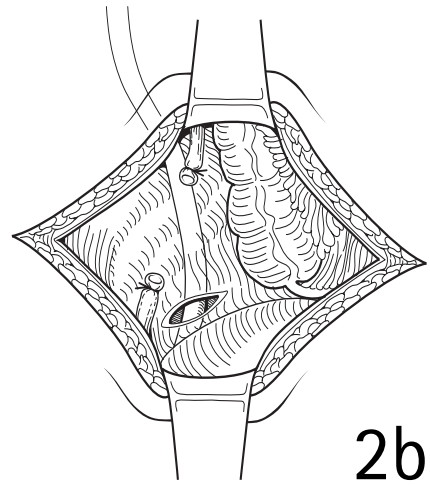
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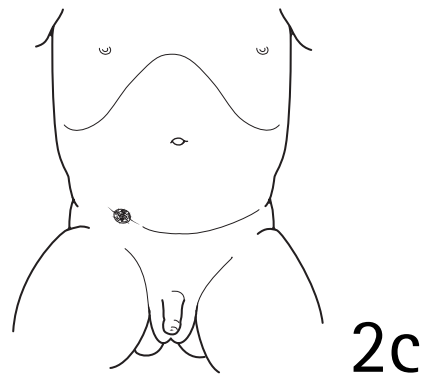
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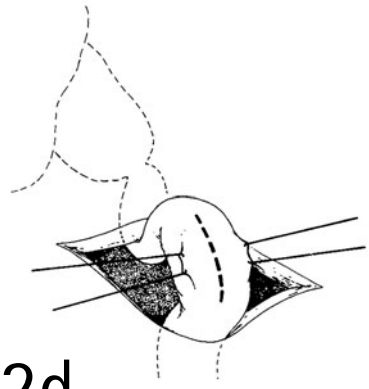

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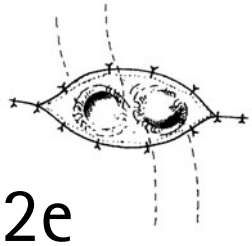

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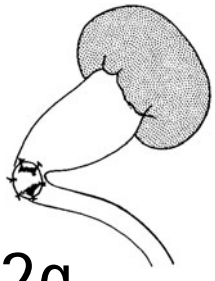


2g

POSTOPERATIVE CARE

The ureterostomy is allowed to drain freely into the nappy.

COMPLICATIONS

Stomal stenosis or stomal prolapse may occur. In cases of dilated tortuous and adynamic upper tracts, good drainage may not be achieved with a ureterostomy and an indwelling catheter or intermittent catheterization of the stoma may be necessary.

Ileal conduit

Because of the high incidence of complications, an ileal conduit diversion is very uncommon nowadays in children. However, in very carefully selected cases, this may be a useful option, for example in children with neuropathic bladder or pelvic malignancy who, along with their carers, may prefer this to major urinary tract reconstruction and continent diversion in the short to medium term.

3a–f

TECHNIQUE

POSITION

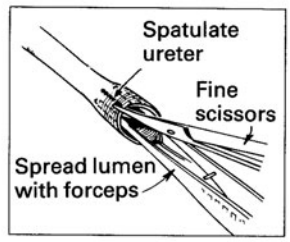
The patient is placed in a supine position.

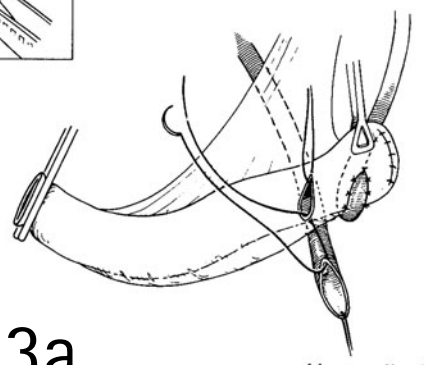
INCISION

The ileum can be approached through the same incision as that for the primary procedure if the diversion is being performed concomitantly. If being performed in isolation, a lower midline incision may be preferable.

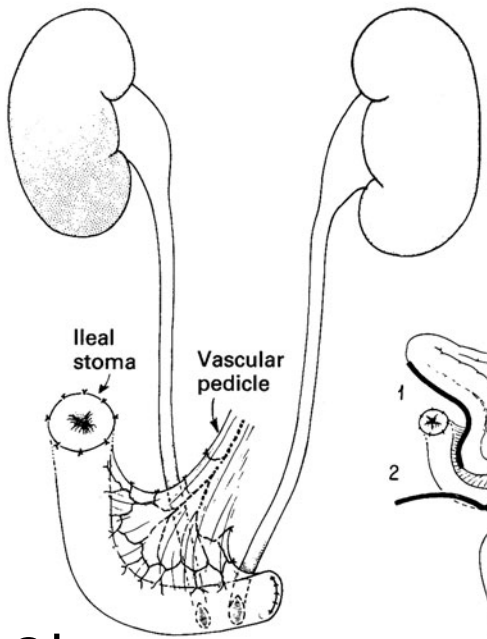
PROCEDURE

A short, 6–10-cm, segment of ileum on its mesentery is isolated and the proximal end is over-sewn with a 4/0 or 5/0 PDS extramucosal, single-layer, interrupted or continuous suture. Intestinal continuity is restored with 5/0 PDS extramucosal, single-layer interrupted or continuous suture. The ureters are spatulated (a) and implanted in the conduit in a submucosal tunnel, and the anastomosis is completed with 6/0 PDS interrupted sutures (b). The distal end of the ileal loop is brought out at a suitable pre-marked position in the right iliac fossa and everted to create a nipple (c). The mucocutaneous anastomosis is completed with interrupted 5/0 or 6/0 PDS sutures. Two 4 Fr or 6 Fr feeding tubes are left indwelling within the implanted ureters and exiting via the stoma. If the ureters are dilated, the spatulated ends may be anastomosed with 6/0 PDS to form a common channel (d), which is in turn anatomosed to the proximal end of the conduit with 4/0 or 5/0 PDS sutures (e and f).





3a



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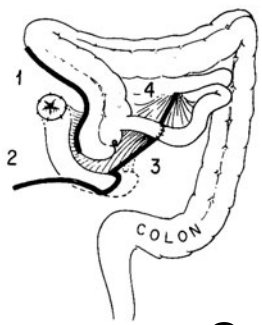

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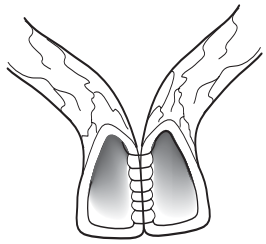

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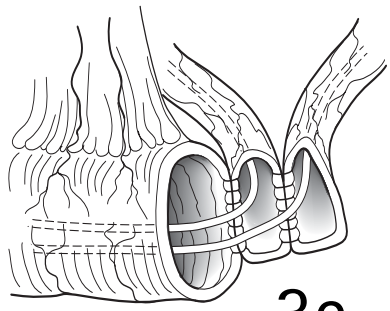

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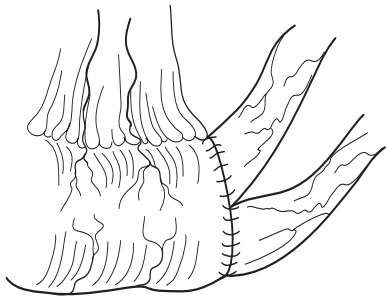


3f

POSTOPERATIVE CARE

The feeding tubes are removed 10 days to 2 weeks after surgery. Close monitoring of the upper tracts is essential.

COMPLICATIONS

Problems with ureteral stenosis or vesicoureteric reflux may occur along with deterioration of renal function. Therefore close follow-up is mandatory.

Colonic conduit

4a–d

TECHNIQUE

POSITION

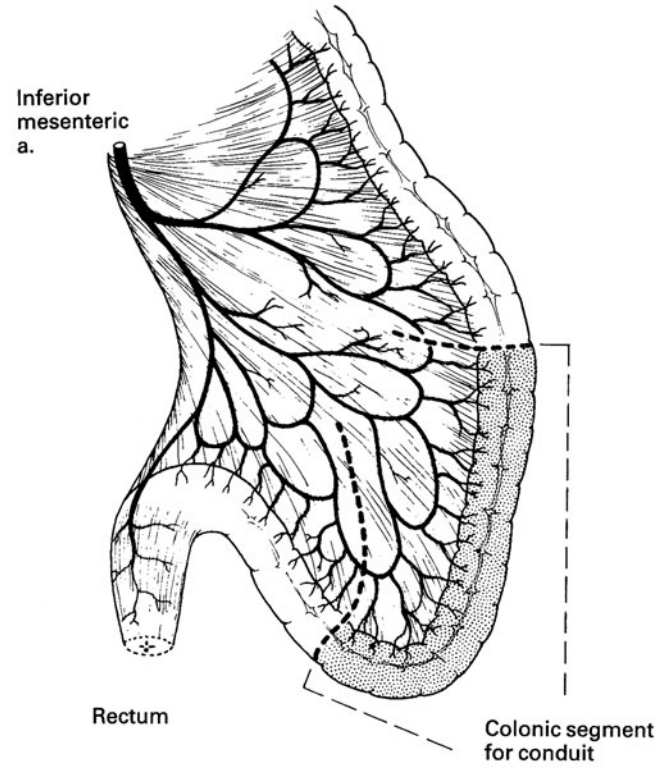
The patient is placed in a supine position.

INCISION

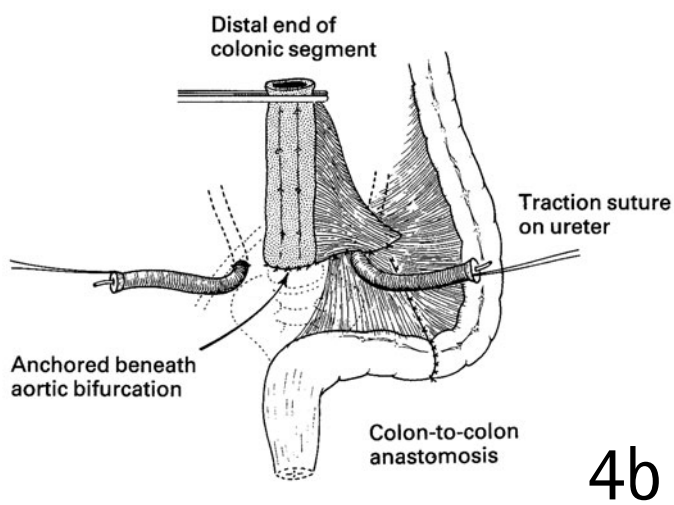
A lower midline incision is preferred.

PROCEDURE

A suitable segment of colon, preferably the sigmoid colon, is isolated on its mesentery and intestinal continuity is restored with 5/0 PDS sutures (a and b). The distal end is over-sewn and the ureters are spatulated and implanted in the conduit in a non-refluxing manner (c). The distal end of the conduit is brought out as a stoma at a predetermined, pre-marked site on the abdominal wall and everted. Mucocutaneous anastomosis is performed with absorbable sutures of 5/0 Vicryl or 5/0 PDS sutures (d).



4a



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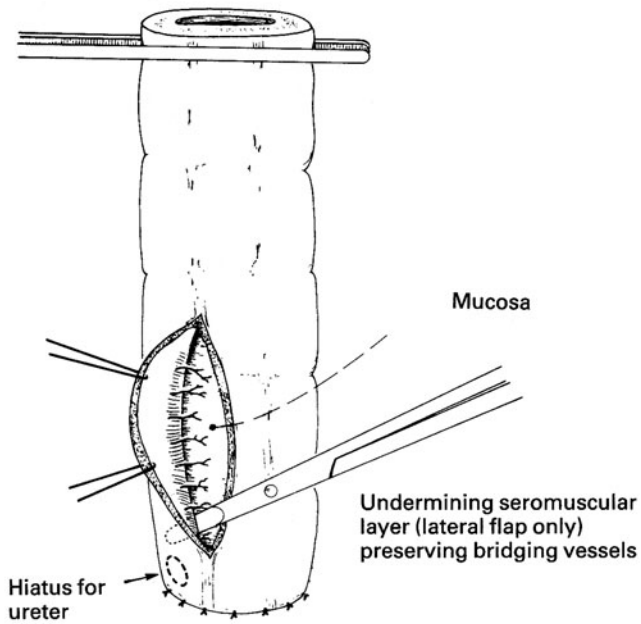

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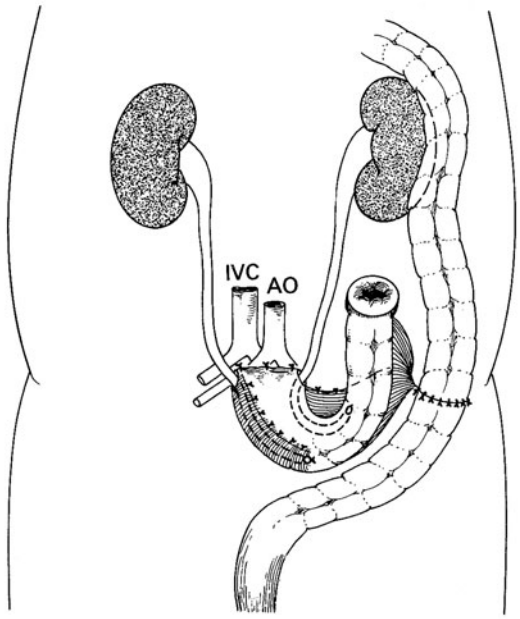


4d

POSTOPERATIVE CARE

As with ileal conduit, careful monitoring of the upper tracts is essential.

COMPLICATIONS

These are similar to those of an ileal conduit and include ureteral stenosis and kinking and deterioration of the upper tracts.

CONTINENT URINARY DIVERSION

Surgery of the reservoir

ILEOCYSTOPLASTY

A detubularized segment of ileum is usually used for bladder augmentation. In cases where the mesentery is too short to reach the pelvis, e.g., gross kyphosis, colon may be used. A bladder outlet procedure may be performed simultaneously to increase outlet resistance. We do not routinely advocate preoperative bowel preparation for an ileocystoplasty, as the contents of the ileum are fluid and in our opinion do not need mechanical cleansing.

5a-g

TECHNIQUE

POSITION

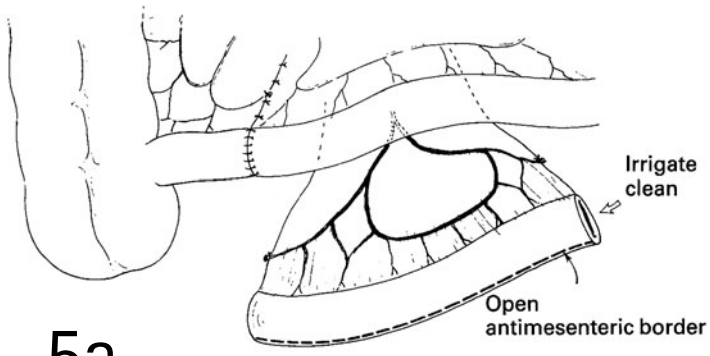
The patient is placed in a supine position.

INCISION

A pfannenstiel or midline incision is preferred.

PROCEDURE

Initial extraperitoneal mobilization of the bladder is carried out circumferentially down to the region of the bladder neck. A small opening is made in the posterior aspect of the reflected peritoneum and the cecum and terminal ileum are delivered. If an appendicovesicostomy is to be performed, mobilization of the appendix and mesentery can be performed at this stage. A suitable segment of ileum approximately 25 cm long and 20 cm from the ileocecal junction is isolated on its mesentery and intestinal continuity is restored with 5/0 PDS sutures (a). The ileum is detubulularized along its antimesenteric border (b). The peritoneum is closed, thereby extraperitonealizing the segment of ileum and bladder. The bladder is opened from one ureteric orifice to the other in either the coronal or sagittal plane (c). The appendix is re-implanted in the bladder at this stage and a clam cystoplasty is performed with continuous 3/0 absorbable suture (d). Just before completion of the anastomosis, a large-bore 16 Fr or 18 Fr Foley catheter is inserted suprapubically. If an appendicovesicostomy has been created, an indwelling catheter is left within this conduit. If the bladder is very small, thick walled and contracted, it may be excised, leaving only the bladder plate comprising of the trigone and the ureteric orifices. In this case, the ileal patch may be reconfigured as a pouch (e–g), which can be sutured to the bladder remnant.



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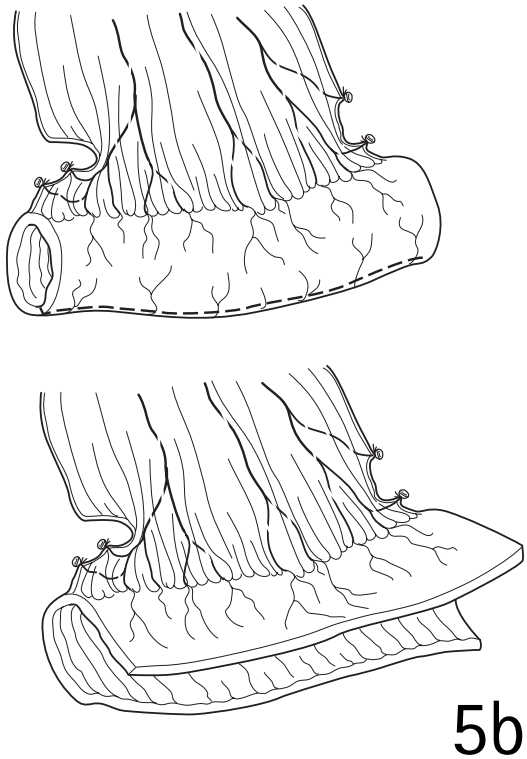

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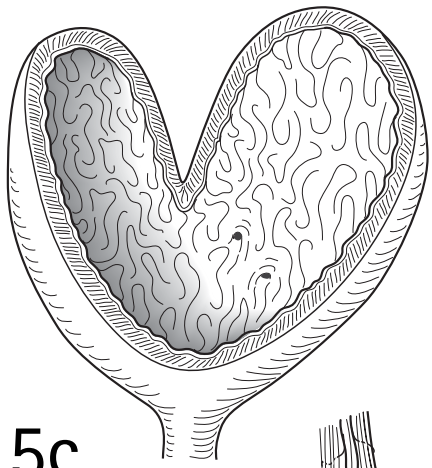

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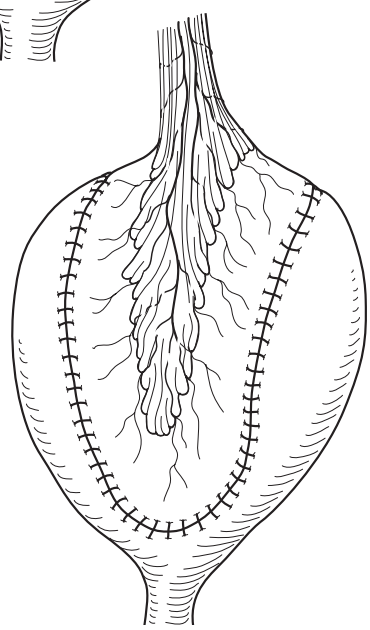

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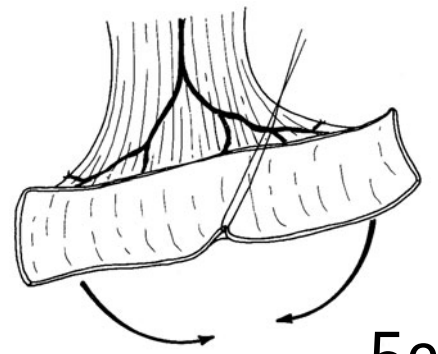

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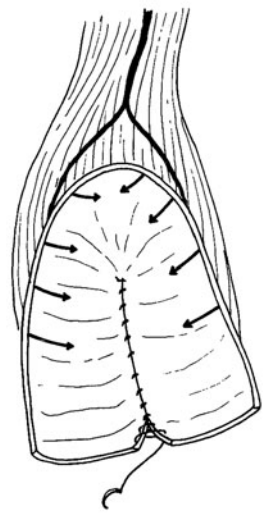

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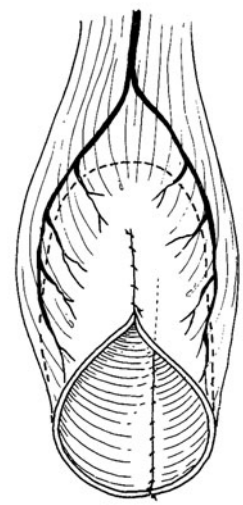


5g

POSTOPERATIVE CARE

The suprapubic catheter is left on free drainage for 3 weeks with bladder washouts as required. A regime of clamp and release is then instituted prior to commencing clean intermittent catheterization either urethrally or via the Mitrofanoff channel. All patients should have periodic renal ultrasound scans and regular estimation of biochemical parameters, including acid–base status.

COMPLICATIONS

The patient and carers must be warned of the potential risks of this procedure. These include mucus blockage, urinary infections, the development of bladder calculi, persistence of a high-pressure neoreservoir, metabolic and acid–base disturbances, and the possibility in the long term of the development of adenocarcinoma.

COLOCYSTOPLASTY

The segment used may be detubularized sigmoid colon, the transverse colon, or the ileocecal and ascending colon. The principles of the technique are similar to those described above, as are the postoperative care and complications.

The Mitrofanoff principle

In 1911, Coffey desribed the flap valve technique of ureteral re-implantation in ureterosigmoidostomy. This principle was utilized by Mitrofanoff in 1980 to create a continent, catheterizable appendicovesicostomy. The main components of the Mitrofanoff technique are:

- a narrow, supple conduit brought to the skin as a catheterizable stoma;
- an antirefluxing connection between the conduit and the reservoir (flap valve);
- a large, low-pressure urinary reservoir;
- an antirefluxing mechanism between the upper urinary tract and the reservoir;
- intermittent catheterization to allow effective, regular, low-pressure emptying of the reservoir.

Appendicovesicostomy

6a,b

TECHNIQUE

POSITION

The patient is placed in a supine position.

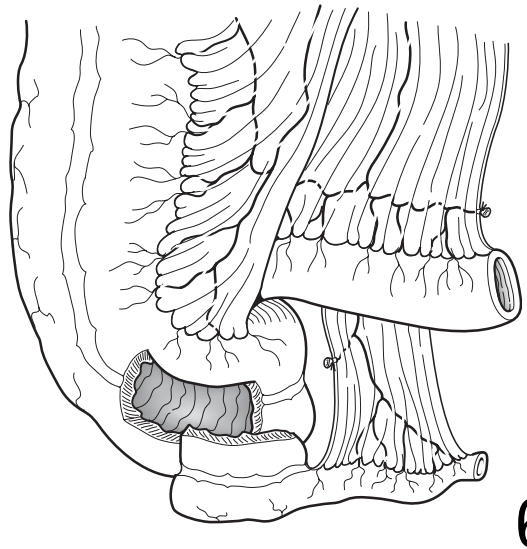
INCISION

A pfannenstiel or lower midline incision is used.

PROCEDURE

Assessment is made of the length of the appendix and its suitability as a conduit. The appendix is mobilized with its mesentery and detached from the cecum, which is closed with absorbable sutures. A cuff of cecum may be mobilized with the appendix and tubularized to gain additional length, if necessary (a). The tip of the appendix is incised and a 12 Fr feeding tube is inserted to confirm patency and diameter of the lumen. The lumen is irrigated with dilute betadine solution. The distal end of the appendix is then tunneled into the bladder in a submucosal tunnel for 3–4 cm to maintain a 5:1 ratio and ensure a competent flap-valve mechanism (b). This is then anchored to the muscle and mucosa of the bladder using absorbable suture. The rest of the circumference of the appendix is sutured to the bladder mucosa using absorbable sutures. The appendix is also anchored at its entry into the bladder by a few interrupted sutures.

The proximal end of the appendix is brought out as a stoma by the shortest and straightest route possible to enable easy catheterization. The site of the stoma will therefore be variable. The umbilicus may be used to site the stoma in certain children for easy access, or a VQZ stoma technique for abdominal Mitrofanoff stomas can be used.



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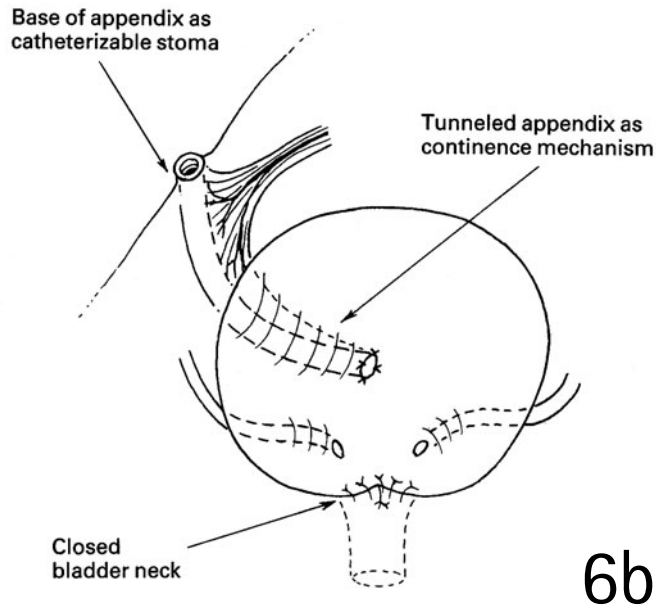


6b

POSTOPERATIVE CARE

An indwelling 10 Fr or 12 Fr Jacques catheter is left for 3 weeks and is followed by clamp and release and clean intermittent catheterization under nursing supervision.

COMPLICATIONS

Problems with the conduit include stomal stenosis, difficulty in catheterization leading to the creation of a false passage, and the development of calculi.

Ileovesicostomy

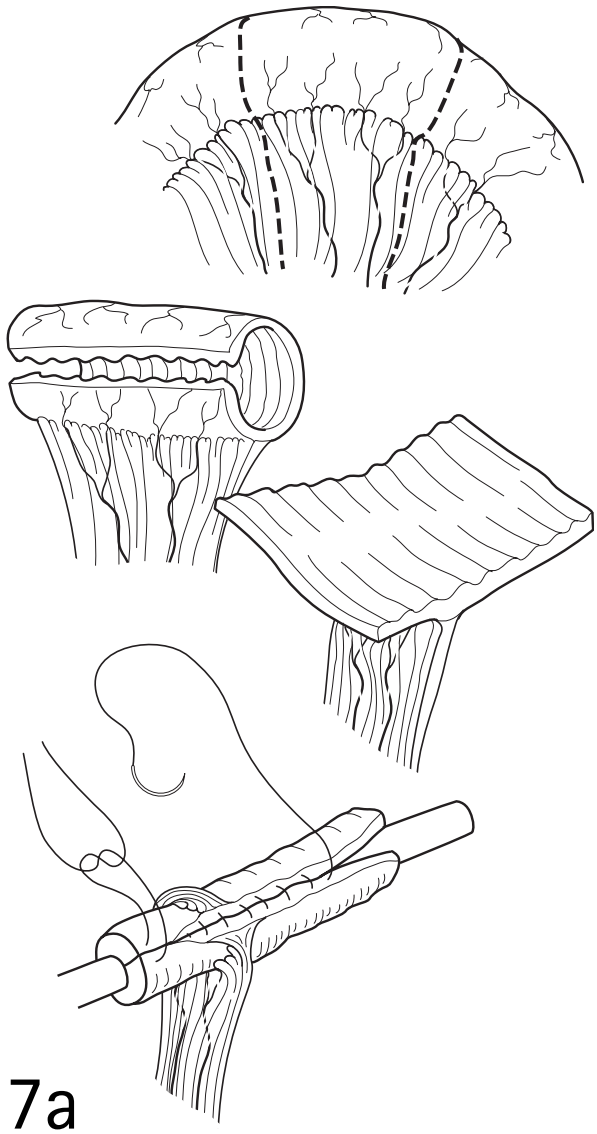
Where the appendix is absent or not suitable, a segment of ileum can be utilized to create a transverse ileal tube. This is the preferred second-line option for creating the Mitrofanoff channel, especially when ileum is used for bladder augmentation.

7a–c

TECHNIQUE

PROCEDURE

A segment of ileum approximately 1.5–2 cm in width adjacent to that used for the cystoplasty is isolated on its mesentery and opened longitudinally on its antimesenteric border. The position of the mesentery along the tube can be moved depending on the position of the longitudinal opening. This tube is then tubularized transversely over a 12 Fr catheter (a). In patients with a long distance between the reservoir and the abdominal wall or the umbilicus, two adjacent segments can be isolated, tubularized, and anastomosed (Full Monti – illustration b). A similar technique has been described by Casale using a single piece of bowel to gain additional length (c). The tube is then tunneled into the reservoir as described above to create the Mitrofanoff flap valve.



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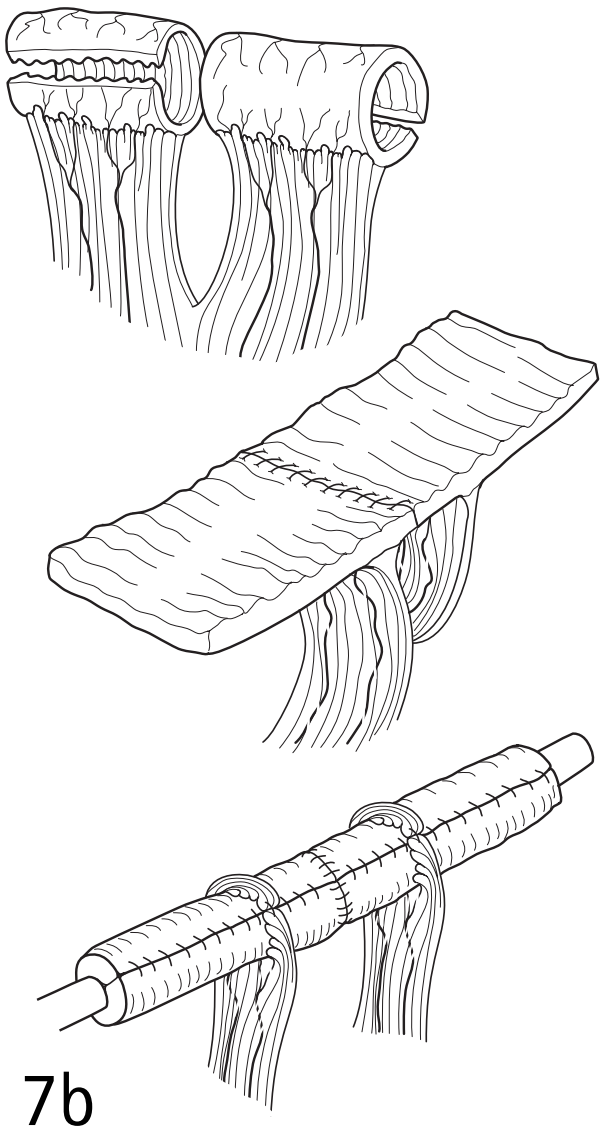

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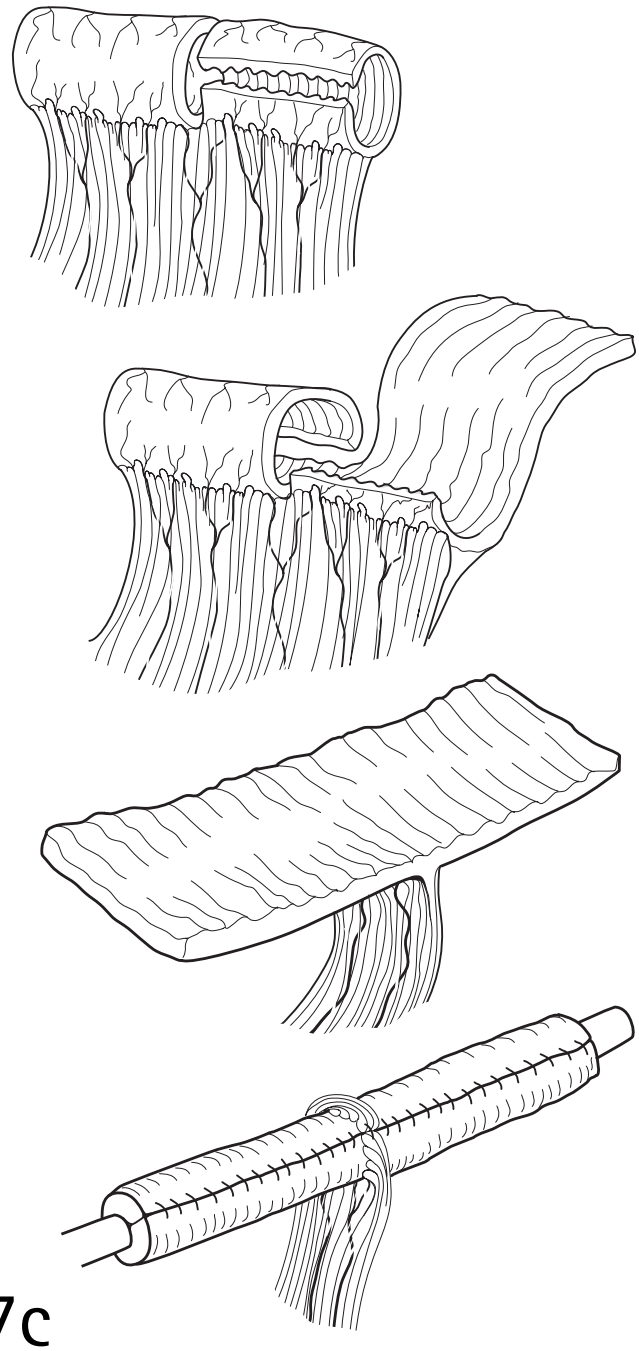


7c

POSTOPERATIVE CARE

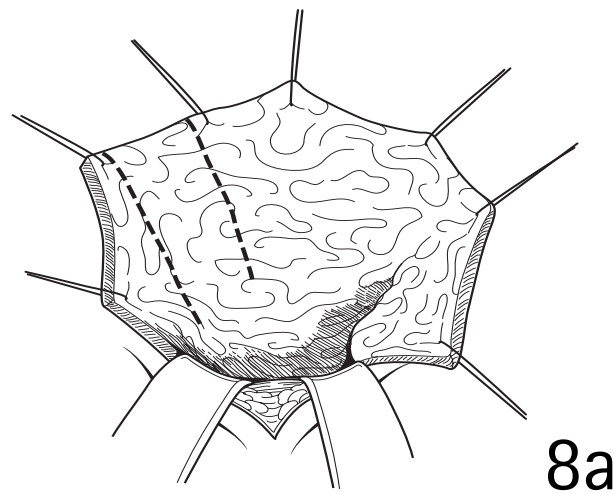
As with the appendicovesicostomy, an indwelling catheter is left in situ for 3 weeks before instituting clamp and release and clean intermittent catheterization.

COMPLICATIONS

Due to the elasticity of the ileum, more problems may be encountered with catheterization than with the appendicovesicostomy.

Detrusor tube vesicostomy

This technique involves a segment of tubularized bladder with an antireflux flap valve constructed primarily of mucosa. Patients must have a large bladder capacity to be considered suitable.

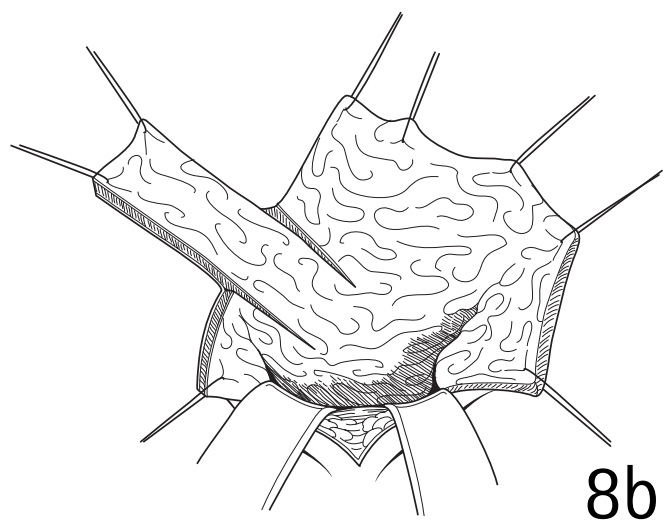

8a

8a–f

TECHNIQUE

PROCEDURE

A broad-based rectangular flap (approximately 3 cm × 7 mm) of full-thickness detrusor muscle and mucosa is raised on its vascular supply from the superior vesical pedicle. This is tubularized in two layers (mucosa and muscle) over a 12 Fr catheter. The proximal mucosal end is further tubularized within the bladder for about 2 cm and covered by laterally raised adjacent mucosal flaps.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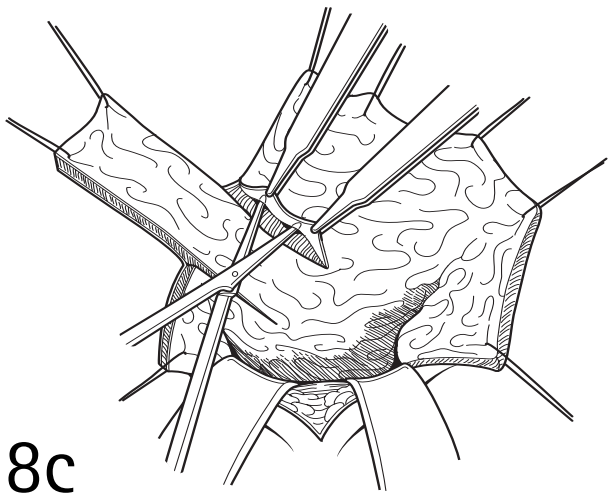

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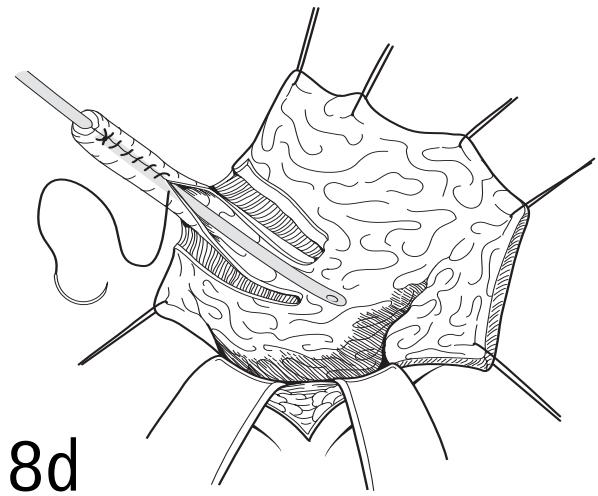

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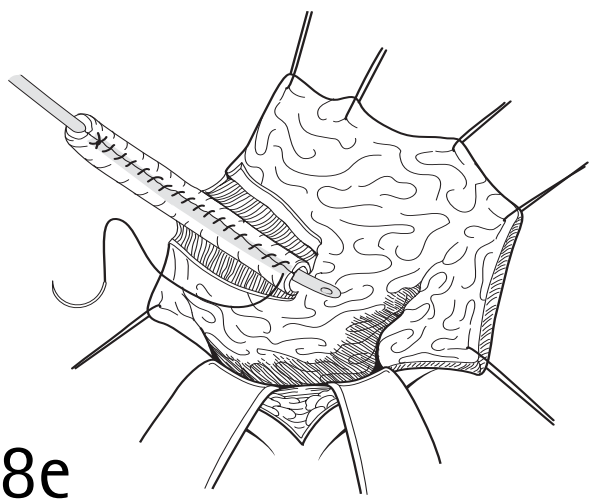

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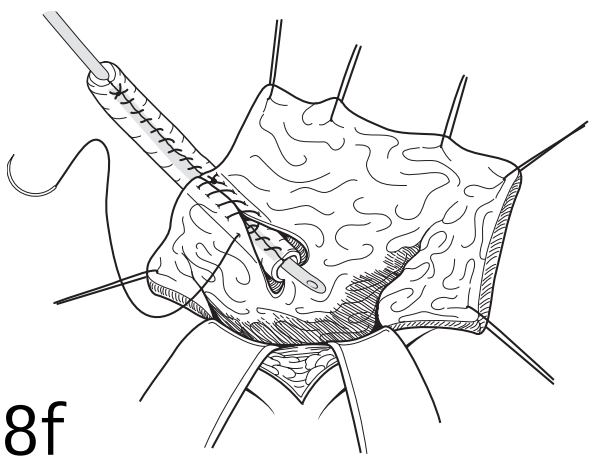


8f

POSTOPERATIVE CARE

Postoperative care is similar to that for appendicovesicostomy.

COMPLICATIONS

Stomal stenosis is the main complicating factor.

The VQZ stoma

The VQZ plasty for the cutaneous opening into a Mitrofanoff conduit has several advantages over the standard V flap or flush stoma in that it is discrete and there is no exposed mucosa and hence less likelihood of contact bleeding.

9a–k

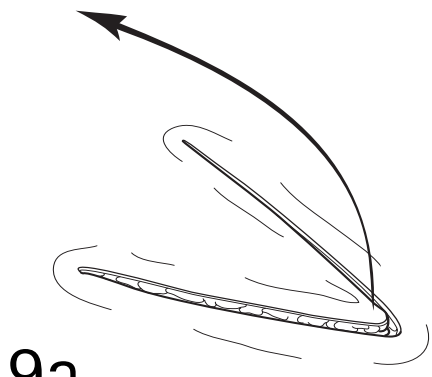
TECHNIQUE

POSITION

The patient is placed in a supine position.

INCISION AND PROCEDURE

A V flap is raised with its base in the region of the exiting appendix (a). A generous opening is made into the layers of the anterior abdominal wall and the appendix is delivered. The abdominal incision is then closed. The appendix is incised along its antimesenteric border and the flap is sutured in place with 5/0 PDS along its entire length up to its angle on its superior border but stopping 5–10 mm short on its inferior border (b–d). The Q flap is then raised and rotated and sutured into place along the rest of the appendicular margin and the remaining inferior border of the V flap (e–g). The mucosa is therefore entirely covered. A standard Z plasty is performed to rotate the adjacent skin to cover the defect (h–k). Subcutaneous 3/0 PDS sutures may be required at the angles of the Z plasty, which will be under some tension. The rest of the skin is closed with 5/0 or 6/0 PDS sutures.



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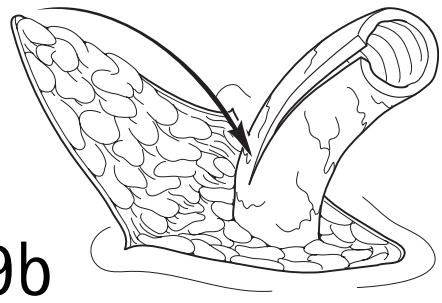

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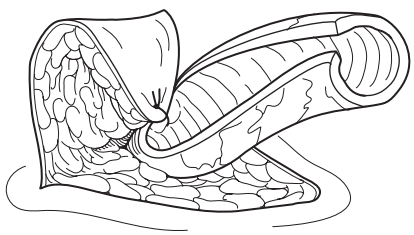

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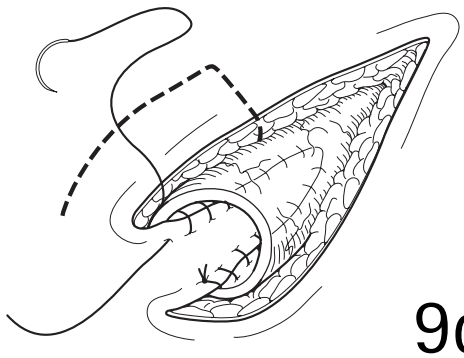

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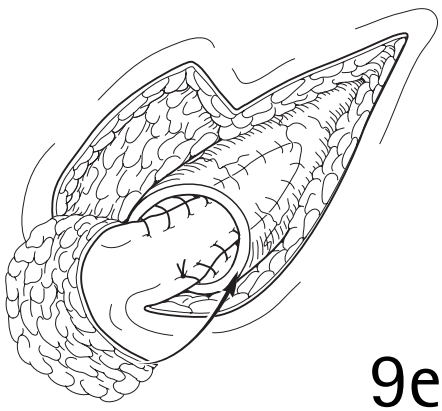

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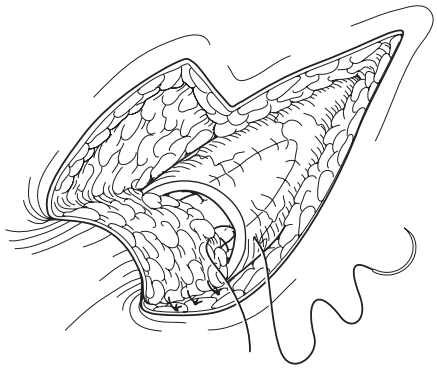

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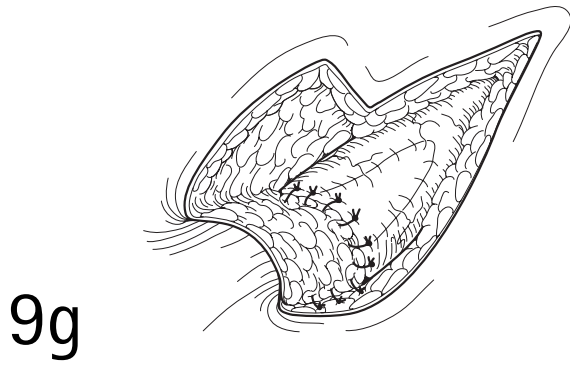

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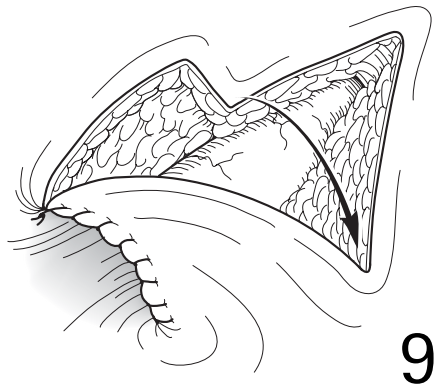

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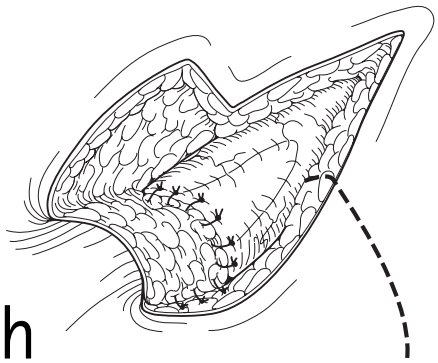

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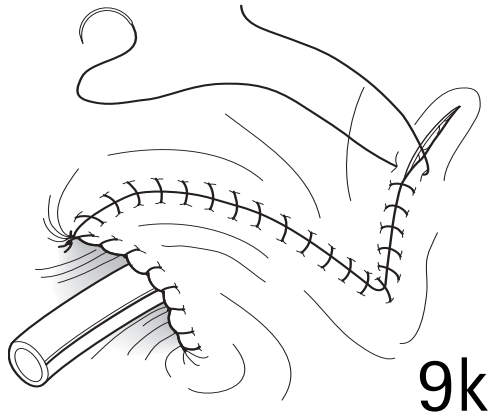

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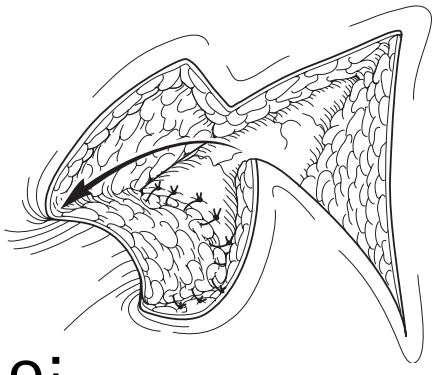

9i

POSTOPERATIVE CARE

The wound should be left open to allow regular inspection of the stoma site.

COMPLICATIONS

Wound infection and wound dehiscence may occur. Stomal stenosis may require further skin-level revisions.

CONCLUSION

Numerous techniques are available for urinary diversion in children. The choice depends on careful patient selection and requires a close working relationship between multidisciplinary teams including urologists, nurse specialists, physiotherapists, psychologists, and nephrologists. The ultimate goal remains the preservation of renal function by any technique that is appropriate for, and acceptable to, the individual child and his or her carers.

FURTHER READING

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82

Surgery of renal calculi

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ETIOLOGY

The majority of renal calculi in children were considered to be of infective origin. A recent review suggests a shift in the epidemiology of renal stone disease in the UK, with a metabolic abnormality detected in 44 percent of children; 30 percent were classified as infective and 26 percent idiopathic. Hypercalciuria (57 percent) was the most common metabolic abnormality, followed by cystinuria (23 percent), hyperoxaluria (17 percent), hyperuricosuria (2 percent), and unclassified hypercalcemia (2 percent).

The organisms most commonly associated with infective calculi were the urea-splitting *Proteus* and *Escherichia coli*. Infective calculi are usually soft, containing organic matrix, and may be poorly opacified. They usually consist of calcium magnesium and ammonium phosphate.

Overall, boys are more commonly affected than girls and tend to present at a younger age – average 36 months for boys and 48 months for girls. Presenting features are macroscopic hematuria, urinary tract infection, or abdominal pain. Obstruction related to calculi may result in pyonephrosis, perinephric abscess, or progressive pyelonephritis.

PREOPERATIVE

Radiology

A plain abdominal radiograph is usually the first investigation and should include the whole urinary tract. In children, bowel gas may make interpretation difficult.

An ultrasound scan is an excellent minimally invasive investigation, which will reveal stones in the kidney or bladder. Dilatation proximal to an obstructing stone will also be demonstrated, and the presence of debris will raise the suspicion of infection and the need to relieve the obstruction. The ultrasound will also give an assessment of the size, shape, and number of calculi in the kidney or bladder.

A combined intravenous pyelogram and di-mercaptosuccinyl acid (DMSA) scan performed simultaneously is a most effective investigation to give an outline of the pelvis and calyces and also to show the differential function of the kidneys. With these detailed investigations, a decision can be made as to the method of treatment, which could be extracorporeal shock-wave lithotripsy (ESWL), percutaneous nephrolithotomy (PCNL), or ureteroscopy.

A micturating cystourethrogram is not essential in the assessment preoperatively and may only be performed postoperatively when the patient is stone and infection free. It is not a mandatory investigation.

Urinalysis

Microscopy and urine culture are performed. Urine is sent for determination of pH, calcium, urate, oxalate, cystine, and creatinine. Stones are analyzed for chemical composition. These investigations may be performed before surgery or much later when the child is completely stone free.

Plasma

Creatinine, urea, potassium, sodium, chloride, bicarbonate, magnesium, calcium, phosphate, alkaline phosphatase, albumin, and urate tests are performed.

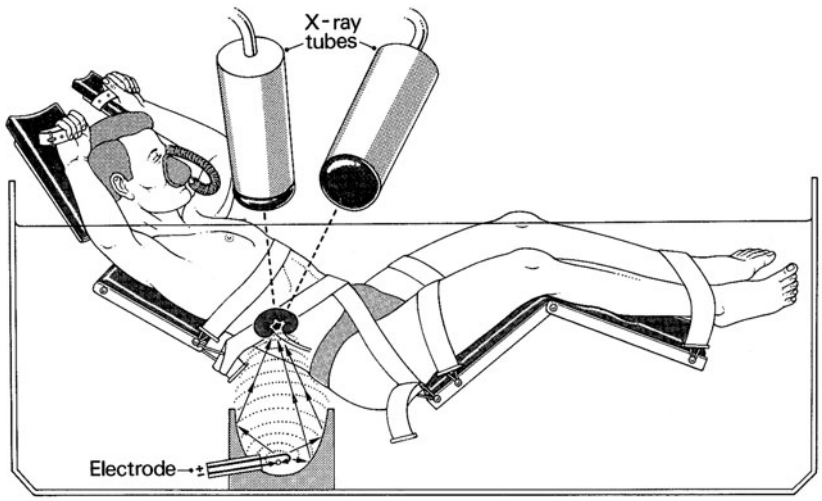
The findings of nephrocalcinosis, bilateral calculi, or recurrent calculi suggest a metabolic abnormality. Nephrocalcinosis may be associated with renal tubular acidosis, hyperoxaluria, and hypercalcemia. Uric acid and cystine calculi are mildly radio-opaque. Xanthine and dihydroxyadenine stones are radiolucent.

OPERATIONS

The more traditional forms of surgery for renal calculi have been superceded by ESWL and PCNL in the adult population. As instruments improve, more children are being treated with these modern techniques.

Extracorporeal shock-wave lithotripsy

1a,b The principle of shock-wave lithotripsy is to disintegrate a stone with the mechanical stress of a shock wave. The shock wave was originally produced by discharging electrical energy into a water bath with some subsequent focus of the wave onto the stone (Illustration a). The patient was placed in a water bath, two X-ray cameras detected the position of the stone, and, with the use of a computer, the shock wave from the probe was directed through the water onto the stone. Other methods of inducing shock waves include rapid vibration of piezoelectric crystals or electromagnetic diaphragms. The newer, second-generation machines do not require a large water bath, and may induce less energy in the shock wave and therefore less pain. The pediatric patient may be treated under local analgesia without general anesthetic because of improved accuracy of focusing of the waves onto the stone and less discomfort (Illustration b). In practice, however, it is difficult to maintain the child in one position during treatment, and general anesthetic is utilized in young children and babies. The long-term effect of ESWL on the developing kidney is unknown.



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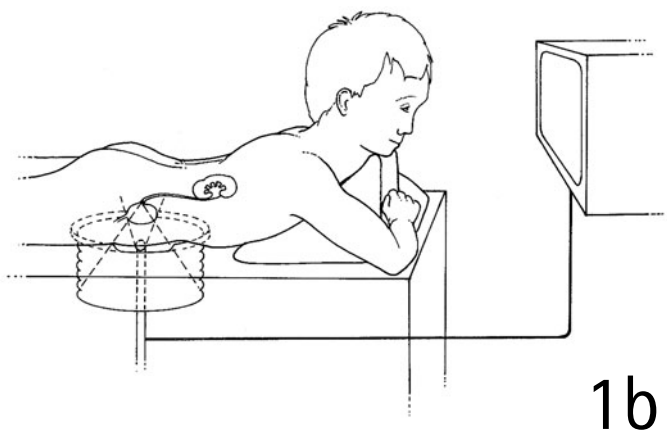
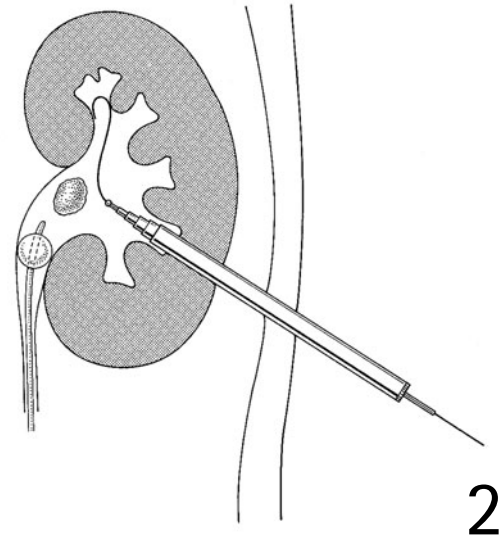


1b

Percutaneous removal

2 Percutaneous nephrolithotomy is utilized to disintegrate and remove (1) multiple, large, or staghorn calculi, (2) calculi in patients with gross spinal deformities where focusing the shock waves onto the stone is difficult, and (3) moderate-sized calculi without disintegration and risk of residual fragments.

The benefits of this surgery increase as the child enters adolescence, as its main advantage is the avoidance of a large, muscle-cutting incision.



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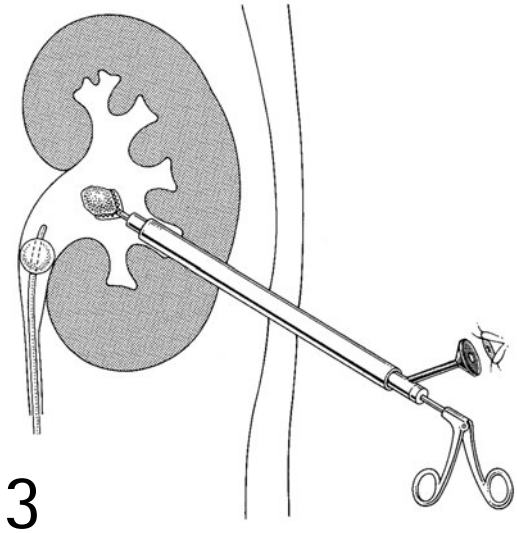
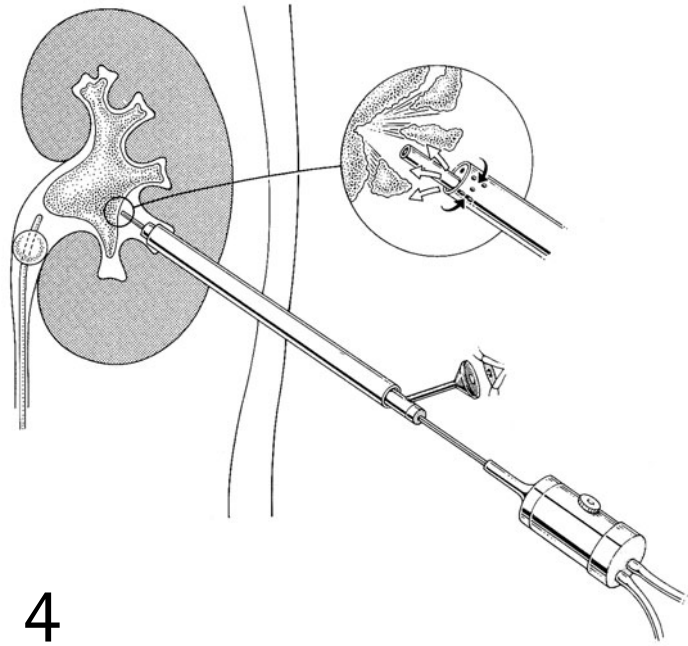


3

3 A nephroscope in a sheath with grasper to extract the stone.



4

4 A staghorn calculus can be disintegrated under direct vision using ultrasound, laser, or lithoclast probes.

POSTOPERATIVE

Nephrostomy drainage is required for 24–48 hours.

Open surgery

For those centers that do not have this equipment, the operation is performed in children using the traditional surgical approach.

PREPARATION OF THE PATIENT

If possible, the urine should be rendered sterile before the operation. An appropriate antibiotic should be administered in the perioperative period.

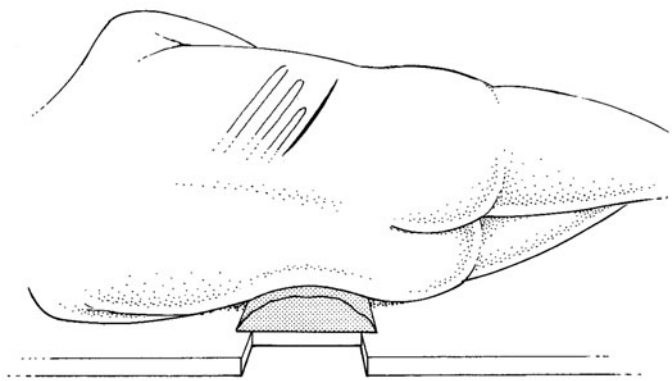
ANESTHESIA

General anesthesia with an endotracheal tube, relaxation, and artificial ventilation are required. Excessive respiratory movement should be avoided to reduce the movement of the kidney and the intrusion of the pleura into the operative field.

POSITION OF PATIENT

The surgeon should personally supervise the positioning of the patient on the operating table.

The patient is placed in a full lateral position with the lower ribs positioned over the table break or adjustable bridge. The degree of break or bridge elevation will vary with the size of the child. For infants and small children, loosely packed sandbags or a foam rubber pad may be more suitable. A degree of head-down tilt is convenient and aids venous return. The patient is secured to the table with non-elastic zinc oxide strapping passed over the pelvis and secured to the table on either side. Further strapping of the shoulder may be required in older children, whereas in smaller children a foam pad or sandbag under the dependent side of the chest may aid stability. The child should be held firmly with the back vertical while the strapping is applied, following which a little lateral roll of the table towards the surgeon may be helpful.



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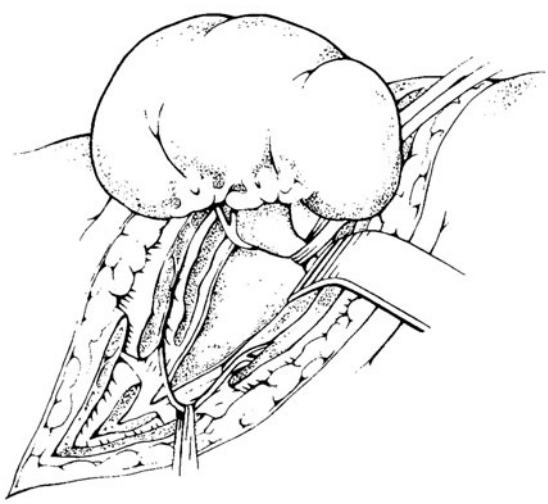
INCISION

5 An incision extending from the tip of the twelfth rib is suitable in most cases. In older children, a supracostal or twelfth rib approach may give better access.



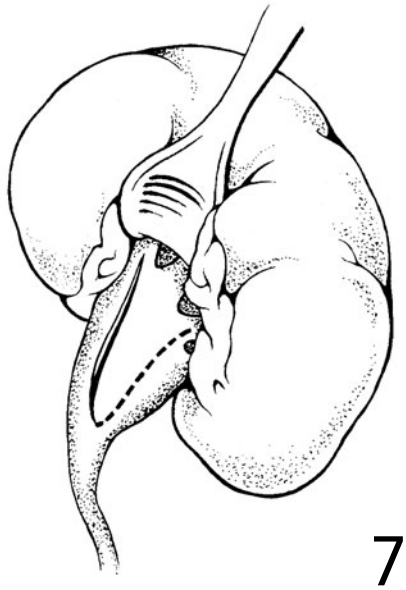
6

EXPOSURE OF THE KIDNEY

6 The incision is deepened using cutting diathermy, the peritoneum being pushed forward with the fingers before completing the anterior portion. The subcostal nerve is identified and preserved. Gerota's fascia is incised longitudinally and a finger is swept over the kidney surface to free it from surrounding fat. The ureter is identified and secured with a sling. In most children, the kidney may now be delivered into the wound and the posterior surface exposed.

INCISION INTO RENAL PELVIS

7 The surface of the pelvis is freed of fatty tissue and the parenchyma retracted. Formal dissection of the renal sinus is not usually required in children. With a large extrarenal pelvis, a vertical incision may be employed. If the pelvis is small, an oblique incision extending up toward the infundibulum of the upper calyx gives better access and may be continued into the lower calyx to raise a triangular flap. Stay sutures are applied to the margins of the incision.



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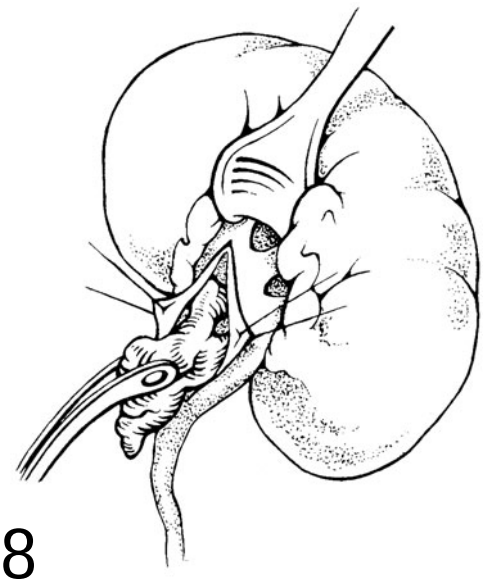


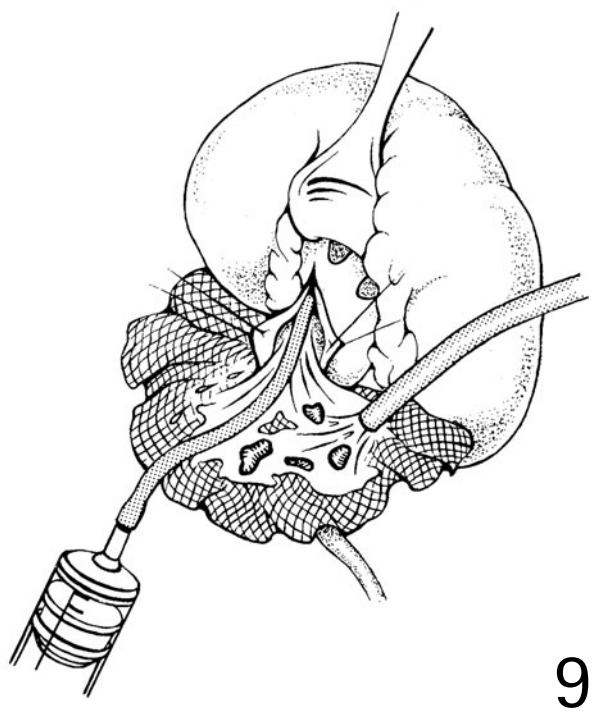
8

REMOVAL OF STONES

8 A stone in the renal pelvis will now be visible and can be lifted out gently with stone forceps.

IRRIGATION OF PELVICALYCEAL SYSTEM

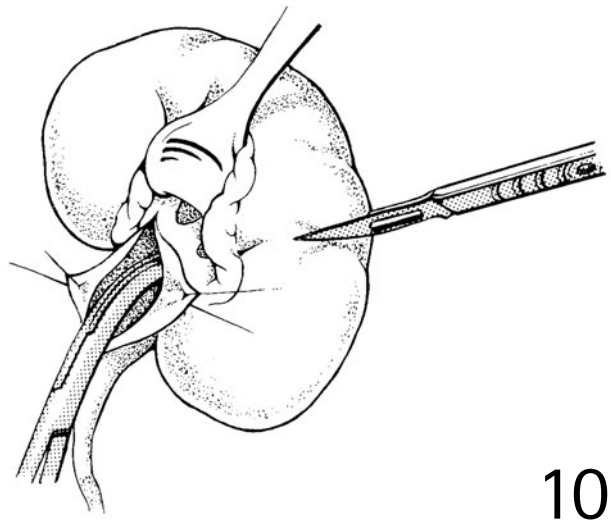
9 Gauze swabs are now placed around the pelvis to catch small stones and debris to allow suction without fatty tissue occluding the sucker. A soft catheter with an end hole rather than side holes is introduced and the calyces are irrigated systematically with normal saline. Stones and debris are carefully removed and any lost into the wound must be retrieved to prevent confusion on later radiographs. A radiograph of the exposed kidney is then taken to confirm complete clearance. A marker should be included in the film to assist orientation. Intraoperative ultrasound may aid in the detection of residual calculi.



9

REMOVAL OF CALYCEAL STONES

10 Calyceal stones may be removed via the renal pelvis with curved stone forceps. A calyceal stone can be identified using stone forceps, palpation, or ultrasound probe, and a nephrotomy incision directly onto the stone permits easy removal.



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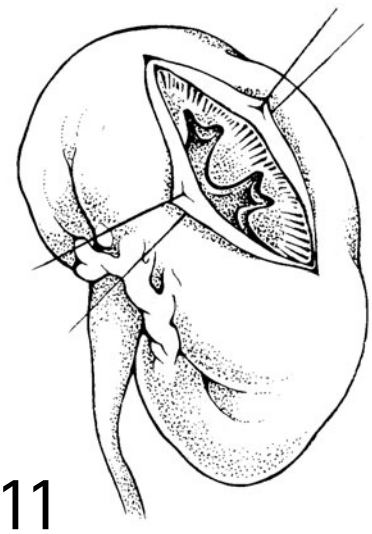


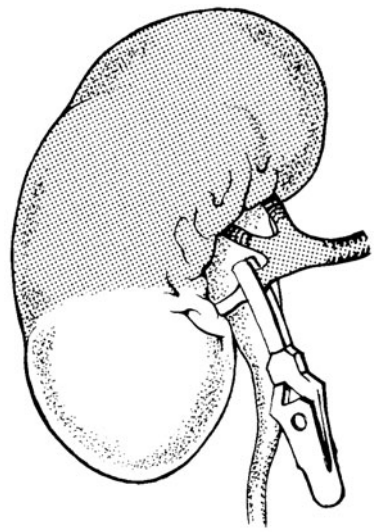
11

EXPOSURE OF SEVERAL CALYCES

11 Large staghorn stones may require the exposure of several calyces. A bulldog clip is applied to the renal artery or the whole renal pedicle is occluded with a soft sling. The longitudinal incision of the posterior surface parallel to the lateral margin of the kidney gives good access. Following removal of the stones, the clamps are released intermittently to allow identification and under-running of major vessels. The calyces are approximated with interrupted 4/0 or 5/0 absorbable sutures and the kidney parenchyma is opposed with loosely tied horizontal mattress sutures through the capsule. The kidney swells on removal of the clamps and if these sutures are too tight they will cut out.

LOWER POLE CALCULI

12 The lower branch of the renal artery is readily identifiable and may be occluded with a bulldog clip. Intravenous methylene blue following occlusion may aid demarcation.



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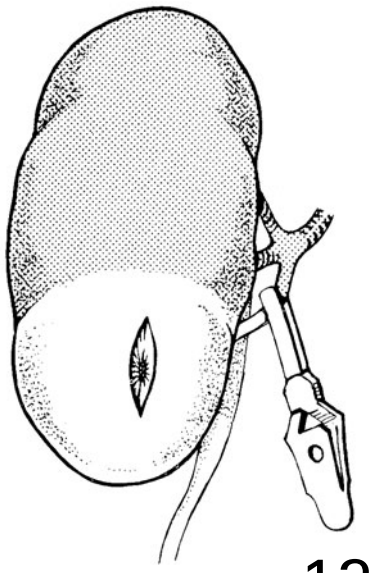
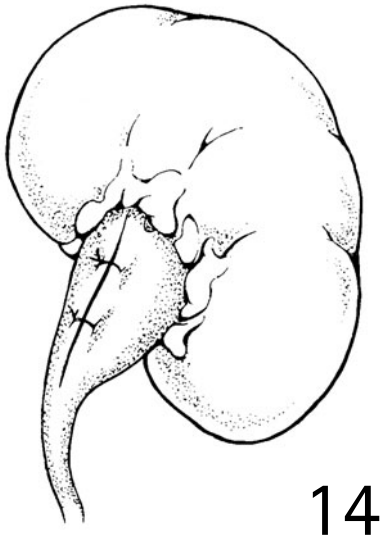


13

INCISION INTO LOWER POLE

13 Simple incision into the lower pole calyx may then be performed. Lower pole partial nephrectomy is rarely required in children.



14

CLOSURE

14 Following radiographic confirmation of complete clearance, the incision in the renal pelvis is closed with interrupted 5/0 or 6/0 absorbable sutures. A drain is positioned adjacent to the renal pelvis and Gerota's fascia is reconstructed over this area. The wound is closed in layers with absorbable sutures.

The stones should be sent separately for analysis and culture.

POSTOPERATIVE CARE

Intravenous fluids are administered for 24 hours. If the peritoneal cavity has been opened during the course of the operation, fluids may be required for a longer period.

The drain may be removed after 48 hours if there is no urine leak, or shortened at 5 days if urine leak has occurred.

Antibiotics are administered throughout the postoperative period and continued prophylactically if vesicoureteric reflux is present. Where a delayed cystogram is planned, reflux must be assumed to be present until the examination has been performed.

Follow-up

The patient should remain on low-dose antibiotics until follow-up investigations, which include plain X-ray, ultrasound, and intravenous pyelogram (IVP)/DMSA, confirm the complete absence of calculi.

In children with no metabolic abnormality whose kidneys have been completely cleared of calculi, review is required only for a period of 2 years. Provided the urine remains sterile, recurrences are rare. Surgery may occasionally be required to correct vesicoureteric reflux.

FURTHER READING

Coward RJM, Peters CJ, Duffy PG, et al. Epidemiology of paediatric renal stone disease in the UK. *Archives of Disease in Childhood* 2003; **88**: 962–5.

Thomas DFM. Stone disease in children. In: *Essentials of Paediatric Urology*, 1st edn. London: Martin Dunitz, 2000, 113–25.

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83

Posterior urethral valve

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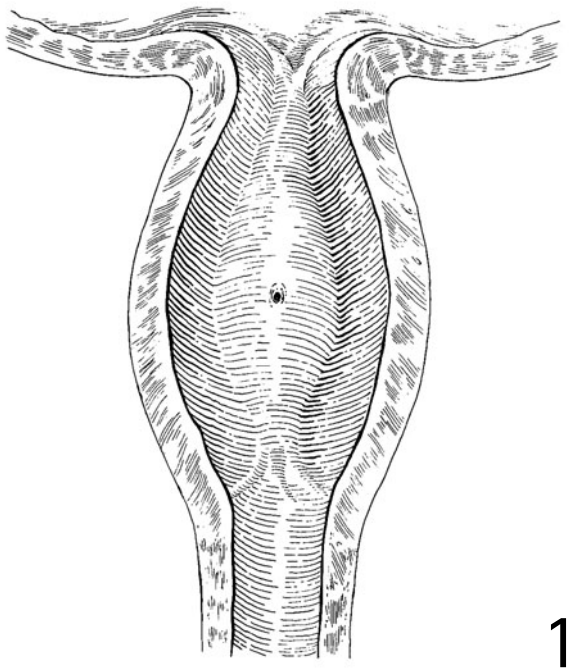
PRINCIPLES AND JUSTIFICATION

1 A posterior urethral valve is a single structure that originates from the inferior margin of the verumontanum. Although its embryology is uncertain, it lies in the position of the infracollicular folds, which can often be discerned in the normal posterior urethra running downwards from the verumontanum toward the bulb on either side of the midline.



1

2a,b When exposed at autopsy through the anterior urethral wall, a posterior urethral valve appears as two separate leaflets, but on endoscopy these are seen to fuse anteriorly to form a curtain where most of the obstruction occurs.

Most valves are thin, filmy structures that balloon downwards during voiding, but a few are thicker and more rigid, forming a transverse obstruction in the mid-posterior urethra. As all true valves originate from the inferior aspect of the verumontanum, Young's classification should be regarded as only of historic interest.

Minor degrees of valve are sometimes encountered, in which the two leaflets blend with the lateral urethral wall. They are more properly regarded as prominent infracollicular folds. It is unlikely that they ever cause symptoms or obstruction, and they do not require treatment.

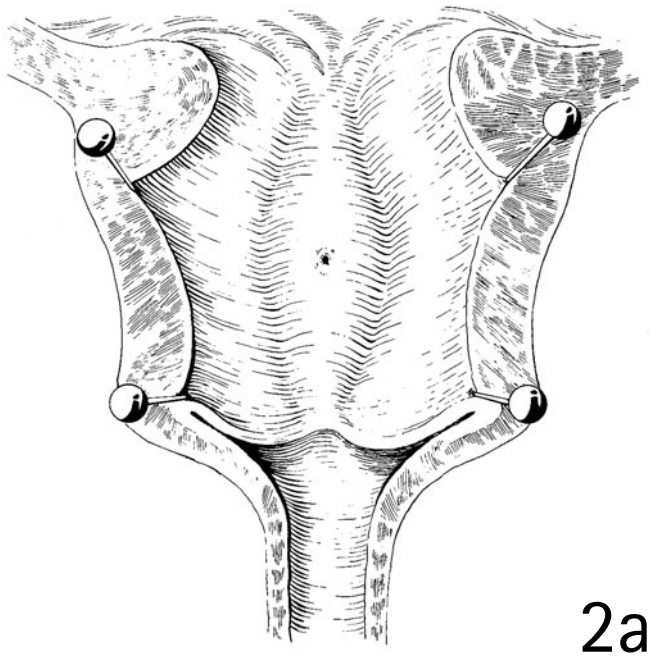
Above the valve, back-pressure effects are nearly always present; these are a widely dilated posterior urethra, a thick-walled and usually trabeculated bladder, widely dilated tortuous ureters, and bilateral hydronephrosis, which is usually symmetrical. Vesicoureteric reflux is common and often associated with a variable degree of dysplasia of the affected kidney.

The bladderneck is always thickened as part of detrusor hypertrophy, but this hardly ever causes obstruction or requires treatment.

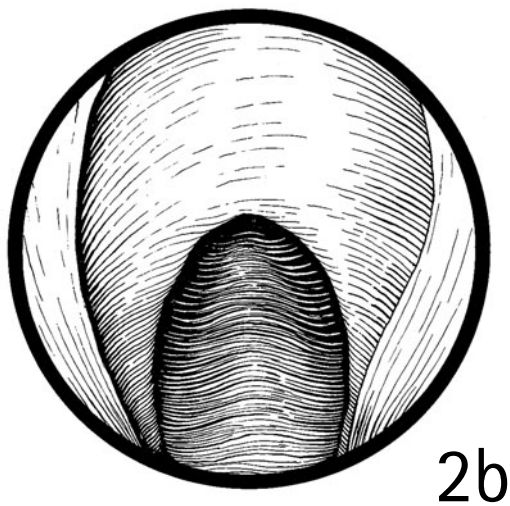
Nowadays, the diagnosis is usually made either before birth as a result of antenatal ultrasonography, or immediately afterwards because of a persistently palpable bladder. Urinary ascites is an occasional presentation in the first weeks of life.

Infants in whom the diagnosis has been missed usually present with urinary infection and acute or chronic renal failure. This is generally accompanied by hyperkalemia and a severe metabolic acidosis, which may lead to respiratory arrest. Water and sodium balance is often also profoundly disturbed. Septicemia is common and may be complicated by a consumptive coagulopathy. Older boys may also present with urinary infection, but often the main complaint is of a poor stream with straining or urinary incontinence.

The diagnosis is usually suspected on clinical grounds and supported by the ultrasonographic findings of a widened posterior urethra, distended bladder, and dilated upper urinary tract.



2a



2b

PREOPERATIVE

On suspicion of the diagnosis, an 8 Fr plastic infant feeding tube should be passed transurethrally and secured for continuous bladder drainage. Self-retaining catheters should be avoided, as the hypertrophied bladder tends to clamp down around the balloon and obstruct the ureters.

It is essential that the bladder drains well, and failure to do so is usually because the catheter has curled up in the dilated posterior urethra. Withdrawing the catheter for a few centimeters and re-passing it with a finger in the rectum will usually ensure its passage through the hypertrophied bladderneck. Persistent difficulty can usually be resolved by injecting a few milliliters of contrast medium through the catheter and manipulating it under fluoroscopic control.

A full blood count including platelets, plasma electrolytes, creatinine, and acid–base status should be determined, and severe derangements, particularly hyperkalemia or a severe metabolic acidosis, should be corrected as a matter of urgency. An assessment should also be made of the infant's state of hydration. In difficult cases, the aid of a pediatric nephrologist should be sought.

When the urine appears to be infected, both a blood sample and a urine sample should be sent for culture, following which ampicillin and an aminoglycoside or a third-generation cephalosporin should be started intravenously. When septicemia is suspected, blood coagulation studies should also be carried out.

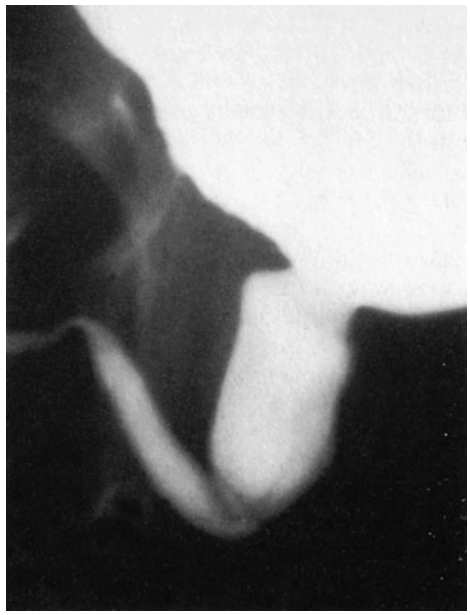
All infants with any respiratory distress should undergo chest radiography to exclude a pneumothorax secondary to pulmonary hypoplasia.

In most cases, the above actions will result in a rapid improvement in the infant's metabolic state and general condition. Those infants who remain in a toxic state or whose plasma creatinine does not begin to fall within 24 hours, despite correction of other metabolic abnormalities, should be considered for percutaneous drainage of both kidneys.

3a,b The presence of a posterior urethral valve should be confirmed by micturating cystourethrography, but this should be delayed until urinary infection has been brought completely under control and metabolic disturbances have been corrected.

A voiding film taken in the steep oblique projection during full micturition is necessary to demonstrate the valve, which appears like a spinnaker sail billowing out before the stream. Distal to this, a thin stream will be seen emerging from the posterior margin of the obstruction. Dilatation of the urethra proximal to the valve is essential to the diagnosis, and signs of bladder wall hypertrophy are usually also present. A very lax valve may occasionally prolapse down as far as the bulbar urethra (Illustration b). Here, the posterior run-off may not be readily apparent, but the filling defect caused by the valve leaflets can usually be made out running down from the verumontanum.

3a

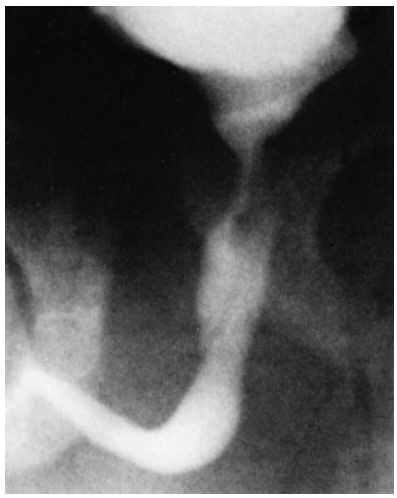


3b

4a-h A variety of other conditions may masquerade as a posterior urethral valve on the cystogram, and failure to recognize them often leads to inappropriate treatment. Among these are prominent infracollicular folds, which can sometimes be made out on a good-quality study in normal children (Illustration a).

Hesitant voiding in a normal baby may cause an abrupt change in caliber of the posterior urethra, while extrinsic compression by the pelvic floor may cause one or more concentric indentations in the urethral contour (Illustration b). Neither of these is associated with evidence of obstruction above the lesion, however, and both should be regarded as normal variants.

A neuropathic bladder may closely simulate a posterior urethral valve (Illustration c), but the thin stream below the obstruction will be seen emerging from the center of the external urethral sphincter rather than from the posterior margin, as seen with a valve. In such cases, the spine should be carefully examined and other evidence sought of a neurologic deficit in the perineum or lower limbs.



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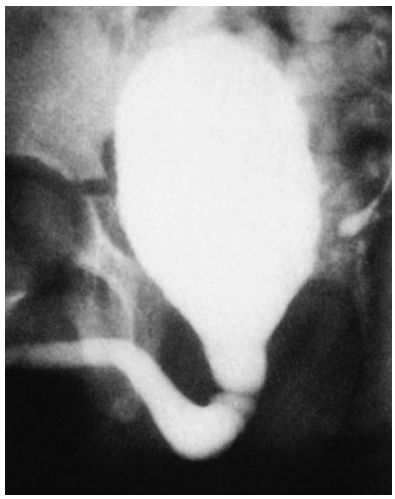

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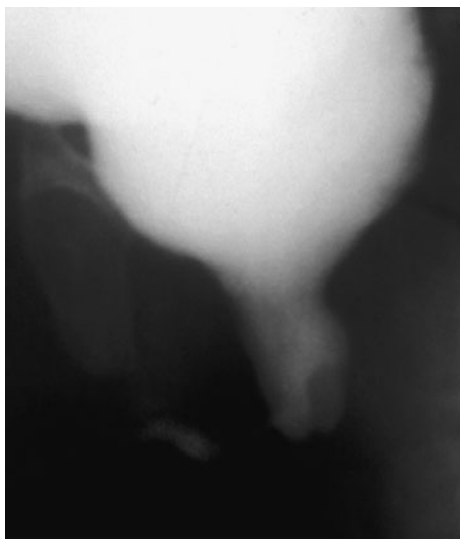
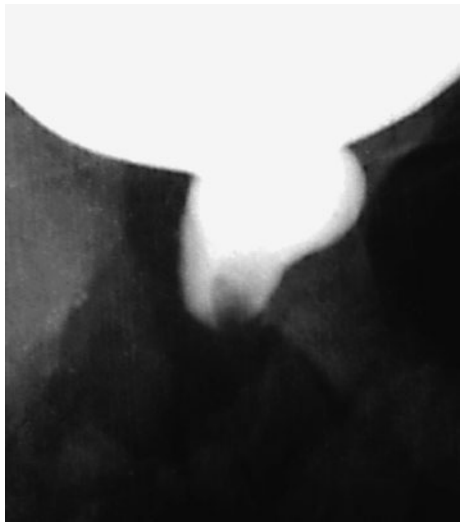


4c

A posterior urethral stricture may cause a similar appearance (Illustration d), but this will usually be associated with a history of urethral or pelvic trauma.

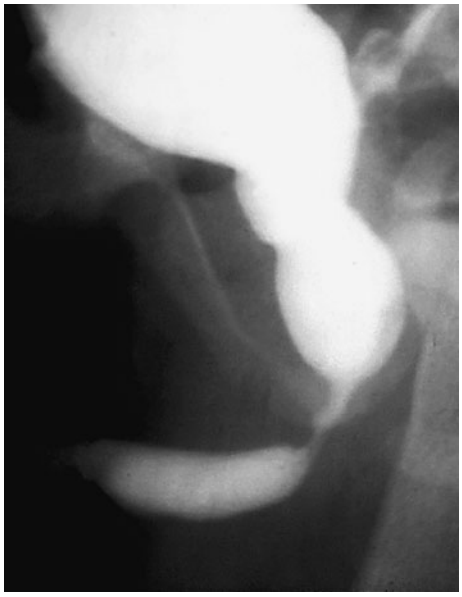
The prune-belly syndrome may closely mimic a posterior urethral valve (Illustration e), but the correct diagnosis should be suspected from the appearance of the bladder, which lies horizontally and is invariably smooth walled, and the dog-leg configuration of the posterior urethra, which often bears a utriculus masculinus.

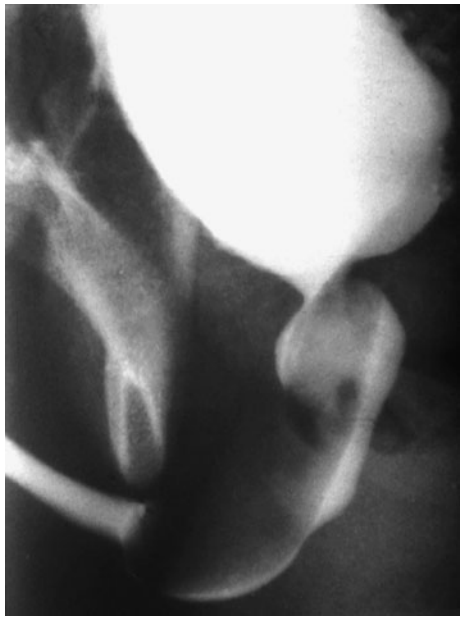
A distended, non-visualized ectopic ureter opening into the ejaculatory duct may distort and partially obstruct the posterior urethra and thus simulate a valve (Illustration f), while dilatation of the posterior urethra may also be caused by a prolapsed ectopic ureterocele (Illustration g) or posterior urethral polyp (Illustration h). Careful examination of these films, however, will usually reveal a filling defect, leading to the correct diagnosis.



4f

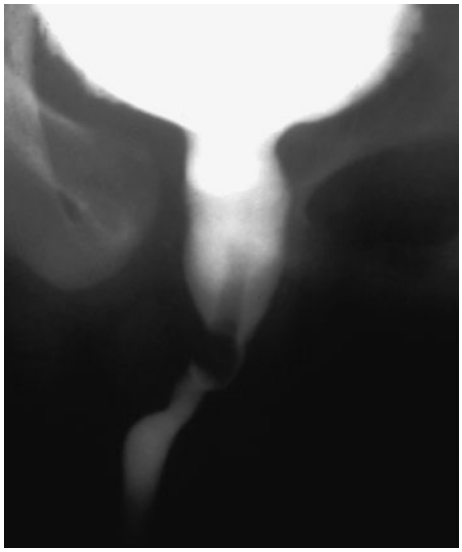
4d





4g

4e



4h

OPERATION

Resection in full-term infants and children

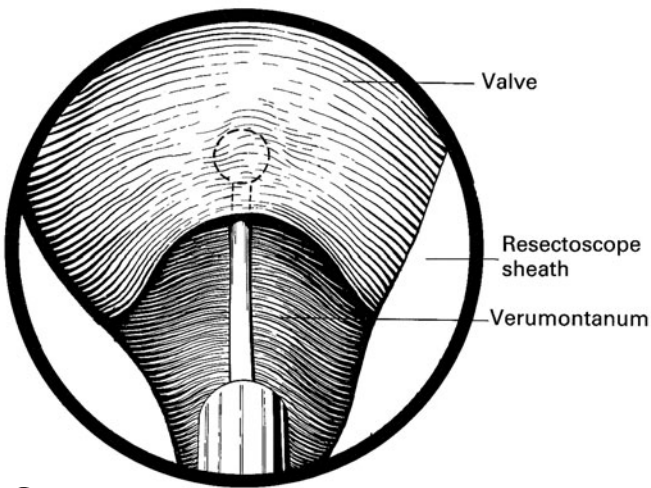
Under endotracheal anesthesia, the intubated infant is placed supine with the buttocks brought well down to the end of the operating table. The legs should be well protected with cotton wool and fixed with crepe bandages, either to pediatric stirrups or in the frog-leg position, taking care to provide ample support to the thighs. The skin is prepared and drapes applied, taking care to exclude the anus from the operative field. Fixing the posterior towel to the perineal skin with three staples or 4/0 nylon sutures will ensure that the anus does not become exposed during subsequent manipulations.

The caliber of the penile urethra should first be checked with a well-lubricated 8 Fr sound, which should be introduced only for 1–2 cm. If necessary, a meatotomy can be performed, but no attempt should be made to dilate the urethra. The diagnosis is then confirmed using a well-lubricated 6.5 Fr or 9.5 Fr cystoscope introduced under vision.

5 Resection of the valve is undertaken using a pediatric resectoscope fitted with a hooked ball electrode. (Storz; Wolf).

The instrument is first assembled and the alignment of the working parts checked using the 0° telescope. The sheath is then dried and thoroughly coated with a water-soluble lubricant, and with its introducer in place is gently inserted through the meatus. The introducer is removed and the instrument reassembled and gently advanced under vision towards the bladderneck. It is usually necessary to angle the eyepiece end of the instrument downwards to allow the beak to move anteriorly and pass through the bladderneck. Once in the bladder, the shape and position of the ureteric orifices are noted and the presence of any periureteric diverticulum recorded.

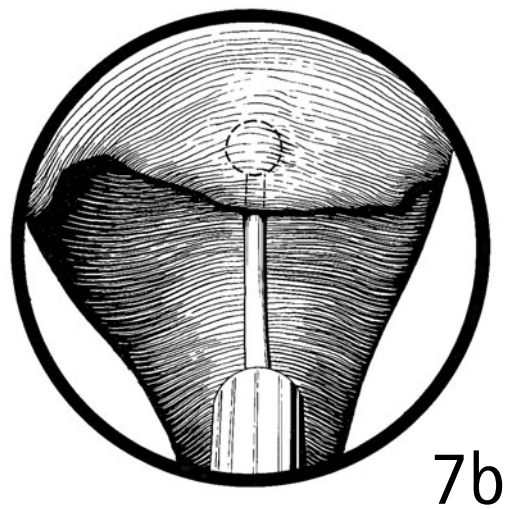
5



6

6 The instrument is now rotated through 180°, and with the irrigation fluid flowing in under low pressure, it is progressively withdrawn. Once through the bladderneck, the ball is run down along the anterior wall of the posterior urethra until, just beyond the verumontanum, the valve suddenly snaps across the anterior portion of the field of view like a curtain. Further withdrawal of the instrument and manipulation of the trigger will cause the ball to engage the valve in the 12 o'clock position. A short burst of cutting current is then applied.

7a



7b

7a,b The instrument is returned to the normal position and advanced under vision back into the bladder. It is again rotated 180° and withdrawn into the posterior urethra to engage the now partially disrupted valve in the 12 o'clock position, where it is further disrupted. This maneuver should be repeated until it is certain that the anterior portion of the valve has been completely ablated.

The instrument is again returned to the bladder and is rotated to engage residual valve tissue in the 10 o'clock, 2 o'clock, 8 o'clock, and finally the 4 o'clock positions. Any remaining freely floating tags do not require treatment.

The resectoscope is removed and the presence of an unobstructed urethra confirmed by manual expression of the bladder. Finally, an 8 Fr feeding tube is passed, placing a double-gloved finger in the rectum, if necessary, to ensure that it is not curled up in the dilated posterior urethra. This is retained in place with a 4/0 nylon suture passed through the prepuce or distal shaft skin and connected to a sealed drainage bag. The tube is removed after 48 hours.

If any significant bleeding occurs, attempts at valve ablation should be immediately discontinued and the situation reassessed after 2–3 days of catheter drainage.

In older children a 13 Fr resectoscope may be used, employing a similar technique.

Resection in preterm infants

It is inadvisable to attempt to pass a 9.5 Fr resectoscope in infants weighing less than 2.5 kg. In such cases, a few days of bladder drainage, using initially a 6 Fr urethral feeding tube, will have the effect of gently dilating the urethra so that a 6.5 Fr or even a 9.5 Fr resectoscope can be safely used. Alternatively, a very small cystoscope may be employed, introducing through the working channel a 3 Fr ureteral catheter with its tip cut off. The exposed end of a metal stylet is then used to coagulate the valve in a circumferential fashion.

Other techniques are also available for resecting the valve in a very small infant, but all have some disadvantages. The Whitaker hook electrode is a slender, insulated metal instrument that can be introduced through the urethra under fluoroscopic control and withdrawn to engage the valve. Short bursts of cutting current may relieve the obstruction, but the procedure is essentially blind and carries the risk of urethral trauma.

A Fogarty catheter with the balloon inflated with 0.1–0.3 mL of water has also been used to disrupt the valve, but carries the risk of avulsion of the urethra.

Access to the valve with the pediatric resectoscope sheath can usually be achieved via a perineal urethrotomy. The small caliber of the urethra and the friable nature of the urothelium, however, render the operation difficult in the neonate and it may be complicated by bleeding, a persistent urinary fistula, urethral diverticulum, or stricture.

An alternative approach is to create a suprapubic cystotomy through which the valve can be resected in an antegrade fashion. Using a 10 Fr sheath and the hooked ball electrode, the valve is first engaged in the 12 o'clock position and coagulated. However, the hypertrophied bladderneck sometimes closes across the telescope lens so that the procedure has to be carried out blindly.

Vesicostomy

In very small infants, it is the author's preference to carry out a vesicostomy rather than attempt to disrupt the valve by the above methods. A few months later, when the infant has reached an adequate size, the valve is coagulated through the urethra using the hooked ball electrode in the standard fashion. The vesicostomy is closed at the same time and the urethral catheter removed 10 days later.

Upper tract drainage

Following relief of the urethral obstruction, correction of metabolic derangements, and eradication of infection, the plasma creatinine will in most cases rapidly fall to within the normal range for the patient's age. When it remains elevated, the possibility of obstruction of the dilated flaccid ureters as they pass through the hypertrophied bladder wall must be considered. In most cases, this phenomenon is transient and, provided that the infant remains well and the plasma creatinine is showing some improvement, an expectant policy may be adopted.

A persistently elevated or rising serum creatinine following successful valve ablation may be the result of infection, hyponatremia, or aminoglycoside toxicity. However, when these causes have been excluded, percutaneous nephrostomies should be carried out. A subsequent fall in the serum creatinine will confirm the diagnosis of obstruction at the ureterovesical junctions. After 2 weeks, the tubes are clamped. If the serum creatinine remains low, they are removed. When this results in a rise of the serum creatinine, Sober Y cutaneous ureterostomies should be carried out to provide optimal tube-free upper tract drainage. The cutaneous limb can then be tied off when the infant is ready to come out of diapers. Alternatively, the ureters may be remodeled and re-implanted, but this operation is rendered difficult by the thickness of the bladder wall and trabeculation, and in inexperienced hands complications are common.

Failure of the serum creatinine to fall after placement of bilateral percutaneous nephrostomies is indicative of irreversible renal dysplasia.

POSTOPERATIVE CARE

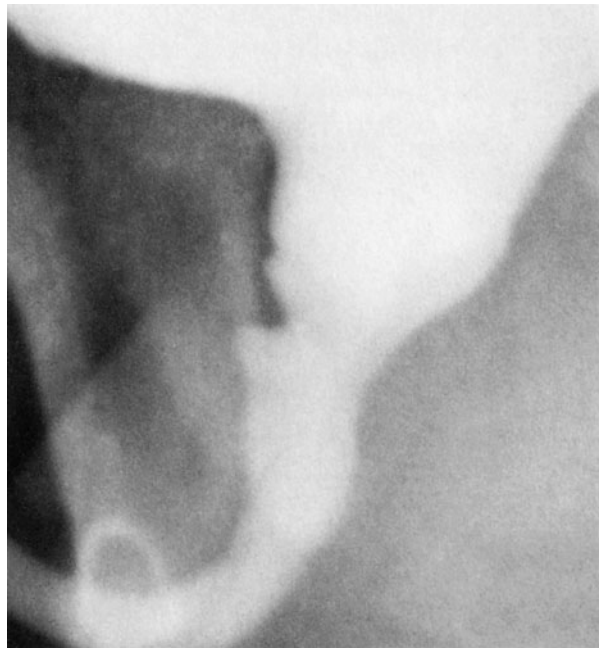
Following removal of the urethral catheter, adequate emptying of the bladder should be confirmed clinically or by ultrasonography. Postoperative antibiotic prophylaxis, usually with trimethoprim-sulphamethoxozole, is continued for 1 month to guard against infection in the healing posterior urethra. In infants in whom preoperative cystography revealed the presence of vesicoureteric reflux, this should be continued for 6 months. Sodium bicarbonate supplements are also often necessary to correct a persistent metabolic acidosis, and these may need to be given for 1 year or more. Polyuria is also common, and the parents should be advised to give supplementary clear feeds early in the event of a diarrheal illness.

At 3 months, the glomerular filtration rate of each kidney is measured by the slope clearance method using ^{99m}Tc-DTPA (diethylenetriaminepenta-acetate), and intravenous urography is carried out. Both of these will serve as a baseline for any future studies. A blood sample is also taken to check the plasma creatinine, electrolytes, and acid–base status.

8 At 6 months, micturating cystourethrography is repeated to confirm adequate resection of the valve and the absence of any stricture of the urethra, and to determine whether any previously noted vesicoureteric reflux is still present. In about one-third of cases, it will be found to have disappeared. When reflux is persistent and unilateral, renal scanning using ^{99m}Tc-DMSA (dimercaptosuccinic acid) is carried out to determine the contribution of the kidney on the refluxing side to total renal function. When this is negligible, nephroureterectomy should be carried out through two incisions to ensure safe ligation of the ureteric stump. Ureteral reimplantation should be considered if the reflux persists and the kidney is useful, particularly when urinary infections supervene.

Urinary infections occurring during childhood after successful valve ablation are often due to incomplete bladder emptying. Double or triple micturition should be tried before instituting clean intermittent catheterizations.

All infants and those older children with impaired renal function at presentation will require close supervision until adult life is reached. A progressive rise in plasma creatinine is often seen during childhood, and in the most severe cases, renal transplantation may be required before puberty. Persistent urinary incontinence is an indication for cystometrography. Bladders showing severe hyperreflexia or very poor compliance generally require augmentation, which should be carried out before transplantation.



8

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84

Hypospadias repair

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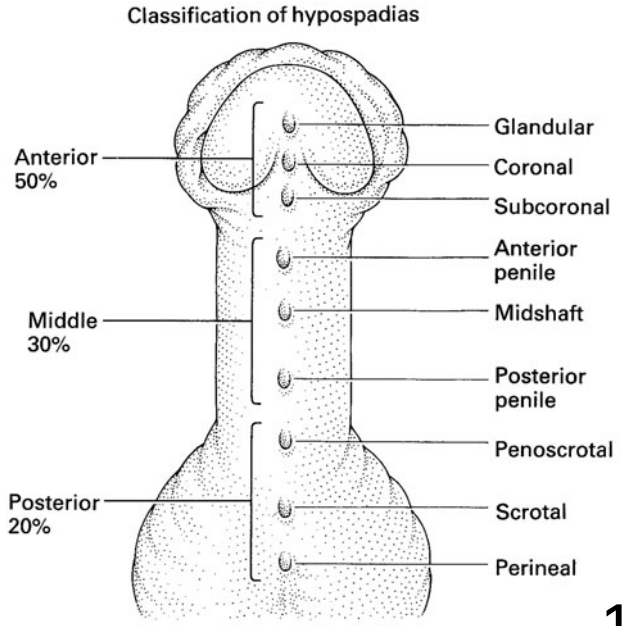
Hypospadias is classically defined as an association of three congenital anatomic anomalies of the penis: (1) an abnormal ventral opening of the urethral meatus, (2) an abnormal ventral curvature of the penis (called chordee), and (3) an abnormal dorsal 'hood' distribution of the foreskin with ventral deficiency. The diagnosis of hypospadias is evident on newborn examination, but in some instances of mild hypospadias such as the megameatus-intact prepuce (MIP) variant, in which the foreskin is normal, detection of hypospadias might not occur until the foreskin is retracted.

1 The most common classification system employs anatomic description of meatal position. Glanular, coronal, and subcoronal positions constitute the majority of cases (approximately 70 percent), but hypospadias can occur in more severe forms with the urethral meatus opening near the scrotum and perineum.



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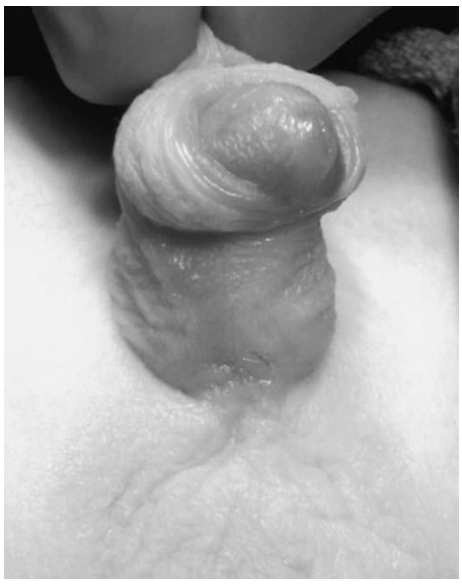


Fig. 84.1 Mild hypospadias in which the urethral meatus is distally located and the penile shaft is associated with mild chordee and ventrally deficient prepuce.

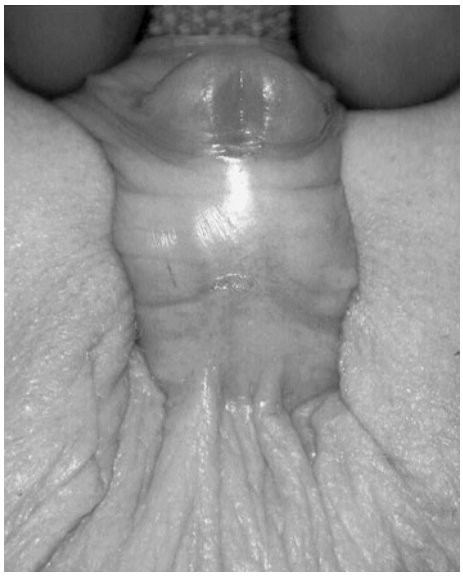


Fig. 84.2 Moderate hypospadias in which the urethral meatus is subcoronal. There is a deep glanular groove distal to the meatus (called the urethral plate), and the prepuce is deficient ventrally.

INCIDENCE AND ASSOCIATED CONDITIONS

Hypospadias has been reported to occur in approximately 1 out of 100–300 live male births. Recent studies have suggested that the incidence of hypospadias may be on the rise. Interestingly, the rate of severe hypospadias seems to have increased three-fold to five-fold over the last two decades.

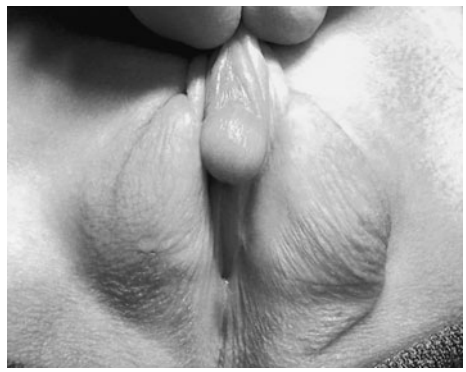


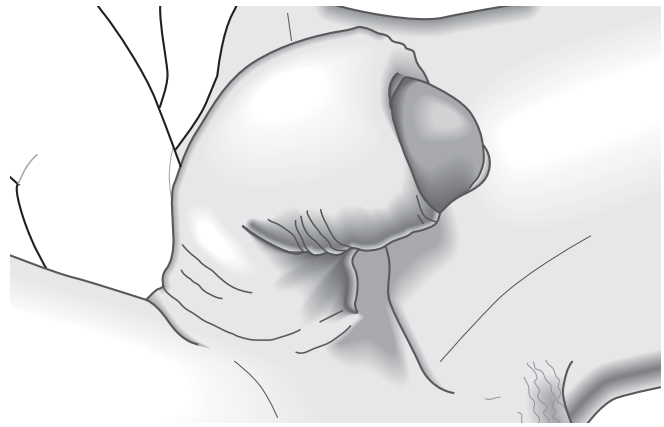
Fig. 84.3 Severe hypospadias with the urethral opening in the perineum.

This trend could reflect more frequent reporting and earlier diagnosis, or it may reflect other undefined biological factors such as environmental endocrine disruptors during pregnancy. Hypospadias is probably an inherited congenital defect, as it occurs in 6–8 percent of fathers of affected boys and in 14 percent of male siblings.

Associated anomalies include cryptorchidism (8–9 percent) and inguinal hernia/hydrocele (9–16 percent). The rate of associated genital anomalies increases in patients with more severe proximal defects. An intersex condition should be considered in patients with concomitant hypospadias and cryptorchidism. Patients with severe hypospadias may also have a significantly enlarged utricle, which may serve as nidus for urinary tract infections and cause voiding difficulties.

Galen (130–199 AD) was the first to use the term 'hypospadias' and the first to emphasize the major significance of associated penile curvature. Early on, the ventral curvature was thought to be caused primarily by fibrous tissues encasing the hypospadiac urethra, thereby tethering and bowing the penis, and this led to the proposal that the most important part of the chordee correction was aggressive excision of so-called 'chordee tissues'. Penile embryology and anatomic studies have shown that the etiology of chordee is much more complex, and it includes (1) abnormal development of the urethral plate (the mucosa-lined longitudinal groove between the urethral meatus and glans), (2) abnormal, fibrous tissue at the urethral meatus, and (3) corporal disproportion between dorsal and ventral cavernosal tissues. The concept of corporal disproportion is particularly important in that a routine division and lifting of the urethral plate is no longer accepted as the principal maneuver in chordee correction. Indeed, the preservation of the urethral plate for neourethra reconstruction along with corporal repair for chordee correction constitutes a fundamental approach in modern hypospadias repair techniques.

2 Although chordee is commonly seen in association with hypospadias, it can occur in isolation with orthotopically located urethral meatus. In some cases, this is caused by a simple skin tethering, representing a simple, straightforward surgical problem. In other instances, the chordee is a form fruste of hypospadias in that more extensive anatomic abnormalities coexist, such as thin, dysplastic urethra and disproportionate corpora, which require a significant reconstruction.



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PRINCIPLES AND JUSTIFICATION

The primary goal of hypospadias repair is to restore normal masculine 'stand-up' urination and sexual/reproductive function. The need for surgical reconstruction is obvious in patients with severe hypospadias and chordee. Early on in the history of hypospadiology (the term coined by the late John W. Duckett Jr for 'the in-depth study of the art and science of the surgical correction of hypospadias'), unsatisfactory results of repair led to the view that mild forms of hypospadias should be managed by 'masterly neglect'. With the advances in anatomic understanding and surgical technique over the last two decades, however, the bar of expectation has been raised high, in terms of both functional and cosmetic outcomes.

PREOPERATIVE ASSESSMENT AND PREPARATION

Preoperative evaluations

In general, the literature does not support routine imaging of the urinary tract with either ultrasonography or intravenous urography for hypospadias patients. In patients with a severe defect such as perineal hypospadias, lower urinary tract evaluation with either endoscopy or voiding cystourethrography will define the utricle size and reveal concurrent anomalies. Routine endocrinologic, cytologic, and other evaluations for intersex in patients with isolated hypospadias are probably unnecessary unless the defect is severe or associated with cryptorchidism.

Hormonal manipulations

There is considerable disagreement regarding the use of hormonal stimulation prior to hypospadias repair for the purpose of penile enlargement. Suggestions were made that preoperative human chorionic gonadotropin treatment may decrease the severity of hypospadias and increase the vascularity and thickness of proximal corpus spongiosum, thereby allowing more simple repairs. Preoperative testosterone was shown to increase penile size as well as improve skin availability and local vascularity. Hormonal supplementation may be useful in children with microphallus and with previously failed repairs, in which local tissues are deficient.

Timing of surgery

A report by the American Academy of Pediatrics suggested that the best time for surgery for hypospadias may be between 6 and 12 months of age. This recommendation was made based on a number of factors, including the psychologic effects of genital surgery in children, improved technical aspects of hypospadias surgery, and advances in pediatric anesthesia. Postoperative management issues such as the care of neourethral catheters and wound dressings are significantly easier and safer in infants as well.

Anesthesia

In young children, general anesthesia with endotracheal intubation is the most reliable approach. Adjunctive analgesia using long-acting, injectable, nerve-blocking agents such as bupivacaine, delivered via either the caudal route or penile block, is safe and efficacious.

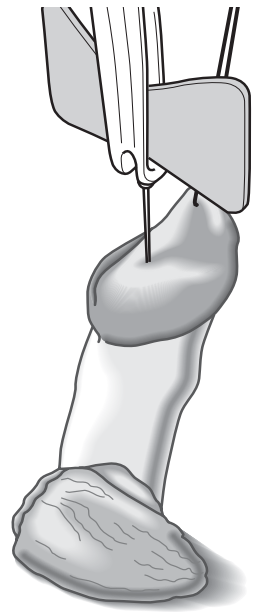
TREATMENT PRINICPLES

Routine perioperative antibiotics are not necessary, unless the urethra and bladder are intubated postoperatively. Hemostasis must be accomplished with precision, applying judicious cauterization, as ischemic tissue necrosis will lead to the breakdown of repair, infection, and urethrocutaneous

fistula. Careful use of vasoconstrictive agent (epinephrine diluted to 1:200 000) around the region of the corona and glans will also reduce the amount of bleeding intraoperatively. A tourniquet may be applied to the base of penis during certain parts of the surgery, but it must not be used for a prolonged time. Most surgeons rely on optical magnification to enhance surgical precision.

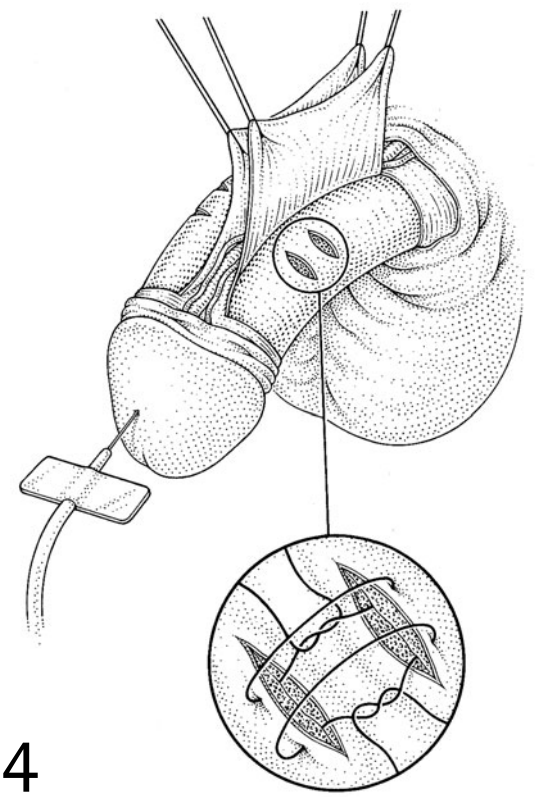
Orthoplasty (chordee correction)

The degree of ventral curvature influences the type of hypospadias repair. Even if the urethral opening is not severely ectopic, the penis may require an extensive reconstruction (even a staged one) in order to accomplish adequate straightening of the phallus. Preoperatively, gentle retraction of the penile base allows decent assessment of the location and severity of overall chordee. At times, release of tethering ventral skin may be all that is required for orthoplasty. In this situation, some of the dorsal prepuce and penile skin may need to be transposed ventrally to make up for the skin deficiency.



3

3 Intraoperative evaluation of chordee using an artificial erection test, as described by Gittes and McLaughlin, is critical. After the degloving of the penile shaft, injection of one of the corpora cavernosa is performed directly using 5–10 mL of saline solution with a small 23 gauge butterfly needle. Alternatively, the needle may be passed through the glans and into the tip of a corpus in order to minimize hematoma beneath the Buck's fascia.



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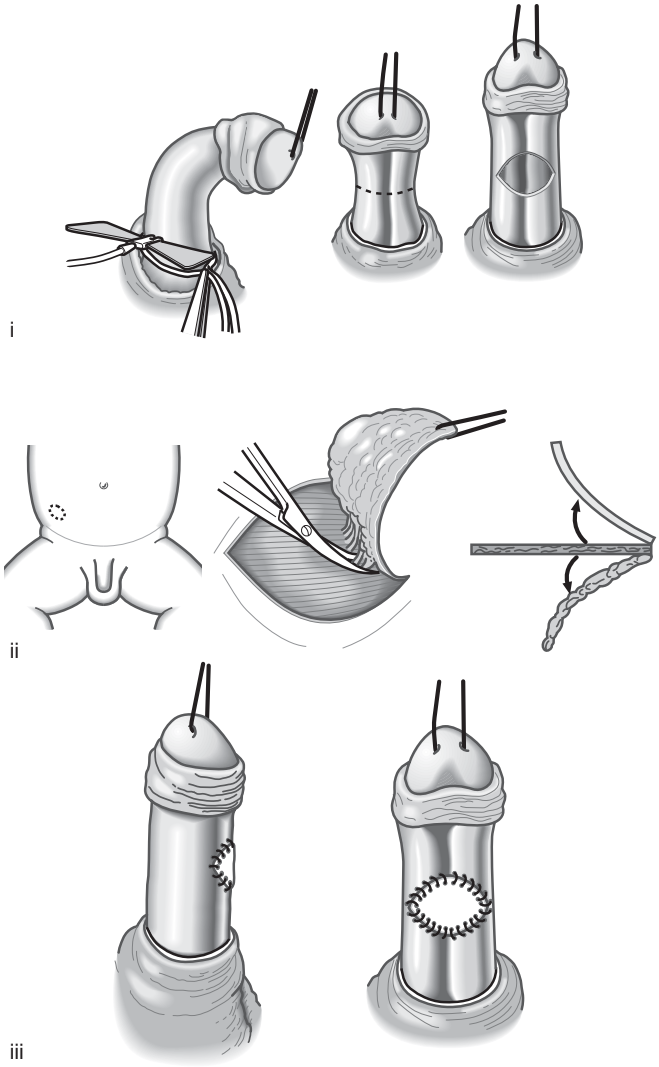
4 One of the most reliable techniques of orthoplasty is dorsal corporal plication, as described by Nesbit. Nesbit's original description involved mobilization of the dorsal midline neurovascular bundle, followed by making an elliptical excision of the dorsal tunica albuginea vertically and approximating these defects horizontally (the Heineke–Mikulicz principle). In an extension of this concept by Baskin and Duckett, two parallel lines of incision are made on the dorsolateral aspects of each tunica albuginea of each corpus at the point of maximal curvature, and the outer edges of the incisions are approximated using buried sutures.

5a–c In patients with severe curvature or with short phallic length, the dorsal plication technique may not be optimal because of potential shortening, and ventral transverse incision of the tunica albuginea and patch grafting may be more suitable. The corporal defect created after the incision is then covered with de-epithelialized dermis harvested from the lower abdominal wall or tunica vaginalis, or a synthetic graft in older patients. At the level of greatest curvature, a deep transverse incision is made ventrally, exposing the cavernosal tissue. This incision must extend from one lateral corner to the other. A de-epithelialized elliptical patch of skin from the inguinal crease is sewn to the tunica albuginea edges using a running 5/0 Prolene suture.



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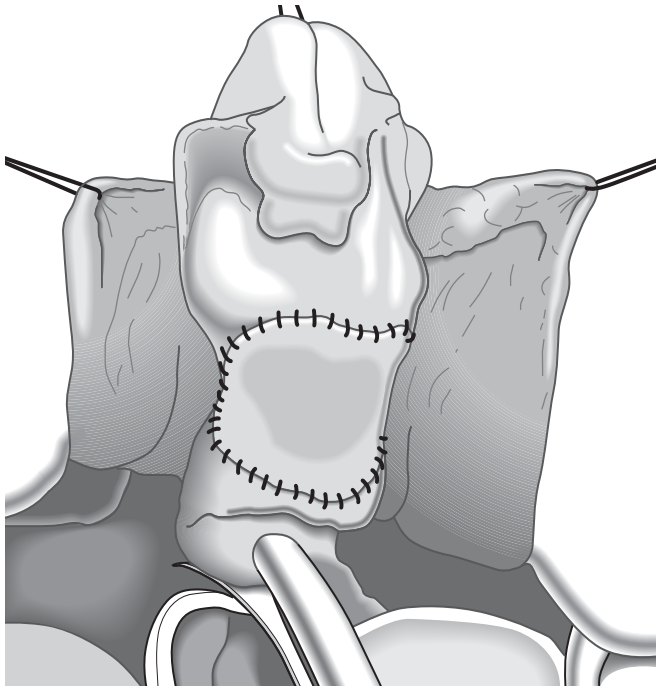

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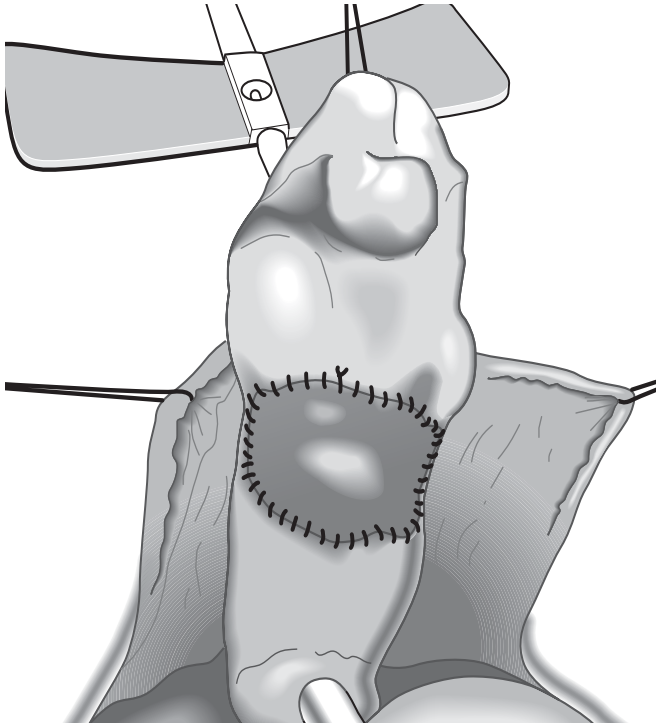


5c

Hypospadiac penis may be associated with counterclockwise penile torsion. In mild cases, this may be corrected simply by rotating the penile skin coverage, but in more severe cases additional correction may be required, such as corporal plication and dorsal dartos flap rotation.

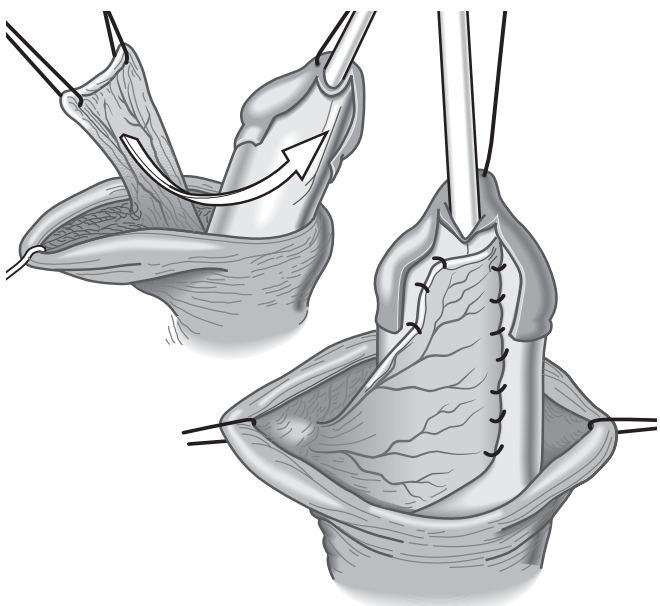
Urethroplasty

An important surgical principle in neourethra formation is meticulous tissue transfer. In urethroplasty, the tissue source may be adjacent tissue, local tissue flaps, or free grafts of either genital or extragenital tissue. The neourethra may be created by simply tubularizing the native urethral plate, especially if there is a deep glanular groove. Biologically speaking, the urethral plate contains epithelial lining with urethral mucosal differentiation and will perform quite well as neourethra, provided that tubularization can occur without suture-line tension and meatal stenosis. Local tissue flaps employed for urethral reconstruction must be thin, non-hair bearing, and easy to handle. They are often derived from distal penile shaft skin or prepuce, and they are called fasciocutaneous flaps because of their reliance upon the dartos fascia serving as the conduit for vascular supply and drainage. The term graft implies that tissue has been excised from one location and transferred to another site, where a new blood supply develops. The initial phase, called imbibition, relies on diffusion of oxygen and nutrients from the graft host site (approximately 48 hours), and it is followed by the second phase, called inosculation, which is the formation of new blood vessels.

Neourethral coverage

It is critical to establish a second-layer coverage of the urethroplasty suture line prior to skin closure in order to decrease the risk of urethrocutaneous fistula formation. Not only does it provide additional protection during the healing of reconstructed urethra, but also the ventrally mobilized flap improves the overall functional and cosmetic outcome. Various vascularized flaps may be used for this purpose, including dartos, tunica vaginalis, and periurethral corpus spongiosum.

6a,b The dorsal prepuce is unfolded, and the subcutaneous dartos flap is sharply dissected off the undersurface of the penile skin down to the base. To prevent penile twisting, the flap may be divided down the middle, preserving the vessels, brought around the shaft to the ventral area, and secured over the neourethra.



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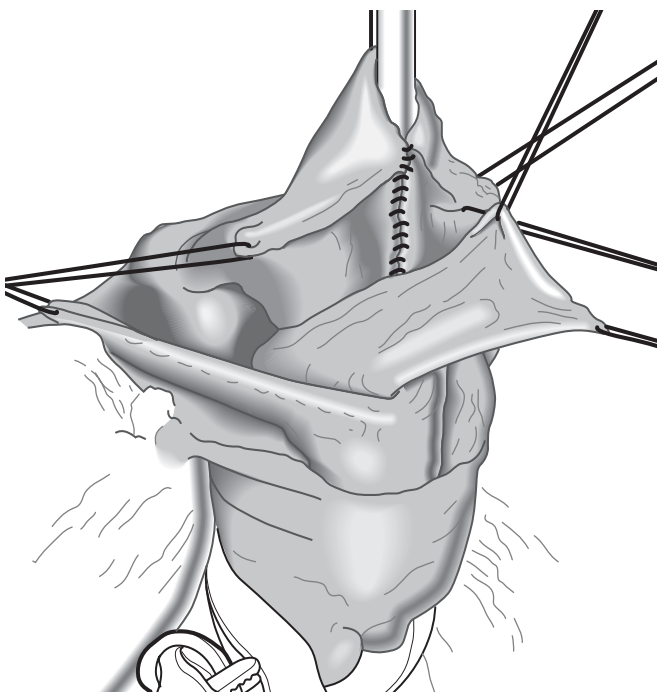
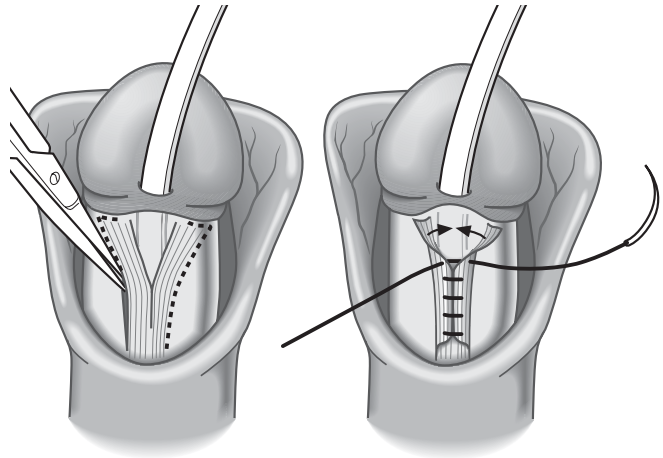


6b

7 In most cases of hypospadias, the corpus spongiosum becomes flat distal to the meatus and fans out around the urethral plate. After the urethroplasty, this layer of tissue can be dissected and approximated over the midline as the second layer coverage. Unlike dartos and tunica vaginalis flaps, the corpus spongiosum used in this way is less likely to cause ventral tethering and penile twisting.



Meatoplasty and glanuloplasty

All attempts at neourethral reconstruction will be for nothing if careful attention is not given to this phase of the hypospadias repair. Glans wings (the lateral flaps of glans tissue that are brought around and approximated over the middle) must be loose with minimal tension, with enough space to accommodate both the neourethra and the second-layer coverage flaps. If the glanuloplasty is performed with tension, it can lead to either a troublesome meatal stenosis or breakdown of the distal repair, resulting in a recurrence of coronal hypospadias or a 'blow-out' type of urethrocutaneous fistula, usually at the coronal location. In all techniques of hypospadias repair, the final meatal caliber must be ample (8 Fr for infants and at least 10 Fr for older children) with generous spatulation. It is best to avoid a circumferential suture line around the meatus, which predisposes to meatal stenosis.

INTRAOPERATIVE DECISION MAKING: USING THE RIGHT TISSUES AND CHOOSING THE RIGHT PROCEDURE

Decision making begins with assessment of meatal location, penile size, curvature, and the quality of ventral skin over the native urethra. Hypospadias in most patients is repairable using well-vascularized local tissues. In mild glanular hypospadias, simple glanuloplasty and meatal advancement (MAGPI) may be all that is required. In other situations, consideration is given to neourethral reconstruction using either a flap technique or urethral plate tubularization. If the urethral plate is healthy with decent width and vascularity, it may be tubularized, either with or without the help of a vertical midline incision technique (TIP/Snodgrass). If it is too narrow or too shallow for effective, tension-free tubularization, a preputial (onlay urethroplasty) or perimeatal-based proximal skin (Mathieu procedure) fasciocutaneous flap is approximated onto the urethral plate as the ventral portion of the neourethra. In some situations, the actual meatal location is distal, but the quality of distal urethra leading up to the meatus is thin and dysplastic, and there is near-absent penile skin ventrally. This type of hypospadias should be treated as a more severe proximal defect.

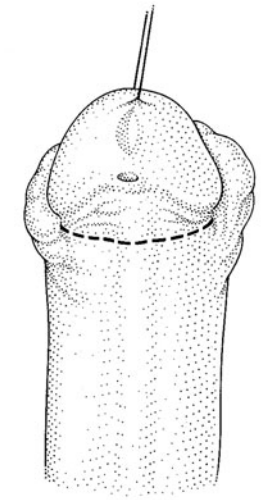
The choice of urethroplasty technique may depend on the degree of chordee. The penile shaft is degloved after making a subcoronal incision, preserving an 8–10-mm width of the urethral plate, except for severe hypospadias such as the scrotal and perineal varieties in which urethral plate preservation is unlikely from the outset. The penis is evaluated for curvature by an artificial erection test. If the curvature is severe (greater than 90°) and there is short phallic length, correcting chordee by aggressive dorsal corporal plication may not be the best option, as it may lead to shortening of the phallus. In this situation, consideration must be given to division of the urethral plate, excision of ventral fibrous chordee tissues, and placement of a corporal patch after a relaxing transverse incision of the tunica albuginea. The neourethra may then be created either at the same time or later using transposed dorsal preputial skin (staged operation), or immediately with an island pedicle flap (transverse island tube urethroplasty) or free graft (oral mucosa graft urethroplasty).

OPERATIVE TECHNIQUES

There is no single, universally applicable technique of hypospadias repair. A surgeon attempting an effective hypospadias reconstruction must be familiar with several different techniques, and understand their pros and cons, as well as their indications for specific anatomic configurations. It is unwise rigidly to apply one or two 'favorite' techniques to all situations. Instead, a given anatomic situation must dictate the procedure to be employed, and the surgeon must maintain flexibility and versatility. It is not possible within the scope of this chapter to discuss all reported techniques of hypospadias repair. The authors therefore present several of their own preferred techniques.

Meatal advancement and glanuloplasty

8a–j For the glanular and some of the coronal hypospadias without significant chordee, MAGPI is a simple yet elegant procedure with an excellent functional and cosmetic outcome. In many cases of distal hypospadias, there is a transverse glanular tissue ridge that separates the true meatus from a distal, blind-ending groove. In the first step of MAGPI, this tissue ridge is incised deeply in a vertical direction, creating a diamond-shaped defect, which is then closed in a transverse direction (the Heineke–Mikulicz principle). This maneuver widens, advances, and flattens the urethral meatus. A circumferential incision is then made in the subcoronal region, and the penis is degloved. After addressing any chordee, the glanuloplasty is performed. After ventrolateral de-epithelialization of the glans tissue proximal to the urethral meatus, glanuloplasty is performed in two layers, while gently retracting the lower lip of the urethral meatus upward.



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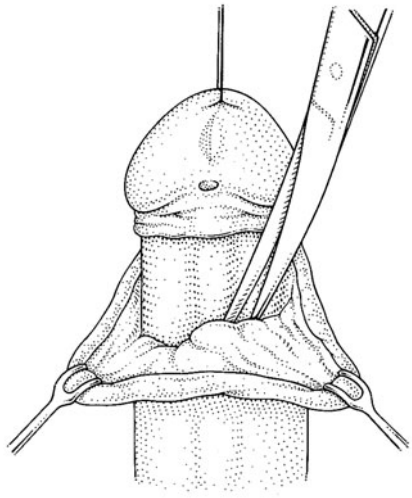

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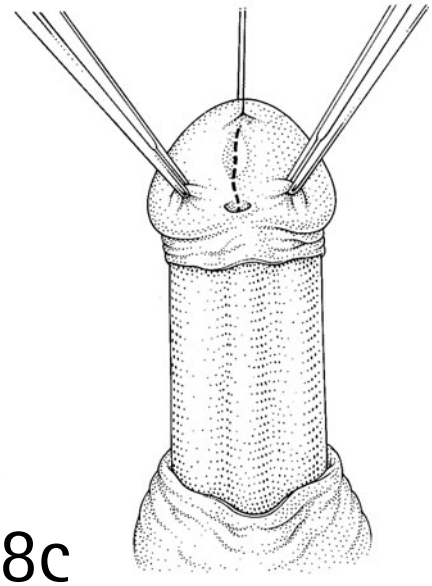

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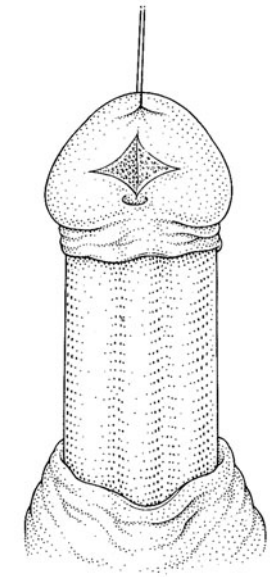

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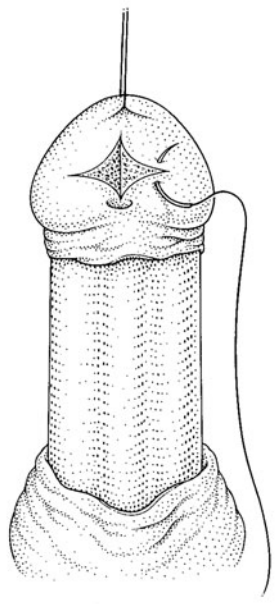

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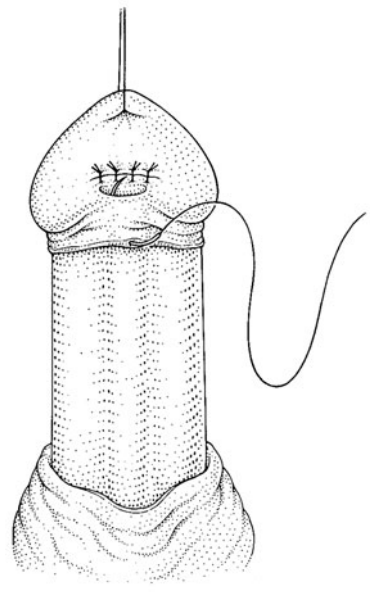

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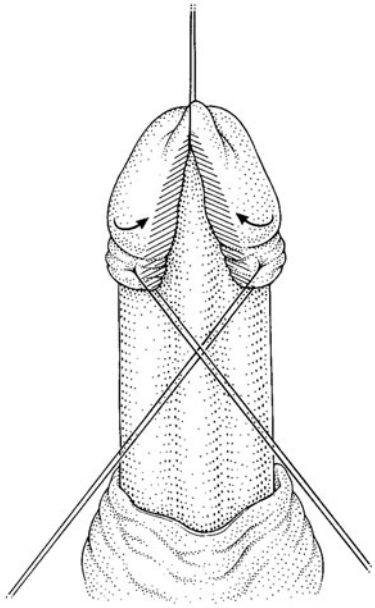

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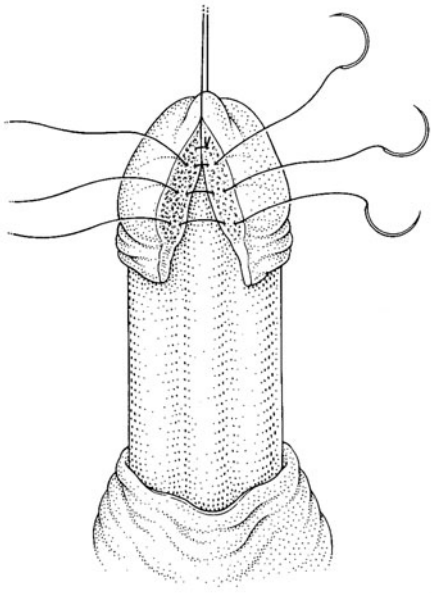

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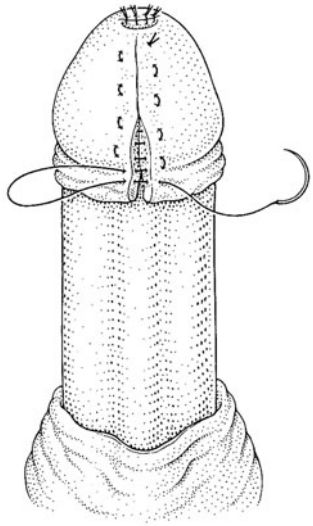

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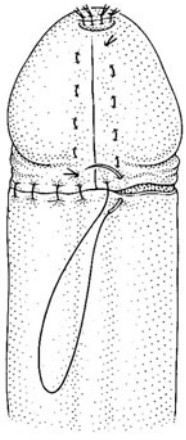
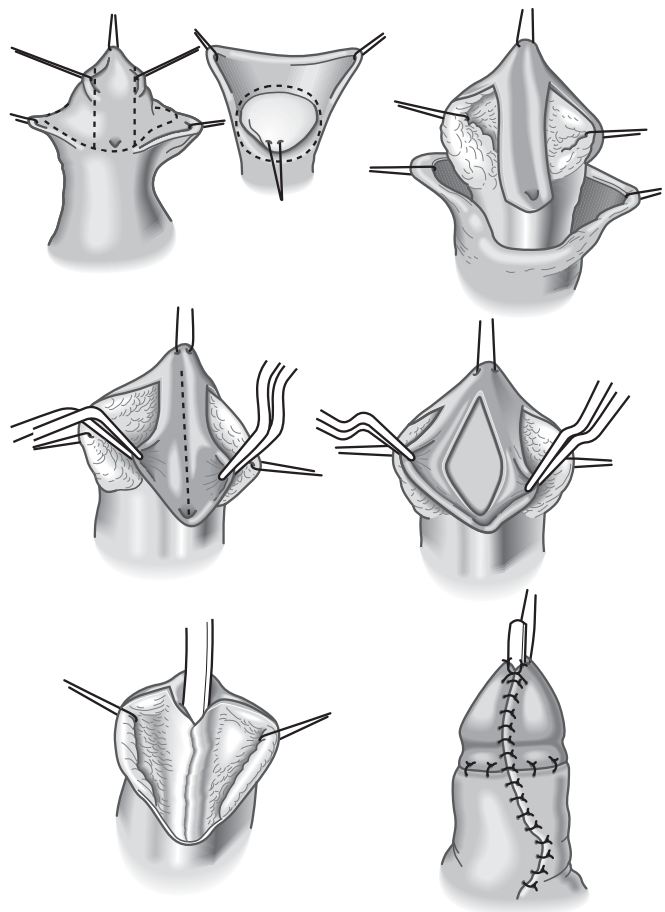


8j

Tubularized incised plate urethroplasty (Snodgrass) repair

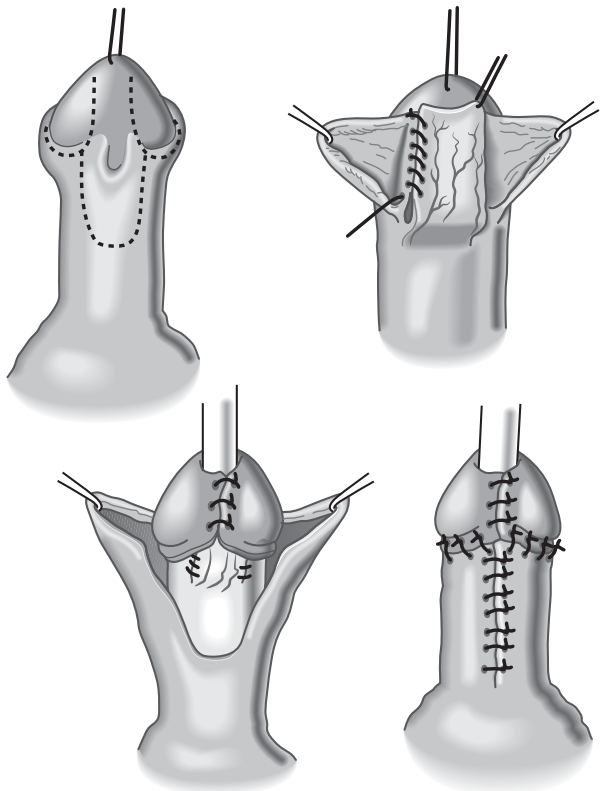
Thiersch and Duplay were the first to describe the technique of urethral plate tubularization in repairing hypospadias. Additional modifications were described by King and Zaontz (GAP procedure), which all result in excellent cosmetic and functional outcome in patients with hypospadiac penis associated with a deep glanular groove. However, urethral tubularization techniques were not deemed suitable for penis associated with a flat urethral plate, as it was feared that there would be excessive tension of the neourethral suture line. Snodgrass combined the vertical distal urethral plate incision technique to relax its tension, as utilized by Rich et al., with the Thiersch–Duplay tubularization technique, to propose the TIP repair technique. Many institutions have reported excellent outcomes with this procedure. Unlike the traditional urethral plate tubularization techniques, TIP repair is versatile for many different anatomic presentations, including proximal, and for previously failed cases. A segment of the urethral plate 8–10 mm wide is marked out distal to the urethral meatus and incisions are made along the lateral borders of the urethral plate. The distal limit of this incision must be carefully planned to avoid ending up with a circumferentially sutured urethral meatus, which may contract and stenose. An incision is made subcoronally and is completed ventrally proximal to the urethral meatus. The penis is carefully degloved down to the base. Ventrally, the skin can be quite thin and adherent to the urethra, and a sharp iris scissor dissection using carefully placed skin hooks for counter-traction provides an optimal visualization of the surgical dissection planes to avoid button holing into a thin native urethra. After addressing any chordee, glanular wings are adequately developed lateral to the urethral plate, so that the subsequent glanuloplasty can be performed without tension. The urethral plate is gently wrapped around an 8 Fr (10 Fr in older children) tube to check for any areas of tension. While providing symmetrical traction and countertraction, a deep midline vertical incision is made with a knife in the urethral plate. The adequacy of hinging is confirmed by again wrapping the urethral plate margins around the tube. The neourethra is then reconstructed using multiple interrupted or running sutures. Again, care is taken to ensure that the newly reconstructed meatus is wide in caliber and without circumferential suture line. A second-layer coverage is then sought with mobilized dorsal dartos, tunica vaginalis, or fanned-out, Y-shaped distal spongiosal tissues. Skin coverage of the phallic shaft completes the procedure.



9

Perimeatal-based flap urethroplasty (Mathieu)

10 Among the more commonly used local flap techniques for coronal and subcoronal hypospadias is the perimeatal-based flap technique of Mathieu. This requires ample penile ventral skin proximal to the hypospadiac meatus. It is begun by measuring the length of the defect from the urethral meatus to the glans. An equal-distance flap is then measured from the meatus toward the base of the penis on the proximal shaft skin. The urethral plate and the matching proximal shaft skin flap are incised to be approximately 7–8 mm wide, and the penis is degloved after subcoronal circumferential incision. Glanular wings are developed deeply in order to perform a tension-free glanuloplasty. If the urethral plate is very flat distally, or if the distal limit of the urethral plate is too ventral, the Barcat balanic groove technique may be employed. In this modification, the urethral plate is dissected off the glans tissue, and after incising the dorsal glans tissue in the midline, the urethral plate is advanced further posteriorly to achieve a more distal neourethral opening. After correcting chordee, the pre-measured proximal shaft skin flap is mobilized carefully and transposed upward toward the urethral plate. This flap is folded over the urethral meatus, and both edges are approximated with precisely placed fine absorbable sutures. After maturing a wide-caliber meatus, a second-layer coverage is performed as described previously. Glanuloplasty and skin coverage complete the procedure.

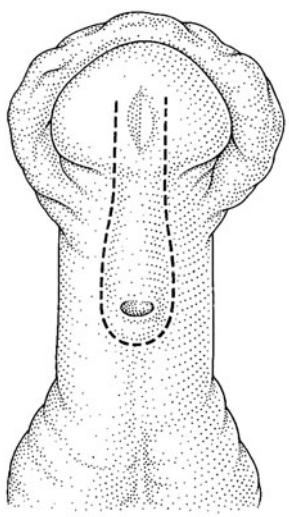


10

Preputial island pedicle flap urethroplasty

11a–f,12a–f The introduction of this method of repair was a huge contribution to hypospadiology, built in part on Hodgson's reconstructions. Duckett, Asopa, and Standoli described preputial onlay urethroplasties almost simultaneously, although the Duckett variant became the popular choice. The penis is prepared by means of degloving, and orthoplasty is performed as described above, preserving the urethral plate in a manner similar to that for the TIP repair. The hood of foreskin is laid out and a rectangular portion is marked on the inner surface with a marking pen to outline the graft for the intended neourethra. In general, this should be at least 8–10 mm wide to produce a reasonable caliber, and a length of 3–4 cm is not an unreasonable expectation for infant hypospadias repairs. The use of gentle, fine, traction sutures to display the foreskin minimizes the trauma to the flap by repetitive forcep grasping. Injecting dilute (1:200 000) epinephrine solution subcutaneously is useful for hemostasis and for separating the inner and outer layers of prepuce. A rectangular flap is developed using sharp knife and iris scissor dissections, leaving it attached to a broad dartos vascular pedicle. This island pedicle flap must be mobilized adequately down to the penile base in order to swing it away from the remainder of the foreskin and bring it around the penile shaft ventrally without twisting the shaft. The native urethral meatus is prepared to ensure that it is vascular, ample, and spatulated. The island flap is then sutured onto the urethral plate using fine absorbable sutures (onlay technique). If the urethral plate is not available or is unsuitable for onlay urethroplasty, the flap is then tubularized over an 8–10 Fr catheter to fashion a tubular neourethra. The neourethral suture line should be positioned dorsally against the corporal bodies to minimize the chance of fistula. A spatulated anastomosis is performed between native urethral meatus and the neourethra. The neourethra is then secured to the penile shaft ventrum with several interrupted, fine, absorbable sutures to stabilize it.

In general, the onlay technique is associated with a lower incidence of proximal anastomotic stricture as compared to the tubular neourethra, but at times the urethral plate may need to be divided from the native urethral meatus in order to perform an effective correction of severe ventral chordee, necessitating the use of a tubular neourethra. The glanuloplasty is performed either by splitting the glans and creating lateral glans wings to bring over the distal neourethra, or by a tunneling technique to bore an ample core through the glans. In either case, the distal meatus is secured to the glans with fine, interrupted, absorbable sutures, and the new urethral meatus must be spatulated and fashioned wide to prevent stenosis. A critical next step is to provide a supporting vascular tissue to cover the neourethra before skin closure using either the remaining penile dartos or tunica vaginalis. Lastly, ventral skin approximation is accomplished using the dorsal remnants of the prepuce.



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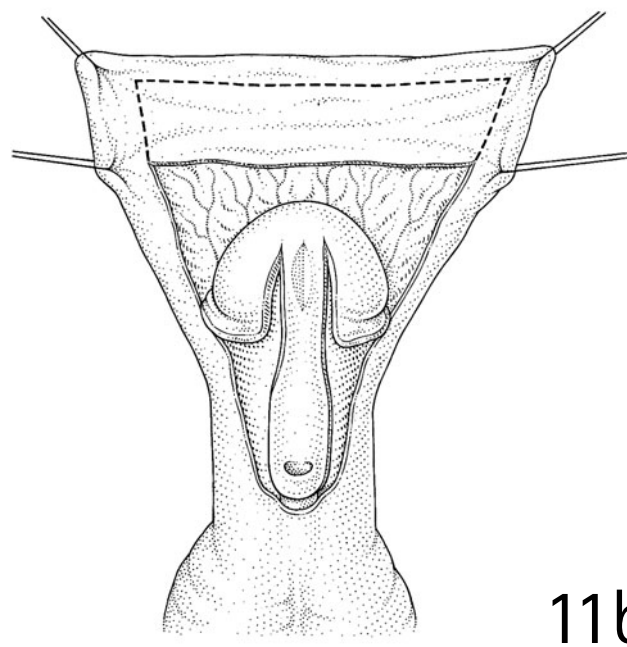

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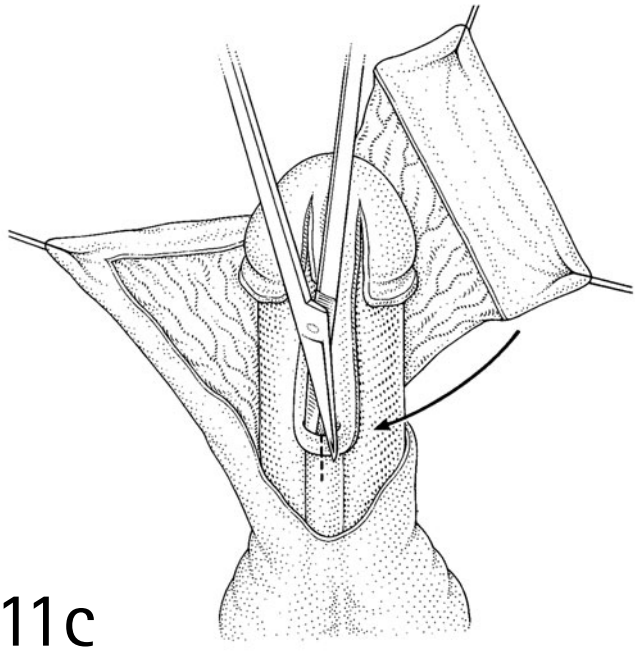

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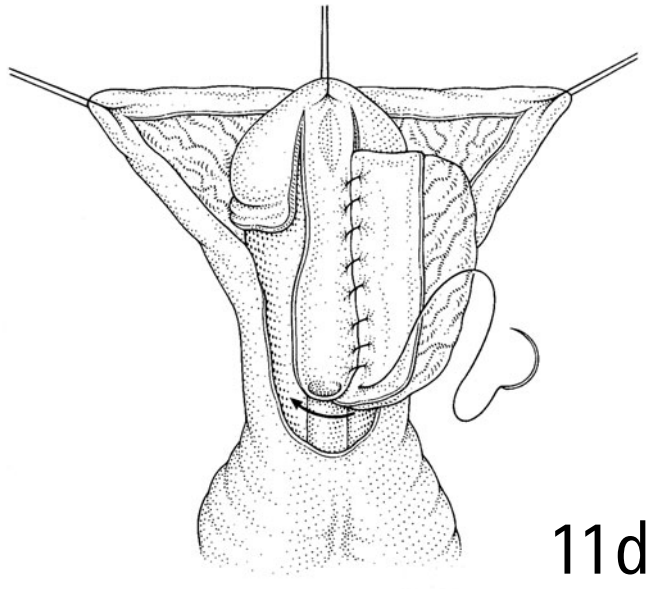

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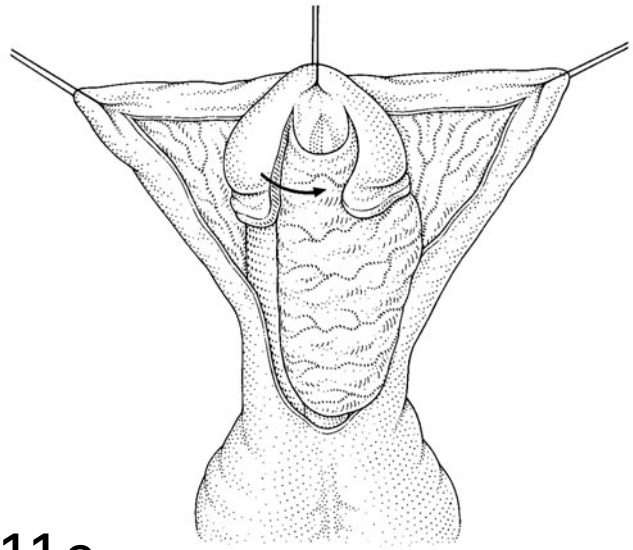

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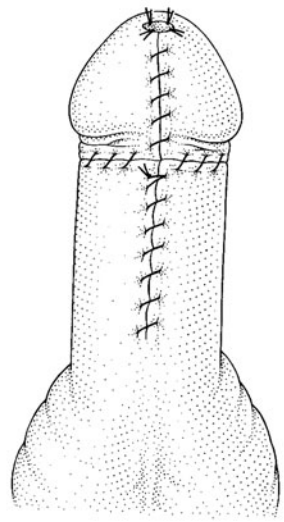

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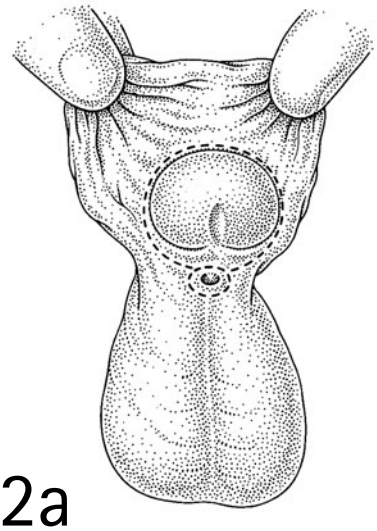

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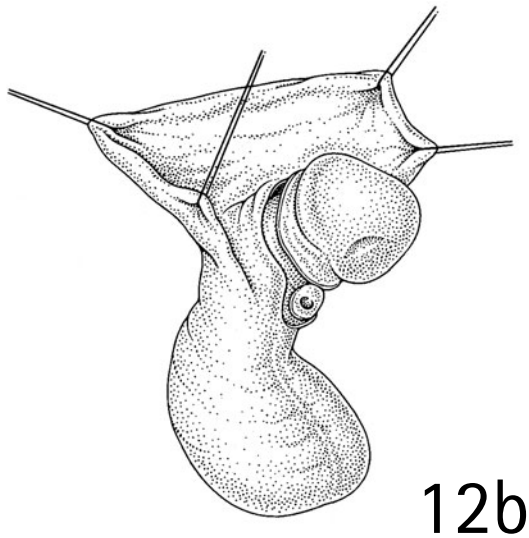

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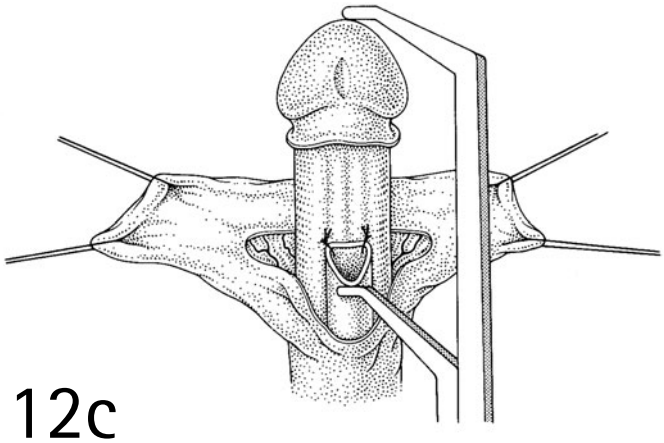

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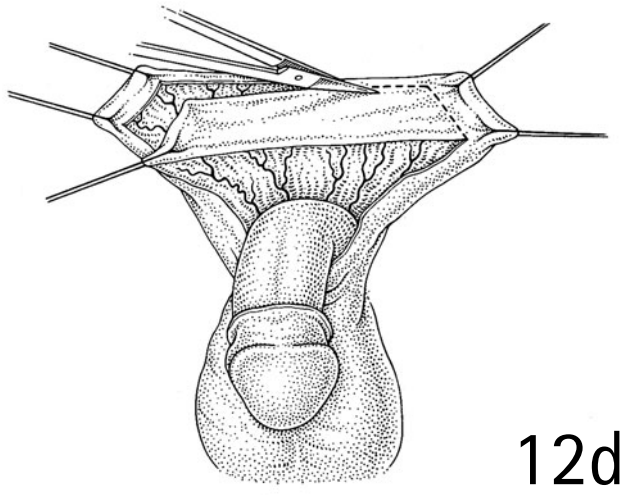

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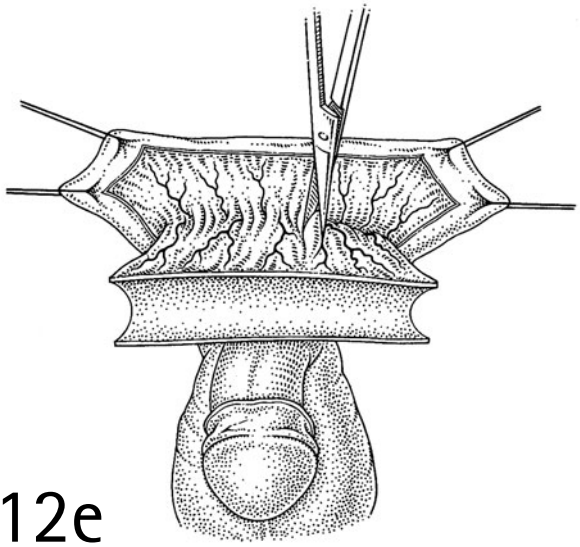

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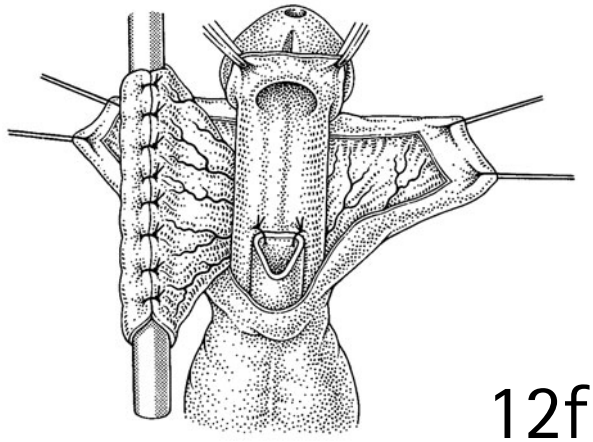
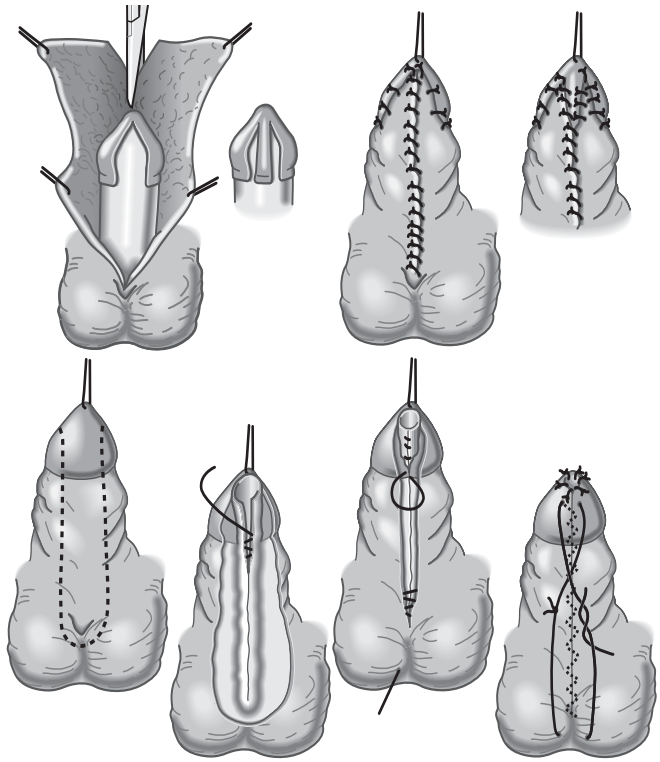


12f

Staged repair

13 Staged repair was once preferred for most instances of hypospadias. Accumulated expertise in hypospadiology relegated it to being used only for the more difficult cases of hypospadias, and some surgeons even favored complex single repairs for all instances of hypospadias. Pendulums do swing, however, and many surgeons prefer to manage patients with complex hypospadias via several dependable, if less heroic, steps. The principles of staged repair are to correct any chordee and other scrotal anomalies (such as penoscrotal transposition) during the first repair, followed by subsequent urethroplasty during another operation. Some patients present with an extreme degree of chordee (more than 90–180°) along with severe hypospadias. In this scenario, even after straightening the penis perfectly at the time of surgery, as proven by an artificial erection test, some regression of chordee becomes evident after a number of months. Thus, for the first stage, an initial orthoplasty is performed along with ventral resurfacing with dorsal preputial skin. At least 6 months later, the patient is brought back for the second stage. At this second operation, the residual chordee (which is invariably much less than at the initial procedure) can be corrected definitively, and the neourethra can then be fashioned by either tubularization of the previously transposed preputial tissue or free graft (which we now favor above an oral mucosal source).



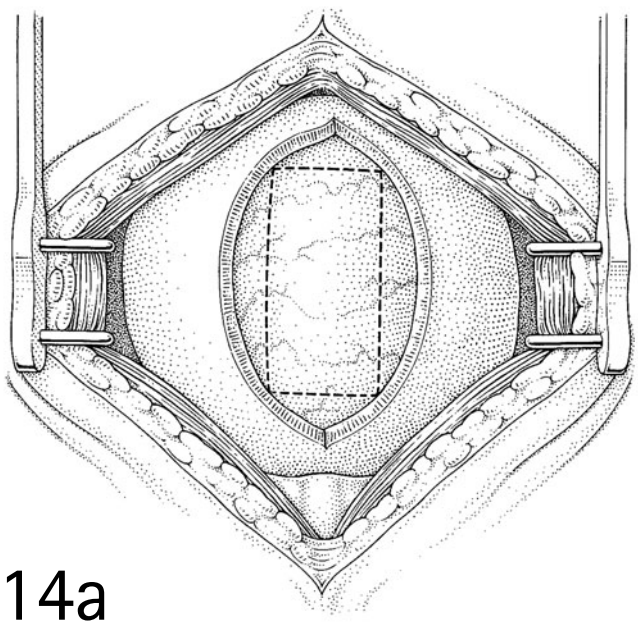
13

Oral mucosa graft urethroplasty

14a–g Humby reported this technique in 1941 in a single case, but somehow the best hypospadiologists remained ignorant of this innovative technique for more than 40 years, until it was rediscovered and resurrected in Italy and France. The next set of papers appeared in the early 1990s, and the use of oral mucosa increased steadily over the following decade in the reconstruction of hypospadias and other urethral problems. This procedure was first applied for hypospadiac 'cripples' who had failed multiple operations and were left with no viable local penile tissues for adequate repair, but its use has now expanded satisfactorily to primary repairs in severe hypospadias. It may be used as a single-stage or a staged repair, depending on the anatomic situation.

The oral mucosa graft may be obtained from either the inner lining of the lower lip or the inner cheek, and sometimes a contiguous combination of both is useful when a longer graft is needed. An 8–10-cm long graft can be achieved with this technique. In the case of shorter grafts, inner cheek is preferable, and during the harvesting one must be careful to avoid injury to Stensen's duct, which is opposite the second upper molar in most patients. The lower lip source has few potential drawbacks. First, the width is limited by the lower incisors and the visible portion of lower lip. One also needs to be careful at the corners of the mouth, where any scarring can have a visible cosmetic impact. However, thinner grafts from the lower lip source may be useful in creating a distal glanular urethra facilitating tension-free glanuoplasty.

The graft harvest site is marked out with a marking pen, retracting the mouth with a combination of retractors and fine traction sutures. Subcutaneous injection of dilute (1:200 000) epinephrine solution is useful for purposes of hemostasis and facilitating the dissection of the oral mucosa. A sharp dissection is performed using a knife and iris scissors, leaving muscle bundles in the mouth. Handling the graft with fine traction sutures minimizes the trauma from repetitive forcep grasping. Once the graft is harvested, it is rinsed multiple times and placed in iced saline solution to minimize desiccation. Bleeders in the graft bed are managed with a combination of direct pressure, low-grade coagulation, and fine suture ligatures. The harvest site may be closed with fine absorbable sutures (such as 5/0 chromic), or it may be left open. No clear-cut advantage has been demonstrated for either strategy, and postoperative morbidity in terms of pain and dietary issues has been negligible. Most patients can resume normal diet in 24–36 hours. The underside of the graft should be carefully trimmed to remove any extraneous adipose and muscle tissues, leaving behind the dermal layer only. Once the graft is prepared, it is rinsed several times and then kept in an iced saline bath. During the entire oral procedure, the initial operative field over the penis and the surgical instruments are segregated before the mouth is prepared and draped. After completing the graft harvest and preparation, the surgeons re-glove and re-gown and return to the original operative field and instruments. The graft is then employed for creating neourethra via either an onlay (if an adequate urethral plate is present) or tubular neourethra technique. Stabilizing the graft and finding a healthy vascular supporting tissue for graft coverage are critical for graft take. Distal glanular urethra and neourethral meatus must be fashioned widely to avoid stenosis. After penile skin coverage, the dressing must be applied loosely, and the patient's activity must be restricted for 24–48 hours to encourage vascular ingrowth into the graft. In general, the neourethra is intubated with an appropriately sized catheter for 10–14 days.



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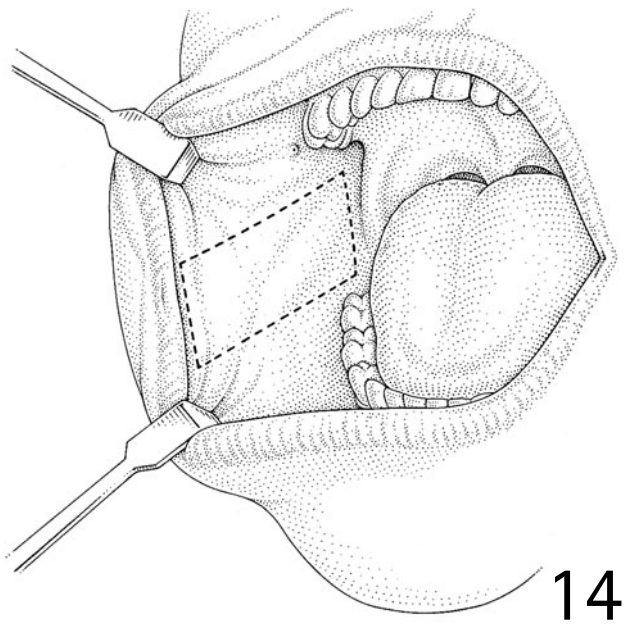

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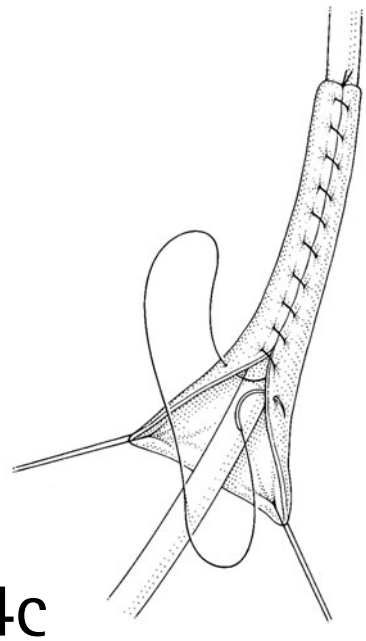

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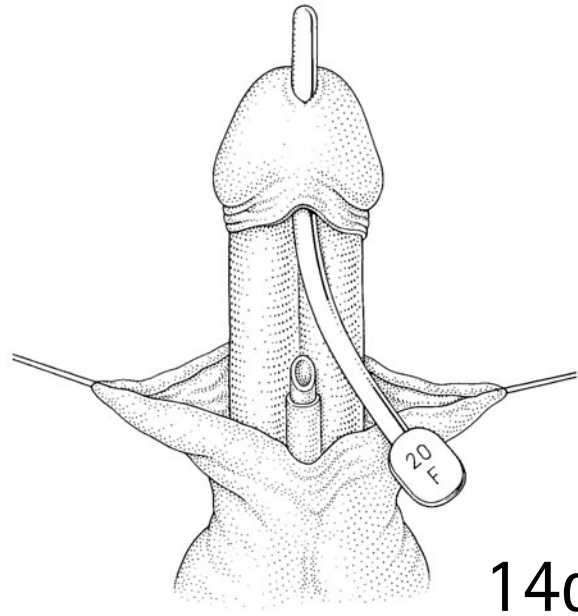

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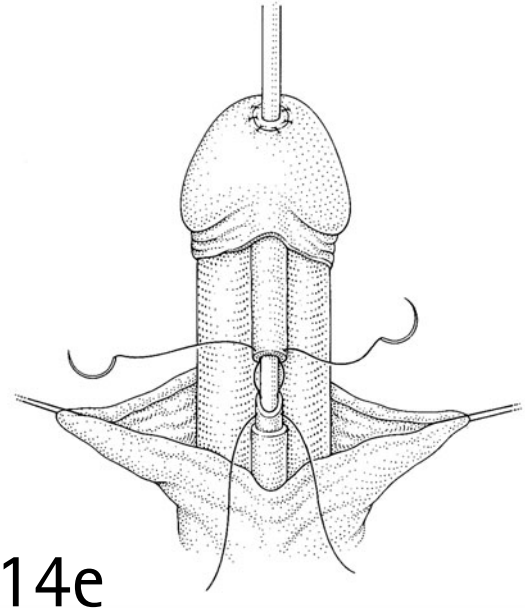

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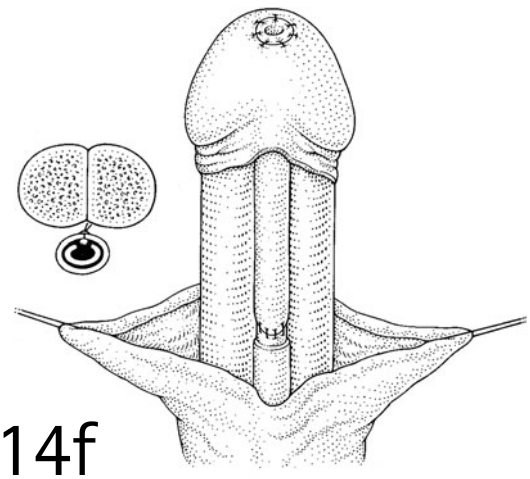

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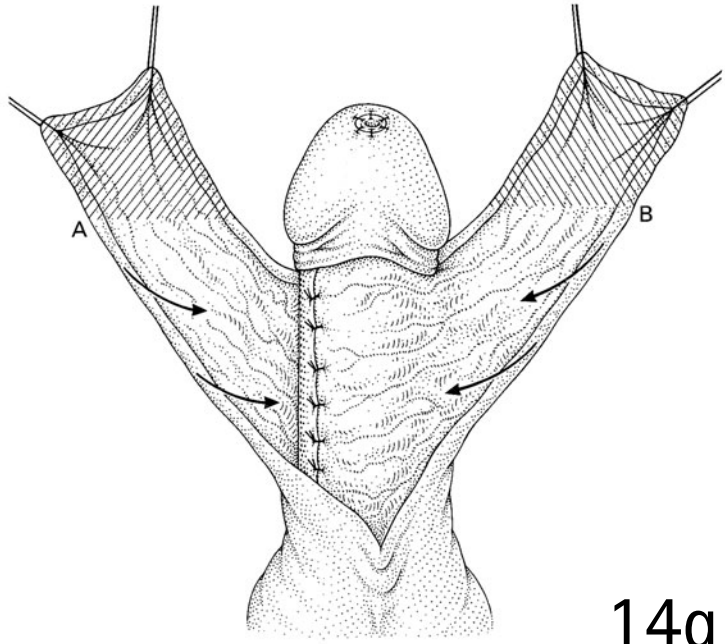

14g

POSTOPERATIVE CARE

Urethral catheter

Intuitively, a newly reconstructed urethra with a long suture line is likely to be better protected during healing with the use of a diverting urethral catheter. It is routine among many surgeons to leave an indwelling urethral catheter postoperatively for 7–10 days. However, recent studies have not proven the benefit of a urethral catheter. In a multicenter report, excellent results were obtained in 96.7 percent of more than 300 patients undergoing mild to moderate hypospadias repair with the Mathieu technique, and the complication rate was not affected by the catheterization status. Others have speculated that the routine use of a urethral catheter may at times increase the chance of complications. Based on these observations, routine use of a urethral catheter may not offer any significant advantage in mild to moderate cases of hypospadias with a straightforward neourethra reconstruction. If we use a catheter, we prefer a soft, Silastic tube of 8–10 Fr caliber without retention balloon, secured at the meatus using sutures. We usually drain the urine via a double diaper technique in infants, in which the catheter is brought through a ventral hole in the inner diaper and is allowed to drain continuously into the outer diaper. With this set-up, fecal material is kept away from the catheter opening, and the double diaper provides secure, additional padding over the genitalia without the worry of the drainage bag pulling on the reconstructed urethra.

Dressing

An ideal penile dressing after hypospadias should be nonadherent, absorbent, and compressive, and yet soft and elastic enough to accommodate postoperative swelling. Several variations have been reported in the literature. Careful application of a secure penile dressing can prevent postoperative complications such as hematoma and edema and may additionally reduce parental anxiety. It is critical to keep the reconstructed meatus moist and free of dried-up secretions by generously applying a petroleum-based ointment for several weeks.

Complications

BLEEDING

In most hypospadias repairs, electrocautery must be used very sparsely, applying precise pinpoint cauterizations. Bleeding from the exposed spongiosal tissues, from the glans, corpus spongiosum, or corpus cavernosum, should not be managed with aggressive cauterization, not only because of its ineffectiveness in stopping the bleeding, but also because of excessive tissue ischemia and injury following such maneuvers. Dilute epinephrine solution, direct pressure, or fine suture ligatures are appropriate for most intraoperative bleeding. Pressure dressings are useful for hypospadias repairs, with the caveat that they should not be so tight as to cause ischemia. Occasionally, a patient will return to the emergency room a day or so after hypospadias repair because of unanticipated bleeding, and in these instances we simply re-apply a pressure dressing. It may be necessary on rare occasions to return to the operating room to evacuate a clot and control a bleeding source. Late bleeding (more than a week postoperatively) is unusual and may occur as the result of trauma. Large, expanding hematomas are best evacuated under anesthesia, with control of active bleeding sites and reapplication of a pressure dressing.

MEATAL STENOSIS

A wide distal anastomosis without a circumferential suture line and with good vascularity to all the involved tissues should minimize subsequent meatal stenosis. In the initial few postoperative months, it is critical to keep the distal meatus continuously moist with petroleum-based ointment. When we see patients back in clinic, we have a low threshold for passing a small-caliber feeding tube to assure patency. When meatal stenosis is suspected, we will often ask the families or the patients to pass a tube on a regular basis to keep it open. Topical vitamin E or corticosteroids may help soften up an incipient stricture. A late stricture may require an anesthetic for aggressive dilatation or urethrotomy. It is important to distinguish true meatal stenosis from distal urethral stenosis, which would require more aggressive management and even a re-do urethroplasty.

URETHROCUTANEOUS FISTULA

A very distal fistula in the glans or corona can be corrected by simply incising the intervening glanular tissue to create a more ample meatus, even if it becomes slightly hypospadiac, as long as there is no chordee. More proximal fistulas are corrected by generous incision to expose the defect and multiple layers of closure, usually using some type of de-epithelialized skin edges to create vascular coverage layers. Some fistulas represent only the tip of an iceberg, as it were, and are indicative of unhealthy tissue over a portion of the neourethra or distal urethral stricture. In these instances, one must consider doing an entire revision of the neourethra with alternative tissue sources such as oral mucosa. Not appreciating the poor quality of the neourethra as the true cause of fistulas will probably lead to their recurrence. In many instances, the sine qua non for a successful fistula repair is 'Make it a big operation to fix a little hole.'

INFECTION

True skin infections with cellulitis are uncommon after hypospadias repair. When we suspect them, however, we institute an aggressive antibiotic treatment with broadened coverage, and we would not hesitate to bring the child back

into hospital for several days of intravenous antibiotic coverage. When children are sent home with a catheter in place, they are routinely given low-dose prophylactic antibiotic coverage daily to minimize colonization and reduce the risk of urinary tract infection until the catheter is removed.

URETHRAL DIVERTICULUM

This complication occurs as the result of distal urethral stenosis and/or insufficient ventral tissue coverage of the neourethra. A small diverticulum may be left alone if a patient is willing to support the sacculed area manually while voiding; however, more substantial urethral diverticuli are best managed surgically, first, by ensuring no distal stenosis and, second, by exposing the diverticulum, removing excess tissue, and re-closing the urethra. The critical step is finding adequate tissue (either dartos or tunica vaginalis) to support the area of the resected diverticulum before closing the skin. Again, if a significant portion of urethra is unhealthy or ischemic, it is better to perform a revision urethroplasty using an oral mucosa graft.

STRICTURE

Urethral stricture may occur at the meatus (meatal stenosis), in the glanular portion of the urethra, or at the proximal anastomotic site. Strictures within the middle portion of the neourethra are less common. Early strictures are best managed by gentle dilatation and intermittent catheterization. More severe strictures, particularly along the course of the urethra, may be corrected by a dilatation technique or internal urethrotomy using either an endoscopic knife or holmium laser. Generally speaking, if the strictured segment is long and surrounded by dense periurethral fibrosis, the above-mentioned minimally invasive techniques are ineffective, and the stricture is prone to recur. In these situations, it is advised that one should consider a revision urethroplasty using oral mucosa. Placing an oral mucosa graft dorsally against corpora cavernosal bodies will minimize the risk of fistula formation.

RE-OPERATIVE HYPOSPADIAS REPAIR

Some patients present with a difficult constellation of failed hypospadias repair, residual chordee, recurrent stricture, fistula, or other functional residua after multiple attempts at repair. These patients have been called 'hypospadiac cripples'. *The fundamental principle of management is thorough assessment of the relevant symptoms and anatomic features of the patient.* Usually this is best done by examination under anesthesia with cystourethroscopy. Our first principle is properly to inform the patient and family about the problems and likely etiologies and to plan for correction and potential complications. Establishing realistic expectations is critical. The second principle is to recognize any related anatomic features such as penoscrotal transposition and chordee, as well as any available viable tissues not only for neourethra formation and penile skin coverage but also vascular supporting tissues for covering the neourethra. At times, planning a staged approach, correcting chordee, and debriding unhealthy local tissues before proceeding with neourethra reconstruction may be prudent. Third, the neourethra must be reconstructed from robust, mucosa-lined tissues with an excellent potential for neovascularity (such as oral mucosa). Stubbornly insisting on using the remnant of fibrotic preputial and penile skin will lead to an inevitable failure and poor long-term outcome. Finally, a period of postoperative catheterization, early reassessment with calibration of the neourethra, and even a confirmation endoscopic examination when appropriate, will be essential.

CONCLUDING REMARKS

Hypospadias repair in all of its possible forms remains a core competence for any surgeon engaged in pediatric urological reconstruction. This work requires technical repertoire, skill, clinical judgment, and expertise, as well as the ability to deal with the emotional needs of the patient and family. These traits are not developed overnight, and their perfection is the epitome of the practice of medicine.

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Orchidopexy

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PRINCIPLES AND JUSTIFICATION

Undescended testis is a common abnormality, affecting 3–5 percent of male infants at birth. In preterm infants, the incidence may be as high as 20 percent or more. A significant number of testes not in the scrotum at birth descend in the first 12 weeks after birth, so that by 3 months of age the incidence of congenital undescended testes is approximately 1–2 percent. Some of those testes that descend late into the scrotum may re-ascend later in childhood, producing an acquired variant of undescended testes known as ascending testes.

Embryology

The urogenital ridge forms on the posterior abdominal wall and contains the mesonephros and its draining duct and the developing gonads. The primitive germ cells migrate from the yolk sac at about the sixth week of gestation, as the gonad in the male develops into a testis. By 7–8 weeks, the testis is producing hormones that control its subsequent descent.

Descent of the testes

Descent of the testes occurs in two morphologically and hormonally distinct phases. The key structure in controlling the process is the gubernaculum, which is the embryonic ligament anchoring the testis and urogenital ridge to the inguinal region. The gubernaculum enlarges in the first phase to anchor the testis near the inguinal region as the embryo enlarges. This occurs between 10 and 15 weeks of gestation. In the second phase, which occurs between 28 and 35 weeks of gestation, the gubernaculum migrates out of the inguinal canal, across the pubic region, and into the scrotum. The processus vaginalis develops as a peritoneal diverticulum within the elongating gubernaculum, creating an intraperitoneal space into which the testis can descend.

The main hormone controlling the first phase of testicular descent has recently been shown to be a homolog of insulin and relaxin, known as insulin-like hormone 3 (Insl3). This hormone is produced by the Leydig cells, and stimulates enlargement of the distal gubernaculum, with some augmentation of its effect by Mullerian inhibiting substance (MIS) and possibly testosterone. In the second phase, testosterone acts apparently indirectly via the nervous system and the genitofemoral nerve, which supplies the gubernaculum and scrotum. Calcitonin gene-related peptide (CGRP) has been identified recently within sensory branches of the genitofemoral nerve and has been postulated to act as a final common pathway for androgenic control of descent. Androgens probably cause masculinization of the sensory branches of the genitofemoral nerve in the dorsal root ganglia (L1 and L2) between 15 and 25 weeks of gestation, and the masculinized nerve then controls the direction of gubernacular migration by the release of CGRP in the periphery.

Animal models of undescended testes, including the androgen-resistant mouse, the flutamide-treated rat, and the mutant TS rat, all show an absence or deficiency of gubernacular migration and an abnormality of CGRP physiology. This suggests that some undescended testes in humans may be caused by physiologic or anatomic abnormalities of the genitofemoral nerve or its neurotransmitter, CGRP. The most common cause of maldescent appears to be failure of gubernacular migration, leaving the testes in the groin, or so-called 'superficial inguinal pouch.' Whether genitofemoral nerve anomalies are primarily anatomic or secondary to abnormal hormone development in mid-trimester is uncertain. In rare instances, undescended testes are caused by recognized anomalies in the hypothalamic–pituitary–gonadal axis or in the secretion or action of Insl3 and MIS. Recognizable

hormonal syndromes, however, are rare causes of undescended testes in clinical practice.

Undescended testes

Congenital failure of gubernacular development or migration leads to arrest of gubernacular migration along the normal pathway or aberrant migration leading to an ectopic undescended testis in the perineal, prepubic, or femoral region. Many undescended testes are located in the superficial inguinal pouch, which is a subcutaneous space just above and lateral to the external inguinal ring and is just the processus vaginalis (with its contained testis) lying at the external ring or a little lateral to it. As this is the most common position for an undescended testis to occupy, it is not categorized as ectopic.

There is controversy about whether 'ascending' testes are abnormal. These testes are located in the scrotum by 12 weeks of age, but later in childhood tend to retract back out of the scrotum and often have an exaggerated cremaster reflex. It may be an acquired variant of maldescent, with the position of the testis becoming relatively higher as the child gets older. Actually, in most cases, the length of the spermatic cord is constant (i.e., it fails to elongate normally), while the distance between the inguinal canal and scrotum doubles with growth of the boy during the first decade. Surgical intervention is not indicated unless the testis is no longer residing spontaneously in the scrotum. If the testis is near the top of the scrotum, regular follow-up is required to make sure that it does not ascend out of the scrotum with the passage of time. This often leads to orchidopexy being recommended in 5–11-year-old boys.

Indications

Children are recommended for surgery for three common reasons: abnormal fertility in the undescended testis, a risk of testicular tumors in adult life, and the obvious cosmetic abnormalities. Although many undescended testes have an associated patent processus vaginalis, this is an uncommon presentation for an inguinal hernia. If an infant presents with an inguinal hernia in association with undescended testis, however, orchidopexy with associated herniotomy is performed immediately. Trauma and torsion to the undescended testis are more common than when the testis is fully descended, although these are unlikely indications for surgery.

Germ-cell development in the undescended testis is normal in the first 6–12 months, but then becomes abnormal subsequently due to secondary degeneration. This is caused by the undescended testes residing at a higher temperature (35–37°C) than when located in the scrotum (33°C). In some instances, there may be an intrinsic anomaly of testicular function.

The risk of malignancy in young adult men with undescended testes is approximately five-fold to ten-fold that of young men with normally descended testes. These risk calculations are not based on current practice, however, but on results from children of a previous generation having orchidopexy at approximately 10 years of age. With the current practice of recommending surgery at a much younger age, it is hoped that the risk of cancer in the next generation will be lower, although this has not yet been proven. The risk of malignancy affects not only the unilateral undescended testis, but also the contralateral descended testis in some patients. Evidence is now emerging that acquired cryptorchidism has a much lower risk of subsequent malignancy.

Recommended age for orchidopexy

To prevent secondary degeneration of the testis with loss of germ cells and a progressively increasing risk of malignancy, orchidopexy is best performed in infancy. Although it is not proven conclusively for humans, it is clear in animal experiments that early surgery is better than a delayed operation. Operation can be performed at any time between 3–6 months and 1 year of age, depending on the experience of the surgeon. Those with less experience in pediatric surgery would be wise to delay surgery to the older end of the range, rather than to attempt orchidopexy in a young infant. For the inexperienced surgeon, the risk of testicular atrophy secondary to surgery may be higher in younger infants. Magnification should be mandatory for surgery in babies less than 1 year of age.

PREOPERATIVE

Secondary preoperative preparation

In older boys with possible ascending or severe retractile testes, a course of gonadatrophins may be appropriate, although this is controversial. In the rare circumstance of bilateral impalpable testes, a human chorionic gonadotrophin (hCG) stimulation test should be performed to determine whether testicular tissue is present at all. Serum levels of MIS/anti-Mullerian hormone will reflect the presence of Sertoli cells. If the hCG stimulation test shows the presence of testicular Leydig cells within the abdomen, the parents should be advised that laparoscopy is indicated at the beginning of the operation to determine the exact site and nature of the intra-abdominal testes.

Anesthesia

As orchidopexy is now a day-case procedure, the type of general anesthesia reflects the need for early mobilization. No premedication is usually required, although an oral preparation such as chloral hydrate would be preferable to intramuscular injections. On admission to the day surgical unit, anesthetic cream containing lidocaine (lignocaine) and prilo-

caine is applied to the back of the hand so that intravenous access can be obtained without pain. An ilioinguinal nerve block, local anesthesia, or caudal anesthesia is provided to control pain for the first 4–6 hours after operation.

OPERATION

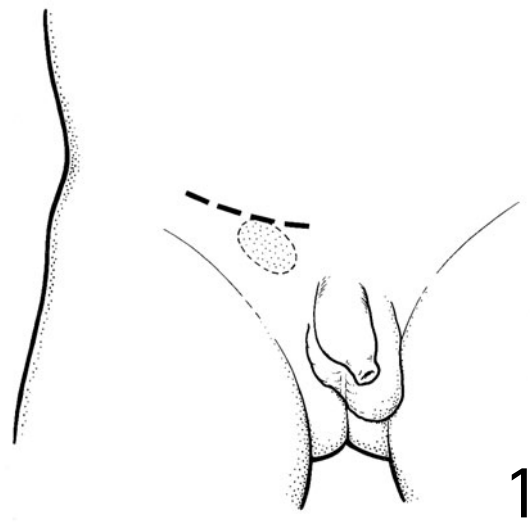
Skin preparation and position of patient

The patient is placed supine on the operating table with the legs slightly apart. Povidone-iodine, or other appropriate antiseptic, is painted on the skin from the umbilicus to below the scrotum and perineum.

Standard orchidopexy for a palpable testis (which occurs in approximately 80–90 percent of cases) involves an inguinal incision, full exposure of the inguinal canal, separation of the processus vaginalis, and mobilization of the testis and spermatic cord. The second part of the operation is the orchidopexy itself, or fixation of the testis in the scrotum.

Mobilization of the testis

1 A transverse skin crease incision is made over the inguinal canal. This incision is usually about one finger's breadth above the base of the penis in an infant. The medial end of the incision is level with the pubic tubercle, while the lateral end is at the mid-inguinal point.



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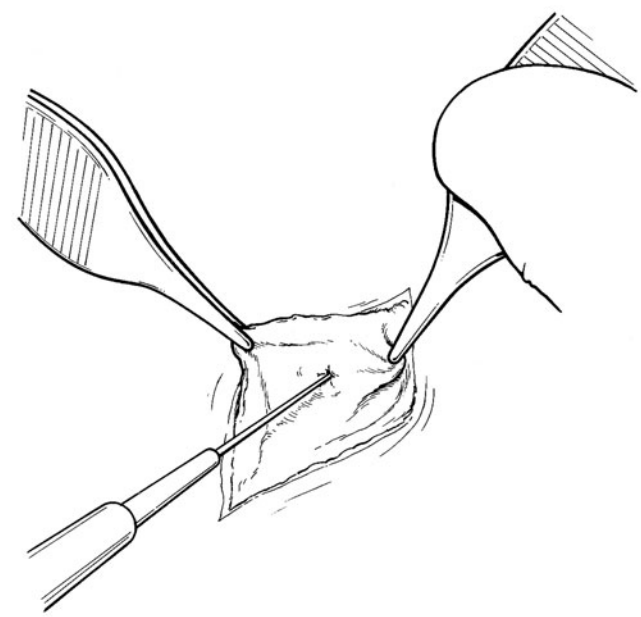

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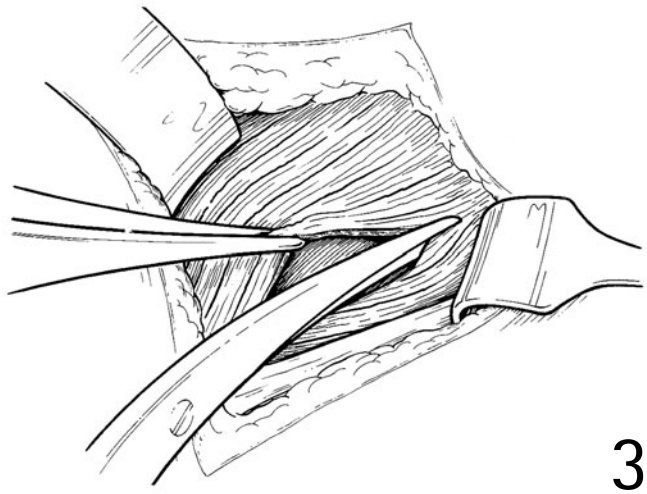
2 The incision is deepened through the subcutaneous fatty tissue with scissors or diathermy. The superficial fascia is in two layers: a more superficial fatty layer and a deeper, well-developed fibrous layer known as Scarpa's fascia. The superficial inferior epigastric vein may be seen in the subcutaneous tissue running obliquely across the incision. Sometimes square-ended retractors can pull the vessel out of the way, and at other times it is best coagulated by diathermy and divided.

3 Once Scarpa's fascia has been divided, the external oblique aponeurosis can be distinguished deep to Scarpa's fascia by the oblique orientation of its fibers, which are absent in Scarpa's fascia. The surface of the external oblique muscle is cleared by placing square retractors under Scarpa's fascia to expose the lower border of the external oblique muscle where the inguinal ligament lies. A sweeping motion with closed scissors parallel to the external oblique fibers will expose the rolled edge of the inguinal ligament and the site of the external inguinal ring, where the spermatic cord is seen bulging.

The inguinal canal is opened with a scalpel incision in the external oblique muscle, with extension of the incision with scissors in line with the external ring. The incision can be extended with scissors by cutting the fibers toward the external ring. The edges of the external oblique muscle are best stabilized with small artery forceps so that they can be identified easily later in the operation. The ilioinguinal nerve will run parallel to the incision, just under the fascia of the external oblique muscle, and should be identified and carefully avoided. Accidental transection of the nerve will produce a sensory deficit in the region of the anterior scrotum. Blunt dissection is used to mobilize the inner layer of the external oblique aponeurosis off the surface of the spermatic cord, and the external ring is opened with scissors if this has not already been done.



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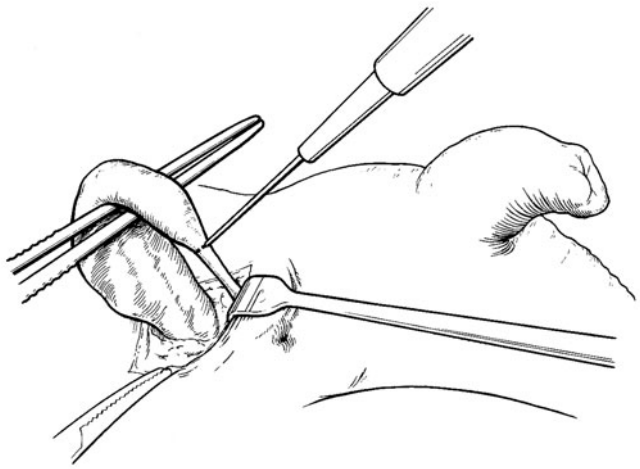

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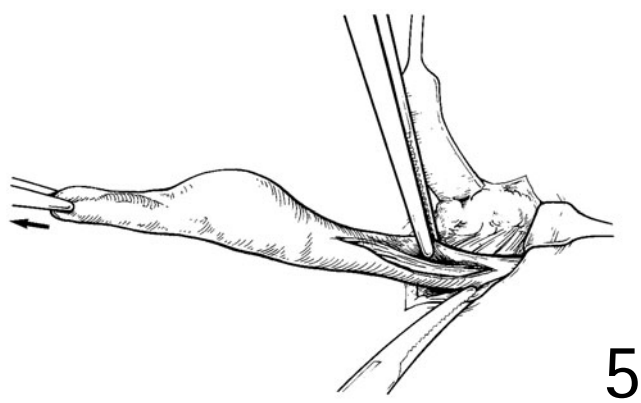
4 The testis and attached spermatic cord are mobilized with blunt dissection and delivered out of the wound. This should identify the abnormal attachment of the gubernaculum, which causes dimpling of the skin above and lateral to the neck of the scrotum. The gubernacular attachment is divided carefully with scissors or diathermy, taking care to avoid any structures within the processus vaginalis, such as the vas deferens, which may extend below the lower pole of the testis. The gubernaculum at this level is usually transparent, with an occasional fine vessel and some fat, and is easy to divide without risk to other structures.

5 Small artery forceps are placed on the gubernacular attachment to the testis and the tunica vaginalis and this is placed on tension. This enables any remaining cremaster fibers surrounding the outside of the spermatic cord to be stripped off with blunt-ended forceps.



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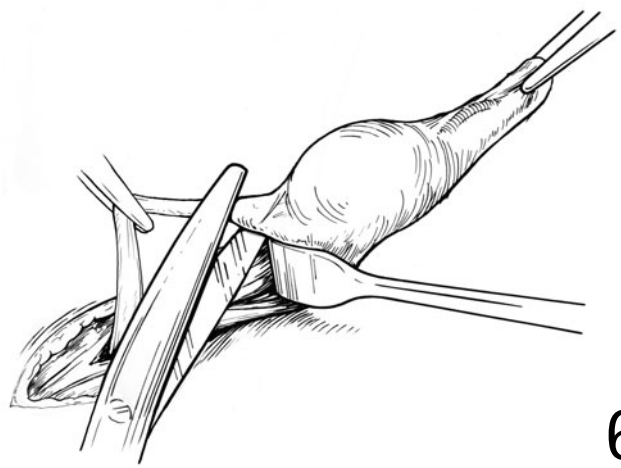
Dissection of the hernial sac

The processus vaginalis is commonly widely patent in the undescended testis. Careful separation of the patent processus vaginalis or obvious hernial sac from the vas deferens and the testicular vessels is an important part of the procedure, as this increases the effective length of the spermatic cord. The hernial sac may be stretched over the index finger while round-ended, non-toothed dissecting forceps gently sweep off the other cord structures, carefully avoiding direct application of the forceps on the vas deferens or vessels. Alternatively, the sac can be held with small artery forceps while the vas deferens and testicular vessels are isolated en masse off the sac. It is best to attempt to separate the hernial sac completely without opening it, as an unrecognized tear may extend through the internal ring into the peritoneum, making closure of the hernial sac difficult. It is absolutely essential that the testicular vessels, and particularly the vas deferens, are visualized clearly before the sac is divided. It is important to note that the vas deferens is closely adherent to the posterior surface of the sac before dissection.

6 With the entire cord held on traction, the testicular vessels and vas deferens are separated from the hernial sac with a small retractor and adequately identified before clamping the hernial sac with artery forceps. The sac is then divided with scissors immediately distal to the artery forceps.



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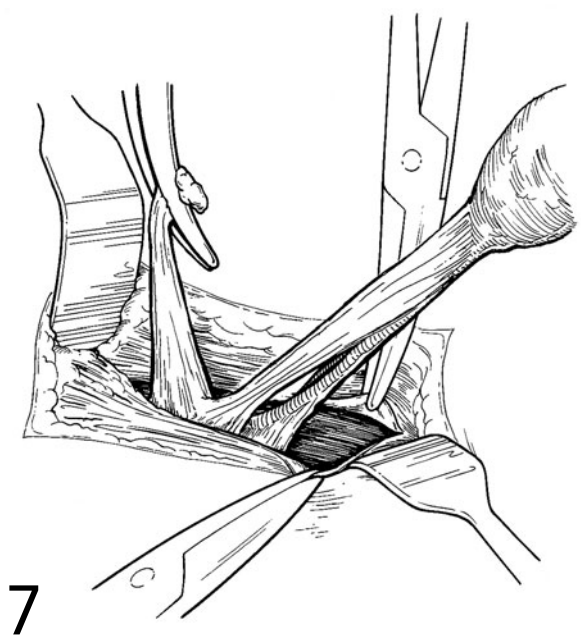

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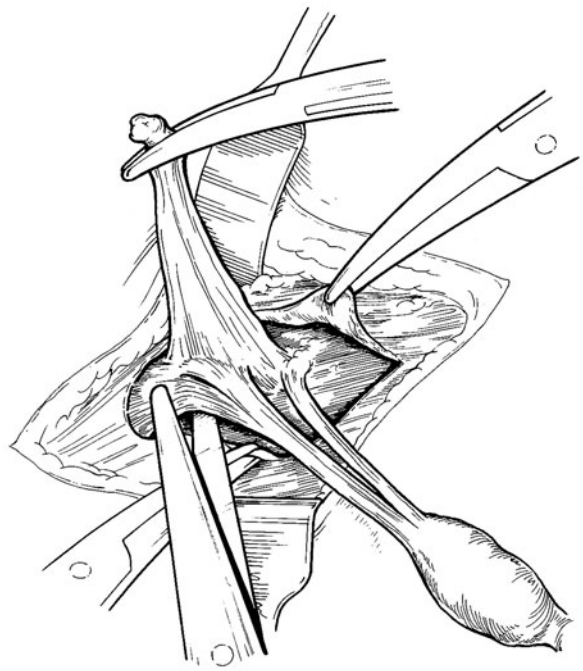
7 The divided processus vaginalis is pulled cranially to put the membrane under tension, and the testicular vessels and vas deferens are separated with blunt dissection from the posterior surface of the sac right up to the internal ring. At the junction between the processus vaginalis and the peritoneum proper, the translucent processus vaginalis becomes an opaque, white membrane with a triangular widening of the base. At this level the vas deferens curves medially around the edge of the transversalis fascia, adjacent to the inferior epigastric vessels. By contrast, the testicular vessels pass cranially on the lateral side of the internal ring and disappear into the retroperitoneal space.

8 If the spermatic cord is not long enough for the testis to reach the scrotum, the retroperitoneal plane behind the processus vaginalis above the internal ring is developed, and a small Langenbeck's retractor is inserted to pull the peritoneal membrane anteriorly. This reveals the testicular vessels passing cranially in the retroperitoneum. The vessels tend to follow a gentle convex curve laterally, and there are a number of lateral fibrous bands attached to the vessels. These should be divided by sharp or blunt dissection once the testicular vessels themselves have been identified. Continuous, gentle traction on the testis allows the testicular vessels to be seen and preserved.



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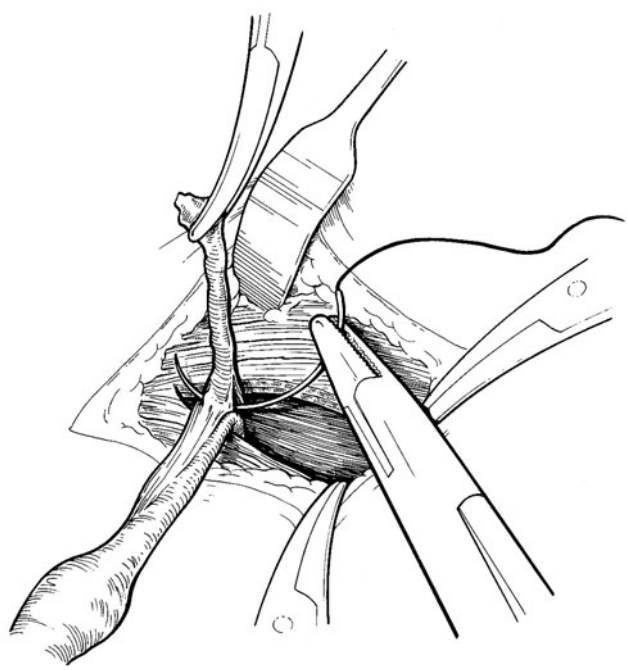

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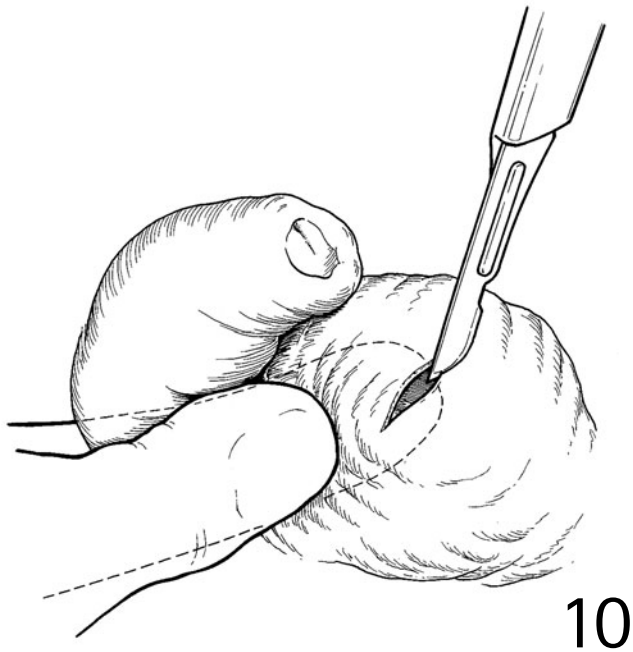
9 There should now be adequate length in the vas deferens and the vessels to allow the testis to reach the scrotum. The processus vaginalis is now twisted up to the internal ring to make sure that it contains no intraperitoneal contents, and it is transfixed and ligated at this level. At this point, if the length of the vas deferens is found to be a limiting factor in the position of the testis, the inferior epigastric vessels can be divided electively to gain an extra 0.5–1 cm.

10 Traction on the testis is now stopped and a finger is introduced through the incision and down to the scrotum, breaking down any fascial layers near the neck of the scrotum so that the tip of the index finger can reach the mid-scrotum. The scrotal skin is immobilized between the index finger internally and the thumb externally. A scrotal incision is then made, going just through the skin but not through the deeper tissues. This incision can be either horizontal (the author's preference) or midline in the scrotal septum. Horizontal incisions are associated with less bleeding.



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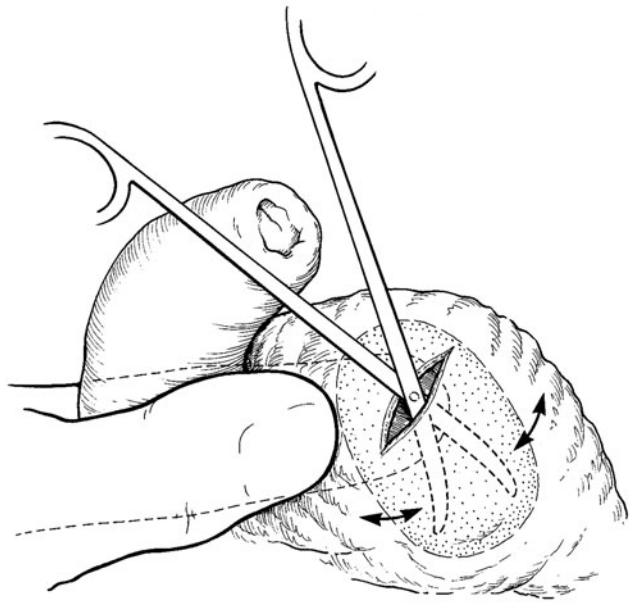

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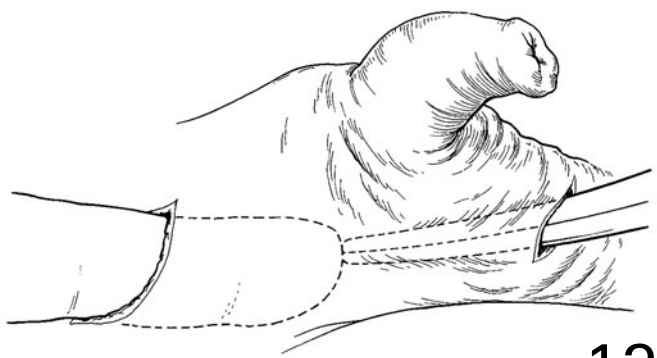
11 While the index finger is still inside the scrotum and the thumb is immobilizing the scrotal skin, fine artery forceps or a scissors are used to develop a subcutaneous pouch, which is usually just deep to the dartos muscle. This should be developed inferiorly more than superiorly, so that the external incision is placed near the cranial end of this subcutaneous pouch. Any troublesome bleeding is controlled by diathermy before proceeding further. This is an important step, as a scrotal hematoma around the testis will inevitably become infected and lead to wound breakdown.

12 Fine artery forceps are placed through the incision in the scrotum and pressed against the index finger internally. The index finger then guides the artery forceps back to the inguinal incision where the tip of the artery forceps is pushed through any residual fascial plane.



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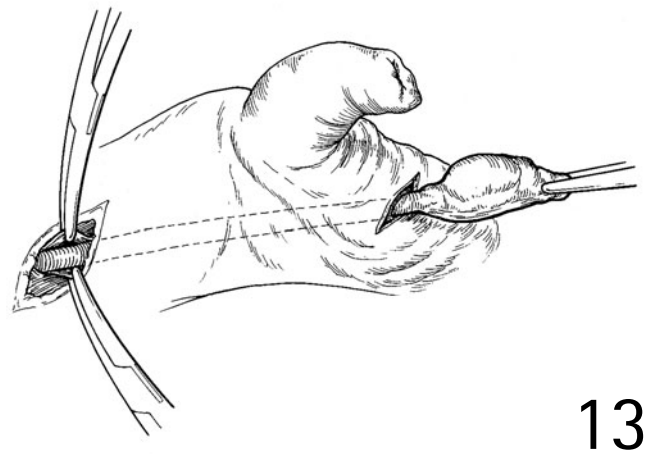
13 The artery forceps, which have been pushed up through the scrotal incision to the inguinal incision, then grasps the gubernacular attachments of the testis, making sure that the cord structures are not twisted. The testis and attached structures are then drawn gently down through the track made by blunt dissection and pulled through the 'button-hole' in the subdartos fascia and delivered through the scrotal incision.



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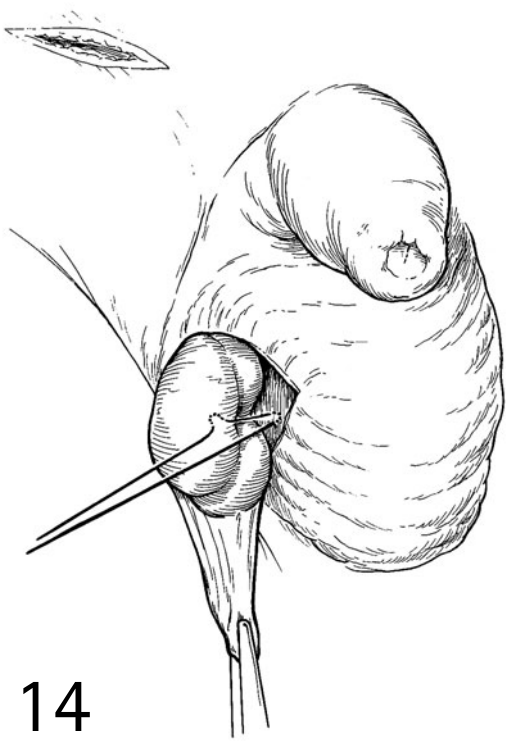

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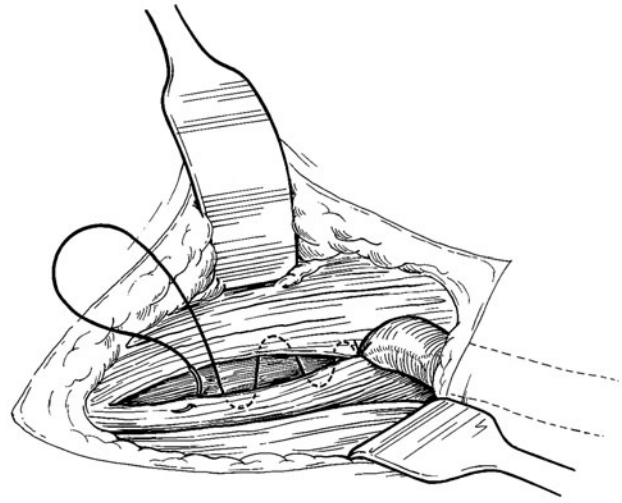
14 If the hole made by the artery forceps through the fascial planes between the two incisions is not too large, the testis will sit comfortably in the scrotal pouch like a button through a button-hole. If there is any concern that the testis may retract through a larger defect, it can be anchored to the midline scrotal septum with a 3/0 absorbable suture that passes through the tunica albuginea but does not need to pass right through the body of the testis. To enable this maneuver to be accomplished effectively, many surgeons deliberately open the tunica vaginalis so that the anatomy of the testis can be defined precisely and recorded. In addition, any testicular appendages can be excised at this time. The testis, epididymis and adjacent coverings can then be placed in the subcutaneous scrotal pouch using blunt forceps.

15 The scrotal incision is closed with a 4/0 subcuticular absorbable suture.

15

16 The surgeon now returns to the inguinal incision, where the external oblique aponeurosis is reconstituted with one to three interrupted sutures or a short continuous suture of 3/0 absorbable material.



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The retractors are removed from the wound, and the fibrous subcutaneous (Scarpa's) fascia is carefully identified and picked up with toothed forceps and closed with one or two interrupted sutures. The skin is approximated with a 4/0 subcuticular suture. The inguinal incision can be covered with a sterile, semipermeable, adhesive film dressing, which provides a waterproof and childproof covering for the wound for the first 7–10 days. A similar dressing can be applied to the scrotal incision, or alternatively this can be sprayed with a plastic skin spray. Depending on the anesthesia used, local anesthetic infiltration to both the inguinal and scrotal wounds can be performed near the end of the procedure.

POSTOPERATIVE CARE

The patient is discharged from hospital the same day, unless an overnight stay is necessary, such as may be the case with bilateral impalpable testes. Most boys return to normal activities within 2–3 days, although they may need to refrain from active sport for 1–2 weeks.

The dressing is removed and the position of the testis checked after 1–2 weeks and again at 6 months after surgery.

Complications

Wound infection and hematoma are the two most common complications. Hematoma can be avoided by meticulous hemostasis with diathermy at the time of surgery. Wound sepsis can be avoided by the placement of waterproof dressings on both incisions, which remain in place for at least 1 week. The risk of testicular atrophy, which is determined at 6 months after surgery, should be less than 5 percent; in most series it is 1–2 percent. There is a small risk of retraction of the testis into the groin, particularly if there is significant sepsis or hematoma, or postoperative trauma.

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Testicular torsion

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Torsion of the testis must be the first consideration in any child or young adult with acute scrotal pain. There are two peaks in the incidence: in the perinatal period, and between the ages of 10 and 25 years.

DIAGNOSIS

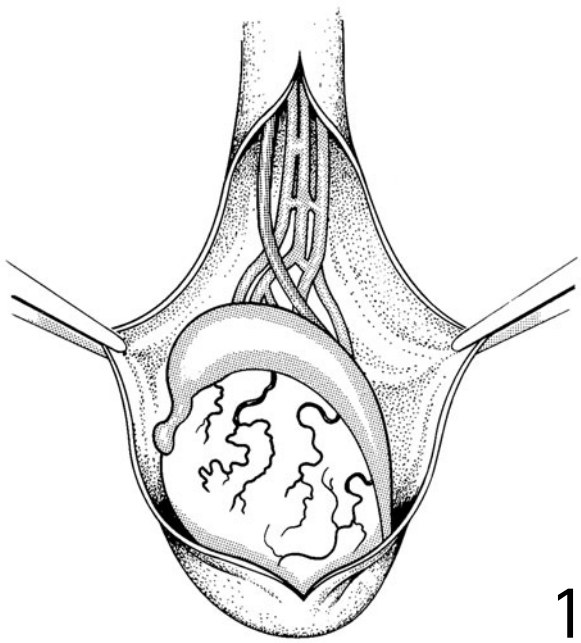
Neonatal torsion

Neonatal torsion accounts for 12 percent of all cases of childhood torsion. Neonatal torsion is almost always extravaginal. There are a few reports of sequential contralateral torsion and there are reports of salvage of the testis, especially if the neonatal examination was normal. Therefore it has recently been proposed that urgent surgical exploration with contralateral testicular fixation should be undertaken.

An intrauterine or perinatal torsion may present as an impalpable testis. Contralateral testicular fixation has also been advocated for these cases to avoid loss of the remaining testis.

Intravaginal torsion of the testis

If the tunica vaginalis invests the whole of the epididymis and the distal part of the spermatic cord, the testis is in effect suspended in the scrotal cavity like a bell clapper and is free to rotate within the tunica vaginalis. This situation may be indicated by an abnormal horizontal lie of the testis. Testicular maldescent may also predispose to torsion. The torsion is usually toward the midline septum, that is, the right testis rotates in a clockwise direction and the left in an anticlockwise direction from the examiner's point of view.



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Recurrent torsion

There is a history of episodic testicular pain, with or without swelling of the testis. The testis may lie more transversely in the scrotum, but may otherwise appear completely normal. However, the history alone indicates that both testes should be fixed by early surgery.

DIFFERENTIAL DIAGNOSIS

Torsion of a testicular appendage

Torsion of a testicular appendage presents at 4–10 years of age. Careful palpation will reveal a tender nodule associated with the upper pole of the testis; the lower part of the testis is not tender. On transillumination this nodule may appear as a dark spot. If the diagnosis of torsion of a testicular appendage can be made on clinical assessment, neither hospital observation nor operation is mandatory.

Acute epididymo-orchitis

Unilateral epididymo-orchitis is common in adults but rare in children, in whom it is likely to be associated with infection or anomaly of the urinary tract.

Mumps orchitis

This rarely, if ever, occurs before puberty, and appears within 3 days to 1 week after the onset of parotitis. It is usually bilateral.

Idiopathic scrotal edema

In this condition the erythema and swelling of the scrotal skin spread into the groin, perineum, or base of the penis. Pain is not a major feature. The skin may be tender, but the underlying testis and cord are normal. The peak incidence is between 4 and 6 years of age. The diagnosis can usually be made clinically; ultrasound may have a limited role in confirming this, and all that is required is reassurance. However, if there is any doubt, scrotal exploration should be undertaken.

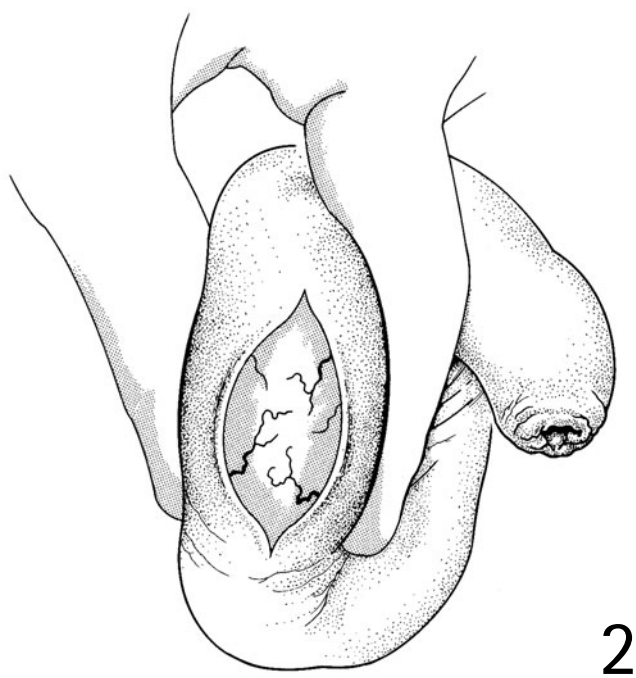
Incarcerated inguinal hernia

The symptoms and signs of torsion of an undescended testis closely resemble those of an incarcerated inguinal hernia – a painful, tender swelling in the groin – but are associated with an empty scrotum. Since urgent surgical exploration is mandatory for both conditions, differentiation is, perhaps, unnecessary.

OPERATION

Timing

In most cases, the diagnosis of acute torsion of the testis must be made clinically unless techniques such as Doppler ultrasonography or radioisotope scanning are immediately available. Any delay in operation once the diagnosis is suspected will prejudice the survival of the testis.



2 The testis is most easily delivered from the scrotum through an incision over its longitudinal axis, or, more cosmetically, obliquely in the line of the scrotal rugae. A third option is an incision through the midline raphe.

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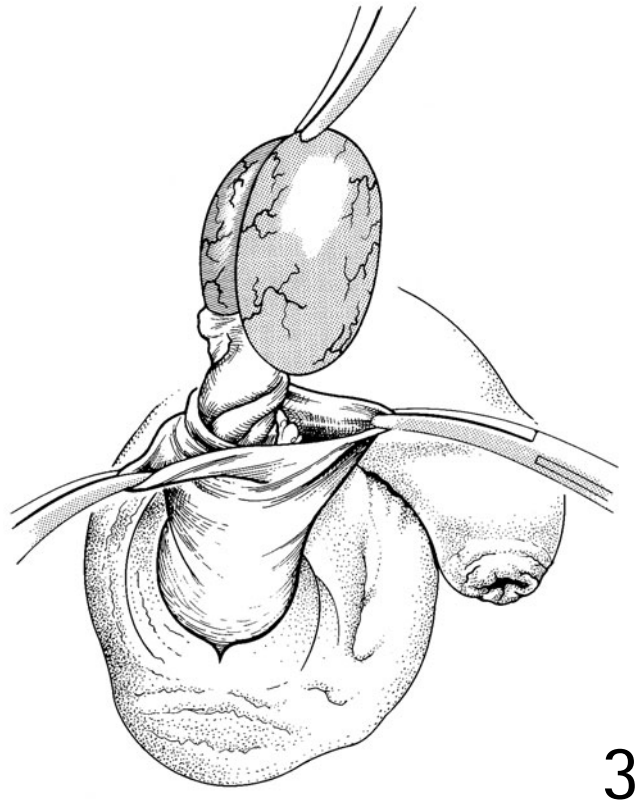
3 The testis is delivered from the tunica vaginalis and the cord is untwisted.

The tunica albuginea is incised in order to release the pressure on the underlying tubules and to assess viability.



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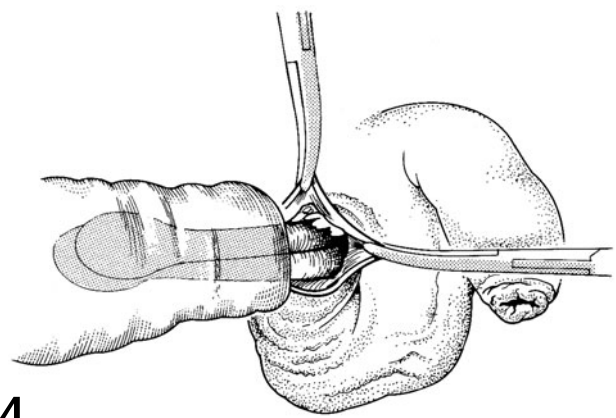

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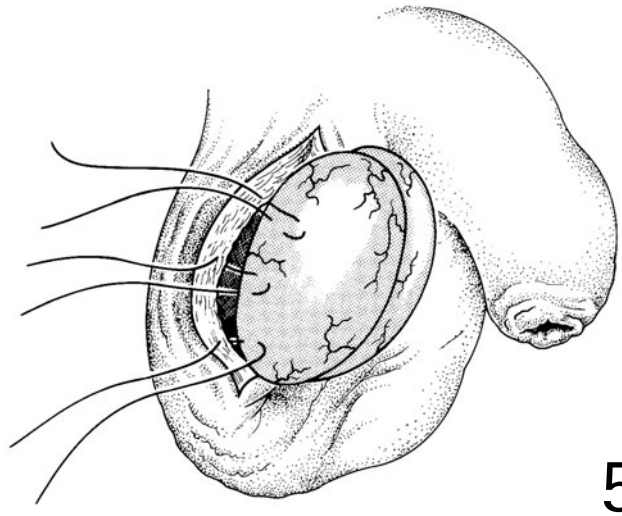
4 The untwisted testis is wrapped in moist, warm swabs and its color carefully observed. While waiting to confirm whether perfusion has been re-established, the contralateral testis may be fixed.

Conservation or removal of the testis

If the testis is completely black and necrotic and is deemed non-viable, it should be removed. The spermatic cord is ligated within the scrotum with a strong absorbable suture and the testis excised. If there is any question that some perfusion might be re-established, the testis should be conserved. It is imperative to fix the contralateral viable testis.

Fixation of the testis

5 The testes should be everted from the tunica vaginalis and fixed by three 6/0 monofilament sutures placed between the tunica albuginea and the lateral wall of the scrotum at the upper and lower poles and the equator. It is controversial as to whether these sutures should be absorbable (polydioxanone – PDS) or non-absorbable (polypropylene – Prolene).



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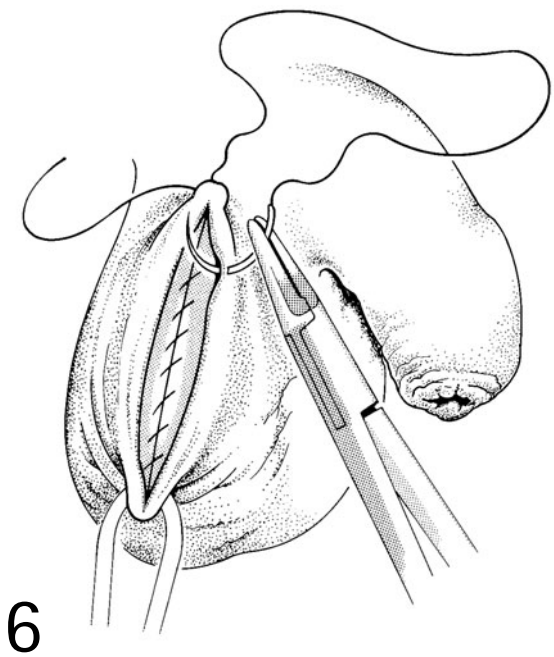

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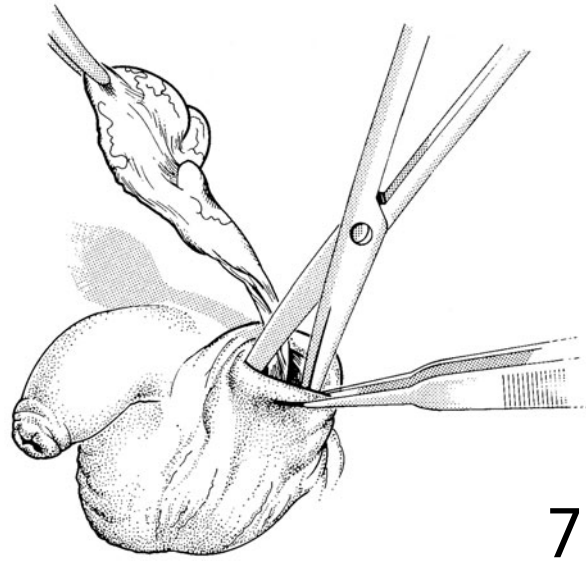
6 The wound is closed using continuous or interrupted 4/0 to 6/0 sutures, depending on the age of the child. Short-lasting absorbable sutures such as Vicryl Rapide or Monocryl should be used. If the incision was made through the midline raphe, a layered closure to recreate the midline septum is advisable. (In pubertal boys, some form of scrotal support may be appreciated.)

Alternative method

Experimental evidence suggests that fixation of the testis in a scrotal pouch may cause less damage to the testis and, more importantly, offer better fixation, preventing subsequent torsion.

7 The testis is delivered through a horizontal scrotal incision and the tunica vaginalis is everted. The testis is retracted superiorly onto the abdominal wall. A fine-toothed pair of forceps lifts the inferior margin of the scrotal skin incision. An adequate scrotal pouch can be made by blunt dissection. The testis – still everted from the tunica vaginalis – is placed in the scrotal pouch. The scrotal wound is closed using continuous or interrupted 4/0 to 6/0 short-lasting absorbable sutures such as Vicryl Rapide or Monocryl.



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Circumcision, meatotomy, and meatoplasty

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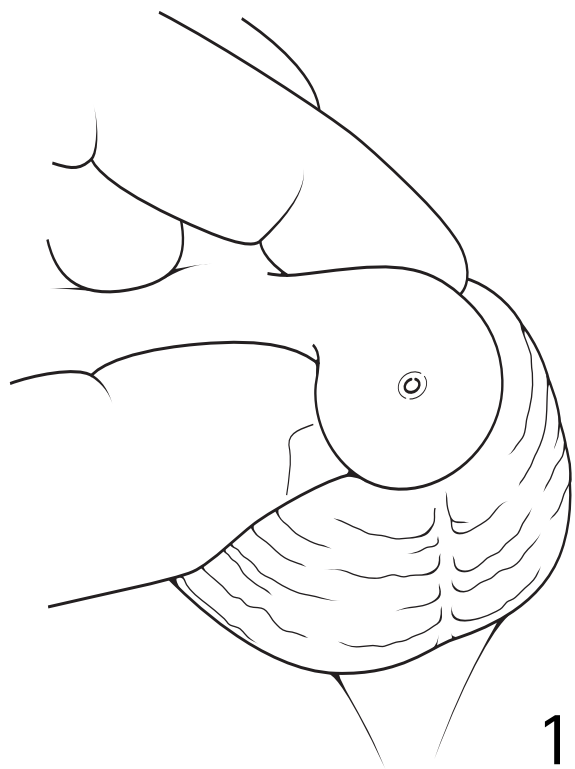
PRINCIPLES AND JUSTIFICATION

Indications

Most circumcisions are performed for non-medical reasons, namely for religious beliefs or cultural practices. Jewish boys are circumcised on the eighth day of life, and Muslim boys are usually circumcised during childhood. In addition, many boys are circumcised in the neonatal period, particularly in the USA.

The only true medical indication for circumcision is a pathological phimosis, which is usually related to balanitis xerotica obliterans (BXO). Other indications include recurrent balanoposthitis and in boys at risk of recurrent urinary tract infections due to an abnormal urinary tract (e.g., vesicoureteric reflux and posterior urethral valves).

Contraindications for circumcision include hypospadias and buried penis. Meatal stenosis can arise following circumcision in between 0 and 11 percent of cases, particularly in boys with BXO. The majority of these patients can be treated by a simple meatotomy, but recurrent meatal stenosis may require meatoplasty.



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PREOPERATIVE

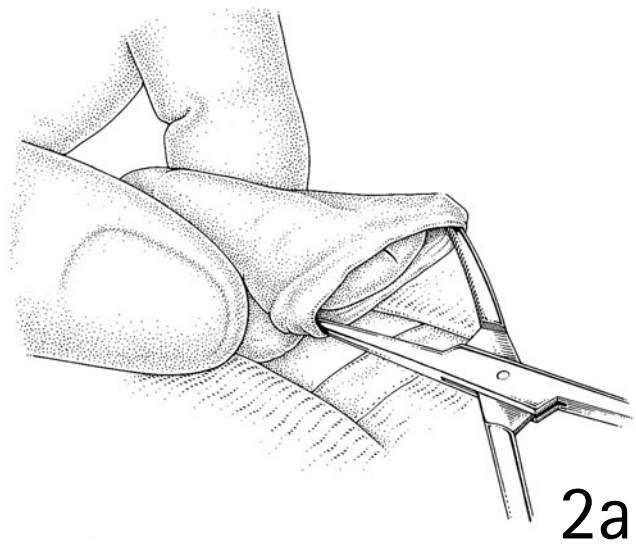
Anesthesia

Neonatal circumcision is usually performed under local anesthesia using a penile block. Older boys require general anesthesia with the addition of caudal anesthesia for postoperative pain relief.

OPERATIONS

Sleeve circumcision

2a–c The phimotic foreskin is dilated with artery forceps and completely retracted. Congenital preputial adhesions are divided and the glans is cleaned with antiseptic solution. A surgical pen is used to mark the line of cicumferential incision just proximal to the coronal sulcus, leaving a cuff of inner preputial skin below the glans.



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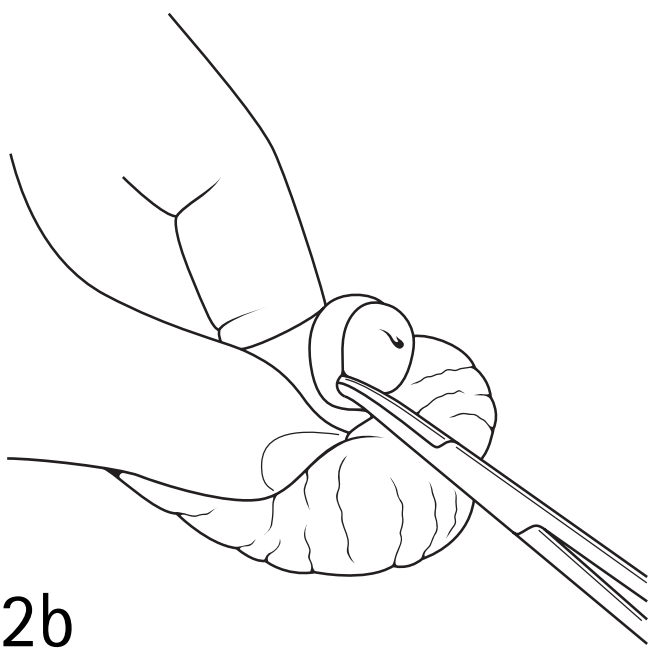

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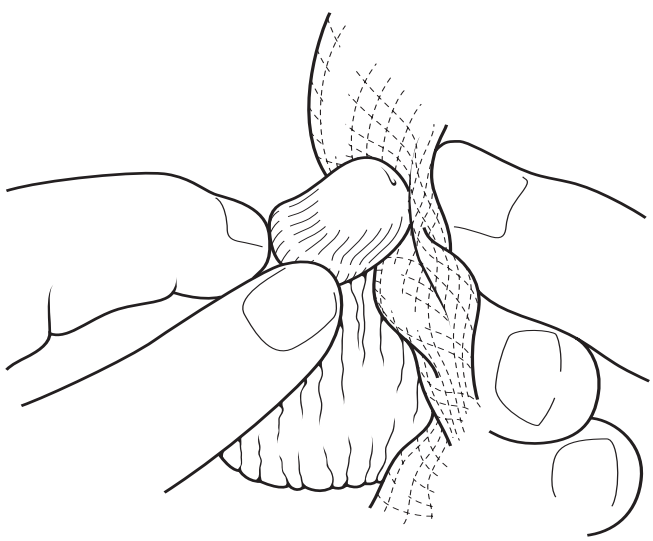
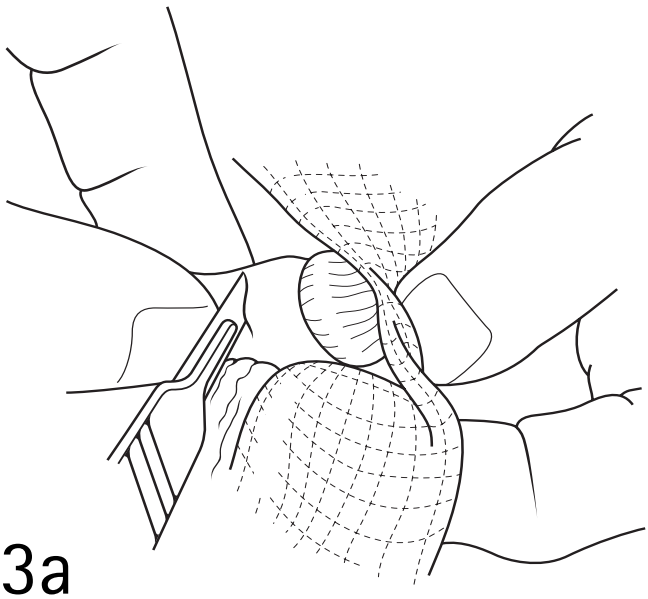


2c

3a–f The incision is then made with a scalpel knife to sufficient depth to allow the penile skin to be retracted proximally. The foreskin is elevated by placing one artery forceps ventrally and one dorsally. A circumferential line of incision on the penile shaft skin is marked with a surgical pen, allowing sufficient skin to cover the full length of the penis without tension. The skin is incised along the line with a scalpel knife. The excess foreskin is removed by dividing the subcutaneous layer between the inner and outer skin layers with bipolar diathermy.



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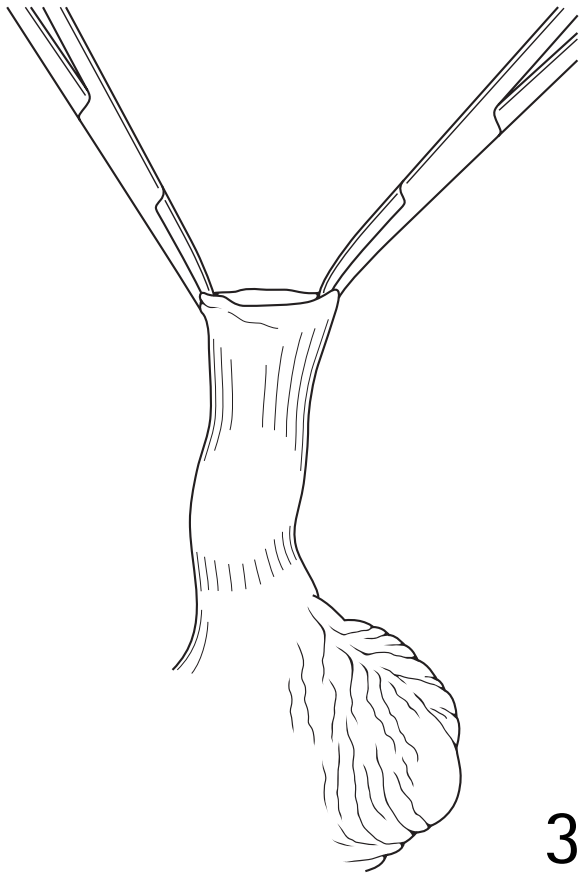

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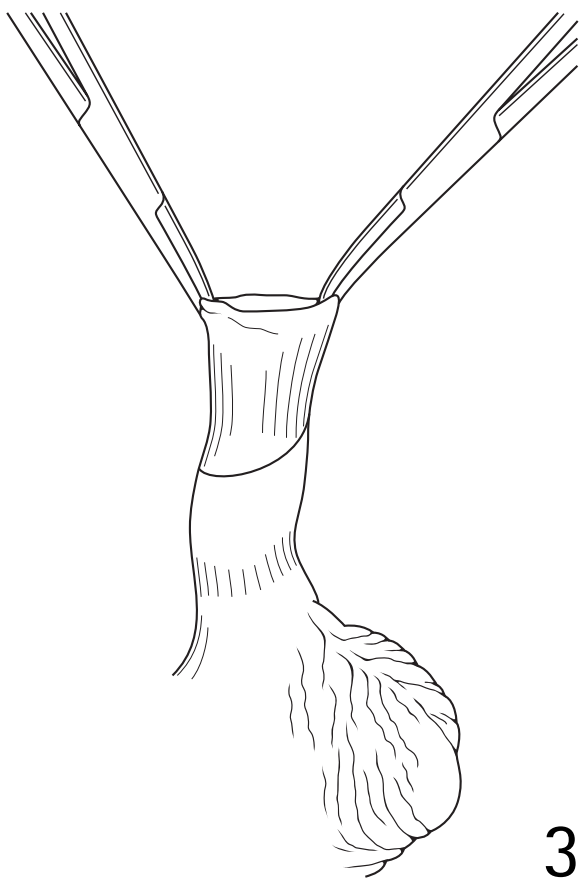

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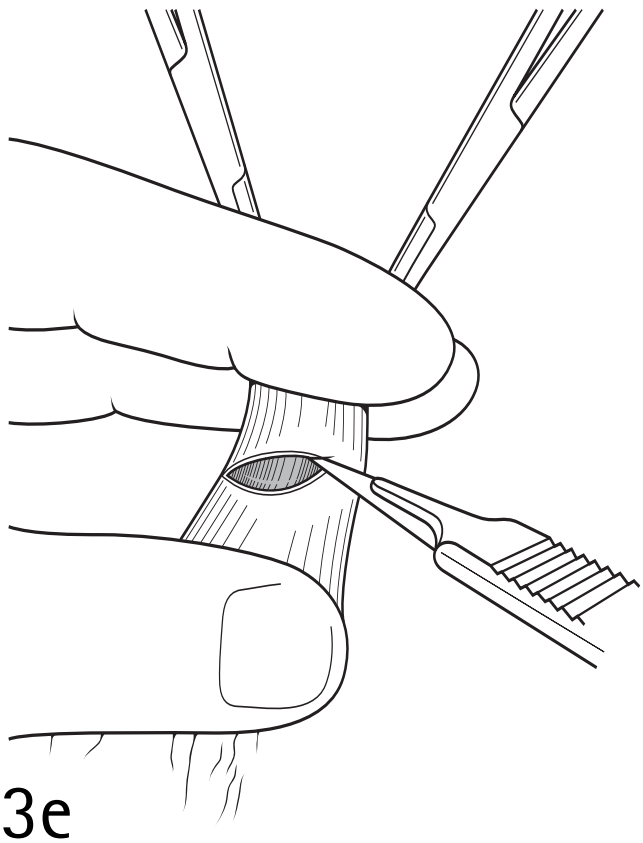

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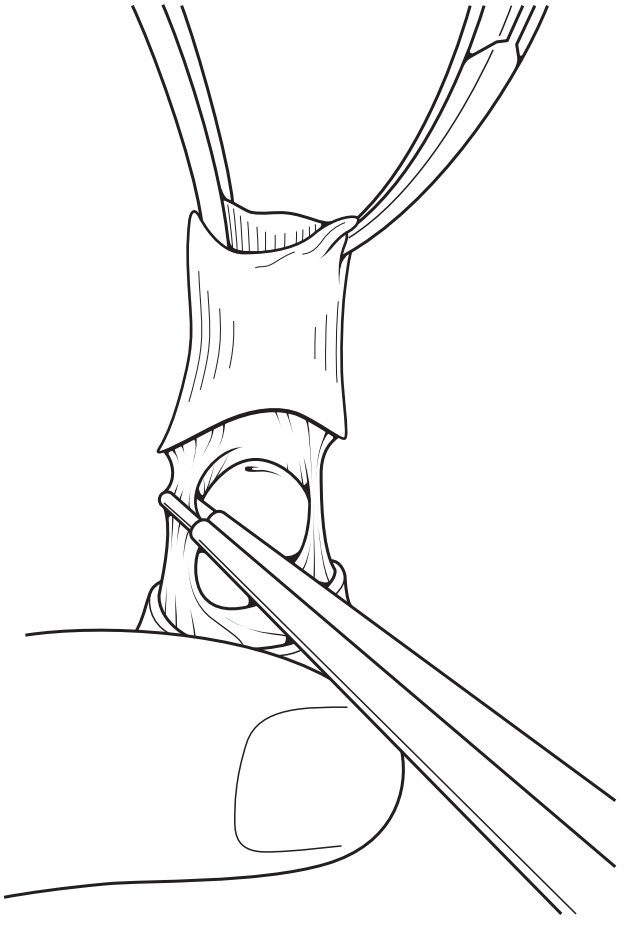
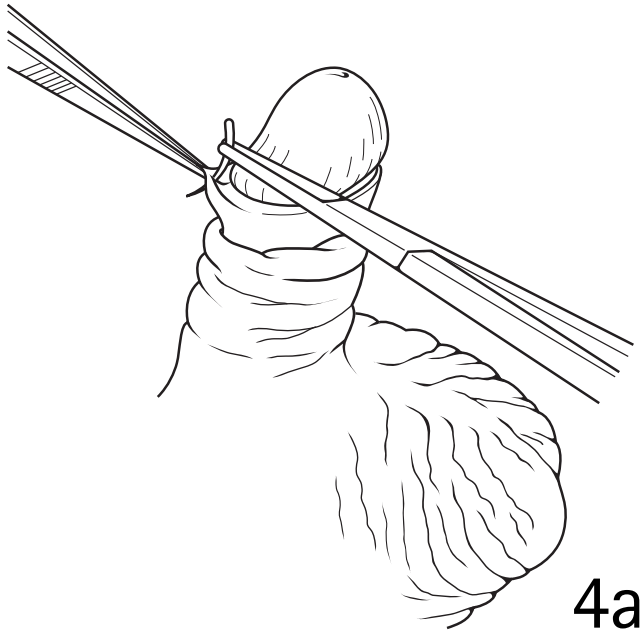


3f

4a–c The outer skin is retracted and any bleeding points are coagulated with bipolar diathermy. The wound is closed with an interrupted absorbable suture, such as 6/0 Vicryl Rapide or Monocryl. The application of chloramphenicol ointment twice daily for 7 days may help to prevent infection and stops undergarments adhering to the penis.



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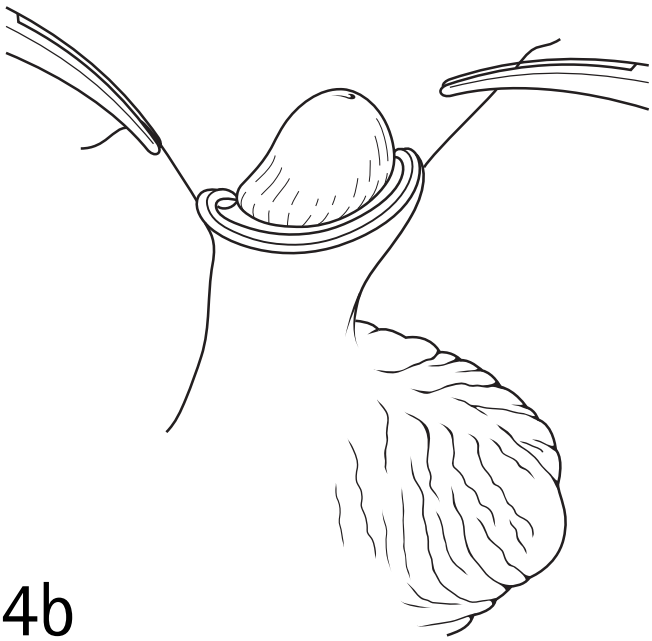
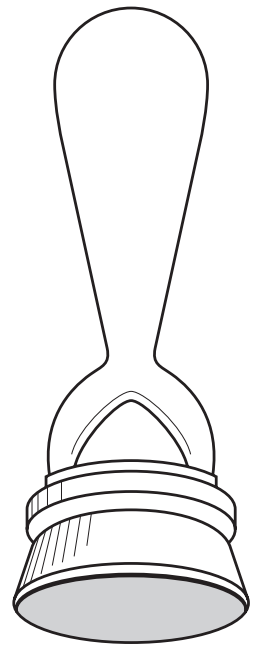


4b

4c

Plastibell circumcision

5a The Plastibell device is a development from metallic devices used as templates for ischemic necrosis of redundant foreskin in circumcision. It is well established as a quick and safe method in neonates and is widely practised.



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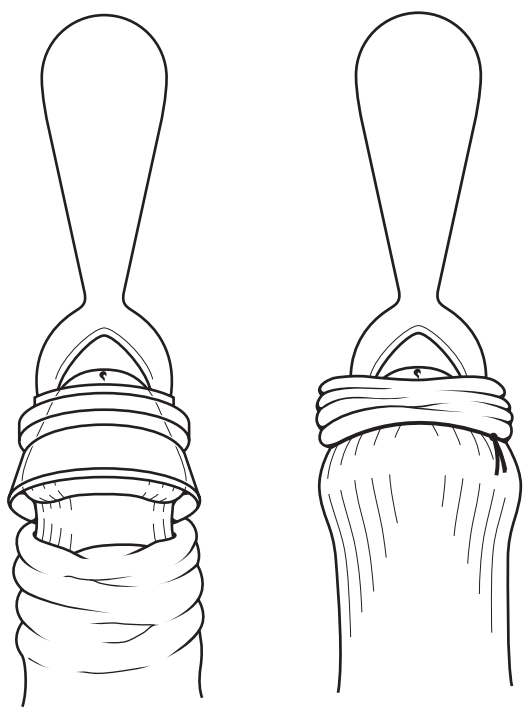

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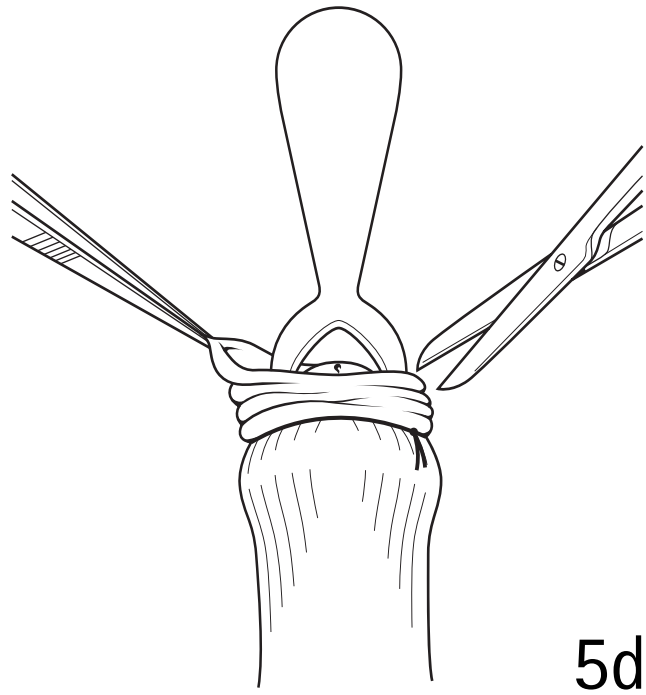
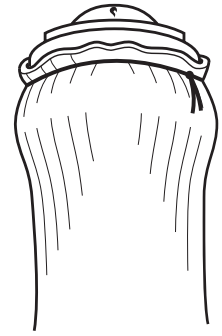
5c

5b,c The preputial opening is dilated and the adhesions between the prepuce and the glans are freed with a probe. The prepuce is fully retracted to expose the coronal sulcus, which should be cleaned of smegma. An appropriate-sized bell is placed over the glans as far as the preputial reflection. A correctly sized bell should stay in position comfortably and without pressure on the glans. A suture is tied around the prepuce onto the outer ridge of the bell as tightly as possible.



5d

5d,e After about a minute, the preputial skin becomes insensitive and is trimmed off immediately distal to the ligature with iris scissors. The handle is snapped off, leaving the bell in position. Skin necrosis will occur at the site of the suture and will allow the bell to separate and fall off, after a mean of 9 days. In the meantime, the child voids through the open end of the bell.



5e

COMPLICATIONS

Circumcision is often regarded as a minor surgical procedure, usually delegated to junior surgical staff. However, there are probably more complications associated with this operation than with more complex urological procedures.

HEMORRHAGE

Careful attention to hemostasis using bipolar diathermy during surgery will prevent primary postoperative hemorrhage. If postoperative hemorrhage does occur, a pressure dressing may arrest the bleeding in the first instance. If in doubt, it is best to return the child to the operating theater and deal with the bleeding point under general anesthesia.

INFECTION

Infections after circumcision are not uncommon, with an incidence of around 5 percent. These infections generally respond well to oral antibiotics and regular bathing.

URETHROCUTANEOUS FISTULAS

Fistula formation is a rare complication of circumcision and is usually seen at the level of the coronal sulcus. The two most common causes are overzealous use of diathermy and a suture placed too deeply to stop bleeding from the frenulum. Surgical repair should be performed 6 months after the original circumcision.

REMOVAL OF TOO MUCH OR TOO LITTLE SKIN

Removal of too little skin can cause annular scarring at the line of excision and recurrence of the phimosis. In such cases, a further circumcision is required. Removal of too much skin may give the appearance of a buried penis, but only rarely is skin grafting required to achieve a satisfactory appearance.

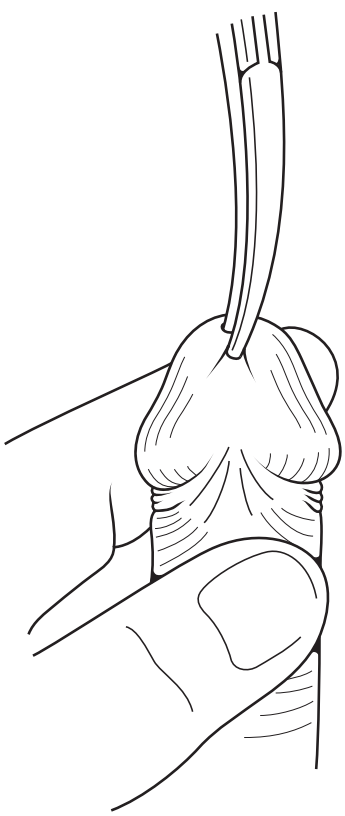
MEATAL STENOSIS

Meatal stenosis is the most significant complication, reported in up to 11 percent of cases. In neonates, it may be the result of ammoniacal dermatitis due to contact with wet nappies. In older children, it is usually the result of the BXO, which affects the prepuce causing the original phimosis. Such cases are troublesome to treat but usually respond to meatotomy and/or meatoplasty.

Meatotomy

6a–c The meatus is calibrated with a lacrimal probe to determine the severity of the stenosis. An artery forceps is used to crush the tissue ventrally through the meatus for a period of 1 minute. The artery forceps is removed and the crushed tissue is divided with iris scissors. Bleeding is rare with this technique, but if it does occur, it can be readily controlled with bipolar diathermy.

The main complication of this procedure is recurrence of the meatal stenosis, which may require a more formal meatoplasty.



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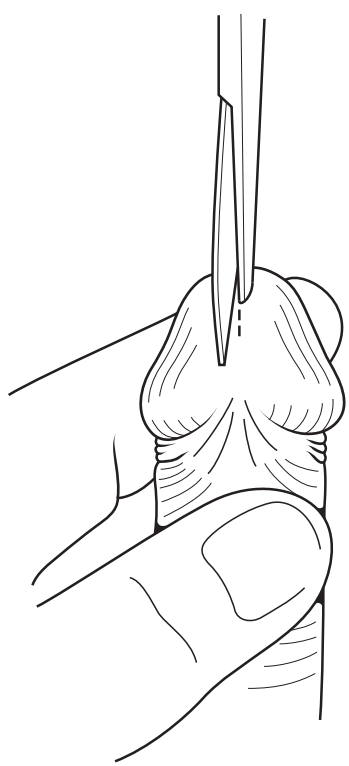

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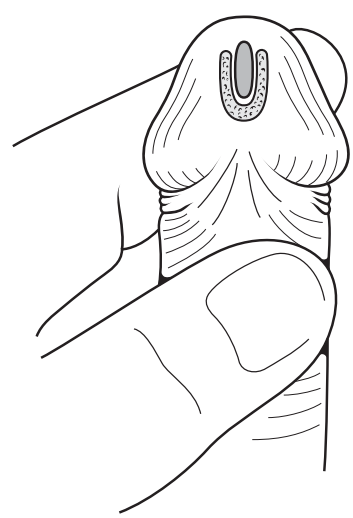

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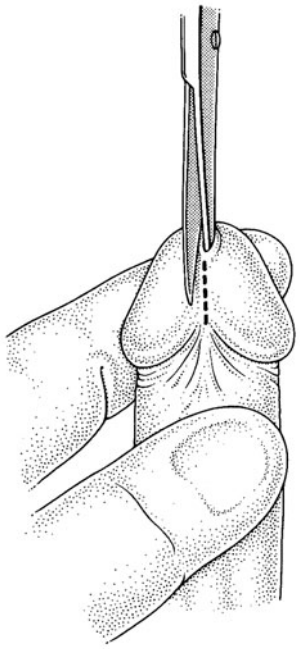
Meatoplasty

7a–c A pair of iris scissors is used to make a generous ventral incision through the meatus. The edges of the urethral mucosa are approximated to the adjacent skin with 7/0 or 6/0 absorbable sutures.

This procedure creates a large meatus with the appearance of a glanular hypospadias. This may cause some deviation of the urinary stream, but it is unlikely to cause functional difficulties.



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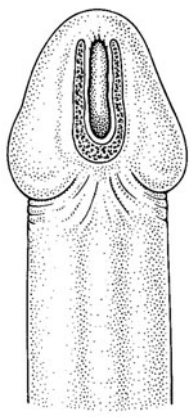

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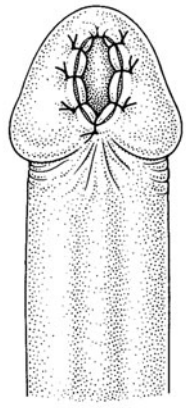

7c

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Bladder exstrophy closure and epispadias

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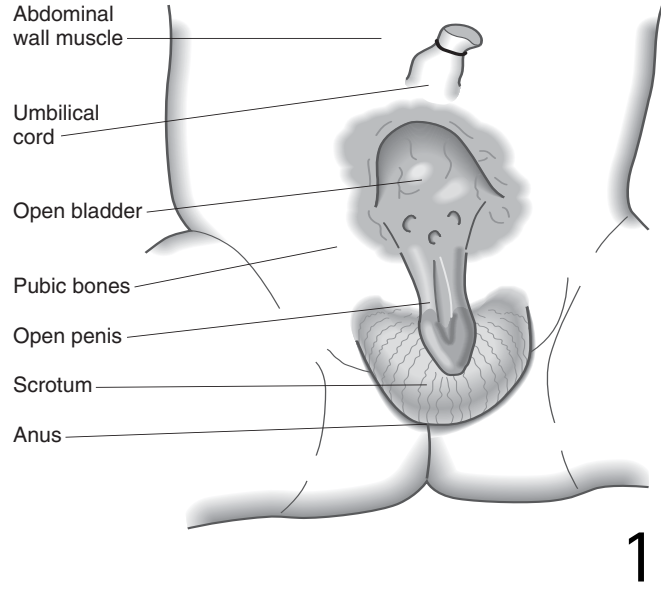
PRINCIPLES AND JUSTIFICATION

Bladder exstrophy has an incidence of 1 in 30 000 live births, with a preponderance of males (males:females 3:1). Primary epispadias has a similar male bias but is much less common, with an incidence of 1 in 120 000. In the UK there are between 15 and 20 cases of primary bladder exstrophy and 4 to 6 cases of primary epispadias presenting per year. The surgical management of this rare group of anomalies tends to be focused on major surgical centers and this has enabled the refinement of different surgical approaches. The aims of exstrophy and epispadias repair are:

- to place the bladder within the abdomen and ultimately to achieve urinary continence;
- to create satisfactory genital appearance and near-normal function;
- to preserve or enable fertility.



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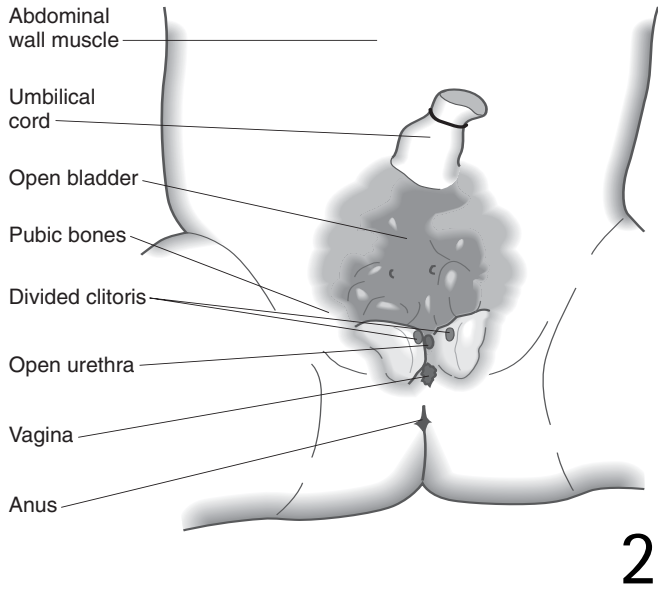
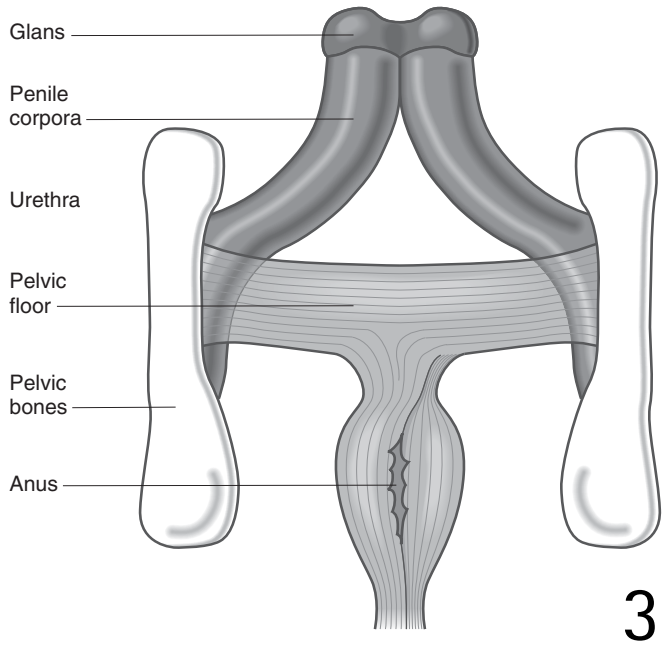


2

1–2 Bladder exstrophy presents at birth with a low-set umbilicus and a split rectus abdominis muscle attached on either side to separated pubic rami. The bladder fills this low abdominal wall defect as an open plate with the ureters draining directly onto its surface. It is continuous with the open urethral plate, which runs down the dorsum of the penile corpora in the male to an open glans. In the female, the urethral plate is shorter and runs between a divided clitoris and the labia minora to the anterior rim of the vaginal orifice.

Inguinal hernias are a common association. Nearly 80 percent of boys have a patent processus vaginalis, and during the primary exstrophy closure it is routine practice to perform bilateral herniotomies. This is less common in girls (incidence of 15 percent) and so the need to perform routine herniotomy is debatable. The perineum appears foreshortened due to lack of anterior structures, principally the anterior pelvic ring. The anus is anteriorly placed, principally because of this lack of anterior structures, although it may be more anteriorly placed relative to the back.

3 Separation of the pubic bones (referred to as the diastasis) varies considerably. In primary epispadias, where only the urethra is open, the recti may be together and the symphysis may actually be joined, whereas in the most severe forms of exstrophy the diastasis in newborns may be greater than 5 cm. The defect in the abdominal wall and the degree of difficulty in closure of the abdomen are directly related to the extent of the diastasis. The proximal ends of the penile corpora are attached on either side to the inferior pubic ramis. Whilst they may be shorter than normal corporal bodies, the separation of the pubic symphysis results in a Y-shaped penis and further limitation of the penile length. The urethra in the male is an open plate on the dorsum of the corpora and it has no relation to any sphincter muscles at birth. A normal sphincter complex is present in the perineum, however, and can be demonstrated by direct stimulation in the area of the prostate gland between the proximal penile corporal attachments.



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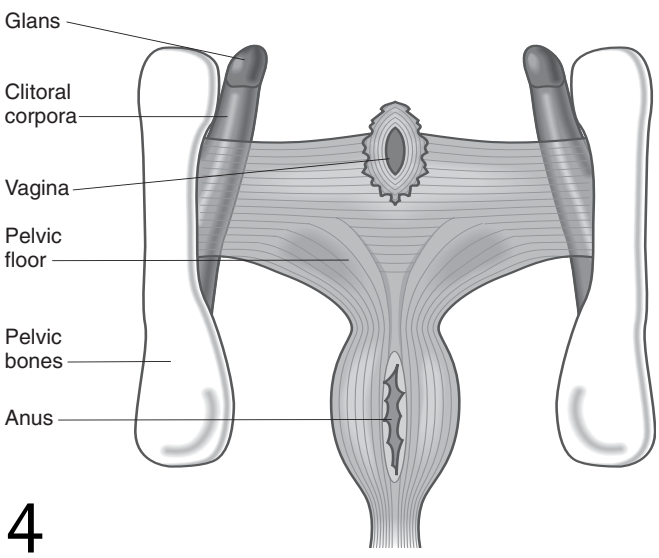


4

4 In females with primary bladder exstrophy there is a similar anatomical arrangement to this. The divided clitoral corpora are attached to the pubic rami with muscle present, running between them. The urethral plate is anterior to the vagina and not associated with this muscle complex.

The inadequacy of the penis has led some in the past to opt for female sex of rearing. Current reconstructive procedures, however, with detachment of the penile corpora, produce greater penile length. As a result, sexual reassignment is never required in cases of primary bladder exstrophy and may now be avoided in many cases of cloacal exstrophy.

Isolated male epispadias varies considerably in severity from that which simply involves the glans of the penis, to a much more severe abnormality involving the entire urethra, bladderneck, and sphincter complex. In all but the least severe cases, patients have stress incontinence, and many bladders fail to achieve capacity. For the majority of these cases, a reconstructive procedure is required, not only for the penis, but also for continence, and we have used the Kelly soft tissue reconstruction for this (see below). In females, the rare anomaly of isolated epispadias includes a bifid clitoris, a short patulous urethra, and a variably open bladderneck resulting a range of incontinence.

The current surgical approach is to close the exstrophy bladder shortly after birth, returning it to the lower abdomen and reconstructing the abdominal wall. The resulting bladder cannot adequately store urine and continence is only rarely

achieved, the child characteristically dribbling continuously and requiring another procedure. A successful primary closure, however, does form a sound basis for the subsequent continence procedure in the form of soft-tissue reconstruction or Kelly's procedure. Through a complete mobilization of the pelvic soft tissues, including the penile corpora, pelvic floor, and bladder, the bladder outlet, proximal urethra, and sphincter mechanism can be reconstructed. The new bladder outlet resistance is a stimulus to its growth and an increase in capacity. These two factors combine to provide continence.

In addition, by mobilizing the penis in boys, greater penile protrusion is possible. In girls, a united clitoris and improved perineal cosmesis are achieved. In males, even greater penile length may be achieved by completely detaching the urethral plate from the dorsum of the corpora and creating a hypospadiac meatus. A subsequent urethroplasty is required to create the distal urethra and a terminal meatus. This procedure has been successful in cases where penile length is significantly impaired and may be a primary indication for the Kelly procedure in patients who have already had a continent urinary diversion. An alternative approach to the penis is to combine the soft-tissue mobilization with a Cantwell–Ransley epispadias repair. The decision to do this is made at the time of operation and depends on penile and urethral length.

As historically patients have not normally been continent following exstrophy repair, bladderneck resistance procedures such as the Young Dees Ledbetter bladderneck reconstruction have been required. In the majority of patients, an additional bladder augmentation cystoplasty (usually an ileocystoplasty) to increase capacity and an appendicovesicostomy (Mitrofanoff channel) has provided a continent conduit for bladder emptying by intermittent catheterization. This will remain the ultimate management strategy in a number of patients, but currently the soft-tissue mobilization is allowing some (at least 50 percent) to void spontaneously per urethra with an adequate native bladder for storage.

As with many conditions in pediatric urology, lack of standardized assessment and nomenclature has led to much confusion about the results of treatment of bladder exstrophy. Our group has proposed a standardized definition of levels of incontinence in bladder exstrophy by which all interventions should be assessed (Table 88.1).

Table 88.1 *Levels of continence in exstrophy*

0	Dribbles urine all the time with no control
1	Able to retain urine with a 'dry interval' Some control but still wearing protection
2	Sufficient dry intervals by day In underwear and not needing protection Wet at night
3	Dry by day and night, no protection or accidents 'Normal' as peers

SURGICAL STEPS TO CLOSURE

The surgical steps may be summarized as follows:

1. Closure of the bladder in the neonatal period within the first 24–48 hours may proceed with approximation of the pubic bones without the need for osteotomy. After 48 hours, osteotomy is advised and a modified Salter anterior iliac osteotomy is currently recommended. Postoperative immobilization of the lower limbs with a frog plaster or a mermaid dressing is recommended whether or not osteotomy has been performed. This immobilizes the baby and greatly facilitates nursing care. In complex cases, restricted handling and continuous suction of bladder urine with a Replogle tube postoperatively are also advised.
2. Late closure usually requires osteotomy, as does reclosure after dehiscence. In these cases, external fixation of the pelvis is recommended.
3. Radical soft-tissue reconstruction (Kelly procedure) is proposed 6–9 months after primary closure, although it can be performed at any age. Bilateral ureteric re-implantation is usually performed at this time, to avoid the compromise to the upper urinary tract that a combination of increased outlet resistance and ureteric reflux may bring.
4. Completion of the urethroplasty in boys is performed in the third year of life or 18 months after the Kelly procedure. This is usually performed in two stages, with skin grafting to create a neourethral plate and glans groove on the penis, followed by urethral closure 12 months later.
5. Primary male epispadias is treated by the Kelly procedure if there is incontinence or poor penile length, or both. The timing of this procedure varies, but it may be performed after 6 months of age. Less severe cases (those that are continent) are treated by the Cantwell–Ransley epispadias repair.
6. Primary female epispadias is usually incontinent and so a Kelly soft-tissue mobilization is used.
7. Bladderneck reconstruction, augmentation cystoplasty, and appendicovesicostomy are available for patients in whom spontaneous voiding continence is not achieved by the above procedures. Continence may develop slowly after the Kelly procedure, so this salvage surgery is usually not performed before the fifth year of life.

The aim of this chapter is to show the basic steps of bladder closure, radical soft-tissue reconstruction, and epispadias repair; however, a detailed guide to the management of the patient with bladder exstrophy can be found in the further reading recommendations.

PREOPERATIVE

In these days of antenatal diagnostic ultrasound, many exstrophy cases are diagnosed in utero with characteristic non-visualization of the bladder, a low insertion of the

umbilical cord, a short, thick phallus, and an irregular lower abdominal wall. Families should be counseled during pregnancy so they understand the implications of the diagnosis and the postnatal surgical strategy. After birth, the bladder is covered with plastic wrap and the baby is transferred to the surgical center. Surgery may be delayed for up to 24–48 hours to allow the mother to recover from the delivery and for a preliminary work-up of the baby, to include renal ultrasound and a blood cross-match. Premature babies may require a longer period of medical stabilization before closure, which can be delayed for several weeks if this is medically indicated.

Vitamin K is given to neonates if it was omitted at birth, and they are started on oral clotrimazole suspension to help prevent fungal colonization of the urinary tract. Referring hospitals and doctors looking after preoperative neonates should be encouraged to use the lower limbs for blood sampling and cannulae. These veins will not be available postoperatively, when the surgical team will need good access via the upper limb veins.

General anesthesia is required with the usual neonatal precautions. A caudal anesthetic is helpful or, preferably, an epidural catheter is inserted to provide excellent peroperative and postoperative pain control in both babies and older children having the Kelly procedure. In some difficult neonatal cases and where abdominal closure has been tight, a period of postoperative ventilation in an intensive care unit is required.

BLADDER EXSTROPHY CLOSURE

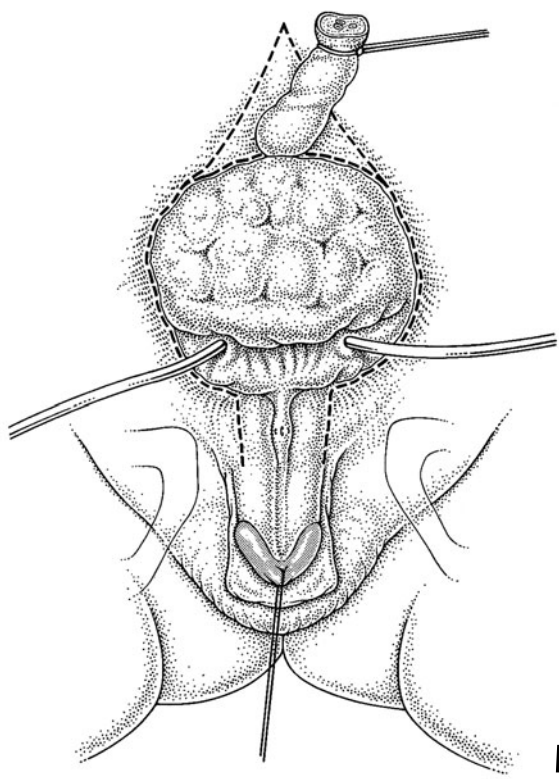
OPERATION

Position of patient

The patient is positioned flat and supine. Total lower body preparation with individual wrapping of the legs allows maximum maneuverability at the time of the surgery and access for the orthopedic surgeons to perform osteotomies if required. Amikacin (7.5 mg/kg), metronidazole (7.5 mg/kg) and ampicillin (6 mg/kg) are given intravenously in the anesthetic room and continued postoperatively for at least 48 hours.

Skin incision

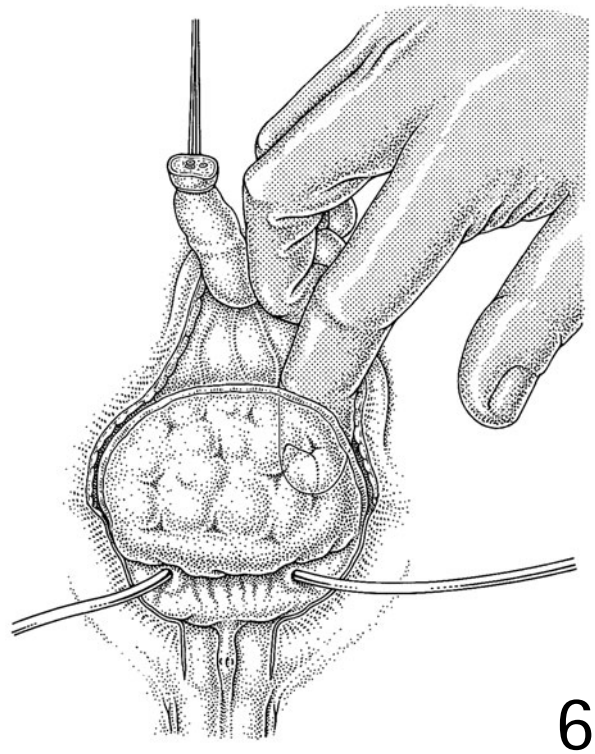
5 The umbilicus is ligated and trimmed but retained. A stay suture is placed through the end of the glans penis to aid its retraction (4/0 monofilament with a round-bodied needle). Incisions are made beginning in the midline above the umbilicus, extending around the margins of the bladder, and forward onto the root of the penis, on either side of the urethral plate as far as the distal limit of the veru montanum. The incisions are deepened with diathermy but the distal incisions may be left superficial at this stage.



5

Bladder mobilization

6 Working from above the umbilicus, the incision is deepened through the midline to expose the umbilical vein, without opening the peritoneum. As dissection continues distally, the umbilical arteries serve as a guide on each side to the extraperitoneal plane. Both are divided, allowing the umbilicus to move upward to a more anatomical position. Careful blunt dissection opens up a plane behind the rectus muscles on each side, in front of the ureter (stent in position for palpation), and down to the pelvic floor at the bladderneck.



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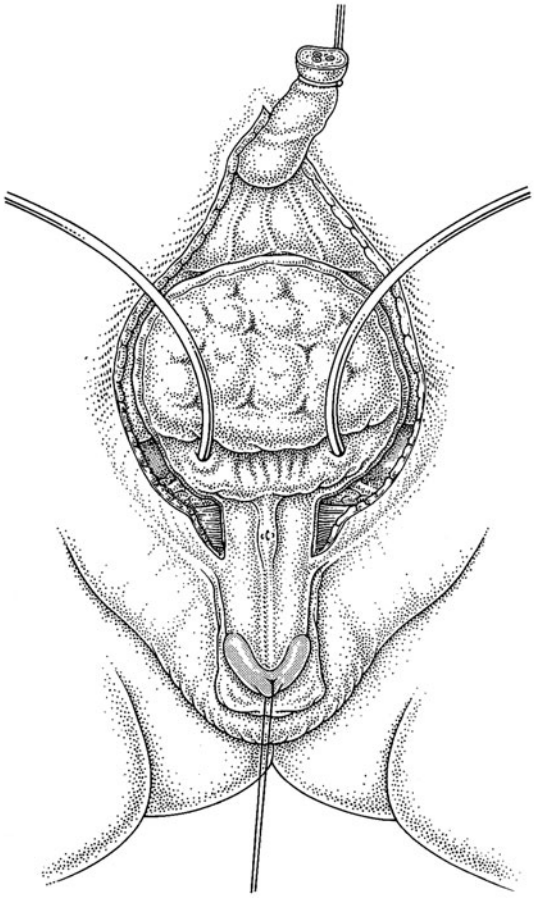


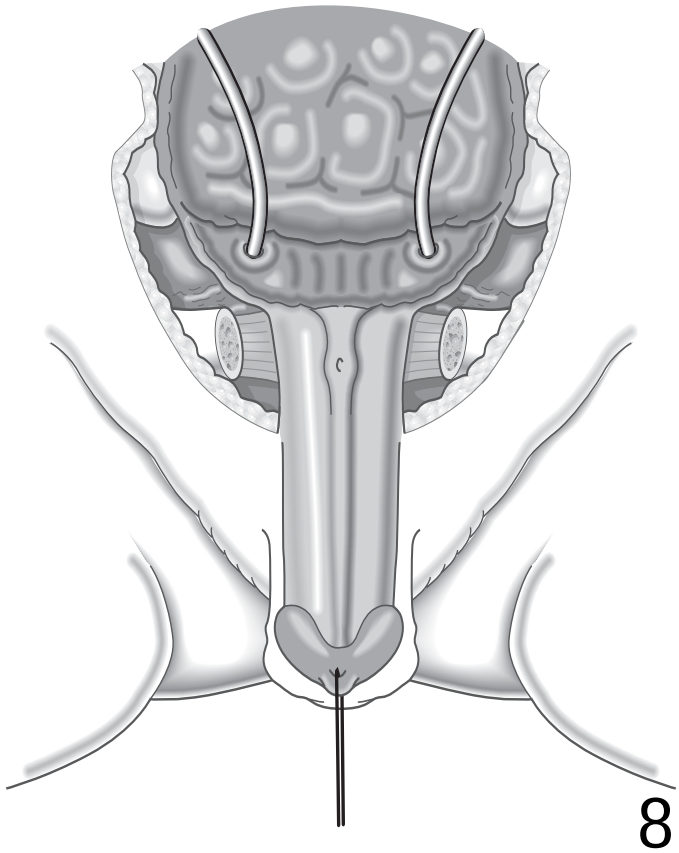
7

Completion of distal incision

7 With a finger in position behind the abdominal wall, the distal incisions may be completed with diathermy to create freedom of the bladder on each side. The intrapubic bar tissue fusing with the bladderneck is now visible and is an important landmark.

Dissection of proximal urethra

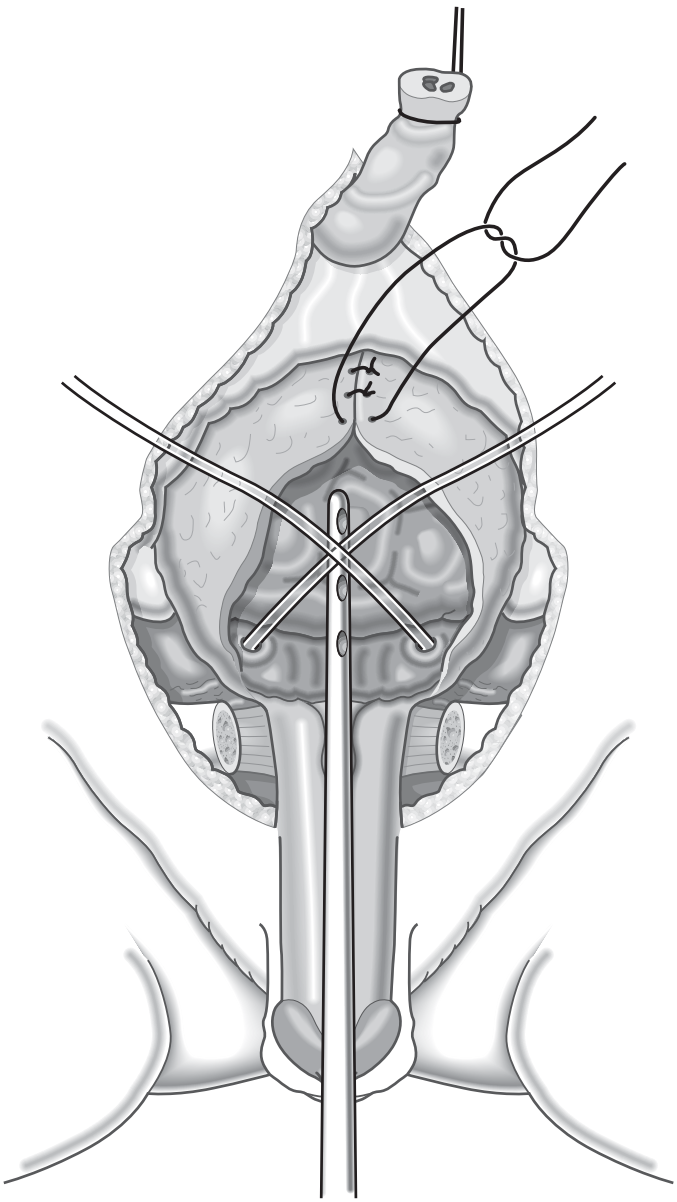
8 The intrapubic bar tissue is divided lateral to the veru montanum to release the bladderneck and allow its placement inside the pelvis. The ends of this tissue and the pubic bones are then available to bring together in front of the bladderneck. Radical corporal mobilization is not performed at this stage, although the incisions may need to be extended distally to facilitate the closure of the pelvic ring. It is preferable to stay above the corporal bodies in patients subsequently having a Kelly procedure.



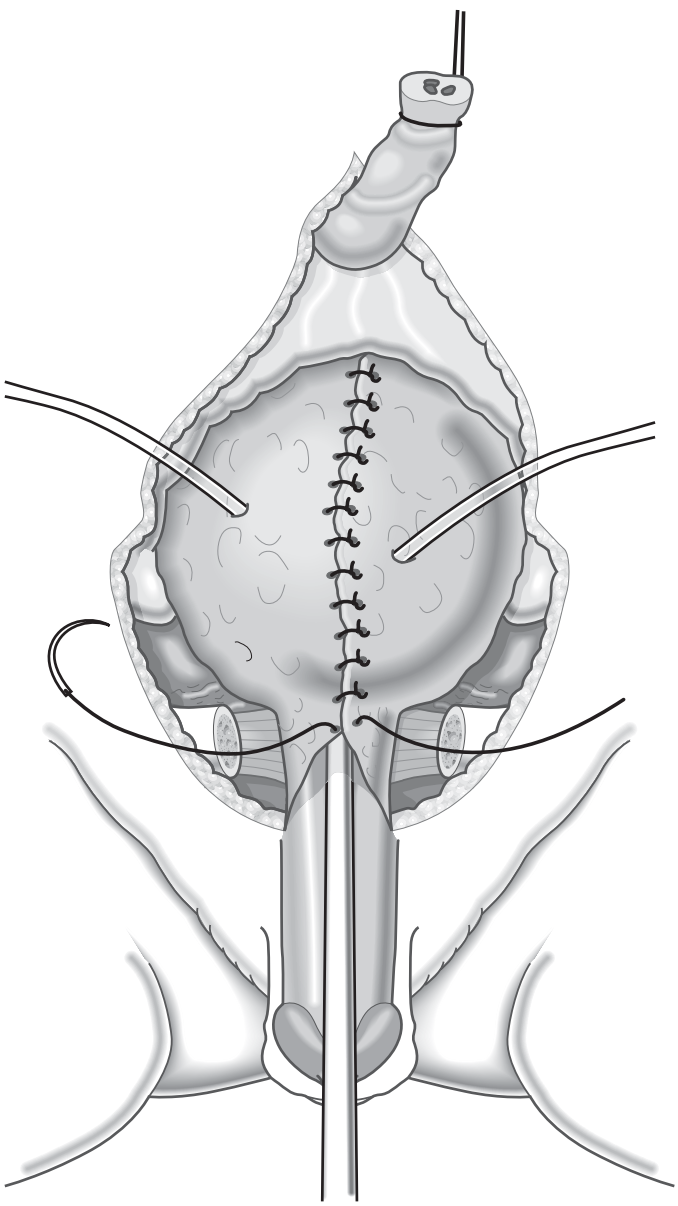
8

Bladder closure

9 The bladder and proximal urethra are now sufficiently free to drop back into the pelvis. An 8 Fr feeding tube catheter with opposed eyes to avoid obstruction is sutured to the bladder wall with 6/0 Monocryl. Bladder closure begins from the apex using interrupted 4/0 Monocryl sutures after trimming the bladder edges. The two ureteric feeding tubes (which have also been secured with 6/0 Monocryl sutures) are brought out between sutures. Closure is continued to the proximal urethra, although the lower sutures may be left untied until pelvic approximation is achieved.



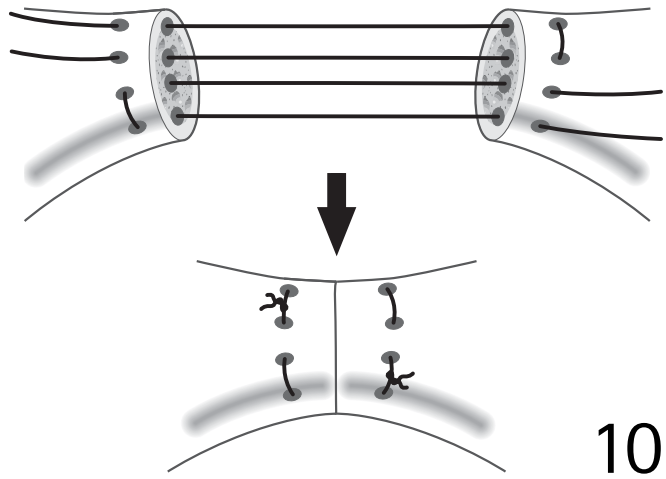
9a



9b

Approximation of pubic bones

10 One or two mattress (0 or 1) polydioxanone sutures (PDS) are laid in position in the pubic bones before abdominal wall closure and are tied once rectus muscle approximation is complete. This helps to prevent a bowstring effect cutting back into the urethra if the bones separate a little postoperatively.



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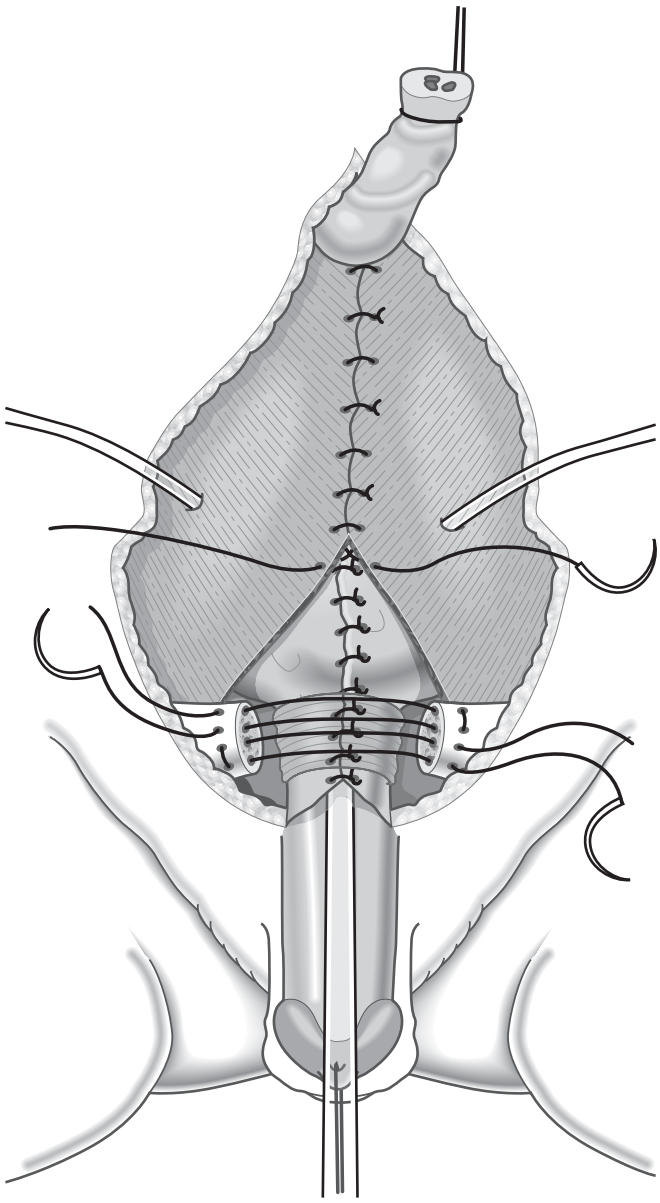


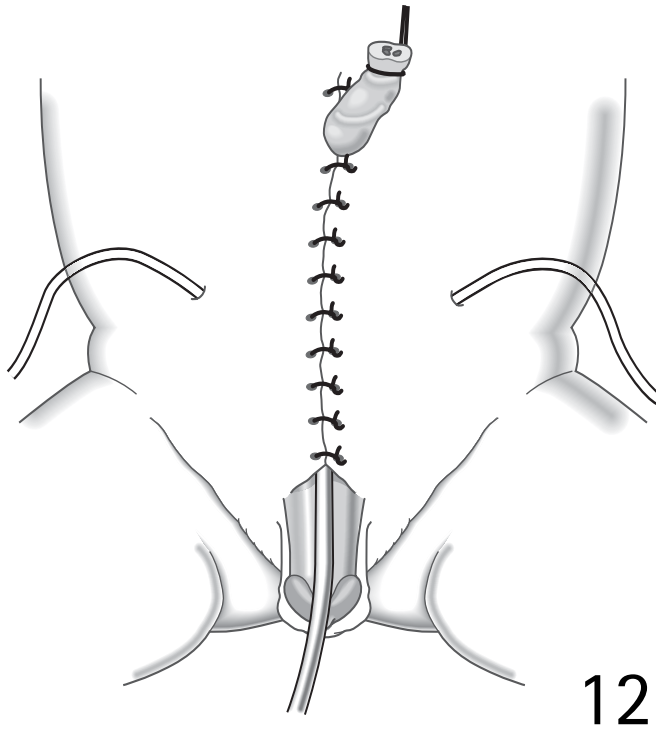
11

Transpositional omphaloplasty and wound closure

11 The umbilicus, suspended on the umbilical vein, is displaced to the apex of the abdominal incision. Closure of the abdominal wound begins by bringing the rectus muscles together, starting from above and proceeding downwards. The aim is to even out the tension of closure throughout the whole length of the wound so that the strain of closure is not taken by the symphysial sutures alone. Interrupted figure-of-eight sutures of 3/0 PDS are used proximally, increasing to 2/0 PDS as the closure proceeds distally. The ureteric stents are brought out on either side of the abdominal wall by cutting off their connectors and threading them through large intravenous cannulae, passed from the outside. Finally, the pubic bones are approximated by tying their sutures. During this maneuver, internal rotation of the hips by the assistant and compression of the pelvis may be helpful.

Skin closure

12 The skin is closed in two layers of interrupted 4/0 Vicryl sutures.



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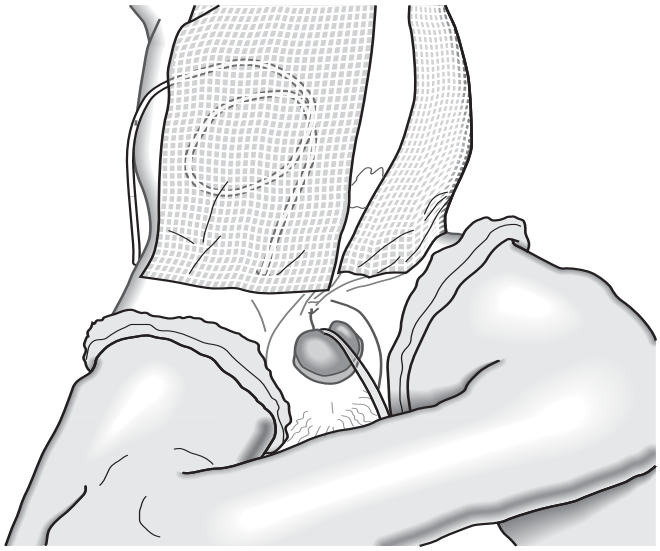


13

Immobilization

13 Either a frog plaster, with internal rotation of the hips and slight adduction with flexion of the knees, or a mermaid bandage is used to immobilize the lower limbs. This is kept in place for 3–6 weeks.

RADICAL SOFT-TISSUE RECONSTRUCTION (KELLY PROCEDURE)

This operation re-opens and dissects the bladder, mobilizes the urethral plate completely from the penile corpora, releases the pelvic floor muscle, and detaches the penis from the pubic rami – preserving its neurovascular bundle (the pudendal pedicle). It allows radical reconstruction of the bladderneck and urethra and re-creates a muscular sphincter. Another major benefit is the improvement in penile length that is achieved. In respect of the penis, there are two choices. Combining the operation with a Cantwell–Ransley epispadias repair leaves the urethral plate attached distally to the glans and enables a single-stage procedure, but at the expense of some penile length. Alternatively, completely detaching the urethral plate frees the penis for more length, but leaves a hypospadiac meatus and the need for a later distal urethroplasty.

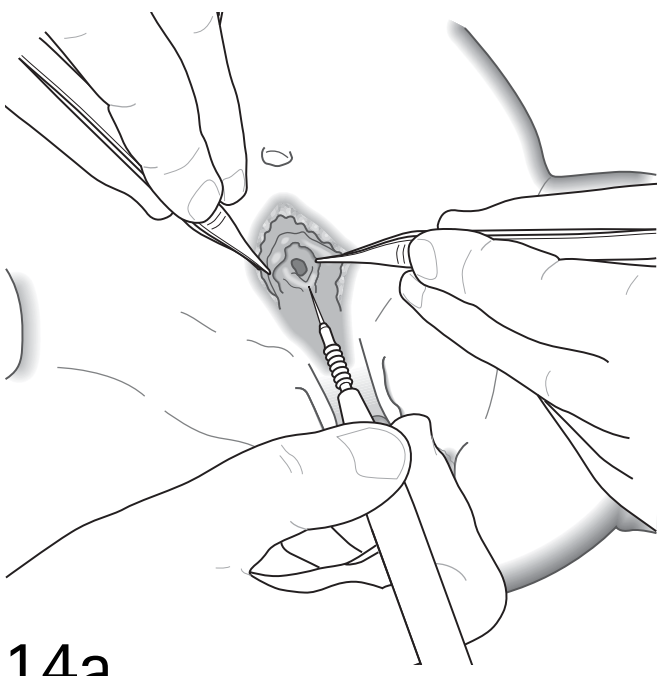
Position of patient

The patient is prepared as for primary closure and is supine on the table with a full lower body preparation and legs wrapped in sterile drapes and in the field to permit full mobility. In addition, a purse-string suture of 0/0 PDS is placed around the anus to prevent fecal spillage during the procedure, which is removed immediately after the operation.

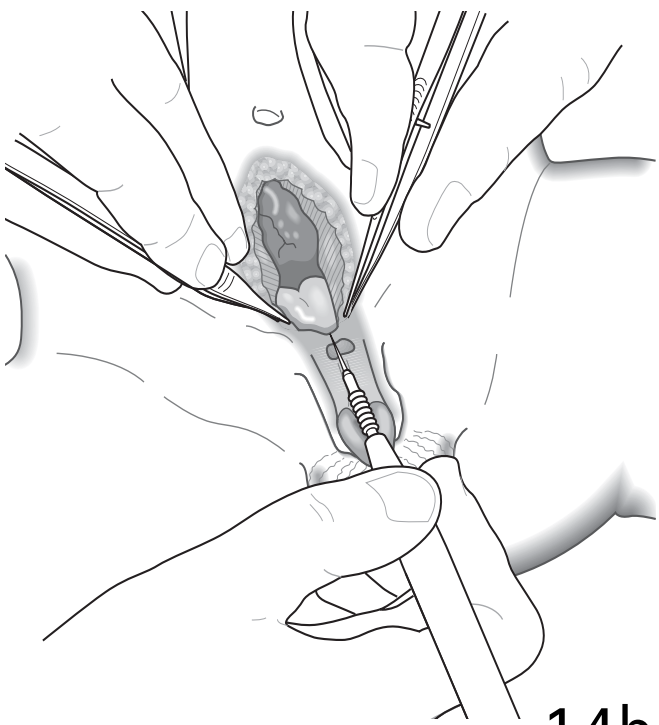
DECONSTRUCTION/DISSECTION

Skin incision

14 A stay suture is placed in the glans as before to retract the penis. The old incision is re-opened, excising any hypertrophic scar, and deepened through the muscle to expose the bladder. The epispadiac meatus is opened to the bladder above, taking care to keep in the midline and avoid damage to the urethral plate.



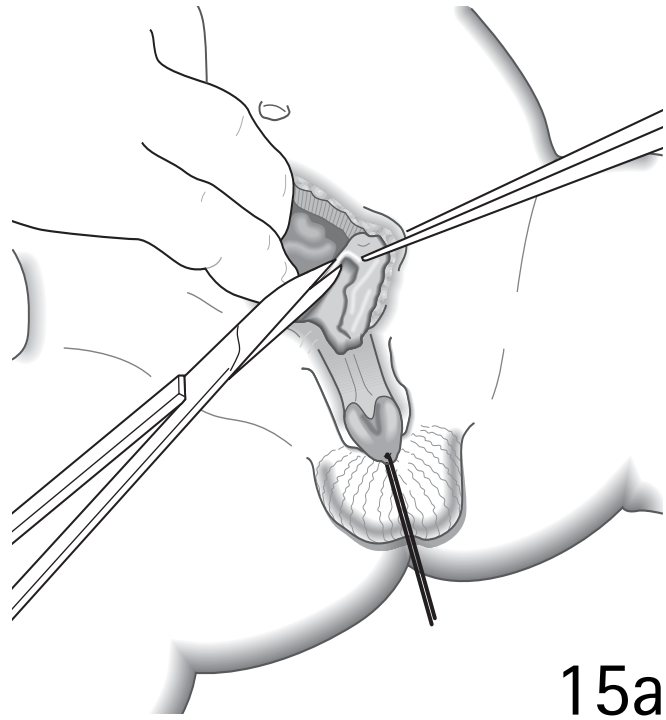
14a



14b

Bladder mobilization

15a,b The lateral edges of the bladder are freed from the abdominal wall on either side and the bladder is re-opened anteriorly in the midline. The lateral edges of the bladder are dissected free to the level of the veru montanum – as in primary repair.



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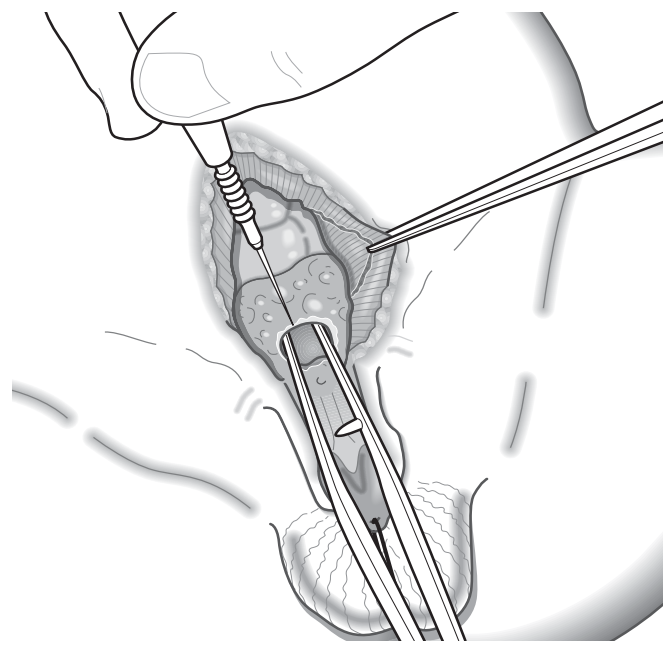
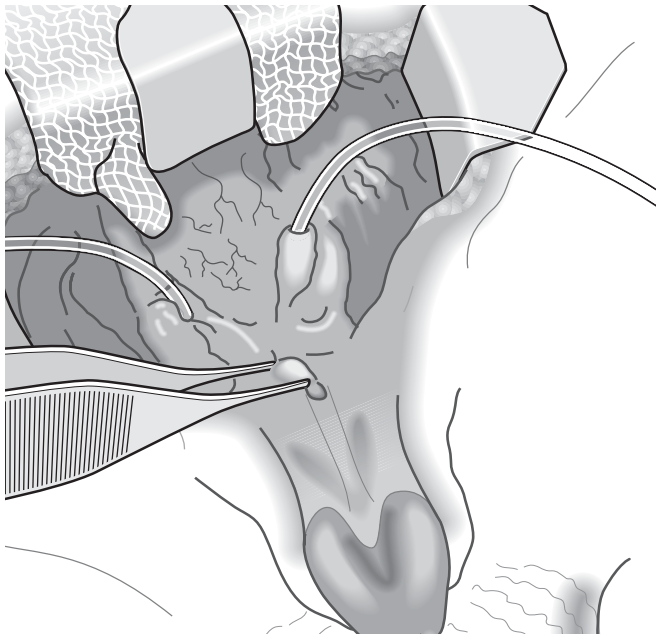


15b

Ureteric re-implantation

16a–c A small Finochetto retractor is used to separate the pubic rami. Two 6 Fr feeding tubes are placed as ureteric stents and the ureters fully dissected through the bladder, keeping close to the ureters. A Cohen-type ureteric re-implantation is completed using a transverse tunnel.



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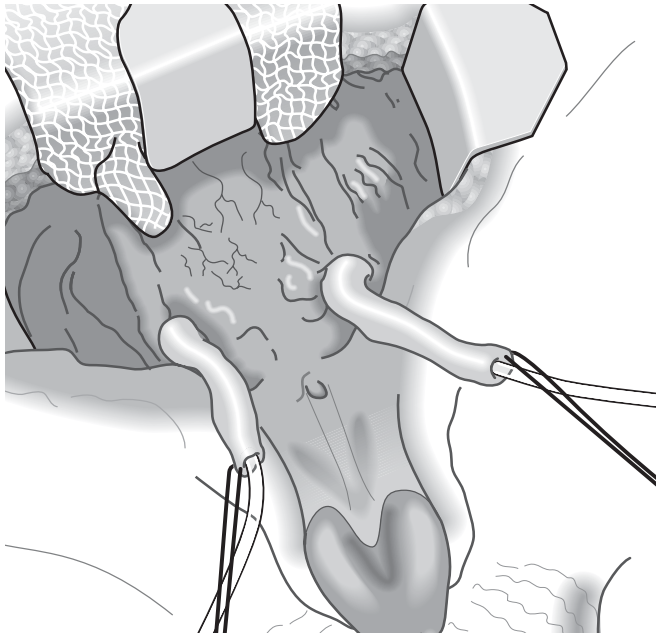

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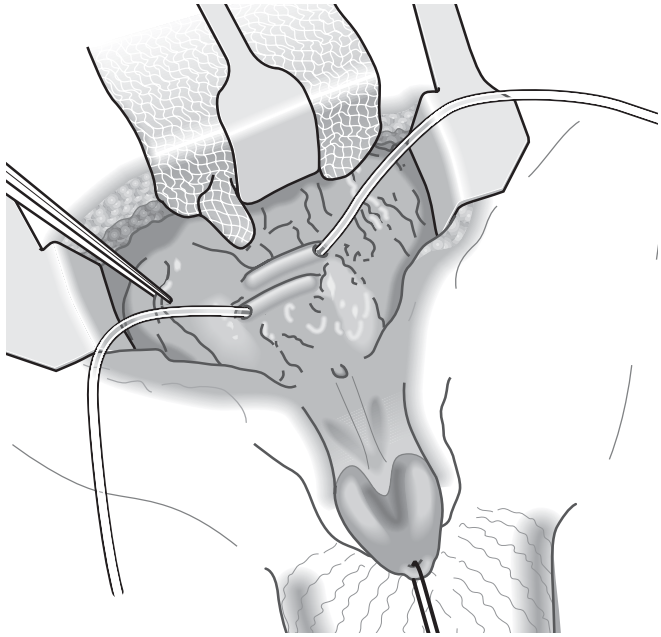
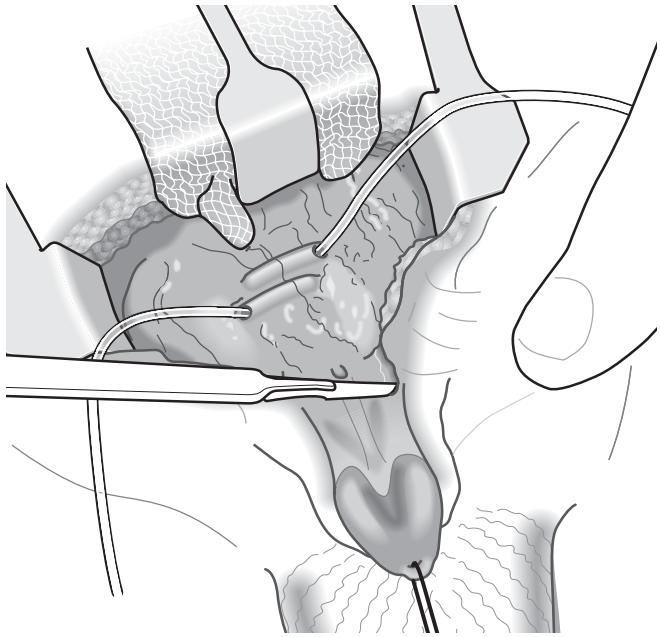


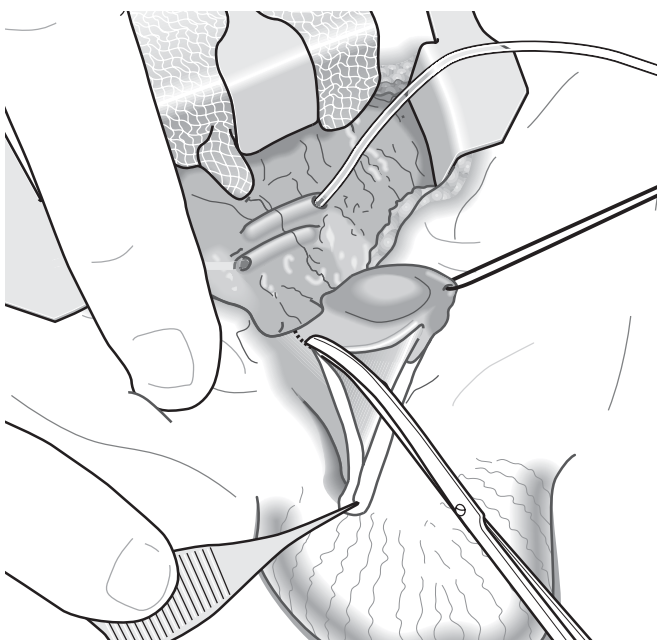
16c

Urethral and penile skin dissection

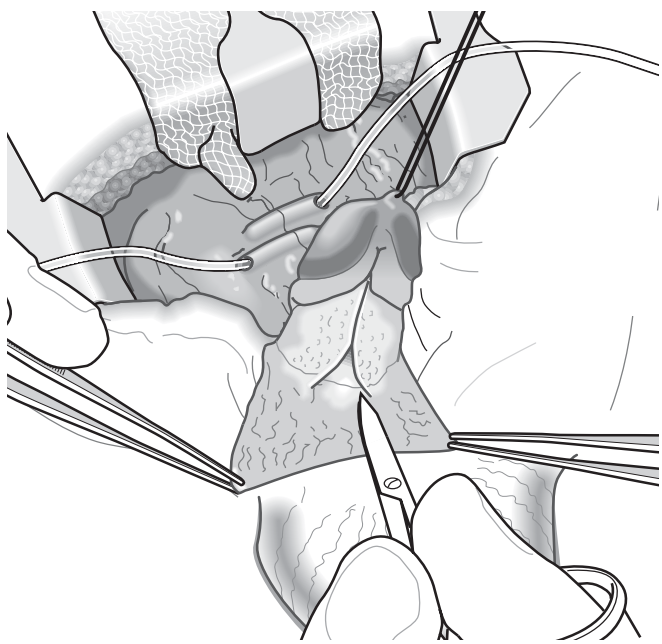
17a–c The urethral plate is marked out from either side of the veru, distally to the edge of the corpora, and incised. Ventrally, the penile skin is marked and incised to leave a cuff beneath the glans. The skin is lifted off the ventrum and sides of the penis and dissected proximally to expose the undersurfaces of the corporal bodies.



17a



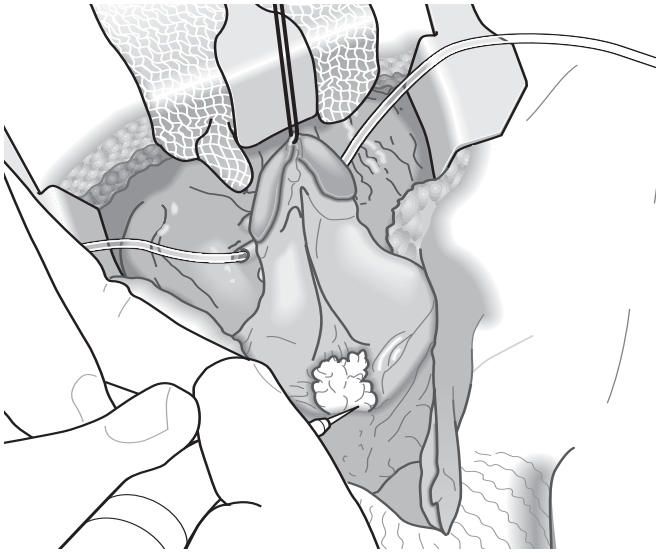
17b



17c

Dissecting the penis

18a–c Dissection continues proximally along the corporal bodies. The separation of the corpora and the prostate gland in between them are identified. Laterally each corpus is traced until its point of attachment to the inferior pubic ramus can be seen and palpated. The perineal muscle can be identified with the muscle stimulator, running between the bases of the corpora.



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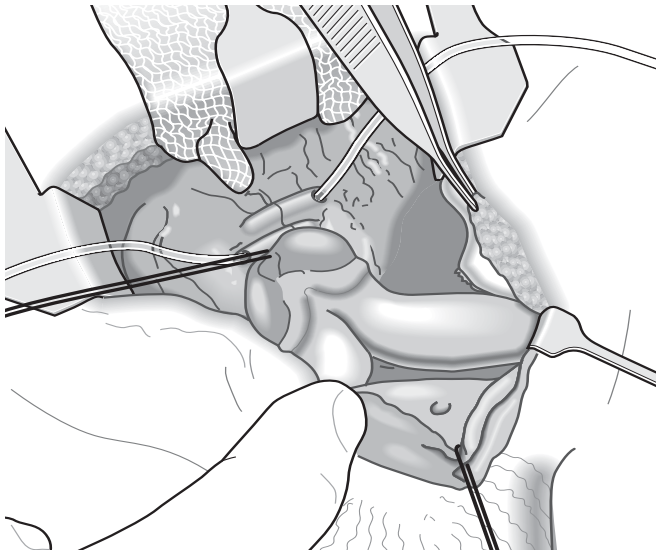
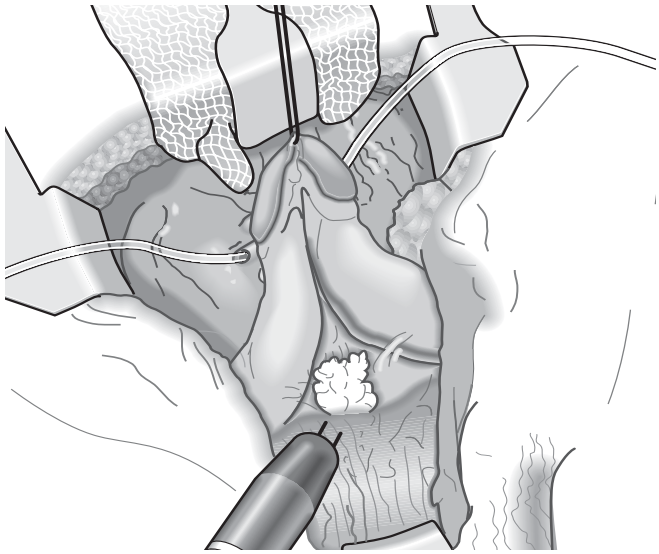


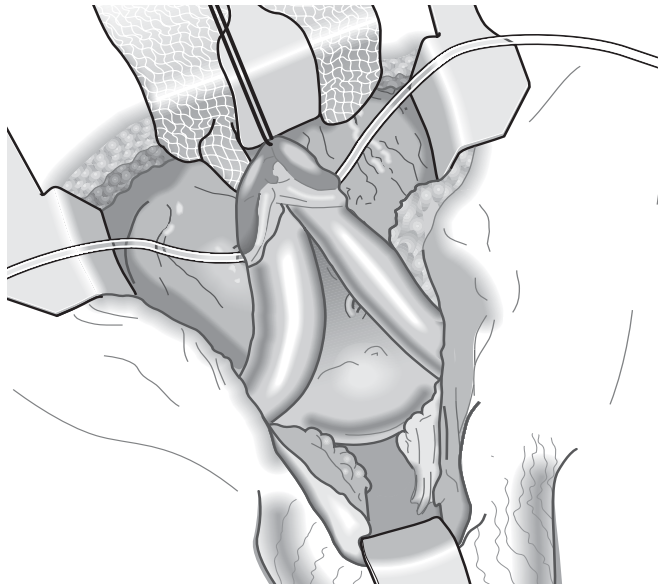
18b



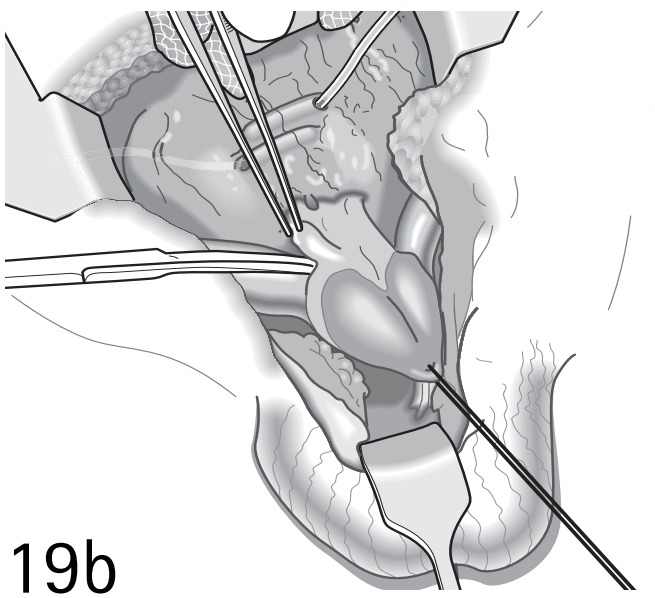
18c

Dissecting the urethral plate

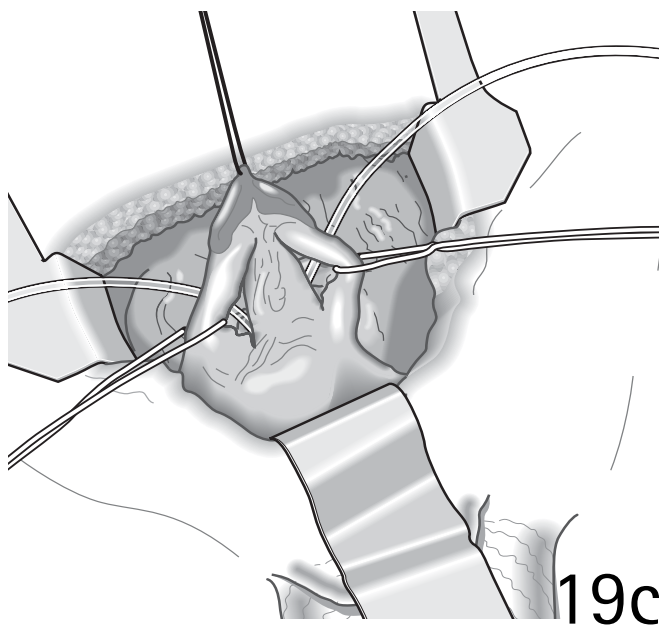
19a–c The urethral plate is dissected from above and below the penis. The tissue below the plate is separated from the corporal bodies by dissecting the bloodless plane between them on both sides, keeping close to the corpora. Dissection is completed anteriorly on either side of the plate, and vessel loops are passed around each corpus. Separation of the plate is completed – distally to the edge of the glans and proximally to the level of the prostate.



19a



19b

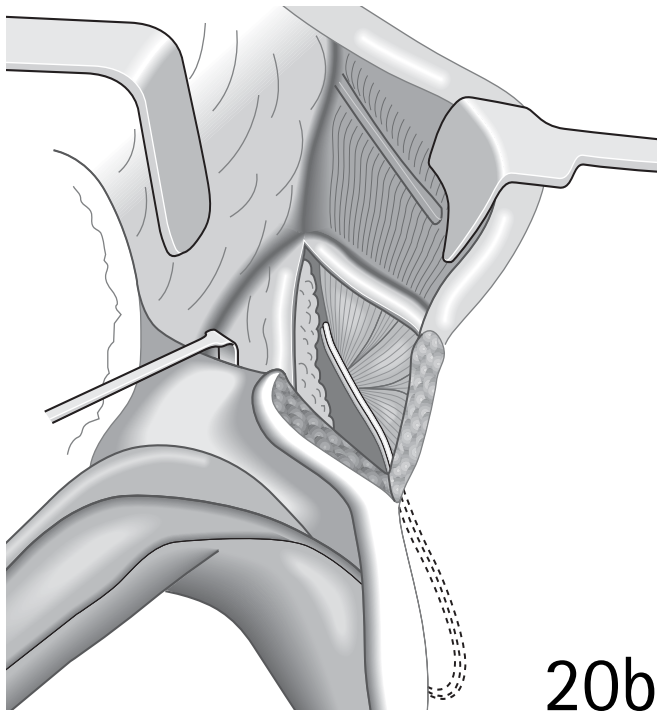


19c

Releasing the pelvic floor from above

20a–c To mobilize the base of the corpora, their neurovascular supply – the pudendal artery, veins, and nerve – must be freed along their course from behind the spine of the ischium from where they run forward in Alcock's canal. This is a fascial channel that runs forward on the surface of the internal obturator fascia, lateral to the ischiorectal fossa, below the pelvic floor.

A retroperitoneal plane lateral to the bladder is developed by forefinger dissection behind the lower rectus toward the pelvic floor. Medial retraction of the bladder and rectum behind it reveals the pelvic floor muscles and a muscle stimulator helps to define their attachment to the obturator fascia (the so-called white line). The muscle fibers are divided about 0.5 cm medial to the white line with bipolar diathermy. This exposes the ischiorectal fat, which can be pushed downwards and medially to expose the fascia over the internal obturator muscle.



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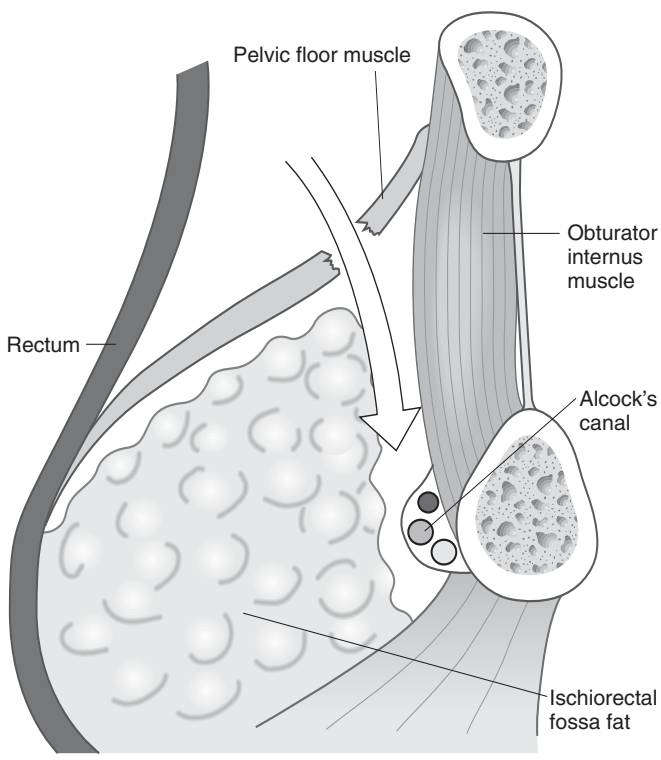
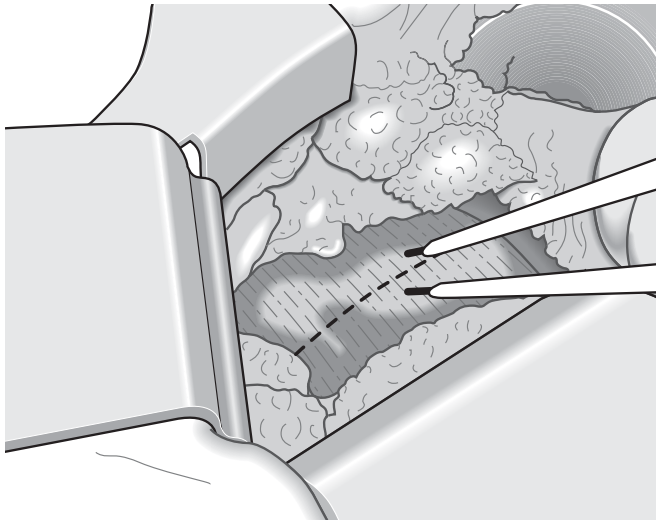
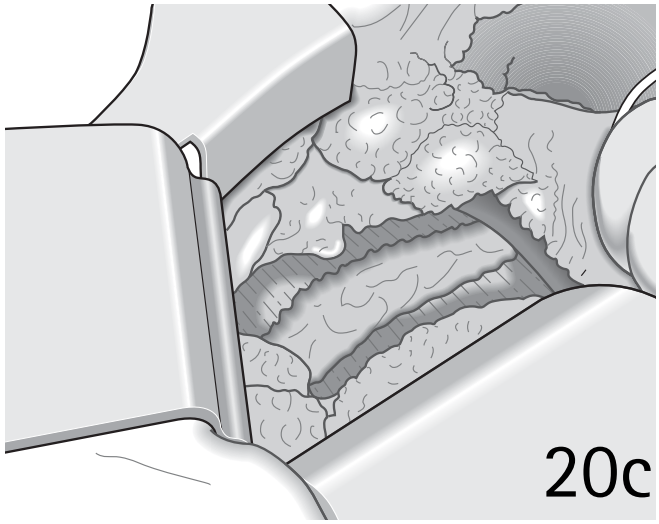


20a

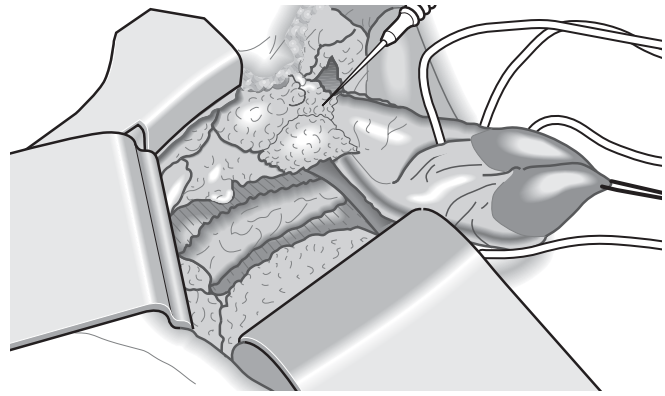




20c

Releasing the base of the penis and exposing the pedicle

21 This dissection is continued anteriorly toward the pubic ramus, where the muscle is often thicker. The corporal body is now released from the bone by first incising along the inferior pubic ramus, lateral to its attachment. A combination of blunt and bipolar dissection peels the periosteal attachment of the corpus off the bone. By careful dissection from above and below, the corpus is freed and the pedicle can be demonstrated running from behind and along it. Once it is seen, any fascial attachments lateral to the pedicle are divided, exposing fat and releasing the neurovascular bundle and the corpus medially.



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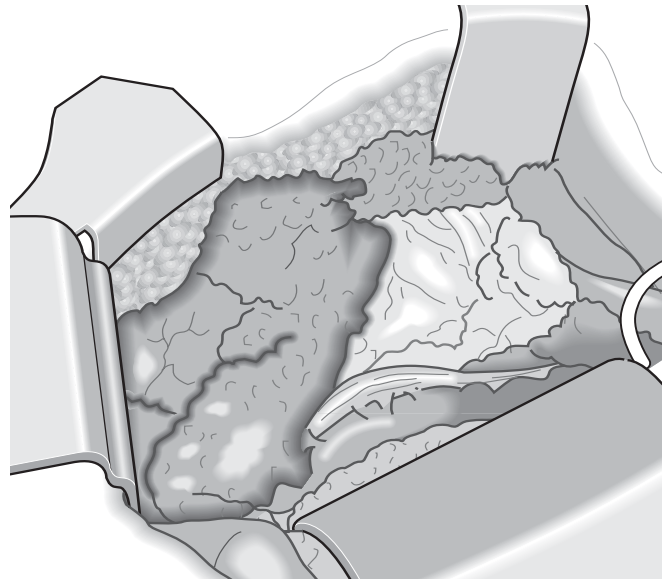
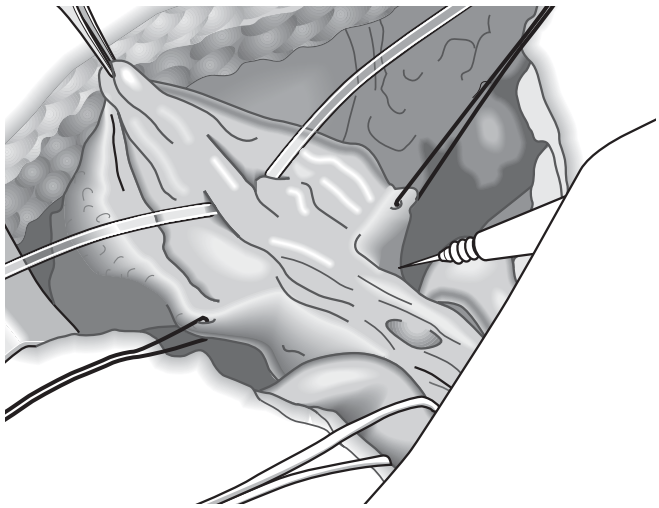


21b

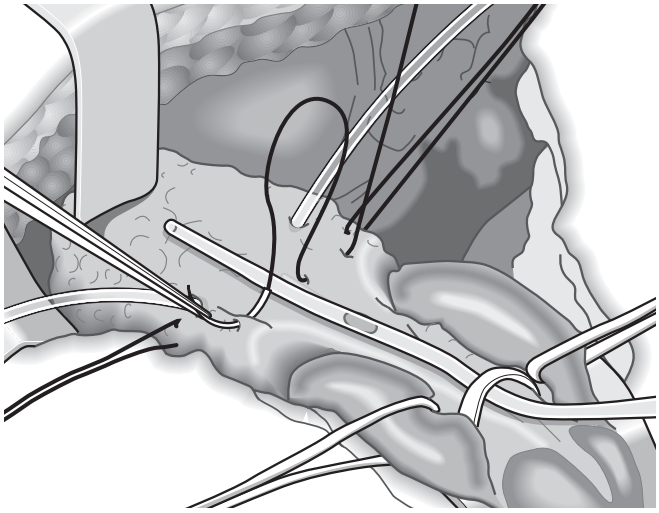


22a

RECONSTRUCTION

Bladderneck, bladder, and proximal urethral closure

22 The veru montanum is identified proximally on the urethral plate. The urethral plate is continued proximally and the area of the bladderneck is identified by marking and excising two triangles of bladder mucosa on either side. The bladderneck is reconstructed around a 10 Fr silicone stent whose proximal end is secured to the bladder mucosa with a fine, absorbable suture. Two layers of interrupted 4/0 Monocryl sutures are used and these are continued proximally in a single layer to close the bladder. A 12 Fr Malecot catheter is inserted into the bladder to be used for suprapubic drainage and the two stents are brought out through the midline closure. The closure is continued anteriorly along the urethral plate.



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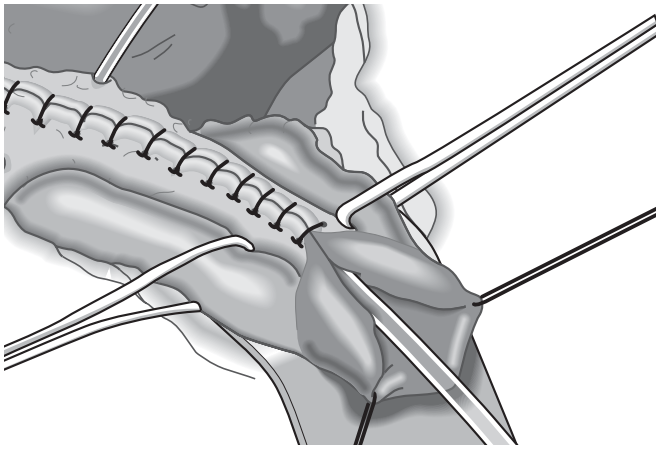


22c

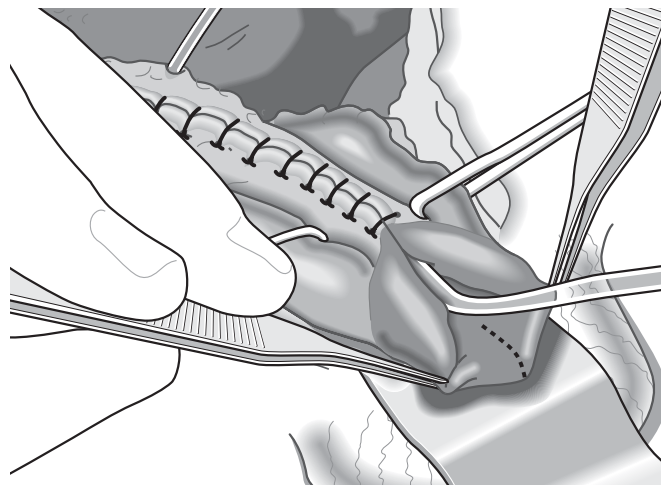
CLOSURE OF URETHRA AND EPISPADIAS REPAIR

ALTERNATIVE 1: CANTWELL–RANSLEY TYPE OF REPAIR

If the penis is a good length and protrudes well, a decision may be made at this stage to perform a one-stage repair, including complete repair of the epispadias. It is important to note that most experience has been gained with the second alternative (creation of a hypospadias) and the long-term results – especially for continence – are still to be evaluated.

'IPGAM' maneuver

23 The distal urethral plate at the tip of the glans is incised longitudinally and the incision is closed transversely with fine, absorbable sutures. This 'IPGAM' maneuver brings the urethral meatus more ventrally on the glans.



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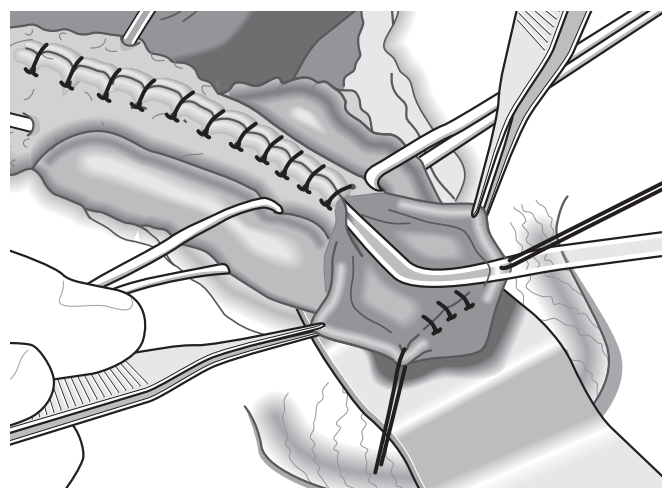

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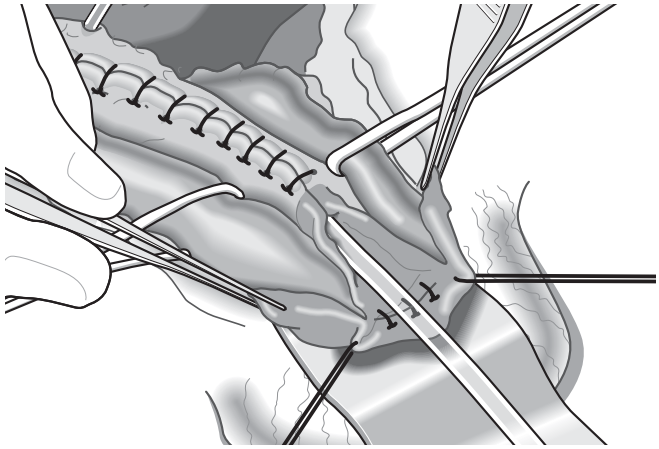
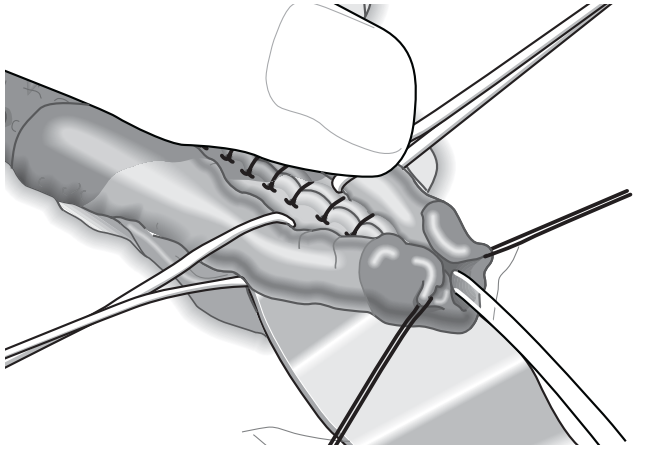


24a

Closure of the urethra and glans

24 Two triangles of mucosa are marked and excised from either side of the glans, preserving the central strip of urethral plate. Proximally, the plate is lifted from the corpora to provide greater mobility, which will allow the urethra's displacement ventral to the corpora and closure of the glans behind it. The plate is then closed with interrupted 5/0 PDS sutures. The gland is reconstructed dorsal to it with two layers of interrupted, subcuticular, 5/0 PDS.



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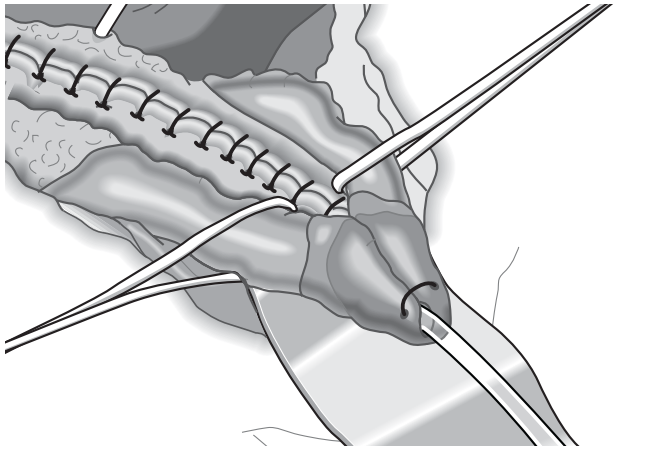
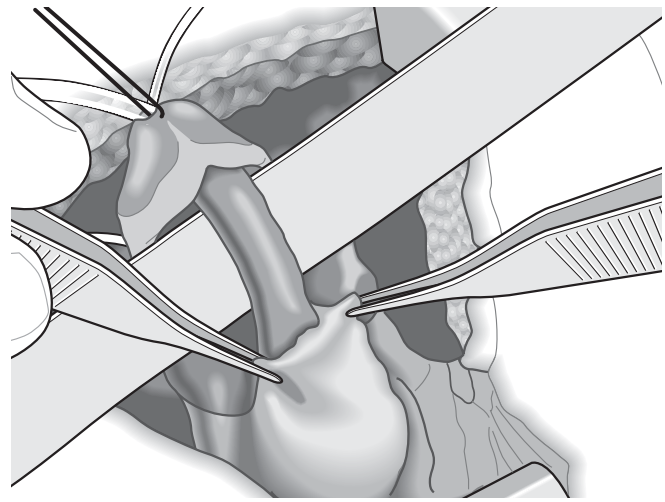


24c

Creation of the sphincter

25 The urethra is now free to bring ventral to the corpora, and this can be demonstrated by inserting a copper retractor. The muscle between the corpora, at and below the level of the prostate, is again identified with the muscle stimulator. A sling of this muscle is wrapped around the urethra by passing two 4/0 Monocryl sutures on either side from behind. The wrap is not circumferential and the sutures should be tied dorsally – snug but not tight. The two corporal bodies are now sutured together in the midline with 4/0 Monocryl. External rotation of each at this time will help correct the dorsal chordee.



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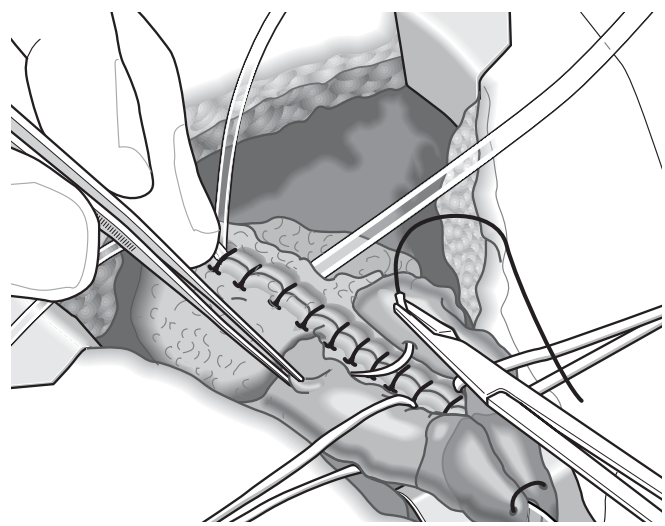
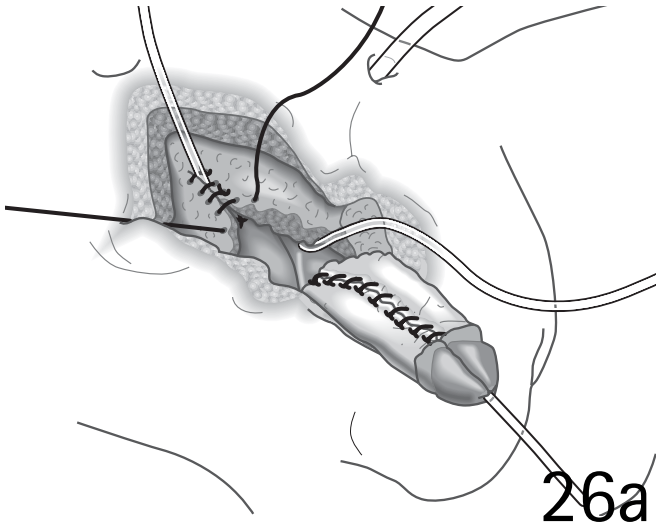


25b

Closure

26 The abdominal wall is closed from above with serial figure-of-eight sutures of 2/0 PDS – as for primary closure. The stents are brought out through the wound, whilst the Malekot is tunneled and brought out to the left of the midline. Two 0/0 or 1/0 PDS sutures are placed at the lower end of the wound to re-close the symphyseal area. The base of each corpus – corresponding to the area previously separated from the bone – is sutured to the lower end of the abdominal closure with 3/0 PDS sutures. Care must be taken to avoid damaging the pedicle at this point.



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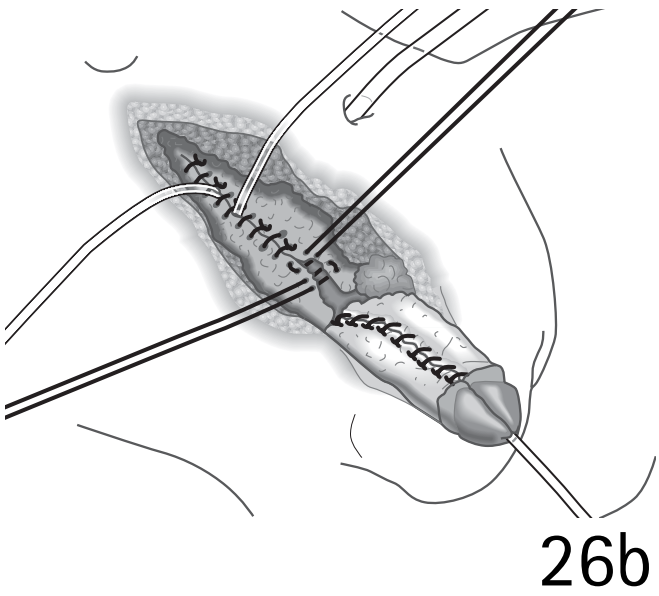

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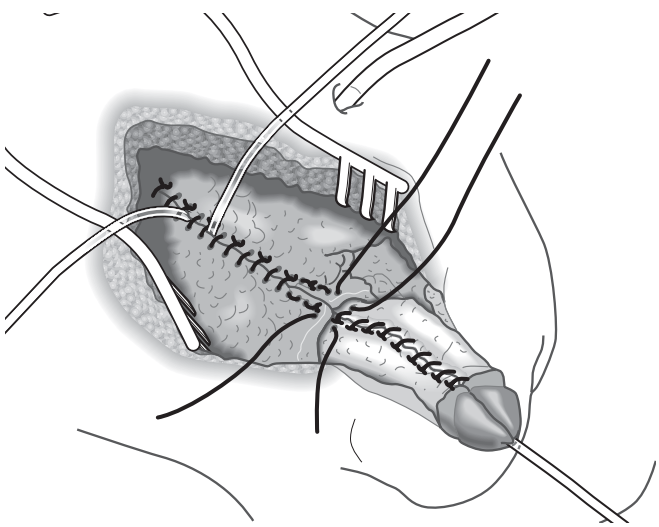
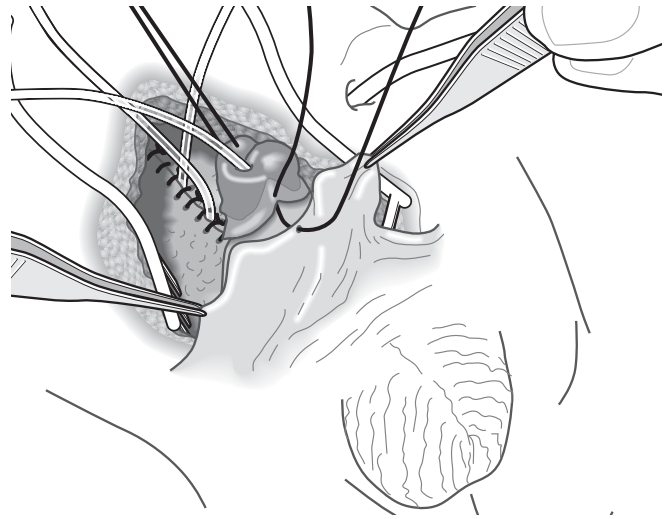


26c

Penile skin cover and skin closure

27 The penile skin flap is now lifted and flattened. It is incised in the midline and the apex of this incision is sutured to the frenular area of the skin cuff with 6/0 Monocryl sutures. The penis is wrapped and the skin closed dorsally in the midline or with z-plasties. The edges are trimmed before the circumcision wound is closed with 6/0 Monocryl. The abdominal wall subcutaneous layers and skin are closed with layers of absorbable sutures.



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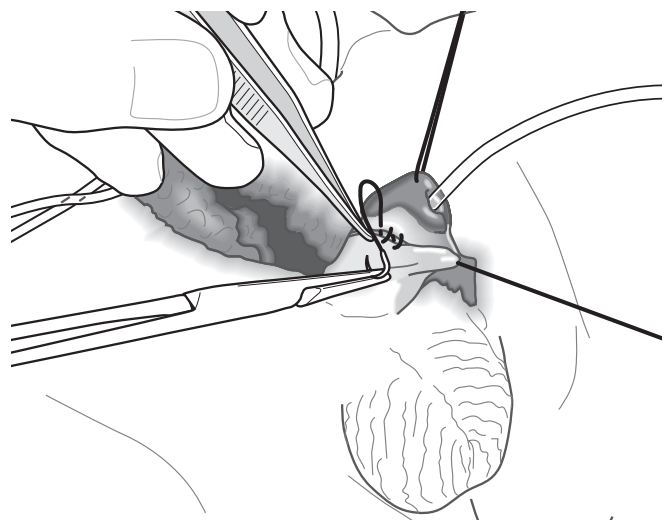


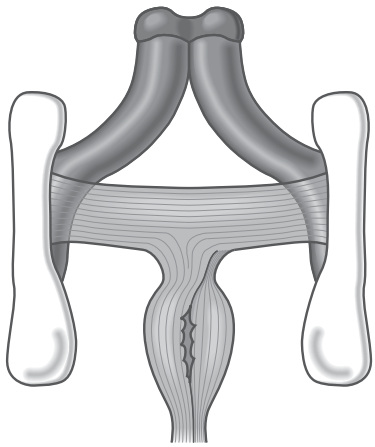
27b

ALTERNATIVE 2: CREATION OF HYPOSPADIAS

Where the penile length is compromised (as is often the case) and in particular when the urethral plate seems to limit its length, this alternative approach is used. In practice, surgeons have more experience with this approach and the published data on continence are principally from these patients. Subsequent procedures are required to create the distal urethra – with or without skin grafting.

Lifting of the urethral plate from the corpora and glans

28 The urethral plate is separated completely from the corpora, including its glanular portion. This is tubularized throughout its length using interrupted 5/0 Monocryl sutures. The neourethra is then dropped between the two corporal bodies. From the ventral aspect, the perineal muscle is identified and wrapped around the urethra using interrupted 4/0 Monocryl sutures. The wrap may not be complete and the sutures are snug but not tight. The corpora are joined dorsally and the penile shaft reconstruction is completed as above, with corporal rotation to correct the chordee. The glans halves are joined in layers of 5/0 PDS. The urethral opening is secured to the ventral shaft with 5/0 PDS. When the shaft shin is reconstructed, a midline opening is made to accommodate the hypospadiac meatus, which is secured around its circumference with absorbable sutures.



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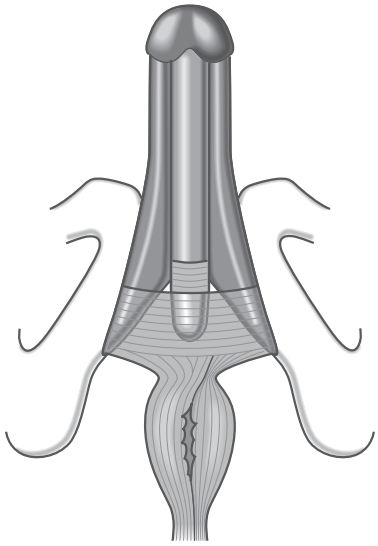

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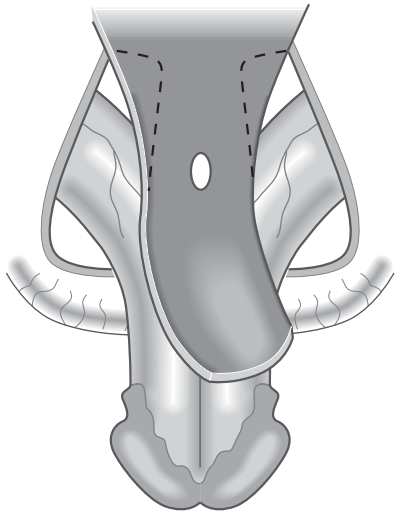

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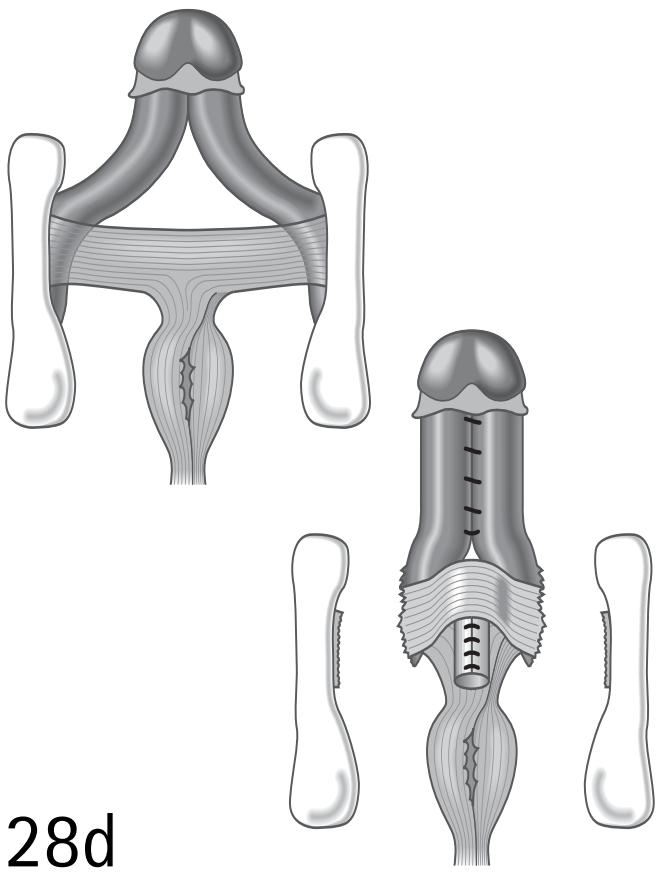

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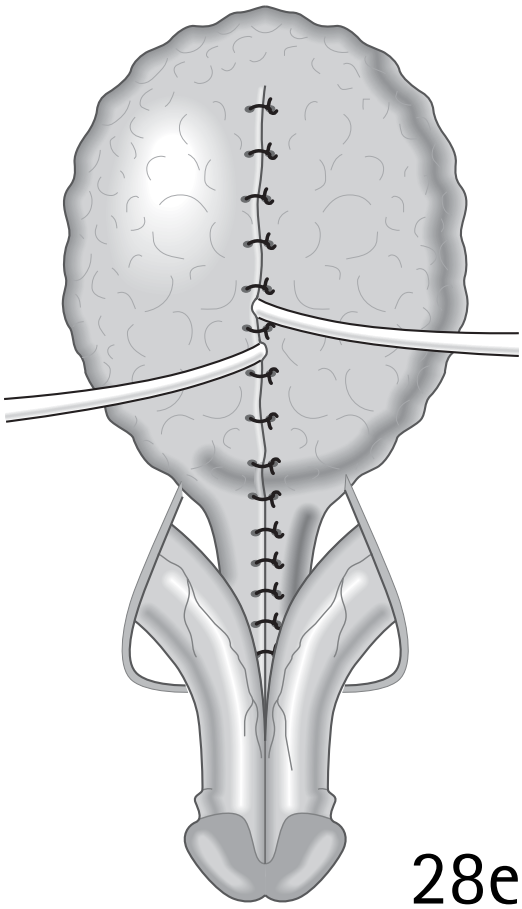
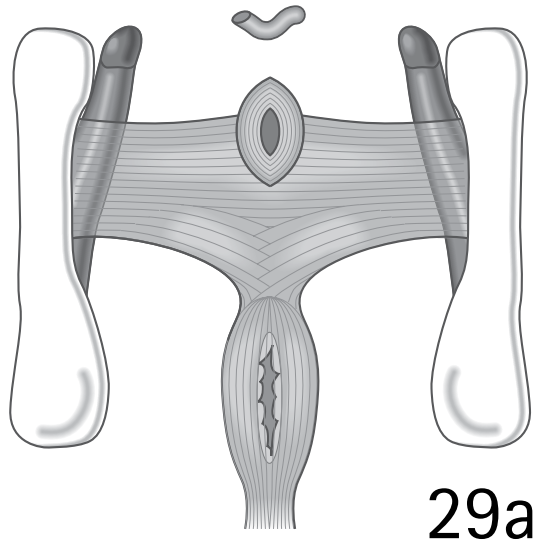


28e

FEMALE EXSTROPHY/EPISPADIAS REPAIR

29 In females after successful primary closure in exstrophy or primary epispadias, soft-tissue reconstruction is used to create continence and also to improve genital appearance. The clitoral corpora are dissected as for the penile corpora, with the labia minora skin flaps left attached around the glans for subsequent reconstruction. The pelvic floor and pudendal pedicles are dissected in the same way and the muscle is identified below and lateral to the vagina – attached to the base of the corpora on either side. At the time of bladder outlet reconstruction, this muscle is brought around the dorsum of the proximal urethra and effectively wraps both the urethra and vagina. At closure, the hemiclitori can be brought together and the labia minora replaced lateral to the neourethral orifice and the vagina. The mons is reconstructed by mobilizing subcutaneous fat from either side.



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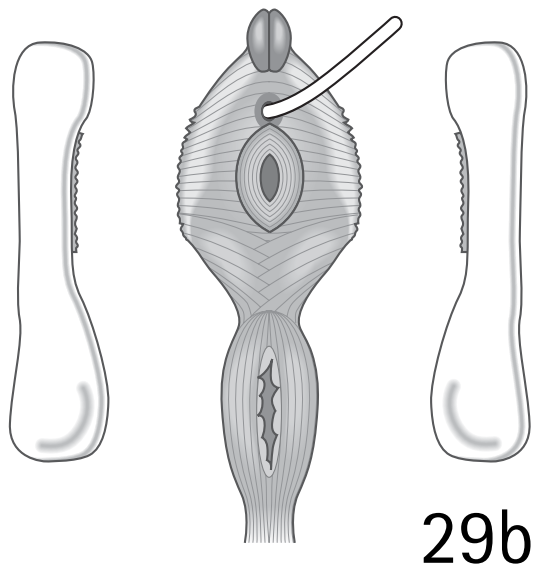

29b

POSTOPERATIVE CARE

After radical soft-tissue reconstruction and epispadias repair, the penis is enclosed in a foam dressing for 1 week. The ureteric stents are left on drainage and removed after 1 week, and the bladder remains on free drainage for 3 further weeks. At this time, the urethral stent is removed and the suprapubic Malekot catheter is clamped intermittently. When this is possible for 3 hours and the child has voided urethrally (checked by ultrasound), the suprapubic catheter is removed. Continence and bladder capacity will evolve with time, and regular evaluations at approximately 3-month intervals are required. Cystoscopic evaluation of the bladder outlet is performed with a fine cystoscope (8 Fr) at 3–6 months.

FURTHER READING

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Kelly JH. Exstrophy and epispadias: Kelly's method of repair. In: O'Neill JA, Rowe MI, Grosfeld JL, Fonkalsrud EW, Coran AG (eds), *Pediatric Surgery*, 5th edn. St Louis, Mi: Mosby, 1998, 1732–59.

Woodhouse CR, Fisch MM, Peppas DS, Reiner WG. Failed bladder exstrophy closure. *Dialogues in Pediatric Urology* 1999; **22**.

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Surgical reconstruction of intersex abnormalities

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HISTORY AND PRINCIPLES

A child with ambiguous genitalia must be evaluated immediately at birth. The gender that is appropriate to the anatomy of the infant must be assigned as early as possible, as new parents are asked about the sex of the child as soon as the birth is known. They must be able to give an answer that is commensurate with the gender assignment that will eventually provide the most satisfying functional result. Surgical correction must then be done at the appropriate time so that confusion about sex is not prolonged, and subconscious rejection of the child is less likely to occur.

Four major categories of abnormality can cause gender confusion at birth: female pseudohermaphroditism, male pseudohermaphroditism, true hermaphroditism, and mixed gonadal dysgenesis. The gonads are symmetrical in the first two, but are not if one gonad has differentiated predominantly as a testis and the other as an ovary, as in mixed gonadal dysgenesis and true hermaphroditism (see MacLaughlin and Donahoe, 2004).

DIAGNOSIS AND PREOPERATIVE ASSESSMENT

Genetic females, no matter how severely virilized, should be raised as females. For genetic males, the gender assignment must be based on the infant's anatomy, that is, the size of the phallus, and not on the 46XY karyotype. If the phallus is inadequate, one should strongly consider assignment to the female gender. The average penile length at 30 weeks' gestation is 2.5 ± 0.4 cm, increasing to 3.5 ± 0.4 cm at term, with a width of 1–1.5 cm. A term size below 2×0.9 cm should cause concern. Exceptions, however, must always be made if the patient presents late and has become fully committed to the male role, or in the event of a diagnosis of 5-α-reductase deficiency.

Female pseudohermaphroditism occurs when a genetic female (46XX) is exposed in utero to androgens, either exogenous or endogenous, as in the congenital adrenal hyperplasia syndromes. The phenotype can vary from clitoral enlargement alone to complete labioscrotal fusion and formation of an entirely normal male penis with a closed urethra formed to its tip. Cortisol deficiency can lead to salt wasting, which can be life threatening unless replacement is instituted. All masculinized females have normal childbearing potential and should be raised as females. The psychologically important clitoral recession should be done as early as possible, particularly if the defect is severe, and is now combined with vaginoplasty as a one-stage procedure.

Male pseudohermaphroditism occurs in genetic males (46XY) with deficient masculinization of the external genitalia due to insufficient testosterone production, conversion, or inadequate target organ response. Many patients with male pseudohermaphroditism have been raised as males. However, if the female gender is chosen, gonadectomy should be done at the time of perineal reconstruction. In the patient with absent or rudimentary vagina, this usually requires only clitoral recession and labioscrotal reduction. The labioscrotal folds should be partially reduced during the first procedure, and dilatation or a substitute vaginoplasty planned for the late adolescent or early adult years. Patients with testicular feminization, in whom an introitus is often present, may have this dilated with bougies at a later age to form a functional vagina.

True hermaphrodites have well-developed, non-dysgenetic male and female gonadal tissue in many combinations, i.e., a testis on one side and an ovary on the other, two ovotestes, or a normal gonad on one side and an ovotestis on the other.

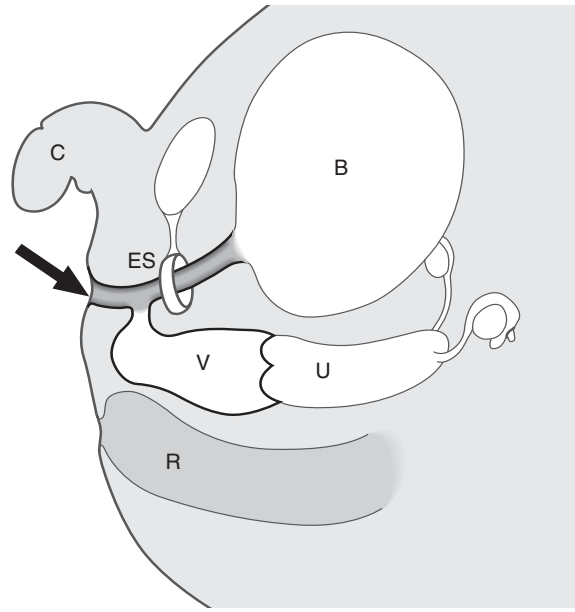
Although 80 percent of these patients have a 46XX karyotype, testicular tissue is present. The patient with a small phallus should be raised as a female; the patient with a large phallus already committed as a male should be raised as a male. Gonads should be bivalved and biopsied longitudinally. The gonadal tissue commensurate with the sex of rearing should be salvaged. Microdissection may be used. Perineal reconstruction should be accompanied by removal of Wolffian structures. If the phallus is adequate for male gender assignment, ovarian and Mullerian structures can be removed, followed by hypospadias repair. Testicular prostheses can be inserted later, should the testicular tissue be inadequate.

Mixed gonadal dysgenesis patients have dysgenetic gonads, retained Mullerian structures, internal and external asymmetry, and a mosaic karyotype, often 45X/46XY. The dysgenetic gonads can develop neoplasms such as gonadoblastoma or seminoma-dysgerminoma. Children with mixed-gonadal dysgenesis should be raised as females, with removal of the gonads, perineal reconstruction with flap vaginoplasty, and estrogen and progesterone replacement at adolescence.

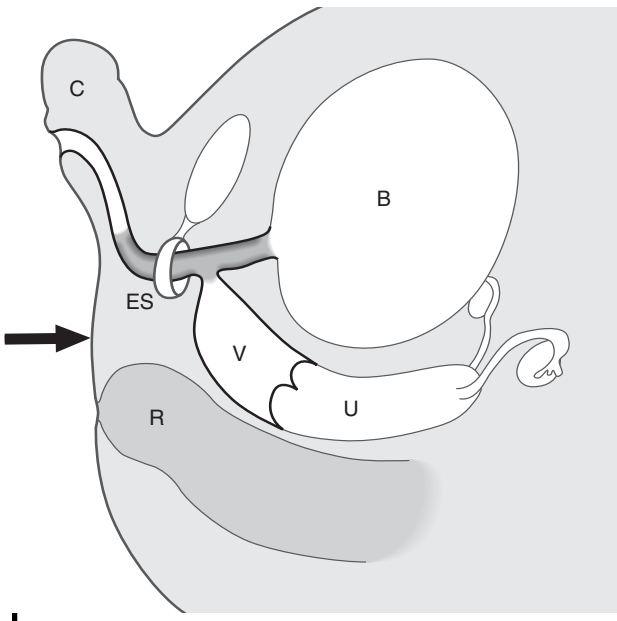
If the patient is already committed to the male role, hypospadias repair will be required. The gonads must be carefully observed for tumor development, which may occur as early as the newborn period.

SURGICAL PLANNING

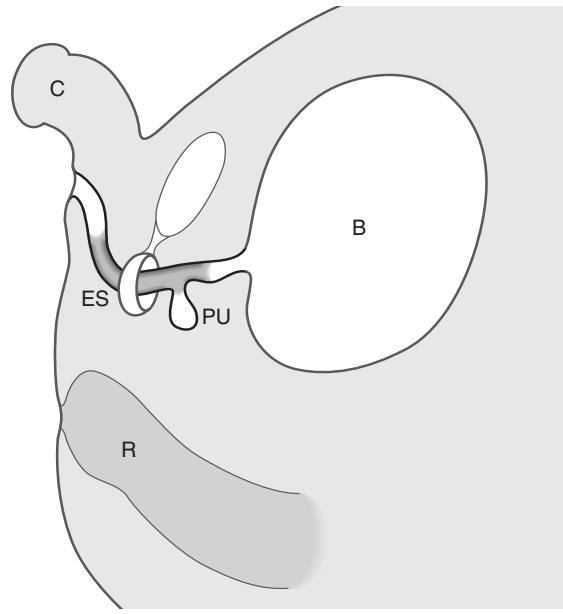
1a–c Cystoscopic evaluation of the confluence level of the vagina and urethra to form the urogenital sinus is crucial. In most cases, the merger occurs distal to the veru montanum and they are considered low (a) and the vagina can be exteriorized by the simpler flap vaginoplasty. The repair can be undertaken at 3–6 months of age, or even in the newborn period if the social circumstances are such that the baby might otherwise be rejected. Babies with the urethrovaginal merger at or proximal to the veru montanum (b) are considered high and require a more complicated pull-through vaginoplasty. If no vagina is found, the vaginal replacement can be planned for late adolescence (c).



1a



1b



1c

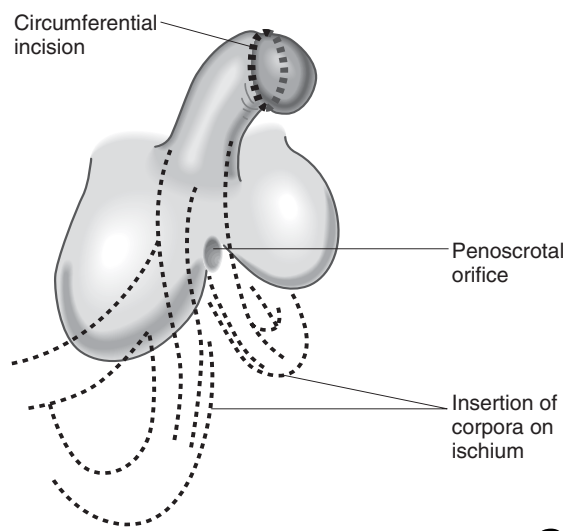
All 46 XX females diagnosed as newborns with female pseudohermaphroditism should be assigned the female gender regardless of the extent of masculinization. Patients with criteria consistent with the female gender assignment should undergo perineal reconstruction, which consists of clitoral recession, vaginoplasty, and labioscrotal reduction. Similar repairs can be used for selected patients who are insufficiently masculinized because of male pseudohermaphroditism, mixed gonadal dysgenesis, or true hermaphroditism. The goal of clitoral recession is to minimize the clitoris while preserving sensation and erection. Exteriorization of the vagina corrects the urogenital sinus defect that results from failure of the vagina to migrate to the perineum. The more masculinized the baby, the more proximal will be the urethrovaginal confluence and the more technically demanding the repair. The labioscrotal folds need to be trimmed, thinned, and elongated to create the appearance of labia majora. The clitoral recession, labioscrotal reduction, and exteriorization of the vagina should, whenever possible, be done together to take advantage of all available redundant tissue.

Most patients reared as males have micropenis and severe chordee; the urethral meatus is situated at the penoscrotal junction between a bifid scrotum, and sometimes a prepenile scrotum is present. Pretreatment with testosterone both enlarges the micropenis and confirms whether the phallus will respond to testosterone. We recommend these repairs to be staged when micropenis is present. Testicular prostheses should, if necessary, be inserted in infancy and changed as the patient approaches adolescence. Mastectomy may be necessary if a male patient develops gynecomastia on approaching puberty.

RECONSTRUCTION FOR FEMALE GENDER ASSIGNMENT

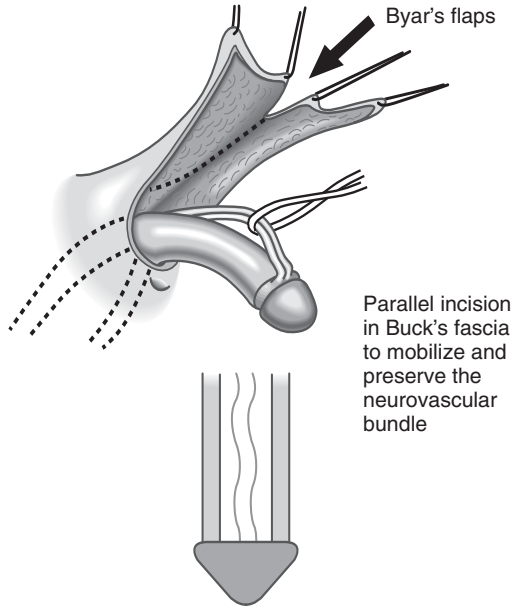
Clitoral recession

2a The appearance of the enlarged clitoris is shown. The corpora cavernosa from the pubic rami join, and form the enlarged glans. A circumferential incision is outlined 2 mm proximal to the corona of the glans. Epinephrine diluted 1:200 000 helps to control the bleeding and facilitates the dissection.

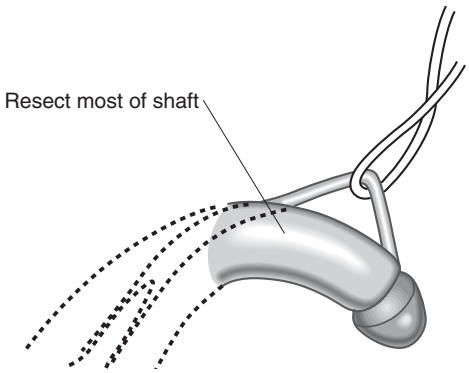


2a

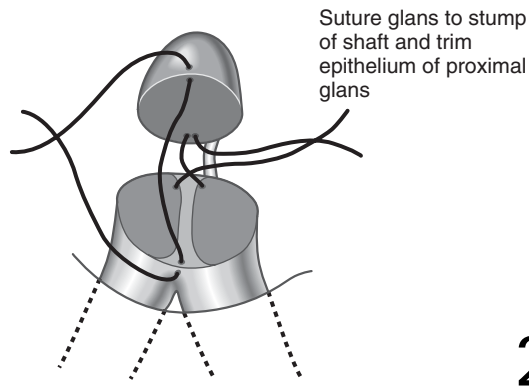
2b–d The shaft is degloved, and the prepuce is divided in the dorsal midline to create Byars' flaps as for a hypospadias repair and is carried anteriorly so that the glans sits comfortably at the mons pubis in the normal female clitoral position. Longitudinal incisions are then made through Buck's fascia on either side of the dorsal neurovascular bundle (b). Dissection through Buck's fascia is done with careful microtechnique and loupes for magnification, and the neurovascular bundle and Buck's fascia are lifted off the corpora (c) and dissected back to where the conjoined corpora divide at the base. The midbody of the clitoris is then resected between the base of the glans and the bifurcation of the corpora (d). Some surgeons prefer longitudinal parallel incisions made on the ventral aspect of the clitoris followed by excision of the cavernous tissue. Hemostasis is maintained over the clitoral corpora using careful cautery to avoid possible impairment of future erectile function. The corpora is sutured to the base of the glans (d) with interrupted 5/0 Vicryl sutures, taking care to avoid injury to the redundant dorsal neurovascular bundle. If necessary, a enlarged glans can be trimmed on all sides except the dorsum. The reduced glans is then sutured at its subcoronal edge to the proximal, separated Byars' flaps and to the ventral mucosa of the urogenital sinus. Excess skin from the clitoral shaft may be moved downward to create the labia minora. This technique aims to retain the important sensitivity of the clitoris.



2b



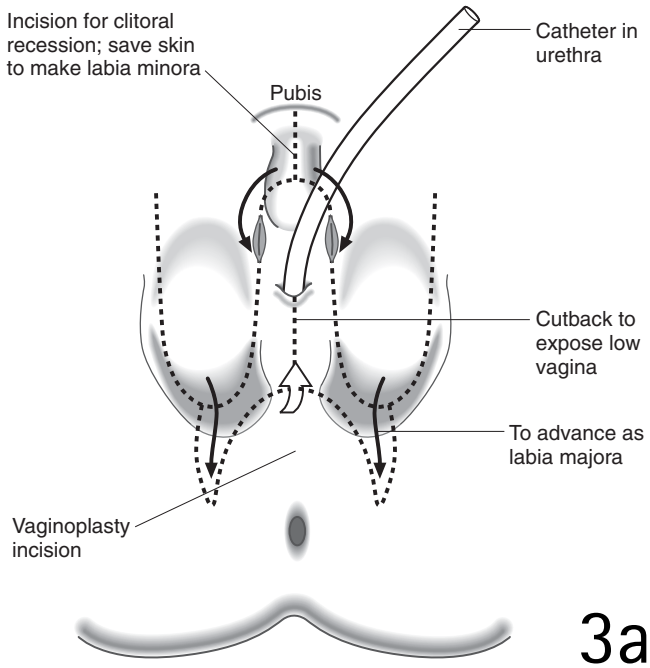
2c



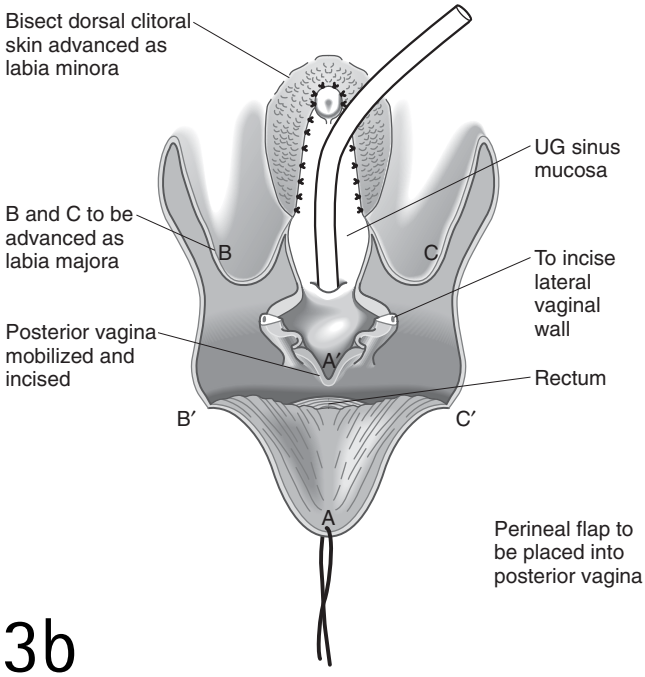
2d

Vaginoplasty

3a If the urethrovaginal confluence is located distal to the veru montanum, vaginoplasty can be performed from the hyperextended lithotomy position, usually at 3–4 months of age. After the clitoris has been recessed and the bisected shaft skin rotated around the repositioned glans, U-flaps are outlined on the labioscrotal folds, their extent depending on the degree of enlargement. An inverted U-flap, outlined on the perineum and broadly based at the level of the anus, is raised. A transurethral catheter is placed to divert the urine.



3a

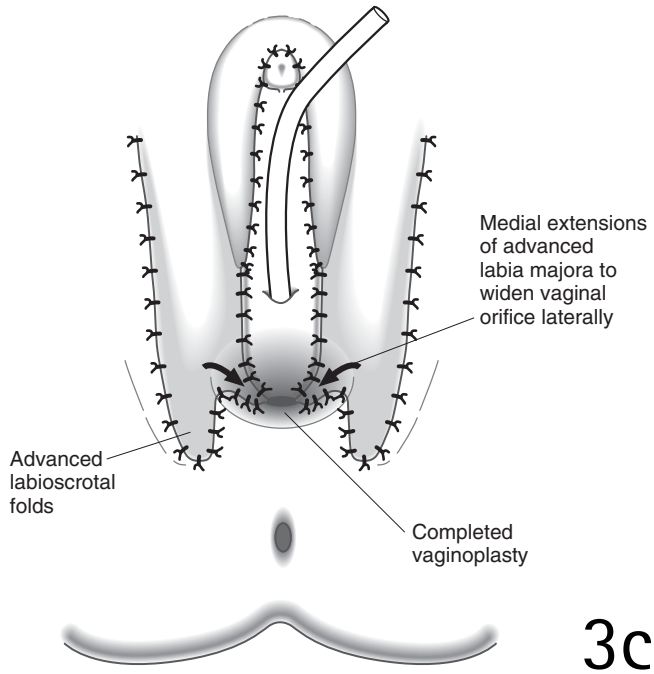


3b

3b The labioscrotal flaps are raised and the shawl of the labia minora sutured medially to the urogenital sinus mucosa. The inverted U-flap based on the rectum is dissected back toward the anus. At this point, a finger is placed in the rectum, which is dissected away from the back wall of the vagina, often as far back as the peritoneal cul-de-sac. It is more acceptable to err on the side of opening into the vagina rather than the rectum to avoid creating a rectovaginal fistula.

The vagina is opened in the midline posteriorly ('cutback'), and the flap is laid and sutured in place with interrupted 3/0 Vicryl sutures. Two small incisions are sometimes made in the lateral wall of the vagina. The labioscrotal folds are then advanced into the side arms of the inverted U-flap created on the perineum (B to B′ and C to C′).

3c This gives an elongated appearance to the refashioned labia majora. The medial portion of the often copious labioscrotal folds is advanced into the lateral incisions on the vaginal wall to enhance the vaginal opening (inset), stenosis of which is one of the commonest long-term complications after these procedures.

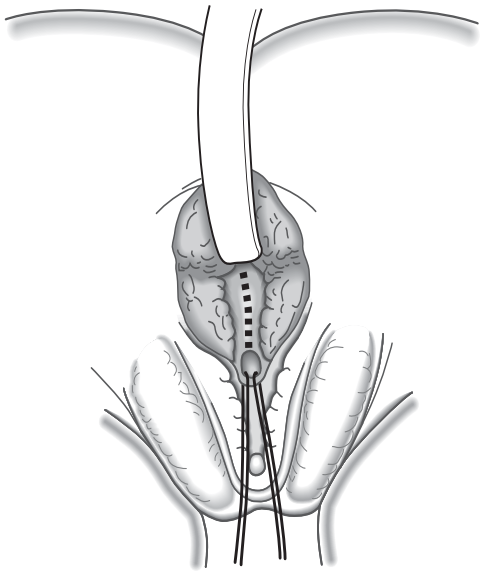


3c

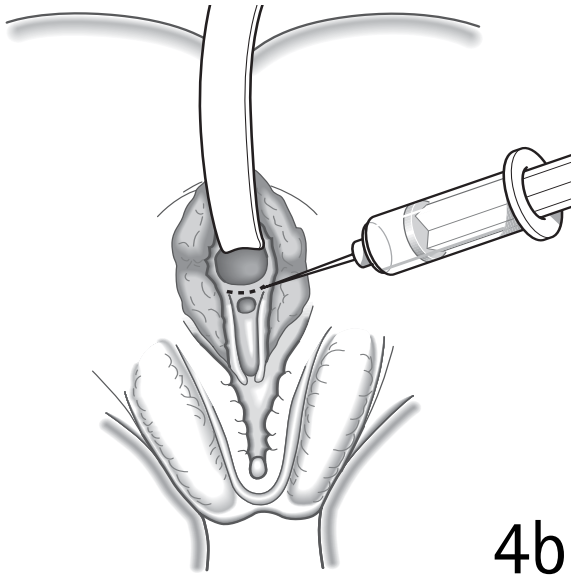
Repair of the high urethrovaginal confluence

4a–f If the urethrovaginal junction is close to the bladderneck, a posterior approach without division of the rectum, as described by Rink and colleagues, is used as it gives a better exposure. The repair is undertaken at about 1 year of age. The entire lower half of the body is prepared from the xiphoid down so that the child can be rotated from prone to supine as necessary.

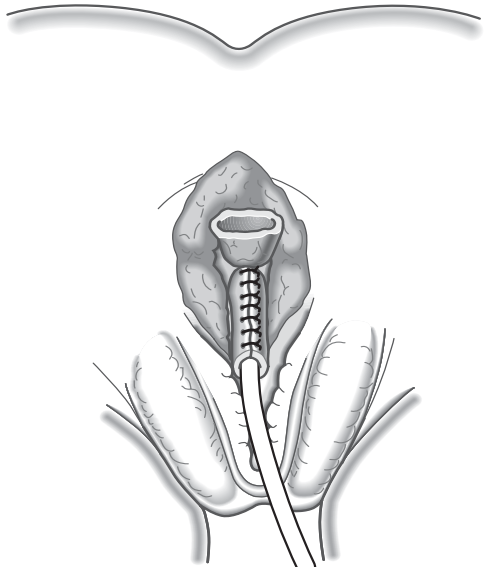
In those patients with a high confluence, the vagina should be mobilized first. After endoscopy and placement of a Fogarty catheter in the vagina and a Foley catheter in the bladder, the patient is turned to the prone position and a posterior inverted U-flap is created based on the rectum. A midline incision opens the urethra up to the urethrovaginal confluence, where it is separated from the posterior rectum (a). The rectum is displaced upward with a narrow retractor and does not require incision or division. After opening the urogenital sinus to the urethrovaginal confluence (b), the distal vagina is opened posteriorly to accept a retractor, which is then lifted upward to facilitate the exposure of the thin mesenchymal layer between the posterior vagina and the anterior urethra. Injection of a 1:200 000 epinephrine solution facilitates the separation of the vagina from the urethra, and a small Beaver blade is used to dissect the common wall between the urethra and anterior vagina (c). The posterior inverted U-flap is turned into the widened posterior vagina (d). Tubularization of the urethra is completed, with the addition of a second layer of sutures and incorporation of labial fat to reduce the incidence of a subsequent urethrovaginal fistula (c and d). A labial flap or an anterior island flap, which for this procedure must now be reconstructed in the midline, can easily reach the anterior vagina. A Y-V plasty is created on the generous labioscrotal tissue, which is defatted, trimmed, and moved posteriorly to create labia majora lateral to the vaginal orifice. The clitoroplasty is undertaken with the patient turned supine, following vaginal mobilization but before the vagina is exteriorized, as the turning may dissrupt this fragile anastomosis. The excess clitoral skin can be brought around posteriorly to fill the gap created by the anterior island flap (e and f). A long neourethra can be opened back to a more normal perineal position and used to create a wet introitus. A suprapubic percutaneous diversion catheter combined with a small silastic urethral catheter may be a better alternative to a small Foley catheter to prevent fistulas. Those cases of intermediate severity in whom the urethrovaginal confluence is distal to the veru montanum may be approached from the hyperextended lithotomy position. The surgeon's judgment is important. However, if the dissection proves to be difficult, the child can be turned prone to complete this dissection.



4a



4b



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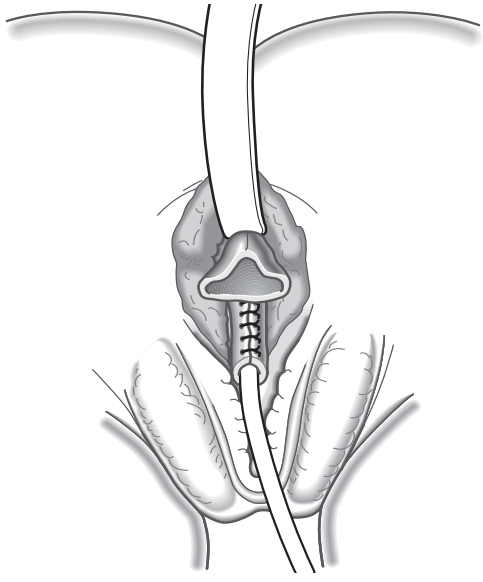
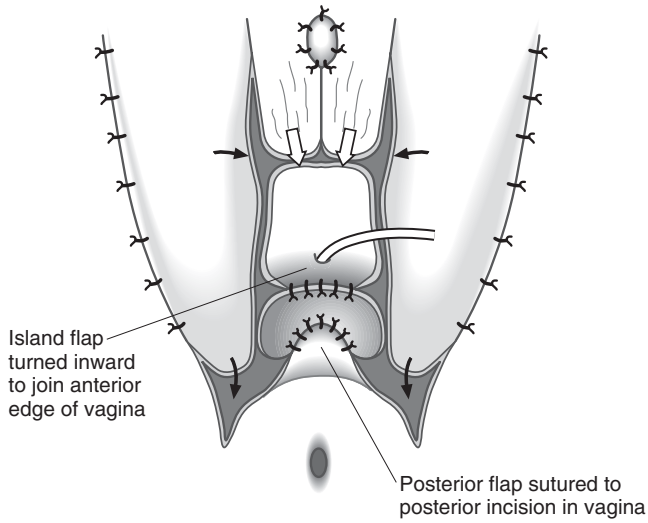


4d



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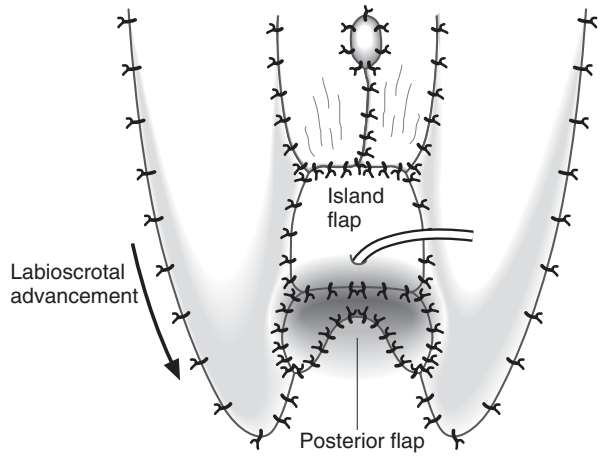
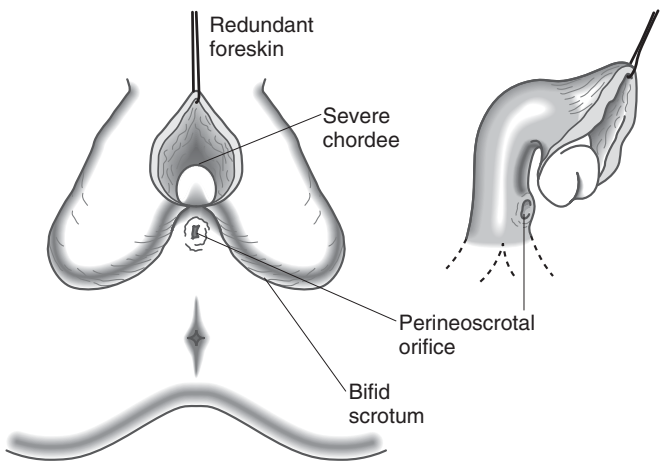


4f

Total urogenital sinus mobilization (TUM), as described by Pena in 1997 for repair of urogenital sinus defects, is another option for patients with a high vagina. The urogenital sinus is mobilized circumferentially to release the avascular fibrous ligaments that hold the urethra and vagina anteriorly to the pubis. The concern about possible damage by this maneuver to the innervation of the external sphincter and its effect on urinary incontinence has been somewhat dispelled, but longer follow-up of these patients is required. The mobilization continues circumferentially until the urethrovaginal confluence can reach the perineum (see Donahoe and Schnitzer, 2006).



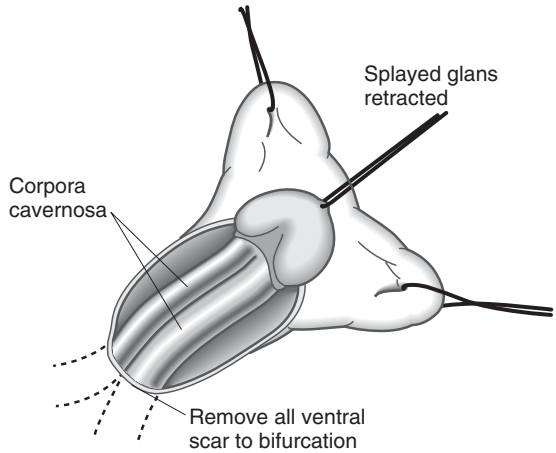
5a

RECONSTRUCTION FOR MALE GENDER ASSIGNMENT

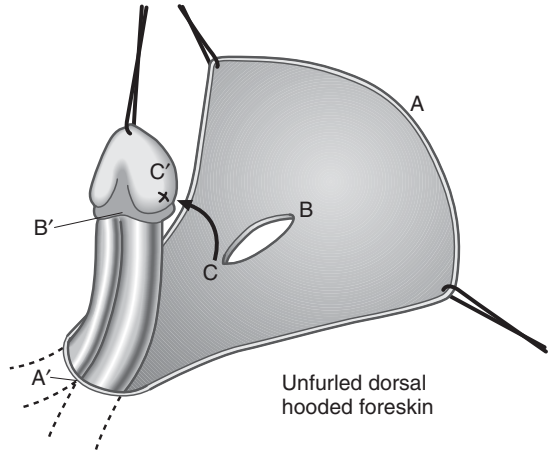
Hypospadias repair: first stage

5a–e In many cases, the surgeon is dealing with micropenis and a bifid scrotum; hence, we suggest staging these hypospadias repairs. In the first stage, the chordee is straightened and the redundant dorsal foreskin is shifted ventrally to close the ventral defect (a). Then it is subsequently used to create the neourethra at the second stage. The foreskin is unfurled and the shaft skin is circumferentially mobilized to the penoscrotal junction. An artificial erection is performed to assess the severity of the chordee. All

ventral scarring (b) from residual atretic corpora spongiosa is removed from the glans to the bifurcation of the corpora cavernosa. We stage the repairs for these children because bleeding can be considerable and can place the neourethral graft at risk. The dorsal hooded foreskin is unfolded (c) to create a single layer, and a longitudinal incision is made to allow the dorsal tissue to be positioned to cover the ventral defect. C is sutured to C′, and the distal unfurled foreskin is rotated over the top of the glans. B is sutured to B′ and A is sutured to A′ on the ventral surface, after the flap has been transposed. There is usually sufficient redundancy for use as a free urethral graft at a later stage of the reconstruction (d). A Foley catheter drains the bladder through the perineal meatus and the shifted foreskin is closed (e).



5b



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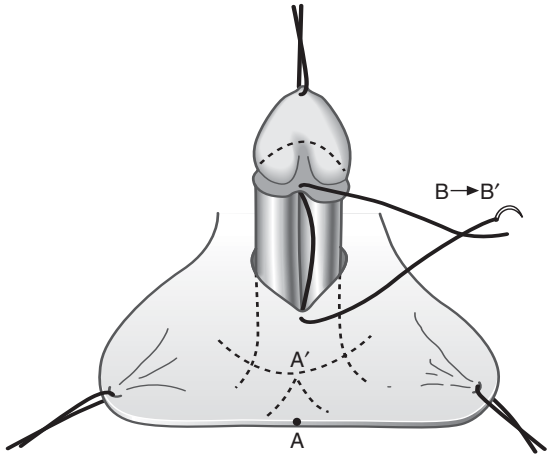
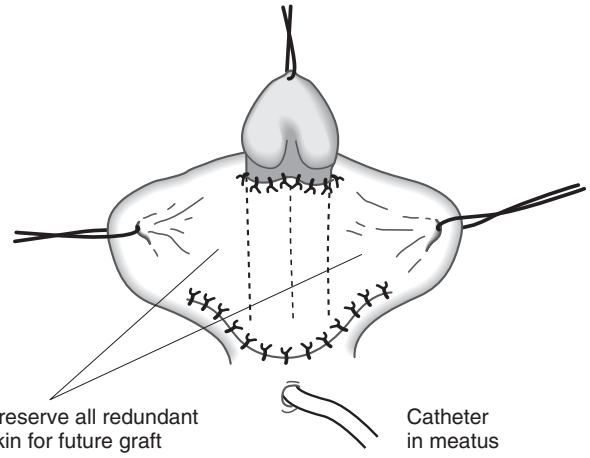


5d

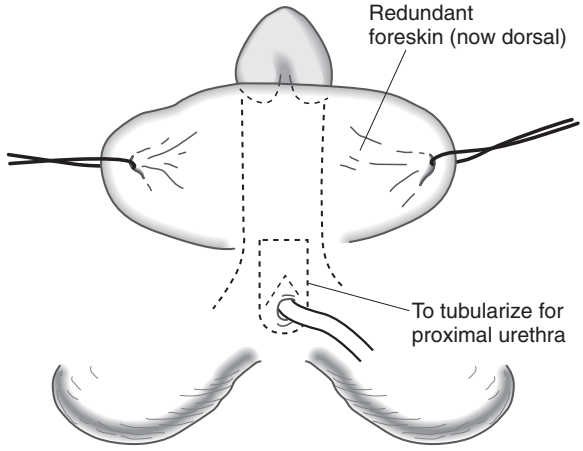


5e

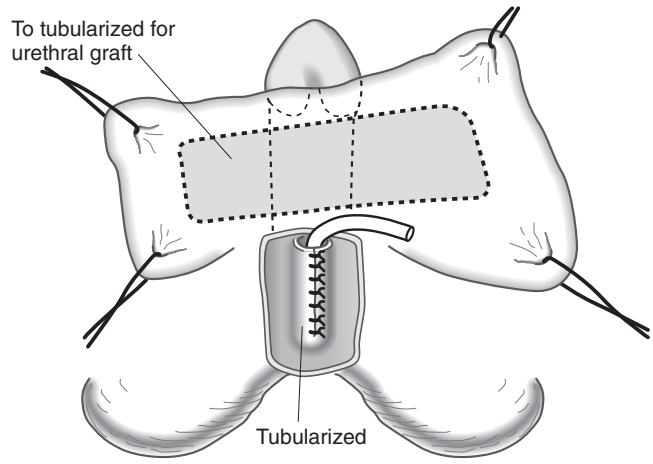
Hypospadias repair: second stage

6a–d Six to nine months later, when the shifted ventral skin feels pliable, the second-stage of a hypospadias repair is performed using composite material to create a neourethra from the perineoscrotal meatus to the tip. The microphallus, which may need to be augmented with two or three doses of testosterone given once a month, is treated with at least a 4-week interval without testosterone to decrease postoperative penile erections and hyperactivity. The transposed preputial skin (a) is used to reconstruct the glans and urethra. A 12–15 mm diameter strip is outlined on the transposed preputial skin (b), removed, defatted, and tubularized with running, inverting, subcuticular 7/0 Maxon or Polydioxanone suture (PDS) (c). The new urethra must be wide enough to avoid strictures; a second layer of subcutaneous tissue decreases the incidence of fistulas. The skin is closed with multiple Z-plasties (d) with interrupted stitches of 5/0 or 6/0 chromic. The repair is performed over a silastic stent, which traverses the complex neourethra, with a proximal urethrostomy or suprapubic catheter to drain the bladder. The penis is wrapped with Xeroform gauze and dry gauze or Tegaderm and fixed upward over the pubis to control postoperative bleeding and edema.

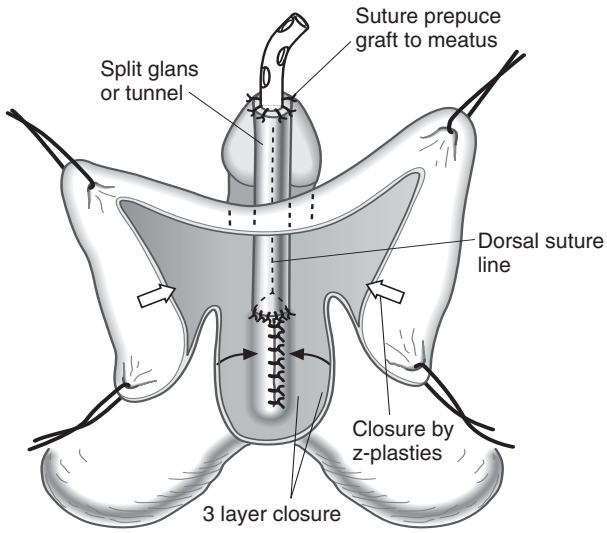
Some surgeons splay a small glans and augment it with transposed foreskin at the first stage. If the phallus is of adequate size, a longitudinal strip (15 mm in width) can be outlined from the perineum to the tip of the glans. This is tubularized and the remainder of the transposed tissue and the glans closed over the neourethra.



6a



6b



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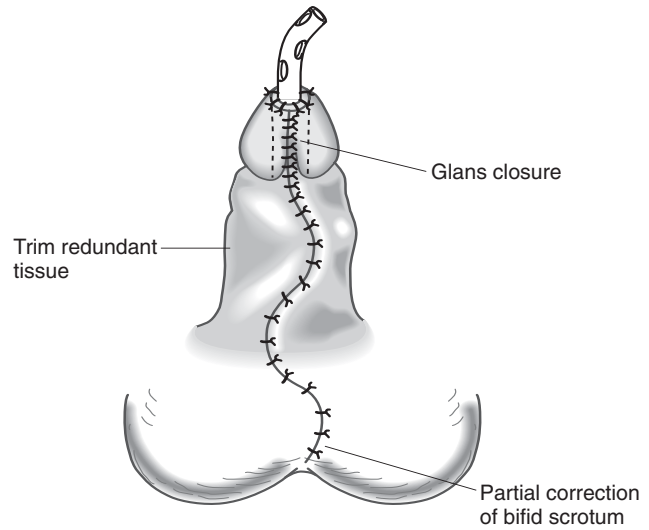
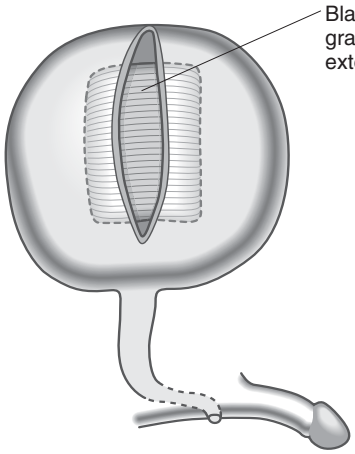


6d

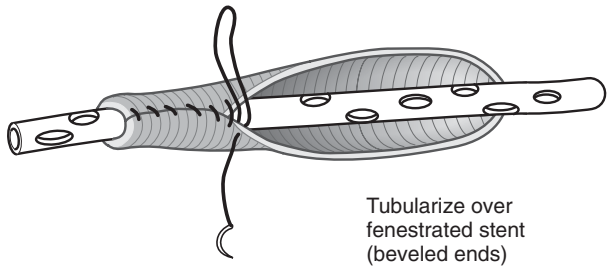
In the rare circumstance in which insufficient tissue is available for the urethroplasty and refashioning of the prepuce, a graft of bladder mucosa or buccal mucosa can be used successfully.

BLADDER GRAFT TO FORM A NEOURETHRA

7a–d If local tissue is insufficient for the creation of a neourethra, bladder mucosa may be used. The bladder is opened in the midline through the muscularis, leaving the mucosa intact. The bladder mucosal graft is then carefully measured, harvested, tubularized over a fenestrated stent, anastomosed to the proximal neourethra in a beveled fashion, tunneled to the meatus, and sutured to the tip. These grafts may stenose at the meatus, and should be dilated with a meatal sound 2 weeks after surgery for another 2–3 weeks. Longitudinal trim may be required later, because these fragile grafts tend to stretch.



7a



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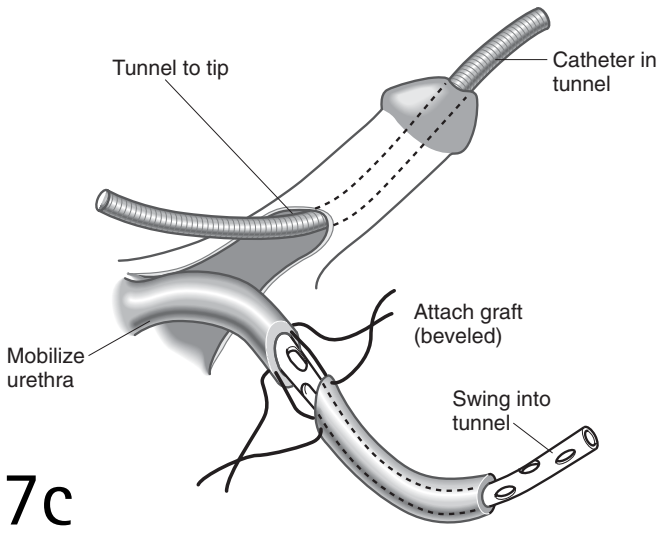

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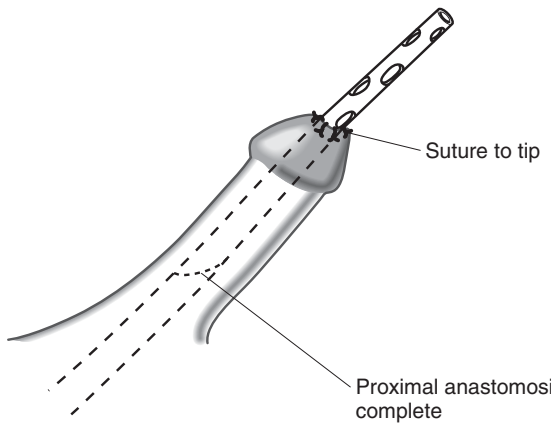

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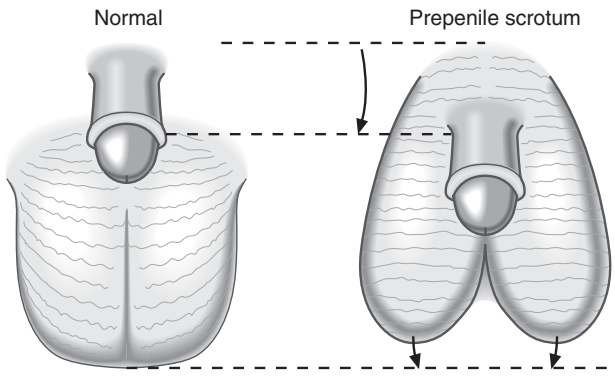
BUCCAL MUCOSA GRAFT

The histologic characteristics of bladder mucosa, ease of its harvest, its compatibility with the urinary system, and the excellent results obtained are responsible for the increased popularity of the buccal mucosa, which could be used as a tubularized graft, as an onlay flap, or for the first stage of a two-stage hypospadias repair. The harvest site has included the lower lip and insides of both cheeks. Buccal mucosa does not stretch and can be harvested one-to-one for urethral size. Meatal problems such as excoriation, encrustation, and protuberance are not encountered as when bladder mucosa is used.

REPAIR OF PREPENILE SCROTUM

8a-f Intersex children often have a bifid, shortened scrotum associated with scrotal tissue anterior to the dorsal base of the penis. This anomaly is repaired by displacing scrotal skin posteriorly and the penis anteriorly. The base of the penis is advanced forward onto the anterior abdominal wall by creating a square, distally based flap, which circumscribes the base of the penis. The flap is dropped distally to restore normal scrotal length. The abdominal wall is then undermined and swung around the base of the penis to join ventrally in the midline. It is important to mobilize the anterior abdominal wall flaps sufficiently so that midline separation does not occur. The scrotum is then closed in the midline and laterally.



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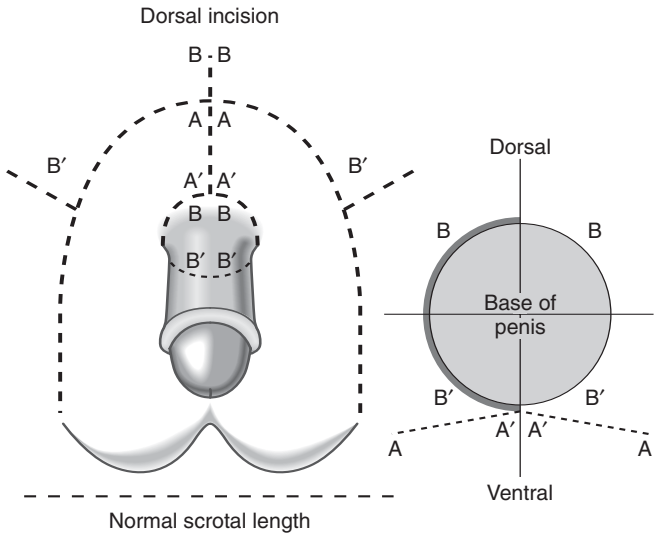

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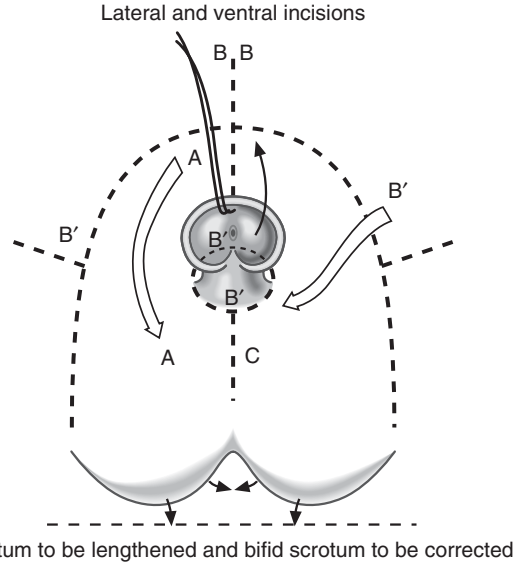

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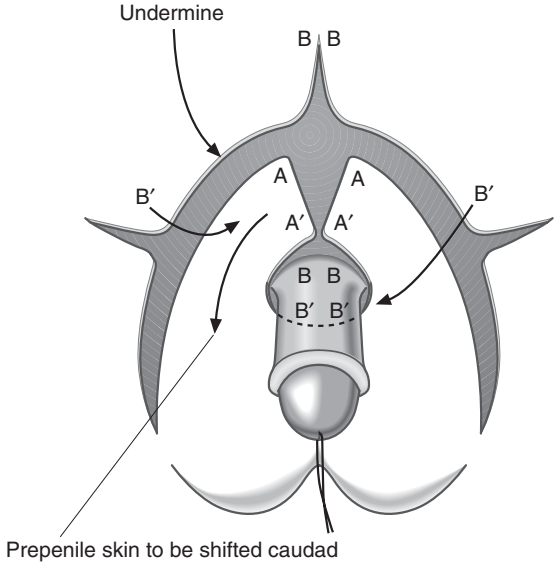

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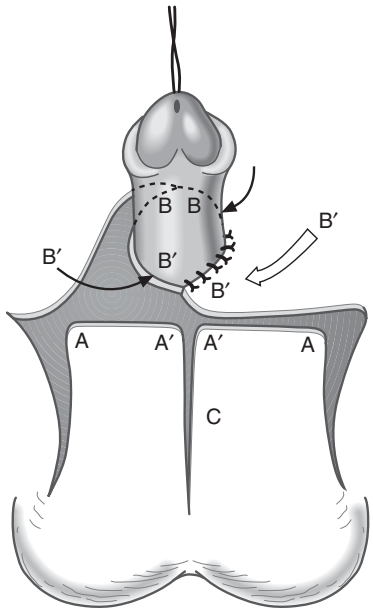

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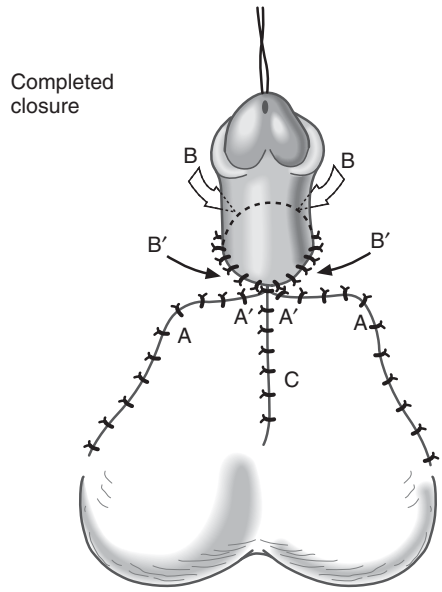
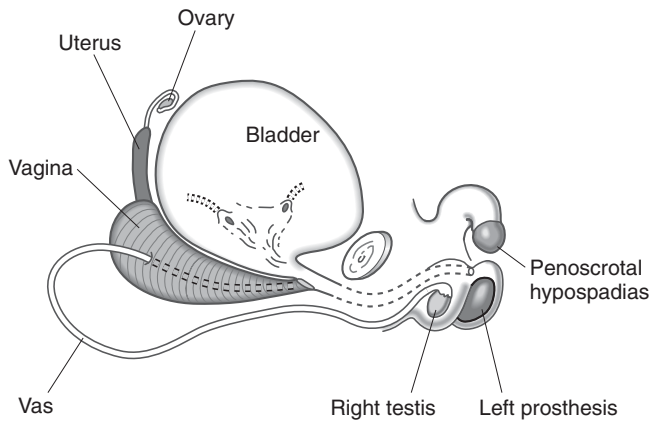


8f

REMOVAL OF RETAINED MULLERIAN DUCTS AND CREATION OF A NEOSEMINAL VESICLE

9a-c In males with retained Mullerian ducts, the vas deferens often lies within the side wall of the dilated vagina; its course, however, cannot usually be palpated in the thickened vaginal wall. Therefore a strip of vagina surrounding the predicted course of the vas is preserved when the uterus and vagina are resected. The vaginal strip is then turned in and tubularized from the proximal vas to the point of union with the urethra using interrupted sutures. If the vas courses in the wall of the uterus, it is not possible to perform this procedure.



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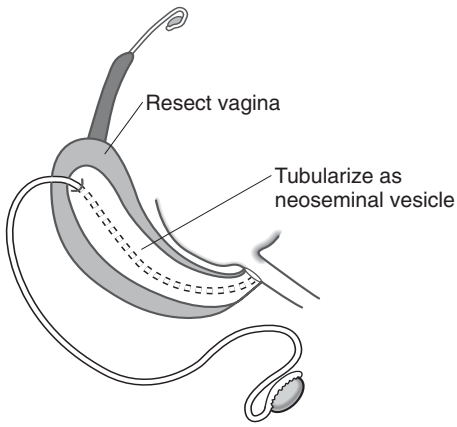

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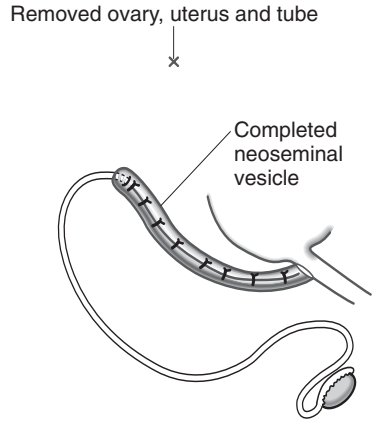

9c

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90

Cysts and tumors of the ovary

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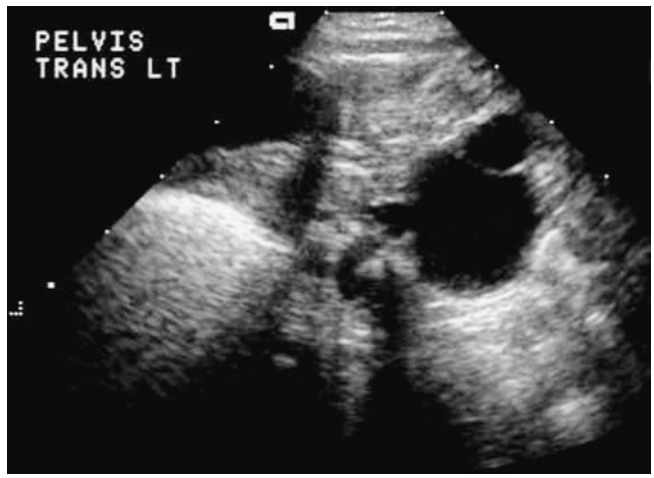
PRINCIPLES AND JUSTIFICATION

Ovarian cysts are extremely common and should be regarded as a separate entity from cystic neoplasms when considering surgical management. Follicular cysts are not neoplastic and are sensitive to hormonal stimulation. They are detected most often after puberty, but are also seen prenatally and in newborn girls due to maternal estrogen stimulation. The majority of neonatal cysts are asymptomatic and spontaneously decrease in size by 3–4 months of age. Torsion may occur antenatally and produce a complex appearance to the cyst. Large cysts may occasionally cause respiratory distress or visceral compression impairing feedings. Symptomatic neonatal cysts require surgical intervention, but techniques designed to preserve normal ovarian parenchyma should be used whenever possible. Simple aspiration is occasionally successful, but high recurrence rates are seen, and cyst fenestration by open or laparoscopic technique is preferred. Cysts in the prepubertal child associated with precocious puberty should be evaluated by the pediatric endocrinologist. Postpubertal cysts may be asymptomatic or may present with pain due to large size, hemorrhage, or torsion. Most simple cysts will resolve spontaneously within three to four menstrual cycles. Indications for surgical intervention include persistent symptoms, pain, or evidence of torsion.

Ovarian neoplasms often have a cystic component. Benign teratomas are most common, but the overall risk of malignancy for ovarian neoplasms in children and adolescents is approximately 20 percent. There are no gross characteristics that clearly distinguish benign from malignant neoplasms and therefore it is best to approach all ovarian neoplasms as if malignancy may be present. Epithelial neoplasms account for approximately 16 percent of ovarian neoplasms in children and most are benign.

PREOPERATIVE ASSESSMENT

Ultrasound is an excellent modality for the initial imaging of ovarian lesions. Simple cysts can be identified and blood flow to the ovary can be determined. Serial examinations can be done for asymptomatic cysts to monitor size.



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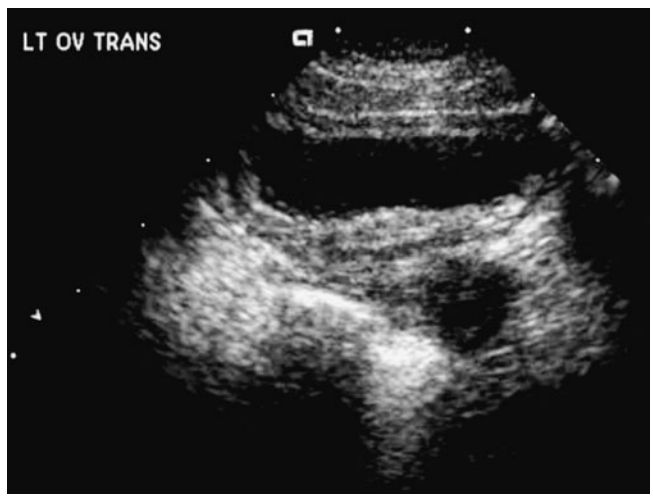

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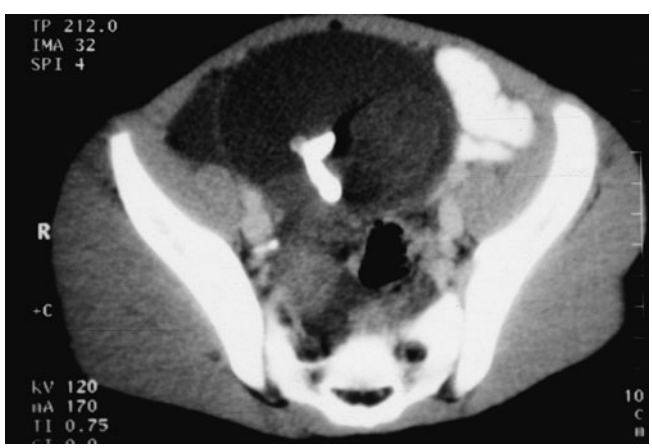
1 Hemorrhagic cysts can be followed with ultrasound to confirm the expected evolution of clot lysis and debris resolution. Early studies may show septation and debris that disappears on follow-up images with decrease in size of the ovary and normal-appearing parenchyma.



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2 Neoplastic masses should also be evaluated by computed tomography scan to assess for adenopathy, hepatic involvement, and the presence of tumor elsewhere within the peritoneal cavity. Most will have a heterogeneous appearance with cystic areas. Calcifications suggest a component of teratoma, but mixed tumors with malignant and benign tissues are common. The tumor markers alpha-fetoprotein (AFP) and beta-human chorionic gonadotropin (βhCG) should be determined preoperatively. Alpha-fetoprotein is elevated in tumors that contain the malignant germ-cell element endodermal sinus tumor, and βhCG is elevated in tumors that contain the malignant element choriocarcinoma. There are no reliable serum markers for malignant tumors composed of only germinoma or embryonal carcinoma. Consequently, the preoperative finding of normal serum markers will not exclude the possibility of malignancy.

Anesthesia

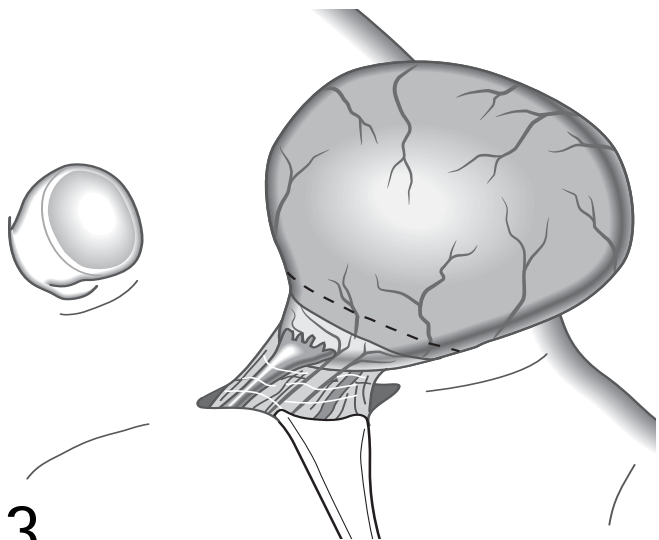
General endotracheal anesthesia with muscle relaxation is required. For both open and laparoscopic procedures, decompression of the bladder with a Foley catheter is recommended to provide optimal access to the pelvis. For laparoscopic procedures, orogastric decompression is also recommended. If a large tumor requiring a generous midline incision is present, a supplemental epidural catheter for postoperative pain management should be considered.

OPERATION FOR CYSTS

Non-neoplastic cysts may be treated by a laparoscopic or open technique.

Procedure

For open cases, a low transverse incision in a natural skin crease on the side of the lesion is convenient. Alternatively, a Pfannenstiel incision may be used.



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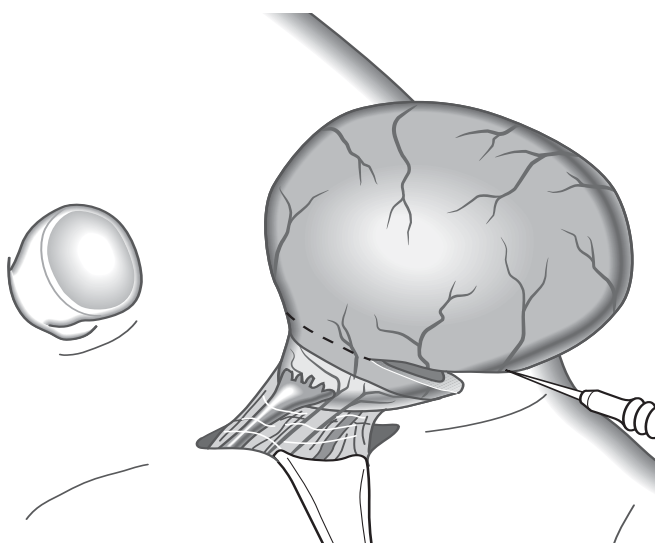
3 The cyst is delivered through the incision. A tense cyst may be needle aspirated to facilitate grasping and delivery through a smaller incision. The ovary is inspected to locate the normal parenchyma and pedicle. The cyst may be enucleated if a plane is easily developed without injury to the ovarian parenchyma.

4 Alternatively, the free margin of the cyst wall is excised with cautery. The ovary is returned to the peritoneal cavity after confirming hemostasis. The fascia is closed, followed by subcuticular skin closure.

For laparoscopic cases, an umbilical port is placed for insufflation and inspection with the camera. Placement of the patient in the Trendelenberg position will allow the bowel to fall cephalad and improve exposure. Two additional 5 mm trocars are placed for grasping and manipulation. As in open cases, a tense cyst may be difficult to grasp. Transabdominal aspiration under direct vision with a long, 18-gauge spinal needle may allow the cyst to be grasped more securely. A portion of the cyst wall is excised with the hook cautery to create an adequate fenestration. The 5 mm ports are withdrawn under direct vision, followed by the umbilical port. The umbilical fascia is closed and subcuticular skin closure is done for all sites.



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OPERATION FOR NEOPLASM

Although some authors recommend a laparoscopic approach for smaller cystic neoplasms, the incidence of rupture of the lesion is higher with laparoscopy for both oophorectomy and enucleation. The risk of malignancy in pediatric ovarian neoplasms and the ability to complete a full staging procedure make an open approach preferable in most cases.

Procedure

The choice of incision depends on the size of the lesion and the surgeon's preference. In general, a lesion with a diameter less than the span of the iliac crests may be successfully approached by a transverse incision in the lowest skin crease. Lesions exceeding this size are best approached by a midline incision tailored to the dimensions of the mass or a Pfannenstiel incision.

Peritoneal cytology

Immediately on entering the peritoneal cavity, the pelvis should be aspirated for ascitic cytology. If no ascites is present, pelvic washing should be done with normal saline.

Inspection of the ovary

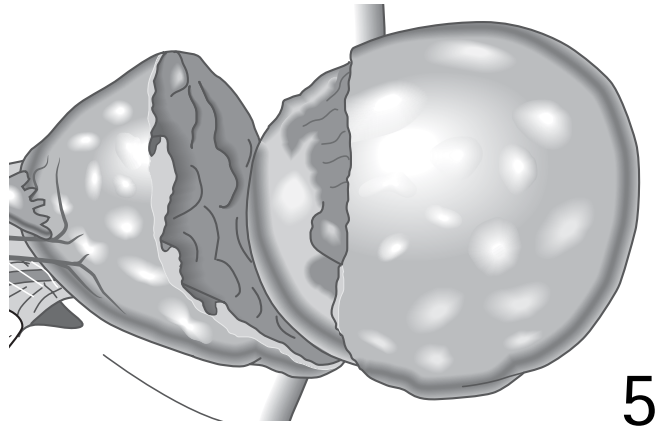
The involved ovary should be inspected for resectability. If preoperative tumor markers are normal and there is a clear plane of demarcation between the mass and normal ovarian parenchyma, enucleation of the mass may be considered.

Enucleation of ovarian mass

5 The ovarian cortex is incised to expose the border between the parenchyma and the mass. Dissection should remain outside the capsule of the neoplasm, with care being taken not to rupture it. The ovarian capsule edges may be re-approximated with absorbable suture to minimize raw surface area.

Oophorectomy

If the tumor markers are elevated or there is no demarcation between normal ovary and neoplasm, oophorectomy should be undertaken. In most cases, the fallopian tube is not involved and may be spared.



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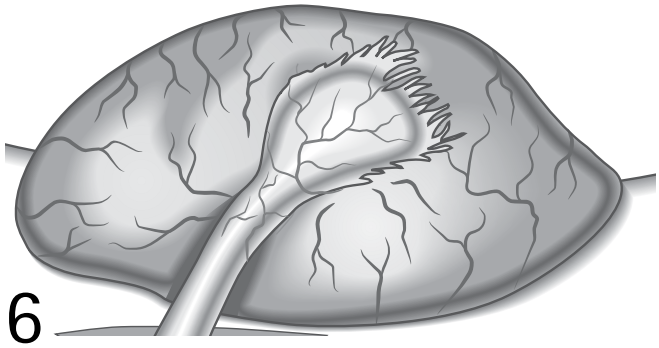

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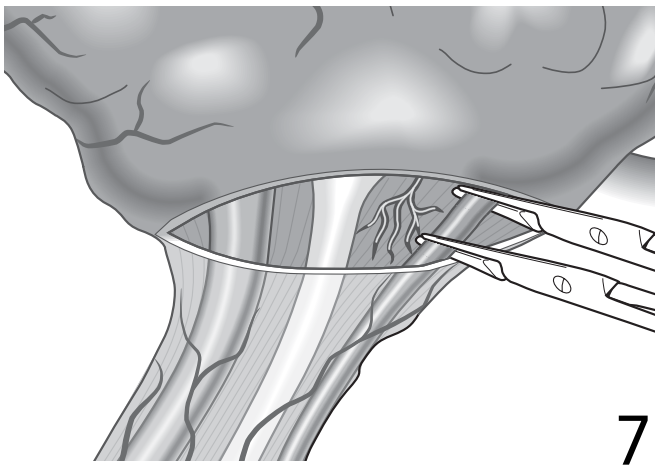
6 With large tumors, the leaves of the mesovarium are widely splayed and the tube may be draped over the mass.

7 The peritoneum of the mesovarium is incised in an avascular plane between the tube and ovary with cautery dissection. This maneuver will expose the vascular pedicle of the ovary. If the tumor is large, the venous plexus is often engorged, and the pedicle may need to be taken in stepwise fashion with pairs of clamps, division of the tissue, and ligation.



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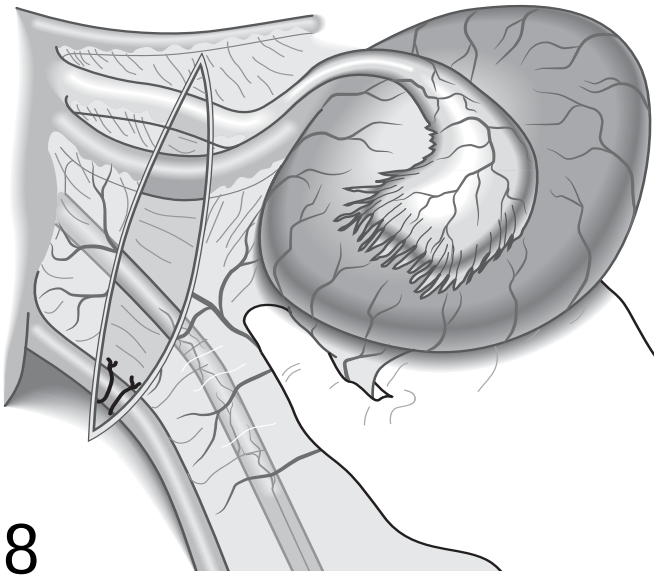

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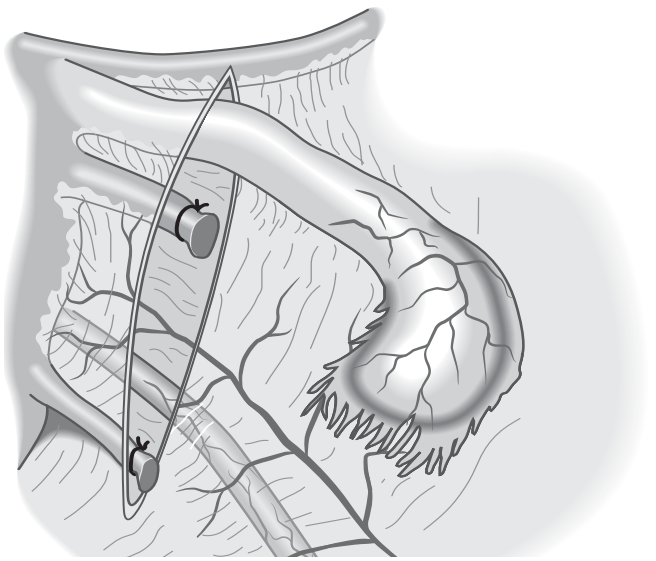
8 The assistant should maintain manual compression of the pedicle as the dissection proceeds. The specimen is passed off the field with its capsule intact.

9 If the tube is adherent, it may be taken in continuity with the ovary. Care should be taken clearly to identify the ureter before proceeding with division of the mesosalpinx.

If the lesion is invading the uterus or adjacent pelvic structures, it should not be resected at the primary operation. A biopsy for tissue diagnosis should be done with a plan for later resection after chemotherapy. Chemotherapy is highly successful in producing shrinkage of tumors to allow later resection without sacrifice of the uterus or other pelvic structures.



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Completion of tumor staging

The remaining staging maneuvers should then be completed for all neoplastic lesions. These include the following:

- Inspection and palpation of the omentum. Removal of any abnormal areas.
- Inspection and palpation of the retroperitoneal lymph nodes from the iliac chains up along the aorta and vena cava at least as far as the renal pedicle. Any enlarged or firm nodes should be biopsied.
- Inspection of the pelvic and visceral peritoneal surfaces with biopsy of any abnormal areas.
- Inspection and palpation of the contralateral ovary with enucleation of any abnormal masses.

Closure

The muscular fascia is closed in layers followed by Scarpa's fascia and subcuticular skin closure.

POSTOPERATIVE CARE

Regardless of the approach chosen, the Foley catheter can generally be removed immediately after the procedure for small to moderate-sized lesions, and rapid advancement of diet is tolerated. For large lesions requiring a generous incision, it may be more comfortable for the patient to leave the Foley catheter in place for 24 hours.

OUTCOME

Fenestration for non-neoplastic cysts has an excellent outcome with minimal long-term consequences. Malignant germ-cell tumors are treated with platinum-based multiagent chemotherapy. Survival with current regimens is greater than 90 percent at 5 years.

All neoplastic lesions, whether benign or malignant, have some risk of contralateral recurrence. These patients should undergo periodic ultrasound surveillance to allow detection of new lesions at an early stage, so that an ovary-sparing procedure may be undertaken, with greater chance of success in preserving endocrine function and fertility.

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SECTION VII

Neuromuscular and Skeletal

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Myelomeningocele

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PRINCIPLES AND JUSTIFICATION

Myelomeningocele is the most severe of the spinal dysraphic states and still the most common, although its incidence is decreasing with improved prenatal maternal nutrition and through the availability of prenatal screening mechanisms. Myelomeningocele is a central nervous system fusion defect. Neural tube defects affect both men and woman and have a slight female preponderance. There have been clusters of increased incidence of myelomeningocele in certain populations, such as the Irish, British, Sikhs, Guatemalans, and the Egyptians of Alexandria. Siblings of an affected child have an increased risk of neural tube defects when compared with the general population. The rate of recurrence ranges from about 1 percent to nearly 10 percent in different series. A clear correlation between maternal diet and neural tube defects has been determined. A seven-fold reduction in neural tube defects has been achieved with folate and vitamin supplementation before and during pregnancy.

Embryology

The central nervous system begins as a focal proliferation of ectoderm. The central groove develops and forms two folds of neural tissue. At approximately gestational day 20–28, the lips of this fold touch and the neural tube fuses. Starting at the center, the fusion of this fold eventually becomes the craniovertebral junction. This fusion proceeds in a caudal and cephalic direction. The caudal section is the last to close. A plane develops between the neural ectoderm that has fused and the overlying superficial ectoderm. Between these layers migrate mesenchymal cells, which give rise to the arch of the vertebrae and the paraspinal muscles. Myelomeningocele represents a failure of fusion of the neural fold, leaving the vertebral arches open and the unfused spinal cord or neural placode either exposed or covered by a thin membrane. It is important to distinguish a myelomeningocele from less severe defects such as meningoceles, which consist only of a spinal fluid-filled sac with meningeal and cutaneous coverings. Meningoceles contain no spinal elements, but can occasionally contain some nerve roots. True meningoceles are rare. Lipomyelomeningoceles are usually covered by well-developed dermal elements and fat, and consist of spina bifida with associated abnormalities of the spinal cord and extension of fat into the spinal canal.

Associated problems

Like most congenital anomalies, spina bifida is a spectrum of disorders. Myelomeningocele represents the most severe of these spinal dysraphic states and includes anomalies of the brain such as the Chiari II malformation and hydrocephalus. It has implications for other organs, such as those of the genitourinary and musculoskeletal systems. Most children with myelomeningocele have some degree of weakness of their lower extremities and many have significant orthopedic problems. As a result of denervation, muscle imbalance ensues and can result in abnormalities at the hip, knee, and foot. Anesthesia of various portions of the skin can lead to pressure sores, particularly later in life. Anorectal neuropathy may cause a variety of defecatory dysfunctions.

Urologic abnormalities are common in children with myelomeningocele and are best managed with intermittent catheterization. Careful follow-up with evaluation of the kidney and bladder function is extremely important.

The cornerstone of the management of children with myelomeningocele is a multidisciplinary team that treats and assesses the various needs of both the child and the family.

The development of cerebrospinal fluid (CSF) diversionary devices (shunts) in the 1950s has allowed these children to be successfully managed and has dramatically changed the outlook for them. More recently, endoscopic techniques have been developed that allow more physiologic diversionary procedures of the CSF (third ventriculostomy) and these have also been used successfully in children with myelomeningocele. Such physiological diversionary procedures may be particularly important in that they eliminate the need for a mechanical device.

Selection criteria

The controversial discussion about the selection of children for myelomeningocele repair has largely subsided, as more effective means of diagnosis and treatment have been developed. Before the development of successful methods of treating hydrocephalus, selection of candidates for non-treatment was commonly practiced. With shunting devices available, the cognitive outcome for these children has significantly improved and, in most cases, children with myelomeningocele receive early repair. Although discussion among various groups, including ethicists, clerics, jurists, administrators, legislators, and physicians, still continues, most children now receive repair of their myelomeningocele and are treated by a team with a variety of specialists. At best, selection criteria related to which children should not be repaired are inconsistent. Studies that discuss the outcome of such unrepaired children must be viewed cautiously, as many who were initially not treated received later repair, and not all who were left unrepaired died, as had been anticipated. The ease of care of children with myelomeningocele is greatly improved following repair, and thus even chronic care facilities often will not accept children with open defects. Unless associated additional central nervous system defects or other congenital anomalies are of such magnitude as to suggest that meaningful survival is unlikely, repair of myelomeningocele in children is now considered a standard of practice.

PREOPERATIVE

Assessment

The principles involved in the repair of myelomeningocele in children consist initially of a complete evaluation of the child, including detailed physical and neurologic examination. Ultrasonographic imaging of the head, spine above and below the area of the defect, and kidneys can be an important adjuvant to constructing a surgical plan for an individual child. If there are many associated intracranial or spinal anomalies on ultrasonic examination, computed tomographic (CT) scanning or magnetic resonance imaging (MRI) should be performed to evaluate these abnormalities more carefully. Radiographs of the spine can identify additional anomalies such as diastematomyelia, which may require repair at the time of the initial surgery. Blood should be cross-matched and available. Consideration of the timing of surgery must also include assessment of underlying medical issues. The decision as to whether a diversionary procedure for the CSF is necessary at the time of the initial repair must also be made.

Preparation

Infants born with myelomeningocele require the skilled services of a multidisciplinary team. After initial determination of cardiorespiratory stability, and any underlying infections, the child may be transferred to an appropriate institution. The lesion itself should be covered with moist, sterile gauze dressings surrounded by a protective plastic sheet. The use of sponges impregnated with bacitracin ointment covered by a thin plastic layer is recommended to prevent additional skin breakdown. Alternatively, sterile saline-soaked sponges may also be appropriately applied to the myelomeningocele placode.

Emergency operative intervention is seldom necessary in neonates with myelomeningocele; the repair can be safely carried out within the first 48–72 hours after delivery. Delaying closure for more prolonged periods may increase the risk of central nervous system infection and may decrease motor function by increasing the trauma to the exposed neural placode. In one study, deterioration of motor function occurred in children left untreated, and a 37 percent incidence of ventriculitis was found in children in whom repair was delayed.

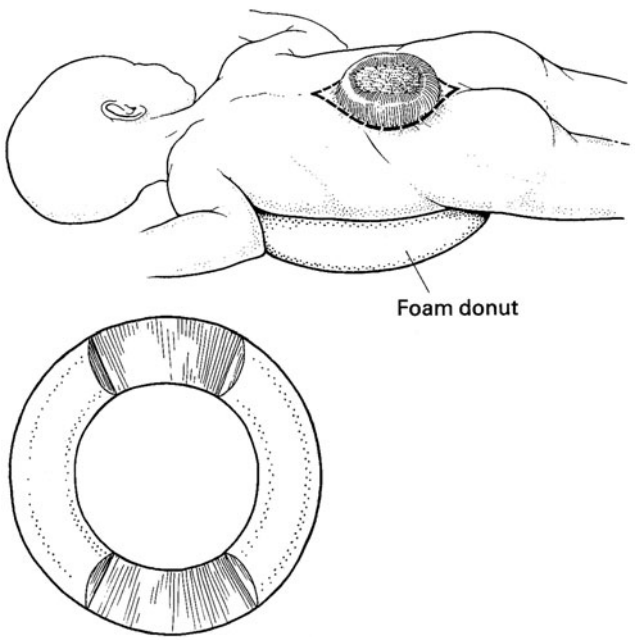
Most myelomeningoceles are in the lumbosacral region, although they can occur anywhere along the spinal cord. Assessment with good spinal ultrasonography can help alert the surgeon to associated abnormalities such as diastematomyelia, arachnoid cysts, and intradural masses such as dermoids, which may complicate the repair of the myelomeningocele.

If significant hydrocephalus is found in the preoperative cranial ultrasound, either shunting or a diversionary procedure of CSF such as endoscopic third ventriculostomy should be considered. If ventriculomegaly is mild to moderate, such a diversionary procedure may not be performed at the same time as the myelomeningocele repair, but may be delayed. A significant proportion of children with myelomeningocele will require shunting in the first days of life. This number has ranged from 60 percent to 80 percent of children with myelomeningocele. It should also be noted that there may be an increased risk of leakage from the repair site in those children in whom shunting is delayed.

Anesthesia

General endotracheal anesthesia with overhead warming lights is appropriate. Occasionally, intraoperative use of a nerve stimulator is necessary to distinguish functional nerve

roots; so paralytic agents must be used appropriately and no longer be present when stimulation is planned. Intravenous lines must be of sufficient size to accommodate blood transfusion. Careful assessment of blood loss is necessary, as blood loss can be significant, particularly when rotational flaps are employed. A Foley catheter is generally used for bladder drainage and proves to be a useful adjunct in keeping the repair site clean and dry during the postoperative period.



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OPERATION

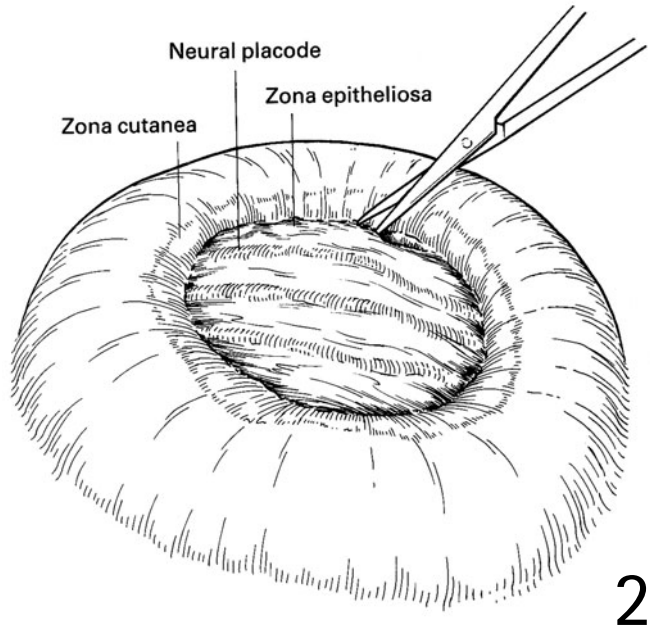
Position of the patient

1 Repair of the myelomeningocele is performed with the child in the prone position. Careful attention to the position of the neck is important, as almost all children with myelomeningocele have a Chiari II malformation and most will also have some aspect of hydrocephalus. Head size may occasionally be quite large in such children. Positioning of the head must therefore be done in such a way as to avoid kinking of the internal jugular veins and undue extension or flexion of the cervical spine. The abdomen must be hanging free so that intra-abdominal pressure is not increased. Increases in intra-abdominal pressure lead to compression of Batson's venous plexus and result in increased engorgement and bleeding from epidural veins. The author has found that a foam-rubber donut that has been cut out at the top and the bottom acts as an excellent bolster. Various bolsters and rolls have been employed, with the goal always to allow the abdomen to be hanging free. Careful attention to positioning of the upper extremities to avoid brachial plexus stretching injury is also necessary. The lower extremities also require careful positioning and padding, as congenital dislocations of the hips and multiple orthopedic anomalies may be present and they make such positioning quite difficult.

The central neural placode and membranous areas are cleansed with sterile saline. The surrounding areas of skin are cleansed with an iodinated solution; iodinated solutions should not be applied directly to the neural placode. Most surgeons find magnifying loupes useful in further repair of such myelomeningoceles. Occasionally, the operating microscope can also be of benefit. Bipolar electrocoagulation should be available, as it is employed throughout the procedure to control bleeding.

Freeing the neural placode

2 Dissection of the neural placode begins at the lateral aspect of the placode at the junction of the zona epitheliosa and the edge of the hemangiomatous skin (zona cutanea). This can be done with sharp iris scissors or tenotomy scissors. A significant amount of yellowish CSF will egress when the sack is opened, which deflates the cystic portion of the myelomeningocele sac. At this point, careful inspection of the interior of the sac is necessary.



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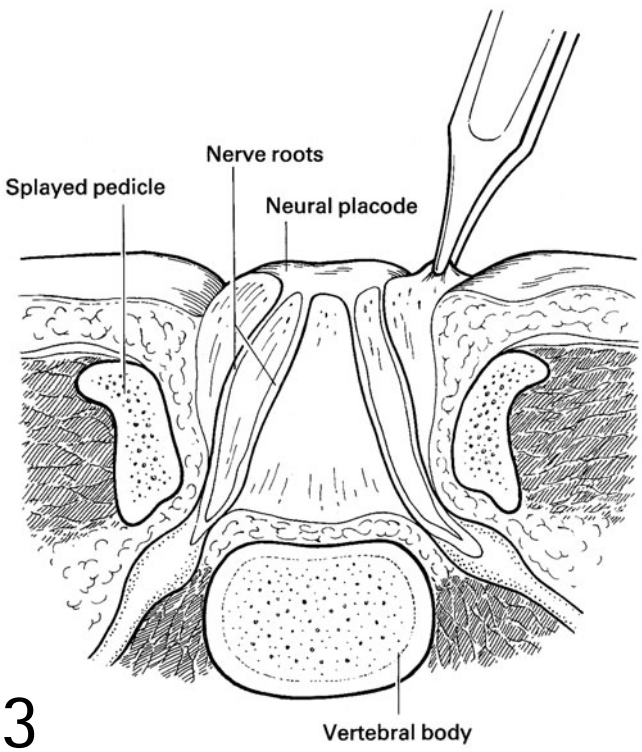

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3 Examination of the contents of the sac demonstrates that the floor of the sac is formed by the glistening white dura, which is adherent to the surrounding fat and mesodermal elements. Medially, nerve roots can be seen passing from the neural placode down to the spinal canal, which is also flattened relative to a normal canal.

The sharp dissection is carried out on either side and then completed at the cephalic and caudal ends. It should be noted that at the upper end the placode is nearly a normal spinal cord and is invested by normal arachnoid and dura and has the typical cylindrical shape of the spinal cord.

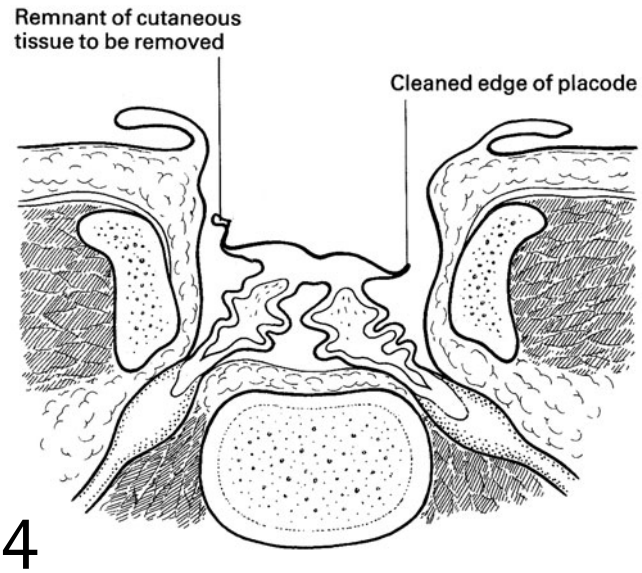
At the upper end of the neural placode, filamentous adhesions may bind the cord/placode to the dura. These should be carefully divided. Dissection of the placode from the zona epitheliosa and the hemangiomatous skin edges is important and it is here that magnification proves to be particularly useful.

Dissection is most difficult at the cephalic and caudal portions of the placode, as there are usually multiple adhesions to both the dura and the surrounding skin. Occasionally, additional laminectomies and dural opening are necessary to achieve adequate exposure of the spinal cord to free adhe-

sions. An important goal of myelomeningocele closure is the untethering of the neural placode as well as repair of the skin and dural defects. Freeing of these adhesions allows the cord to slide more gently within the dural sac. Particularly at the most distal portion of the placode, there may be a very prominent fibrous band, which is a form of filum terminale.

Inspection of the internal contents should include a check for fibrocartilaginous or bony spurs near the level of the first intact lamina. Such spurs, often seen on preoperative radiographs, suggest a narrowed intervertebral space if a vertebral body anomaly is seen or if a midline septum is seen on ultrasonographic examination. A laminectomy at one or two levels above this level may be necessary to visualize and deal with such septae adequately.

Adhesion of the terminal portion of the placode can be quite dense. Where there are significant fibrous bands, use of a nerve stimulator can often help to distinguish these bands from functioning neural tissue.



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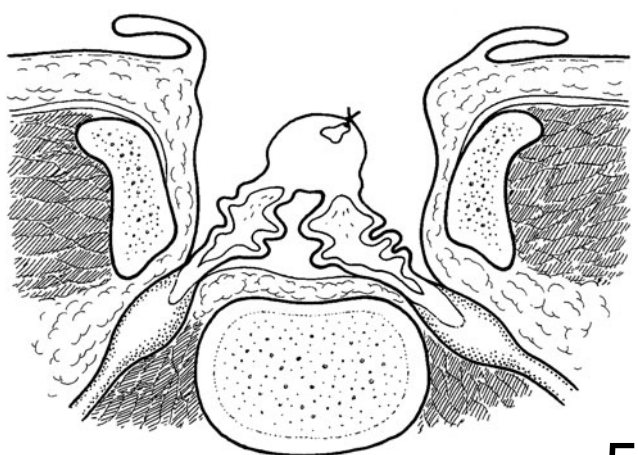
Placement of the neural placode back into spinal canal

4 The edges of the neural placode should be carefully inspected to be certain that there are no dermal elements included in the placode. Pieces of the membranous tissue of the zona epitheliosa and thin parchment skin from the zona cutanea should be sharply debrided from the edges of the placode. The placode, if completely freed of adhesions, should rest within the spinal canal.

Reconstitution of the neural tube

5 Controversy exists as to whether reconstitution of the neural tube is beneficial. If the neural placode is very thin, the edges may be brought together to reconstitute the neural tube. A few sutures of 10/0 polypropylene (Prolene) can be placed into the arachnoid elements at the edge of the placode to reconstitute the tube. It is hoped that the maneuver will decrease tethering and prevent adherence to the surrounding dura. Care must be taken not to place these sutures through neural elements, and reconstitution of the tube should not be performed if the edges of the placode do not easily come together. If such a maneuver will cause undue strangulation or pressure on the placode, it should not be performed.



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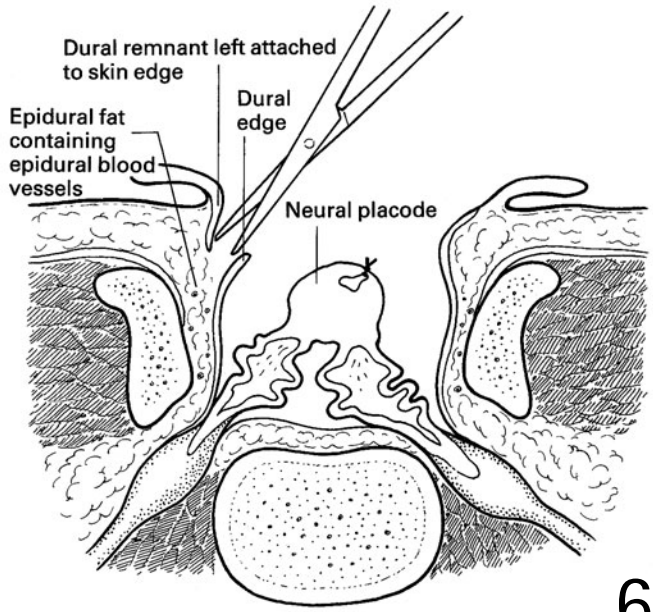
Identification of the dural edges

6 One of the most important aspects of the repair of myelomeningocele in a child is the achievement of a watertight dural closure. The dura (a glistening white structure) is adherent to the edges of the myelomeningocele sac. It is most easily identified by carefully examining the upper end of the sac near where the neural placode is reconstituted into a normal spinal cord. It may not be reconstituted at the lower end, depending on the level and extent of the myelomeningocele defect. The dura is dissected initially at the upper end off the paraspinous fascia. Care must be taken to go high enough up along the walls of the sac to provide sufficient dura to close over the placode and to obtain a watertight closure. It is important that the dural closure is sufficiently capacious to prevent strangulation or vascular compression of the neural placode.



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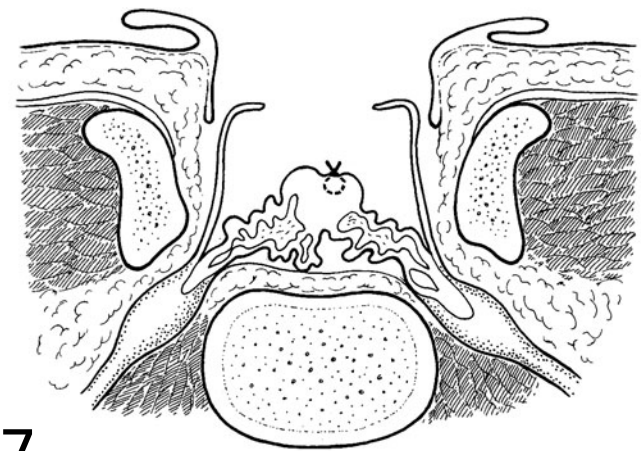

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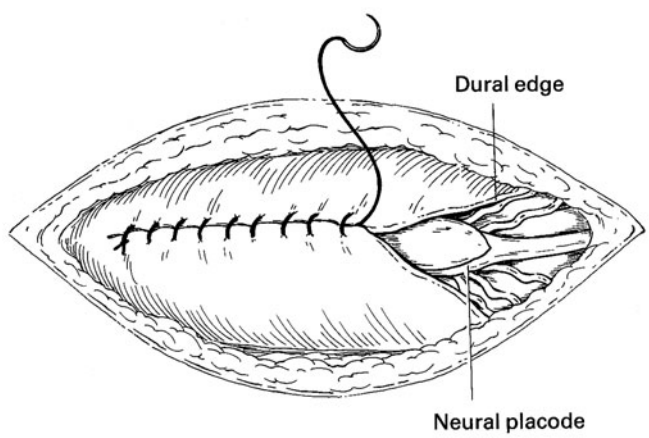
Dissecting of the dural edges

7 A cuff of residual dura is left along the edges and can be used to anchor subcutaneous stitches to assist in the closure of the skin edges. An important landmark in identifying the plane of dissection to free the dura from the surrounding tissue is identification of the epidural fat. The epidural fat is more loosely developed than subcutaneous fat and has within it a rich blood supply including significant epidural veins. These epidural veins can be quite large and should be coagulated where necessary.

Closing the dura

8 The dural edges should be brought together using a running stitch of either 4/0 or 5/0 polypropylene (Prolene) or abraided nylon. If insufficient dura is available, a piece of paraspinous fascia may be used as a patch graft to complete the repair. Dura substitutes such as bovine pericardium may also be used for a patch graft if insufficient fascia is available.



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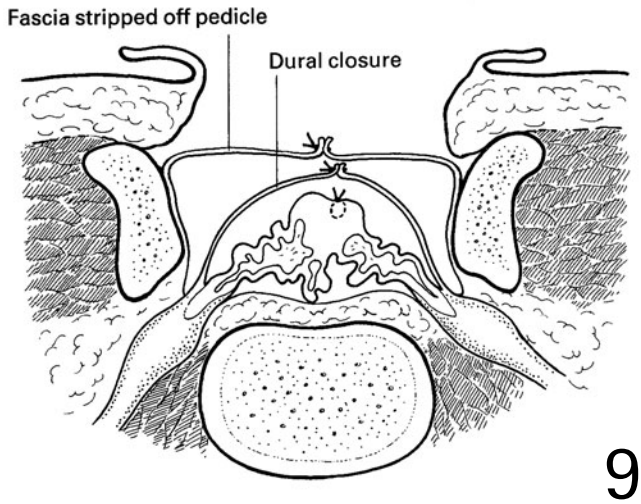
Closing the fascia

9 Where possible, the fascia should be closed over the dura. However, this is not possible in many areas, due to insufficient fascia. In addition, as the pedicles of the spine are widely bifid, the fascia usually ends up lateral to the dura. Splitting of the fascia into superficial and deep layers laterally can provide a sufficient amount of fascia to allow closure over the midline. Here again, closure of the fascia should not cause strangulation of the placode. A complete fascial closure should be attempted, but again, only if it is not likely to cause injury to the placode itself.



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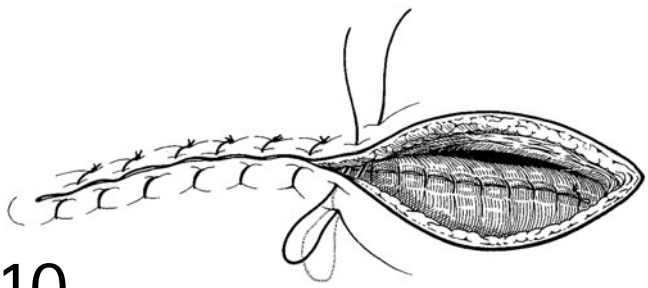
Skin closure

Closure of the skin defect is among the most difficult elements of a myelomeningocele. Serious consideration must be given to employing the talents of a pediatric plastic surgeon when dealing with large defects. Important factors that contribute to the breakdown of the skin repair are the use of poor-quality skin, placing the skin under undue tension at the suture line, and inadequate dural closure or hydrocephalus resulting in leakage of CSF.



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10 All of the skin surrounding the area should be employed, and parchment skin should not be used as it will break down. If the defect in the skin is less than half the width of the back, primary closure can be achieved by carefully undermining the skin. The direction of the closure is not as important as avoidance of pressure and stretching of the skin elements. Undermining the skin edges using blunt finger dissection is useful. Division of the tight fibrous bands tethering the subcutaneous tissue and skin near the iliac crest can be particularly helpful in mobilizing the skin of the lower lumbosacral area.

Occasionally, especially if there is a significant kyphotic deformity, resecting a portion of the pedicles associated with the deformity can be helpful in approximating the skin edges. In more severe cases, kyphectomy may have to be performed to achieve skin closure. This can be done in collaboration with an orthopedic surgeon.

The skin should be closed in two layers, making use of the residual dural cuff that is still attached to the skin edges. Interrupted 3/0 absorbable suture works well in the subcutaneous tissues. Vertical mattress sutures or running baseball-type nylon stitches may also be used on the skin edges. Intravenous fluorescein can demonstrate the blood supply to the skin and may be helpful in identifying any non-viable areas.

Adequate closure may not be possible in a large defect, and the various rotational flaps may be considered. In addition, relaxing incisions may allow closure of the undermined skin.

POSTOPERATIVE CARE

The mortality rates for repair of myelomeningoceles range from 2 percent to 19 percent, but postoperative deaths are exceedingly rare. The early postoperative death rate is about 2 percent and is associated with respiratory failure or severe infection such as meningitis.

The repair site is covered with a light, non-compressive dressing. Plastic drapes should be used to prevent fecal matter

from contaminating the wound. Maintenance of a clear dressing is crucial. The child should be kept off the repair site and can be nursed in a lateral position. The use of a Foley catheter or intermittent catheterization is necessary to prevent stasis and avoid urinary tract infections. Because neonatal ureteric peristalsis may be weak, hydronephrosis may develop, and therefore should be looked for. Ultrasonic examination of the kidneys can be helpful in identifying such problems.

Wound care must be meticulous and the wound must be inspected regularly for areas of breakdown. Bacitracin or Silvadene ointment may be useful in keeping the wound moist and clean. Small areas of wound breakdown will usually respond to local wound management and will eventually granulate in. Wound care is particularly important when lateral releasing incisions have been employed. In most cases, however, rotational flaps created with the assistance of the plastic surgeon should avoid the need for such relaxing incisions.

Leakage of CSF, if present, is usually the result of progressive hydrocephalus. Such diversionary procedures are often necessary within the first days to weeks of life. In those children in whom shunting or CSF diversionary procedures have not been performed, follow-up is mandatory.

Repeated cranial ultrasounds and careful head measurements can identify such progressive hydrocephalus. A ventriculoperitoneal shunt is most commonly employed to relieve such hydrocephalus, but endoscopic third ventriculostomy has also now become more widely used. Recent studies do not suggest any increased risk of infection if the shunt or diversionary procedure is performed at the same time as the repair. However, children with myelomeningoceles are still at increased risk of shunt infections, and physicians must be on the alert for these in the first few weeks after repair.

Occasionally, the Chiari II malformation may become symptomatic during the first few months of life. The Chiari II malformation is characterized by downward displacement of the vermis into the cervical canal, causing compression of the underlying brainstem and spinal cord. It is part of a complex of various congenital anomalies of the brain that can occur in children with myelomeningocele. Symptoms include apnea, stridor, high-pitched cry, and lower cranial nerve paresis. Initial evaluation must be directed toward evaluation of the shunt and treatment of the hydrocephalus. However, if such evaluation proves negative, consideration must be given to early decompression of the Chiari II malformation. It is important to remember that despite adequate decompression of the Chiari II malformation and adequate CSF diversion, a small percentage of children still have intrinsic developmental anomalies of the brainstem that result in brainstem dysfunction necessitating tracheostomy and insertion of a feeding tube.

OUTCOME

Counseling and education of the parents and caregivers of children with myelomeningocele must be considered a vital part of the postoperative care. Multidisciplinary management of these children permits most to lead productive and fulfilling lives. As the technical ability to treat these children and the radiographic capacity to image their associated abnormalities both in utero and postpartum are improved, and, most importantly, as we learn from long-term follow-up with these children, it is clear that the outcome for most is quite good.

Survival for children with myelomeningocele is improving. Most tertiary care centers find that 98 percent of these children survive with aggressive medical management. Associated abnormalities of the central nervous system, particularly complications related to the Chiari II malformation and other systemic anomalies, account for mortality in many of the present-day non-survivors. Shunt failure or acute hydrocephalus also can result in significant morbidity and mortality in these children. Antibiotic therapy has greatly decreased the mortality associated with ventriculitis. Ventriculoperitoneal shunting has successfully managed hydrocephalus, but such shunts can have variable failure, which must be quickly and appropriately diagnosed. McLone has shown that intellectual function is significantly lower in children in whom meningitis has developed in the postoperative period. Some form of learning disability will be present in 70–80 percent of cases, and special education or special programs will be required while in school. Routine evaluation with ultrasound, MRI scan, or CT scans of the head in the first few years of life can be helpful in identifying indolent shunt malfunction. Excessive CT scanning should be avoided, as even the low doses of radiation associated with CT scans may have long-term cognitive consequences for these children. As the children grow older, school performance can be used to follow intellectual function.

Some 80–90 percent of children with myelomeningocele have neurogenic bladder dysfunction. Among the lifelong risks associated with this dysfunction are urinary stasis and infection, trabeculation and diverticula of the bladder, ureteric reflux, hydronephrosis, and renal failure. The use of intermittent catheterization has reduced the incidence of hydronephrosis and urinary tract infections. It has also improved continence, such that 90 percent of affected children can now achieve continence with regular catheterization.

Improvement in urologic management means that urine diversionary procedures are now infrequently employed. Control of defecation can be achieved in 50–75 percent of patients with the assistance of careful dietary management, the use of dietary supplements, and the occasional use of suppositories and enemas.

Multiple orthopedic problems can occur. Scoliosis occurs in 65–75 percent of patients and may require surgical correction. Significant kyphoscoliosis is seen in 5–10 percent of patients and may require surgical correction if respiratory function is impeded. Many lower extremity deformities can occur and often require surgical correction or bracing. The use of orthotic devices in these children requires the specialized attention of a pediatric orthotist familiar with children

who have significant sensory deficits, and therefore may have a greatly increased chance of skin breakdown. Functional outcome can be significantly improved in children for whom multidisciplinary care has been achieved, and a significant number of children may, in fact, become ambulatory.

Careful neurologic monitoring is necessary. Delayed neurologic complications in these children can occur as a result of several problems. Indolent shunt malfunction must always be considered when there is clinical deterioration of any type in a child with myelomeningocele. Chiari II malformation may cause bulbar compression and result in lower cranial nerve dysfunction. It may also result in syringomyelia or cervical cord dysfunction, which may become manifest as upper extremity weakness or numbness. Magnetic resonance imaging has greatly improved our understanding of the Chiari II malformation by allowing us an excellent view of the malformation and the subsequent problems that it may cause. It should be noted that many children with myelomeningocele have significant syringomyelia, which is of no clinical significance, and therefore does not require intervention. However, it must be monitored carefully, as ascending syringomyelia may cause cord dysfunction, and therefore upper extremity dysfunction. Particularly for non-ambulators who are restricted to a wheelchair, loss of upper extremity function can be devastating.

Changes in segmental motor or sensory deficits, evidence of increased spasticity, change in bowel or bladder function, progressive neuromusculoskeletal deformity at the ankle or leg, and progressive scoliosis may occur secondary to tethering of the spinal cord. Because all children with myelomeningocele have evidence of a tethered cord on MRI, careful clinical evaluation must be employed to determine if such tethering is clinically significant and warrants further surgery to untether the spinal cord. It must be remembered that all children with myelomeningocele will have radiographic evidence of a low-lying cord, and this does not equate with clinical dysfunction. In addition, less common causes of progressive deficit may include dermoid cyst formation, diastematomyelia with septae, and arachnoid cyst formation.

It cannot be overemphasized that shunt malfunction is the most common cause of clinical deterioration in children with myelomeningocele. It must always be considered, and shunt function must be carefully evaluated before such deterioration is attributed to other causes, such as Chiari II malformation or tethered cord.

IN UTERO REPAIR OF MYELOMENINGOCELE/ FETAL SURGERY

Because of the improved nature of in utero ultrasound screening, amniocentesis, and the emergence of prenatal MRI scanning, early detection of neural tube defects is increasing. The development of surgical techniques applicable to the fetus and the emergence of drugs for the prevention of premature labor have, over the past 10 years, provided for in utero correction of myelomeningocele. With this particular technique, the concern must largely lie with the safety of the mother, but clearly a significant secondary goal is the safety of and outcome for the fetus. Avoidance of preterm labor is crucial to this outcome. Because of the technical difficulty and fragility of the tissues, the surgery is generally limited to 18 weeks of gestation, and because premature labor increases dramatically after 30 weeks, it is usually not recommend after 30 weeks. In this procedure, the uterus is exposed through a low transverse abdominal incision and the fetus and placenta are positioned in such a way as to provide an optimal view of the myelomeningocele placode. Hysterotomy is performed and the fetus is exposed in as limited a fashion as possible. The goal is to maintain intrauterine volume to prevent placental separation, contraction, and expulsion of the fetus. This is generally accomplished by high-volume profusion of the amniotic cavity with warm Ringer's lactate. As little of the fetus as possible is exposed to accomplish the necessary surgery.

The myelomeningocele closure is really quite similar to that in the already born fetus. The fringe of the full thickness at the region of the zona epitheliosa is incised and a dural as well as fascial closure is performed. Skin closure can be accomplished using absorbable suture. In cases where the defect may be too large to be closed, an acellular human dermal graft can be used to complete the closure, in which case, a secondary closure may be necessary after delivery.

The results of in utero myelomeningocele closure are still being interpreted. There appears to be a decreased incidence of hindbrain herniation as demonstrated by serial MRI scan. It also appears that there may be some slight decrease in the requirement for shunting. There does not appear to be an improvement in neurologic or lower extremity function. Hindbrain herniation was present in 95 percent of patients repaired postnatally. After in utero repair, only 38 percent demonstrated cerebellar herniation. Review of the data from the Children's Hospital in Philadelphia suggests that the overall ventricular shunting rate was 81 percent in children who had their myelomeningoceles postnatally repaired. When these results were compared with a combined series from the Children's Hospital of Philadelphia and Vanderbilt, looking at shunting rates for children who had had in utero repair of their myelomeningoceles, the overall ventricular shunting rate was found to be 44 percent.

The optimal timing for the repair is still also debated. The infants at the Children's Hospital of Philadelphia had their repairs at a slightly earlier gestational age than those at Vanderbilt. Interestingly, the patients who were operated on at less than 25 weeks' gestation had what appeared to be, better lower extremity function than would be expected for the level of their myelomeningocele, whereas those who were repaired later, as was the case at Vanderbilt, had no improvement in their lower extremity function.

These preliminary institutional studies suggest that prenatal surgery for myelomeningocele may have certain clear

advantages. There seems to be a reversal or improvement of hindbrain herniation, a decrease in the need for shunting, and at least preservation if not improvement of lower extremity function, which is somewhat dependent on the gestational age at the time of repair. There is clear evidence of selection bias in each of these studies, and obviously significant risks to both the mother and the fetus with in utero surgery. It is because of this that a clinical trial has now been started, sponsored by the National Institutes of Health and involving three centers in the USA: the Children's Hospital of Philadelphia, Vanderbilt University, and the University of California in San Francisco. Further information regarding the outcome of these trials should be available within the next 1 to 2 years.

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Ventricular shunting procedure

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HISTORY

It is not until the sixteenth century that we have the first references to the condition that we would recognize as hydrocephalus (Versalius, 1514–1564). Robert Wytt's essay on dropsy of the ventricles of the brain (1768) provides a clear clinical description of the condition.

The surgical treatment of hydrocephalus has included ventricular puncture, extirpation of the choroid plexus, and cerebrospinal fluid (CSF) diversion to a variety of body cavities. Drainage of the CSF to the jugular vein via a simple valve housed in rubber tubing was described by Nulsen and Spitz (1952) and was a major landmark in hydrocephalus treatment, but it was the introduction of valved tubing made from durable, biocompatible material (Silastic®) that heralded the modern era of shunt technology and hydrocephalus treatment.

Endoscopic third ventriculostomy entails perforating the floor of the third ventricle via an endoscope introduced via the lateral ventricle and is now an established technique in the neurosurgical treatment of hydrocephalus.

PRINCIPLES AND JUSTIFICATION

Hydrocephalus is the accumulation of CSF that results when there is obstruction to its normal circulation and absorption. As a consequence, the cerebral ventricles enlarge, intracranial pressure increases, and cerebral function may be impaired. There are numerous etiologies, both congenital and acquired, but the principles of treatment remain the same – namely, to divert CSF from proximal to the point of obstruction to some distal site where absorption can take place.

PREOPERATIVE ASSESSMENT AND PREPARATION

Diagnosis

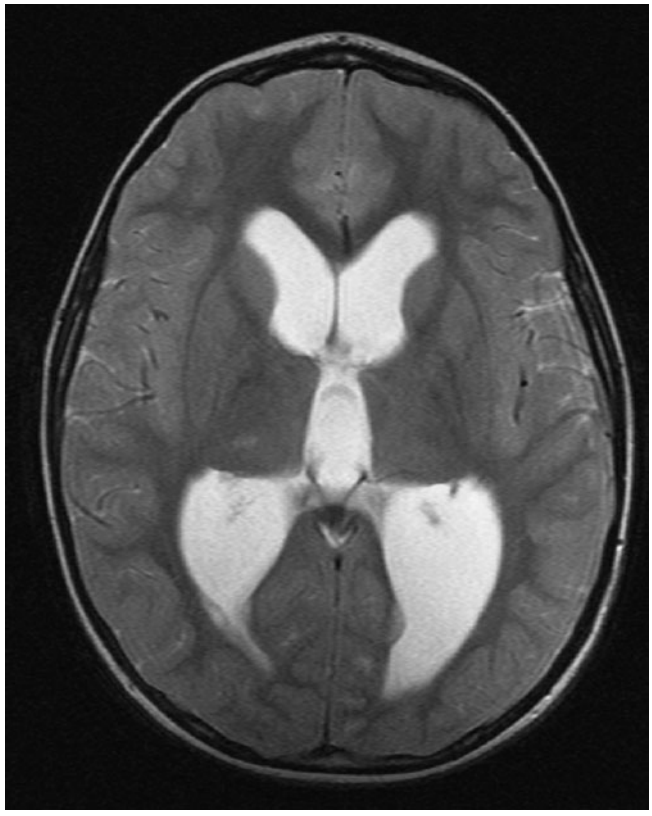
It is mandatory that the results of neuroimaging be interpreted in the light of clinical features before the diagnosis of hydrocephalus can be made and the decision to place a shunt taken. Dilatation of the ventricular system that is static and not associated with raised intracranial pressure may occur in a number of settings, and intervention is not indicated (e.g., parenchymal brain damage, central nervous system (CNS) malformations, post-irradiation, arrested hydrocephalus). Indeed, shunt placement in such circumstances may be harmful and result in symptoms of over-drainage and subdural hematoma formation.

The classical symptoms of raised intracranial pressure, namely headache, vomiting, and drowsiness are often not present, particularly in the neonate or infant, in whom head circumference, anterior fontanelle tension, and general neurodevelopmental progress may be more useful indices of progressive hydrocephalus. Provided there is no overt clinical urgency, a period of observation and sequential imaging may help distinguish between active hydrocephalus and simple ventriculomegaly.

Serial head circumference measurements accurately measured and plotted on a head circumference chart are an essential adjunct to the brain imaging in assessing the necessity for shunt placement in a neonate with post-hemorrhagic hydrocephalus or following closure of a myelomeningocele.

Radiological features

1 Axial imaging, either computed tomography (CT) scan or magnetic resonance imaging (MRI), should be performed prior to shunt placement. This provides clear visualization of the ventricular anatomy, may indicate the underlying etiology, and can be used to plan shunt placement. Ultrasonography is a useful modality with which to monitor ventricular size, but is restricted to infants with a sufficiently patent fontanelle and may not allow adequate visualization of the entire intracranial contents.



1

Once the indication for treatment has been confirmed, the surgeon must decide, first, what type of CSF diversion procedure to perform, and second, what type of device to use.

Surgical treatments for hydrocephalus

VENTRICULOPERITONEAL SHUNT

Drainage is directly to the peritoneal cavity. This remains the commonest technique. In most instances it is quick, effective, and relatively simple to perform.

VENTRICULOATRIAL SHUNT

The distal tubing is placed in the right atrium via a major neck vein. This is a technically more demanding procedure (particularly at revision) and there are well-recognized long-term complications. This technique has therefore become very much second- or third-line treatment.

VENTRICULOPLEURAL SHUNT

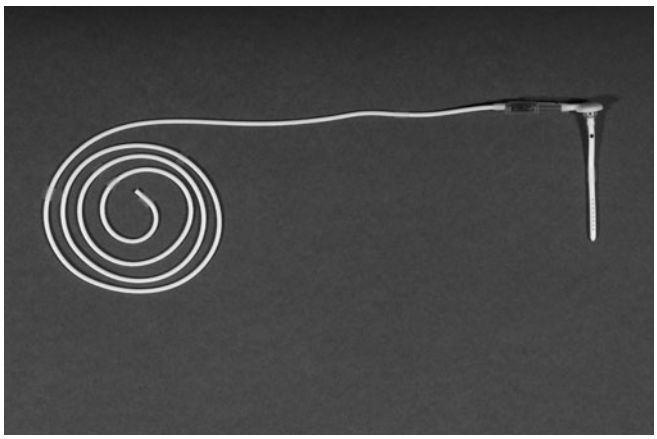
The pleural cavity provides an effective absorptive surface; however, the risks of symptomatic pleural effusion and a higher rate of blockage mean that it is not recommended as a primary treatment.

ENDOSCOPIC THIRD VENTRICULOSTOMY

This is only appropriate for cases of obstructive hydrocephalus, e.g., aqueduct stenosis, or posterior fossa tumor, where the subarachnoid spaces are patent and normal CSF absorption is possible. An endoscope is placed into the lateral ventricle and an opening is made in the floor of the third ventricle to connect the ventricular compartment with the basal cisterns. This is a neurosurgical procedure and is not discussed further here.

Choice of shunt device

2 There are numerous shunt devices on the market; none has any proven advantage. It is strongly recommended that the surgeon becomes familiar with a particular shunt type for regular use, reserving more novel devices (such as programable shunts) for exceptional circumstances. Shunts comprise three components: a ventricular catheter, a valve, and a distal catheter. A reservoir chamber is often included and positioned at the burr hole site or integral to the valve. The reservoir can be punctured percutaneously to obtain CSF in cases of suspected infection or blockage.



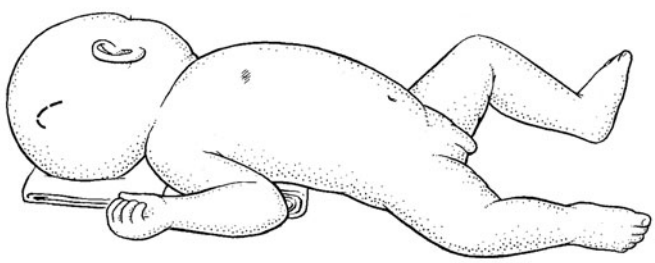
2

Contraindications to surgery

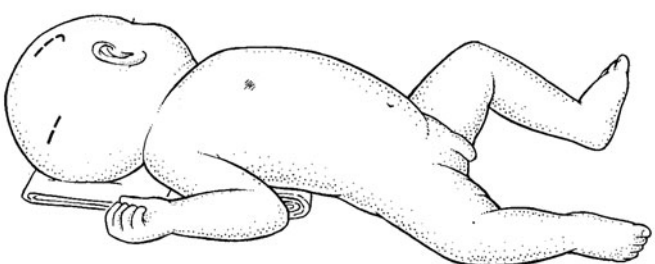
Intercurrent sepsis, particularly CNS infection, should be regarded as a contraindication to surgery. Blood or high levels of protein in the CSF should be considered relative contraindications to shunt placement, as these may increase the likelihood of early shunt failure due to blockage.

Anesthesia

The procedure is performed using a balanced anesthetic technique requiring endotracheal intubation and intermittent positive pressure ventilation (IPPV). Particular attention is paid to the control of P_{CO_2} in view of the presence of raised intracranial pressure (ICP). The proposed shunt position should be discussed with the anesthetist to ensure that vascular access (e.g., intravenous neck lines) and endotracheal tube strapping do not obscure the shunt trajectory. Prophylactic antibiotics are administered at the time of induction of anesthesia.



3a



3b

OPERATION

Positioning and draping

3a,b The importance of positioning the child for ventriculoperitoneal shunt cannot be overemphasized. The head needs to be rotated and the neck extended by placing a sandbag beneath the shoulders to ensure that there is a level trajectory for subcutaneous tunneling between the cranial and abdominal incisions. This position opens out the neck skin creases and reduces the risk of 'buttonholing' the skin during tunneling. Skin preparation should cover the entire area from cranial to abdominal incision; the preparation fluid should be allowed to dry before drapes are applied. An adhesive plastic drape is placed to cover the entire operative field.

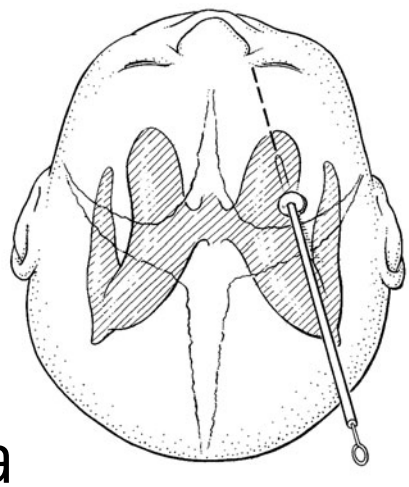
The shunt should be assembled at the beginning of the procedure. This reduces both operating time and the tendency for clumsy manipulations of the shunt in situ. Methods of assembly and testing of the shunt vary for different types of shunt and therefore the manufacturer's recommendations should be followed.

The required ventricular catheter length can be estimated from imaging – the tip of the catheter should sit well into the body of the ventricle.

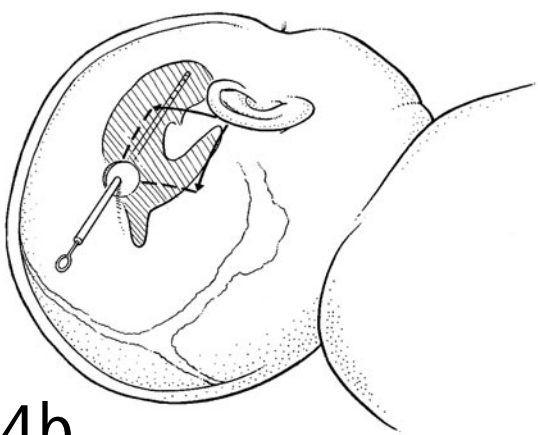
Incisions

Shunt infection is one of the major complications of shunt surgery. Commensal skin organisms are the most common pathogens. The following precautions are recommended to reduce the risk of contamination.

- The cranial incision is semicircular and large enough to ensure that the wound does not overlie the shunt tubing.
- Instruments used in skin opening should be put aside until the time of closure.
- Tissue handling should be kept to a minimum and the wound edges lined with betadine-soaked cottonoid strips.
- The use of the diathermy needs to be kept to a minimum. In the infant, it is very easy to inflict burns at the skin edges.
- A no-touch technique should be maintained throughout the procedure.



4a



4b

4a,b The cranial incision may be frontal or parieto-occipital; there is no clear advantage of one over the other. A frontal shunt will require an additional incision behind the ear to facilitate tunneling to the abdomen. The burr hole for a frontal shunt should be just anterior to the coronal suture and in the line of the pupil. A parieto-occipital burr hole is made approximately 3 cm above and behind the top of the pinna.

The burr hole is made in the standard manner, using power drill or perforator; care is required in the infant due to the thin calvarium. Once the bone is breeched, the burr hole can be enlarged using bone rongeurs. The dural opening should be small, sufficient only to pass the ventricular catheter. Larger openings increase the risk of subcutaneous CSF collections.

Ventricles may be asymmetric and vary considerably in size, particularly in children; the precise site of the burr hole will therefore often be dictated by the underlying ventricular configuration.

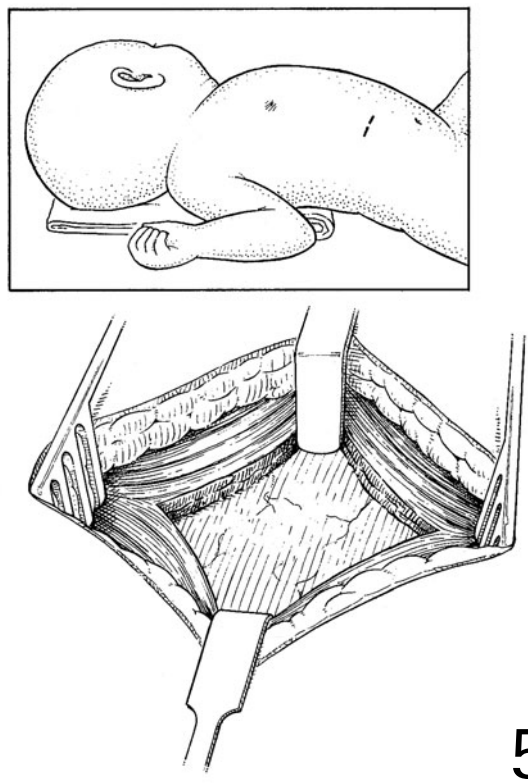
Subcutaneous tunneling

The subcutaneous tunneling device may be passed in either direction. The tunneling is performed deep to the subcutaneous fat but superficial to the deep fascia. Care should be taken to avoid perforating the skin; with one hand holding the device, the other hand can be used to palpate the course of the tunneling device as it advances to the abdomen. Care should be taken to avoid damaging the breast bud in female patients.

Once in position, the shunt tubing can be threaded down the tunneling device, which is then removed.

Peritoneal exposure

5 The peritoneal end of the shunt is usually placed via a minilaparotomy below the costal margin over the rectus abdominis. The rectus sheath is opened, and a longitudinal muscle-splitting technique is used to expose the peritoneum. The peritoneum is then opened; it is important to be quite certain that the peritoneal cavity has been entered to avoid extraperitoneal placement. It is also important to be sure that there is a good flow of CSF from the distal catheter before this is internalized. The entire distal tubing is then fed under direct vision into the peritoneal cavity. A good length of distal tubing reduces the likelihood of the tubing migrating out of the peritoneal cavity as the child grows.



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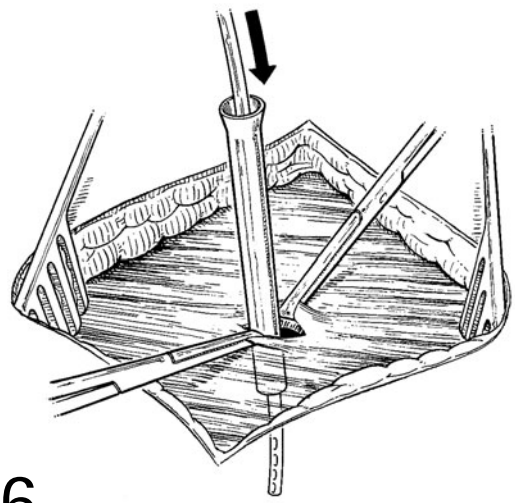


6

6 An alternative method of inserting the peritoneal catheter is by means of a trocar. This is quicker and requires a much smaller incision, but damage to abdominal contents is a reported complication of this technique.

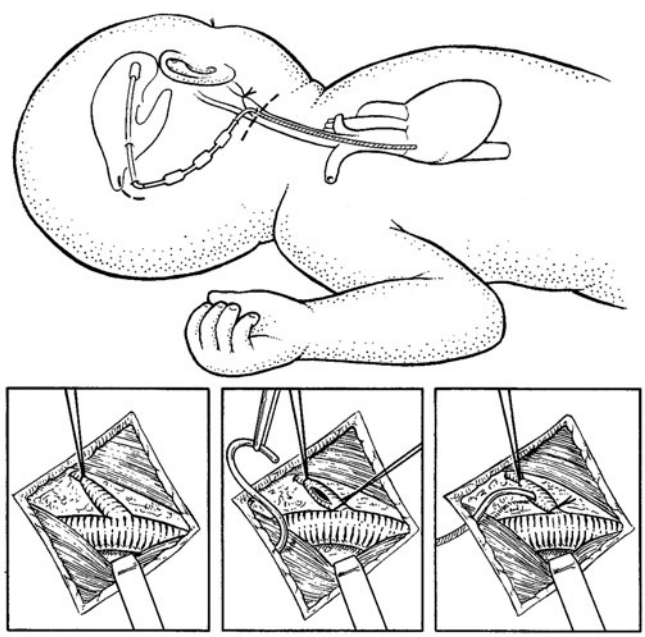
Catheter placement

The ventricular catheter mounted on a stilette is then placed through the dural opening and slowly advanced into the lateral ventricle. As soon as the ventricle is entered, the stilette is stabilized and the catheter is advanced into position. Particularly when the ventricles are small or very asymmetric, it is useful to have rehearsed the catheter placement, predetermining the desired trajectory before perforating the brain substance. For a standard frontal approach, the catheter is passed perpendicular to the skull surface, aiming toward the medial canthus of the ipsilateral eye. In a posterior approach, the catheter is aimed at the midpoint of the forehead. Brisk flow of CSF along the catheter needs to be confirmed before proceeding. If there is no CSF flow or flow is sluggish, the catheter must be re-sited.

If the ventricular catheter is not already attached to the rest of the shunt, this is now done. The distal tubing, now draining CSF, is placed in the peritoneal cavity. The catheter should thread easily. The entire length of distal tubing is placed to allow for subsequent growth of the child.

Wound closure

Meticulous attention must be paid to wound closure. The wounds are closed in layers; an absorbable suture is used to close the skin. Wound infection or CSF leakage invariably results in infection of the shunt.



POSTOPERATIVE CARE

The wound dressings are left undisturbed and the child nursed off the wounds. In a small infant with large ventricles, it is wise to elevate the child slowly over 24–48 hours in an attempt to avoid too rapid decompression of the ventricular system.

ADDITIONAL SHUNT PROCEDURES

Occasionally, drainage into the peritoneum is precluded, most commonly because of adhesions or repeated lower end failure. In this situation, an alternative site for distal drainage must be sought. In each case, the details relating to proximal placement are as described above.

Ventriculopleural shunt

It is important that the upper end of the shunt procedure has been completed and that CSF flow from the distal catheter has been established before the pleura is opened. A bulldog clip is placed on the tubing whilst the distal site is prepared. The lower incision is made at the level of the fifth rib in the anterior axillary line. The intercostal muscles are split just above the rib to prevent injury to the neurovascular bundle. The pleura is exposed and incised; the distal shunt, having been cut to an appropriate length, is then gently threaded into the pleural cavity, avoiding direct trauma to the lung. The anesthetist is requested to induce Valsalva's manuever in an attempt to reduce the risk of pneumothorax. The muscle layer is then closed around the tubing.

Ventriculoatrial shunt

Again, it is important to have completed the upper end of the shunt insertion before the venous system is opened. The neck incision is sited over the anterior border of the sternomastoid muscle. The carotid sheath and internal jugular vein lie just deep to this muscle. It is generally advised that the common facial vein be exposed, mobilized, and divided. The tubing is passed along this tributary into the internal jugular vein and thence into the right atrium. In small children, it is often necessary to insert the tubing directly in the jugular vein. A purse-string suture is placed in the vein wall before the venotomy; the purse string is then tied to seal the opening around the shunt tubing. The tip of the catheter must be in the right atrium and this is confirmed with fluoroscopy. The tubing is then connected to the rest of the shunt by means of a straight connector.

A simpler and more elegant method of placement of the atrial catheter is by a percutaneous technique using ultrasound guidance (see Chapter 3, 'Vascular access').

OUTCOME

Shunt placement results in prompt and sustained control of intracranial pressure in the majority of cases. The two most common shunt complications are infection and blockage.

Shunt infection

Shunt infection occurs in up to 8 percent of cases. Seventy percent of shunt infections will have declared themselves within 2 months of shunt surgery. A raised temperature, irritability, abdominal pain, and any evidence of a wound problem should raise the possibility of a shunt infection. Cerebrospinal fluid needs to be obtained and can be sampled from the reservoir. If infection is confirmed, the shunt should be removed and replaced with an external ventricular drain whilst the infection is treated.

Shunt blockage

Approximately half of the shunts inserted will have blocked on at least one occasion in the 10 years following insertion. Shunt blockage is a surgical emergency. The following points should be borne in mind in planning and performing shunt revision surgery:

- Aspiration of CSF from the shunt reservoir via a butterfly needle may be a useful temporizing measure in an obtunded child with a blocked shunt. Clearly this will not be possible if the proximal catheter is the site of the blockage.
- An apparently normal or small ventricular system on CT scan does not exclude shunt malfunction.
- At surgery, the child is prepared as for a shunt insertion; access to the entire shunt must be possible.
- If in any doubt, the entire shunt should be replaced.
- Exploration of the proximal shunt is performed first. The ventricular catheter is disconnected to see whether there is proximal CSF flow. A saline-primed manometer is connected to the distal system to assess distal run-off.
- Extreme caution should be exercised when removing blocked ventricular catheters. Choroid plexus may be stuck to the shunt and is easily avulsed, resulting in intraventricular hemorrhage. If the catheter cannot be removed easily, a stilette passed down the catheter can be diathermized using monopolar diathermy. This will often shrink away the choroid plexus and make removal easier.
- If bleeding is encountered, the CSF should be allowed to drain freely from the end of the catheter until it clears. If it remains bloodstained, it is safer to leave an external ventricular drain in position for a day or two until the CSF returns to normal.

FURTHER READING

Drake JM, Sainte-Rose C. *The Shunt Book.* Oxford: Blackwell Science, 1994.

Thompson DNP. Hydrocephalus and shunts. In: Moore A, Newell D (eds), *Springer Specialist Surgical Series.* New York: Springer Verlag, 2004.

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SECTION VIII

Trauma

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Surgical treatment of thermal injuries

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HISTORY

Burns are among the most serious of injuries, and although there are still many problems associated with their treatment, substantial progress has been made since Neanderthal man treated burns with plant extract or ancient Romans used a mixture of bran and honey, cork and ashes or vinegar. The modern treatment of burns is a logical exercise in resuscitation, infection control, surgical wound care, pain control, nutrition, and psychological and physical rehabilitation.

The Indian Tilemaker caste is credited with the earliest recorded method of skin grafting, and Reverdin, a Swiss surgeon, performed the first epidermal graft in 1869. Thiersch, in 1874, used more extensive pieces of skin. In Glasgow, Scotland, in 1875, Wolfe utilized a free full-thickness graft to repair a defect on the lower eyelid, and Lustgarten described the technique of early excision and grafting of small burns in 1891. Although allografts have also been used for centuries, Brown of St Louis established the practical aspects of biological skin dressings as life-saving procedures in extensively burnt patients.

In modern times, a combination of early eschar excision and autografting or allografting (cadaver skin), biological and synthetic skin substitutes have substantially transformed burn care management, with a 50 percent predicted survival for a burn of 70 percent total body surface area (TBSA).

PRINCIPLES AND JUSTIFICATION

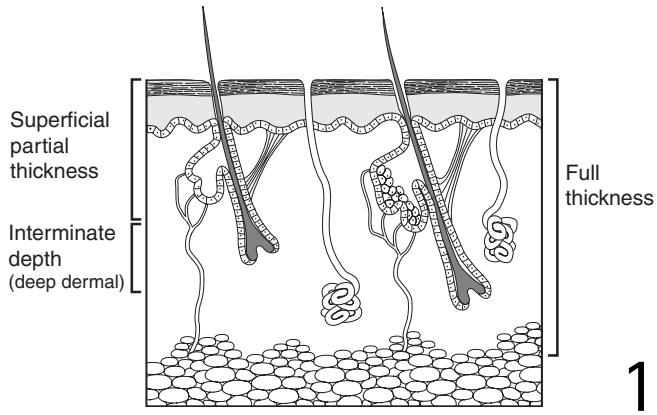
- Superficial thermal injuries that will heal spontaneously within 3 weeks can be clinically identified and are best treated conservatively.
- Hot-water scald burns in children are best left for 2 weeks to assess the need for operative intervention, thereby reducing the area for excision by 66 percent.
- Early excision and grafting should be considered the treatment of choice for all deep burns. Inadequate excision leads to skin graft loss. Exceptions are burns on the face, ears, soles of the feet and genitalia.
- Surgery is an elective procedure in a stable patient.
- The amount of skin to be excised depends on the status of the patient, the burn size, the availability of autografts or allografts or skin substitutes, and the volume of blood loss.
- Once the burn wound has been excised, immediate wound closure with autografts, allografts, or biological alternatives is required.
- Surgical excision may safely be undertaken in the presence of inhalational injury, although operative time and blood loss must be kept to a minimum.
- Surgery should be delayed for burns that are more than 24 hours old on admission. These wounds may be infected and should be swabbed or a quantitative biopsy performed (one biopsy per 24 cm^2 of burnt tissue). Debridement should be carried out, topical therapy applied, and surgical excision postponed for 24 hours or longer.
- Beta-hemolytic *Streptococcus* infection is a contraindication to surgical excision and grafting until the infection has been eradicated.
- Non-life-threatening burns in patients with severe concomitant diseases or injuries should only be excised when the patients are stable and the life-threatening processes have been controlled.

BURN SEVERITY

The severity of a burn is determined by its depth and size, the anatomic site, concomitant disease or injury, and the age and physiologic status of the patient. Skin does not reach adult thickness until puberty; hence, the younger the child, the deeper the burn. Likewise, patients over the age of 60 years develop significant dermal atrophy with involution of skin appendages, resulting in deeper burns and slow donor site healing.

Burn depth

1 No objective clinical methods are available to determine the depth of thermal injury and no standardized method has been adopted. Most burns are a combination of superficial and deeper burns, and the best assessment can be made 2–3 days after the injury when wound evolution has been completed.



1

SUPERFICIAL PARTIAL THICKNESS: DESTRUCTION OF ONLY SUPERFICIAL LAYERS OF THE SKIN

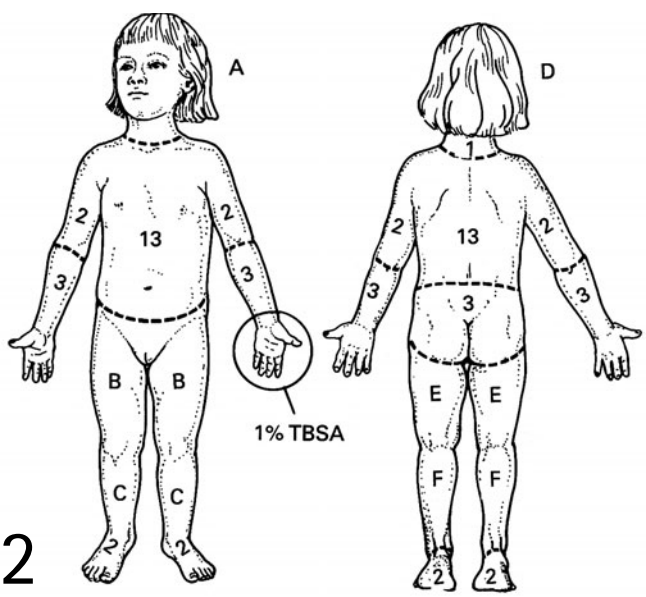
These wounds will epithelialize spontaneously within 3 weeks; excision is contraindicated and the wounds rarely cause functional or cosmetic defects or hypertrophic scars. They may never completely match the color of the unburned surrounding skin. These wounds characteristically have an erythematous, moist, homogeneous surface with blister formation, are painful and hypersensitive to touch, blanch readily, and have a normal to firm texture on palpation.

INDETERMINATE DEPTH (DEEP DERMAL BURN): DESTRUCTION OF EPIDERMIS AND VARYING AMOUNTS OF DERMIS

These wounds are difficult to assess during the first 3 days after injury due to ongoing evolution within the burn wound, which can be modulated by infection and dehydration. These wounds present with a reticulated red and/or white, dry surface and may blister. Capillary circulation may be sluggish or absent when pressure is applied to the wound. Wound pain is perceived as discomfort and the wound is often less sensitive to pinprick than the surrounding normal skin. The burn is depressed in comparison with the surrounding skin. The healing time for these wounds may be variable, but is usually longer than 3 weeks.

UNEQUIVOCALLY FULL THICKNESS: TOTAL IRREVERSIBLE DESTRUCTION OF ALL ELEMENTS OF THE SKIN WITH OR WITHOUT EXTENSION INTO THE DEEPER TISSUES AND STRUCTURES

These wounds will not heal spontaneously within 3 weeks and have unsatisfactory functional and cosmetic results. There is general consensus that the best treatment entails early eschar excision and immediate grafting. These wounds may mimic the appearance of an indeterminate burn and are usually mottled, white and/or red, or charred and dry in appearance, insensitive to pain, and leathery to palpation. Blisters are unusual and if present are thin walled and do not enlarge. Clotted superficial vessels may be visible. The surface of the burn is usually depressed relative to adjacent unburned skin and the appearance of the burn remains static, with little change over the ensuing days.



2

Estimating the extent of the burn

2 The extent of the burn is expressed as a percentage of the TBSA involved. Two methods of assessment are used. The palmar surface of the open hand of a patient amounts to approximately 1 percent of the TBSA and is used to estimate the size of small burns. Different age-related values are used to calculate the percentage area burned in larger burns, because body proportions change with age. The 'pediatric rule of nine' can be modified for different ages.

MANAGEMENT

Emergency management

- At the scene of the injury, smoldering or hot clothing should be removed immediately and adequacy and patency of the airway ensured.
- Hydrotherapy: small burns (<25 percent TBSA) are immersed in cold water (15–18°C) or covered with cold, wet compresses for at least 30 minutes to reduce the depth of injury and relieve pain and discomfort. Larger burns are usually excluded from hydrotherapy, as it may result in hypothermia, and should be covered with clean sheets or blankets.
- No oily substances should be used topically.
- Copious irrigation of the wound with water is indicated for chemical burns; neutralizing agents should not be used.
- In electrical burns, the patient's circulation and ventilation must be evaluated and maintained.
- For suspected inhalation injury, 100 percent oxygen is given by facemask. Progressive airway obstruction may occur over the first hours post-burn. It is safest to intubate early, before extensive head and neck swelling occurs and airway obstruction becomes imminent.
- Oral fluids should be withheld initially.
- Escharotomy should always be considered if there will be a delay before the patient is transported to a specialist burn care unit.

Definitive treatment

MINOR BURNS

Initial therapy of minor burns should include the administration of analgesics, cleaning the wound with bland soap and water or detergent, removal of topically applied agents, and shaving hair where necessary. Dead tissue should be debrided and any tar removed with soft paraffin in a water base. Topical antibacterial agents and occlusive dressings or an adhesive polyurethane sheet should be used to dress the wound and tetanus toxoid should be administered.

FOLLOW-UP THERAPY

The patient should be encouraged to move the affected area. The wounds are either washed daily and topical antiseptic applied, or they are left undisturbed until healed. Prophylactic antibiotics are not usually necessary.

MAJOR BURNS

Thermal injury invariably leads to changes in capillary function, resulting in obligatory isotonic fluid losses and loss of effective circulating fluid volume. If substantial, hypovolemia and burn shock may develop. Patients with deep burns, inhalational injury, or delayed resuscitation have increased volume requirements for resuscitation. The Parkland resuscitation formula (4 mL/kg per percent surface area burn) is still the gold standard. Crystalloids, in particular Ringers' lactate, are the most commonly used. Resuscitation must be initiated as soon as possible. Urine flow of 0.5–1 mL/kg per hour in children or 30 mL/hour in adults is a good clinical indicator of adequate vital organ perfusion. Colloids are not indicated during the first 18–24 hours. However, should serum albumin levels fall below 1.0 g/dL or below 1.5 g/dL in the presence of pulmonary dysfunction or enteral feeding intolerance, supplementary albumin is administered at 1–2 g/kg per day until serum albumin stabilizes at more than 2.5 g/dL.

Other factors that will require ongoing management include the treatment of inhalational injuries, nutrition, pain control, the prevention and treatment of infection, and physical and psychological rehabilitation.

SURGICAL MANAGEMENT

Intraoperative considerations

Burns represent a unique challenge due to the nature of the operation and the concomitiant physiological responses to injury. Important components of anesthetic management are the maintenance of an adequate airway, especially in the presence of inhalational injury, choice of anesthetic agent, maintenance of hemodynamic stability and temperature, the use of pharmacological agents, the position of the patient during surgery, the site of surgery (eschar and donor area), and continual monitoring.

VASCULAR ACCESS

Adequate and secure vascular access is critical. A large-bore venous cannula is usually placed in the subclavian, internal jugular, or femoral vein or another suitable surface vessel. Additional arterial access is established for major burns and when large blood losses are expected.

TEMPERATURE CONTROL

Hypothermia is a significant problem, and preventable measures include an ambient temperature of 28–32°C, heated anesthetic gases, and intravenous fluids at 37°C. Radiant heaters are used throughout the procedure and the patient lies on a covered warming blanket. Exposed areas may be covered with sterile plastic drapes.

MONITORING

Electrocardiogram electrodes may be difficult to site and any accessible area will suffice as the configuration is irrelevant. The electrodes can be secured with surgical clips. A urinary catheter is inserted. Non-invasive blood pressure monitoring with pulse oximetry, a central temperature probe, and urine output at 1 mL/kg per hour are essential. Postoperatively, the

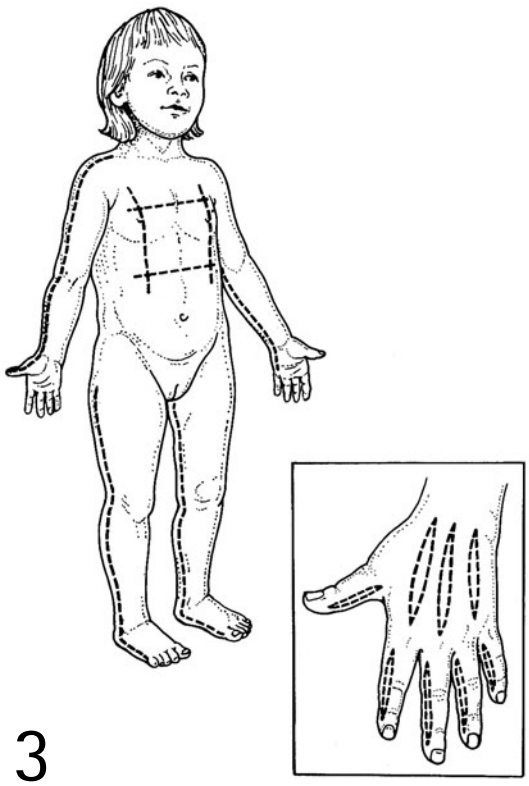
child is kept in the recovery area and constantly monitored until fully awake with satisfactory vital signs.

FLUID REQUIREMENTS

Maintenance fluid is given as isotonic crystalloids at 4 mL/kg per hour and blood loss is replaced volume/volume to maintain the hematocrit above 30 percent. Weighing surgical swabs is a satisfactory measure of blood loss. Blood loss of between 0.4 mL/cm^2 and 0.74 mL/cm^2 of tissue excised can be expected.

PREPARATION FOR SURGERY

The child is thoroughly showered and washed with chlorhexidine or iodine-containing soaps or with 0.025 mmol/L sodium hypochloride solution. Intraoperative antibiotics are given for all excisional procedures when the burn wounds are infected or with large excisions. If indicated, antibiotics should be continued with for 3–5 days post-surgery.



3

Releasing escharotomy (decompression)

3 Releasing escharotomy must be performed where a circumferential deep burn is impeding circulation to more distal parts, especially around the arms or legs or over the chest, where respiration may be impaired.

The releasing escharotomy must traverse the dead tissue as far into the subcutaneous layer as necessary to encounter viable tissue and must extend from normal skin proximally to normal skin distally to prevent deep tissue death. Excessive bleeding can be problematic if the escharotomy is performed incorrectly or too deeply into the adjacent tissue.

PRACTICAL APPROACH TO SURGICAL EXCISION

- Burns < 10 percent TBSA: excision and autografting are performed using meshed 1.5:1 or 2:1 or sheet grafts. Sheet grafts are placed on all vital and visual areas (face, neck, chest, hands) and all small grafted areas.
- Burns 10–30 percent TBSA: excision and autografting.
- Burns 30–40 percent TBSA: sufficient donor sites are usually available to graft the excised bed despite the fact that about 30 percent TBSA is unavailable for donation (face, neck, hands, feet). The grafts should be meshed at 1.5:1 or 2:1, or temporary allografts or a manufactured dermal substitute used if available.
- Burns > 40 percent TBSA: donor sites are limited and it is impossible to cover all the excised wounds with autografts primarily. The preferred method is to perform total or sequential (20 percent TBSA every alternate day) excisions. Skin cover should then be applied (autograft 1.5:1, 2:1, or 3:1, and/or autograft 3:1 with allograft 2:1 overlay, and/or synthetic skin substitutes).

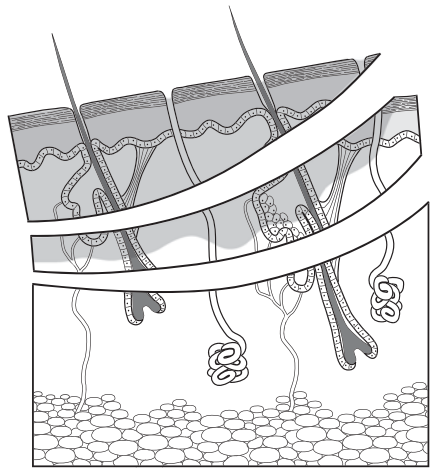
Timing and extent of excision

In general terms, excision is carried out as soon after injury as possible, i.e., as soon as the cardiovascular system is stable and resuscitation is completed, metabolic and physiologic balance is restored, and vital signs, urine output, hematocrit, and albumin levels are satisfactory. This time may vary from a few hours to several days, but a good timing goal is day three. Excision is therefore an elective procedure in a stable patient. The order of priority of areas of major excision (every alternate day) is the posterior trunk, anterior trunk, and clavicular area, one lower extremity, second lower extremity, both upper extremities and hands, and all unhealed areas on the face, neck, and head.

The amount to be excised at each procedure depends on the stability of the patient, the burn size, the availability of autografts and allografts, and the volume of blood loss incurred during the procedure. Most deep burns of less than 40 percent TBSA should be excised and grafted within the first few days, as adequate donor sites are usually available. In large burns the principle is to reduce the burn surface area expeditiously.

METHODS OF REMOVING ESCHAR

Three different methods of removing eschar are used: (1) tangential or sequential excision, (2) fascial excision, and (3) delayed escharectomy.



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Tangential or sequential excision

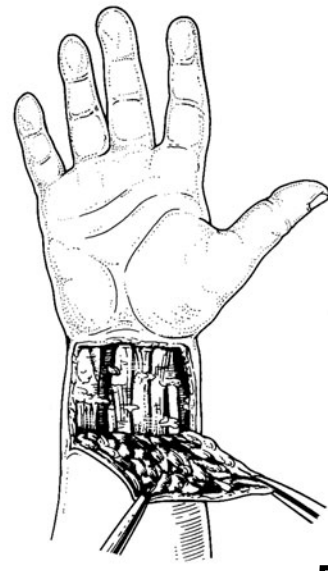
4 This method entails the sequential excision of thin layers of burn eschar until a viable bed is encountered. If done adequately, the minimum amount of living tissue is sacrificed, with satisfactory functional and cosmetic results.

Excision is best performed using a hand-held Humby knife, held at a tangent to the wound surface. Excision over uneven surfaces or bony prominences can be aided by subeschar injection of saline. The appropriate level of excision or end-point is characterized by a shiny white surface with brisk arteriolar or punctuate bleeding or viable yellow, non-hemorrhagic subcutaneous fat globules with briskly bleeding vessels in all areas. Clotted vessels represent nonviable tissue. Dark pink-brown hemorrhagic fat is nonviable and must be removed; residual necrotic areas will jeopardize graft take. Dissection may become difficult once the level of excision has gone beyond the dermis. It may be very important to preserve subdermal fat over bony prominences for esthetic reasons, but grafting onto subdermal fat results in a lower success rate. Blood loss may be substantial and effective control of hemorrhage must be established. It is advisable to limit excision to 25–100 cm^2 to control hemorrhage, and then to proceed with further excision. Tangential excision is best done within the first 1–5 days before hypervascularity and wound infection become established. A maximum of ± 20 percent TBSA should be excised at any one time. Alternatively, extremity excisions should be done under tourniquet; blood transfusions are rarely needed. For stable patients, a hematocrit of 30–36% or Hb 10–12 gm/dl can be accepted.

Once excised to the appropriate level and hemostasis secured, an immediate split-thickness skin graft (autograft) is performed. Sheet grafts are placed on important cosmetic and functional areas. To prevent desiccation of exposed and viable tissue, mesh grafts should not be expanded more than 1.5:1–2:1. If greater expansion is needed, temporary skin substitutes (cadaver or Biobrane) should overlay the autograft. The latter method is also used for all excised and nongrafted areas.

Fascial excision

5 This method is generally reserved for very large life-threatening or deep full-thickness burns. The excision is performed using a combination of sharp dissection, traction, and hemorrhage control. The amount excised at each procedure is determined by the stability of the child, blood loss, and the availability of autografts or skin substitutes. The excision is preferably limited to approximately 20 percent TBSA at any one time. Fascial excision assures a viable bed for skin grafting with moderate blood loss, especially if done under tourniquet control. An excellent graft take may be expected if done within the first few days after injury. By incising at the periphery of the eschar vertically downward to the level of the deep investing fascia, a flap of eschar is raised and the dissection is extended until all the dead tissue down to fascia level has been excised. It is preferable to leave a thin layer of fat over the subcutaneous bony prominences and tendon sheaths. Complete hemostasis with electrocoagulation should minimize blood loss substantially, but bleeding often occurs from the skin edges. Topical vasoconstrictive agents could be applied to the fascia as the dissection proceeds. At completion, the extremity is wrapped with a pressure bandage and elevated for 10 minutes. The excised area is covered with an expanded split-thickness skin graft. If the ratio exceeds 2:1, the autograft should be covered with a 1.5:1 meshed cadaveric allograft. The major disadvantages of this method are that it causes damage to lymphatics and cutaneous nerves, loss of subcutaneous fat, long-term cosmetic deformity, and distal edema.



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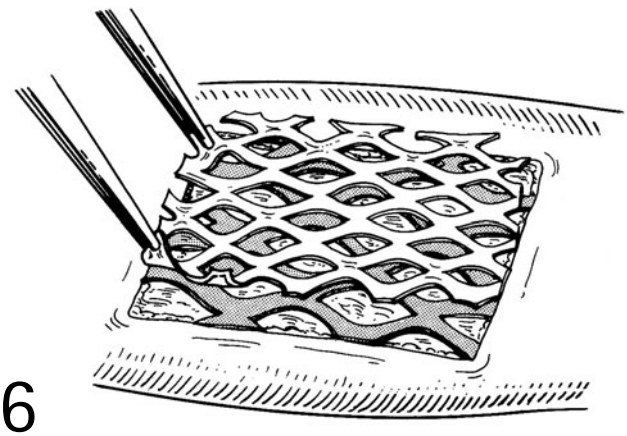
Delayed escharectomy

Delayed escharectomy after 7 days, or following spontaneous eschar separation, allows for the formulation of a bed of granulation tissue. Daily debridement by means of hydrotherapy (showering or bathing) or coarse mesh gauze dressings will hasten eschar separation. The burn wound is ready for split skin grafting when there is a shiny, slightly granular, pink-ish/red uniform bed of granulation tissue with no debris or evidence of infection. This method is most often used for old, neglected burn wounds. Enzymatic debriding agents are seldom used.



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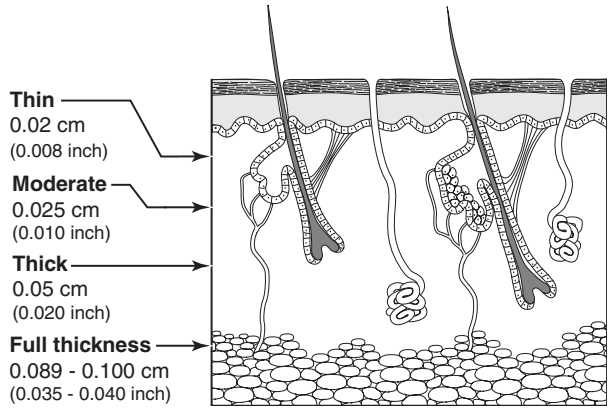
SKIN SUBSTITUTES

6 Allograft is the principal alternative wound closure material and can be used as a (1.5:1.3) meshed graft overlaying an expanded autograft, as a biological dressing, or as a temporary cover on excised wounds in the absence of available autografts for immediate wound closure. Allografts should be removed before rejection becomes evident. In practice, this means removal every 10–12 days and replacement with pool allografts or permanent replacement with autograft when available. If this procedure is followed, the likelihood of rejection and poor recipient bed is reduced. If allografts are left for more than 14 days, removal can only be achieved by excision. Alternative methods (xenografts, cultured keratinocytes, and other biological skin substitutes) are not reliable for routine use and at best function only as temporary skin cover.

Alternatively, the excised wounds can be covered with TransCyte (a fibroblast-derived temporary skin substitute) or Integra (a permanent composite non-cellular dermal substitute) until epitheliazation with a thin autograft can be achieved. Integra may become the gold standard to replace burnt dermal and subcutaneous tissue.

DONOR SKIN PROCUREMENT

All unburned areas can serve as donor sites, although certain areas, i.e., face, hands, perineum, and cervical areas, are unsuitable. The preferred donor sites are the legs, buttocks, and back, and best color match between donor and recipient areas can be obtained from the 'blushing' area of the body for facial and neck burns. Donor skin thickness should be between 0.02 cm (0.008 inch – thin) and 0.025 cm (0.010 inch – thicker). Thicker grafts are more pliable and cause less scarring. Skin grafts are best procured with an electric dermatome.



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Grafts are usually meshed with a Tanner mesher at a ratio of 1.5:1, 2:1, or 3:1, or used as sheet grafts on vital or cosmetically important areas.

Subcutaneous injection of saline and vasopressin will greatly facilitate graft procurement over bony or uneven prominences. Donor sites are usually ready for reuse after 14 days.

HEMOSTATIC CONTROL

Bleeding from excised areas should be minimized by limiting excisional procedures to 25–100 cm^2 at any one time until hemostasis is ensured. Other methods employed are local pressure for 10 minutes, diathermy coagulation, suture ligation, calcium-enriched alginate dressings, and the topical application of sponges soaked in 1:10 000/1:30 000 epinephrine (adrenaline) solution to the excised bed for 10 minutes or subeschar injection of epinephrine. Topical epinephrine should not be used in patients with cardiac disease or arrhythmias. Additionally, extremity exsanquination and pneumatic tourniquet methods can be used.

SKIN GRAFT PLACEMENT

The procured skin is grafted onto the recipient area at the time of eschar excision, directly from the mesh board. Grafts are placed with the shiny or cut surface facing the prepared bed, either longitudinally or transversely over joints. The edges should be approximated or slightly overlapping and secured with surgical clips, sutures, synthetic tissue glue, or fine mesh gauze.

WOUND DRESSINGS

Recipient area

In general terms, the recipient area is covered with an occlusive dressing to prevent infection, avulsion, and desiccation of the graft, and to allow for graft vascularization (3–4 days if grafts are placed on dermis or fascia, and 5–7 days if placed on fat). Both sheet and mesh grafts are covered with a layer of fine mesh gauze, impregnated with topical antibiotics, followed by an absorbable dressing and an elastic dressing and splinting in a functional position where indicated. Small areas can be covered with Adaptic or Vaseline gauze and a dry dressing. Sheet grafts may be left exposed, especially on the face and neck areas. The outer dressings, down to the layer of fine gauze, are taken down the following day to remove any blood clots or wound exudate. Thereafter, the dressings can be left for a few days or changed if there is strike-through bleeding, exudation, or suspected infection. All dressings, clips, or sutures can be removed after 5–6 days and the wounds thereafter protected with Vaseline gauze and bandages until healed.

Donor sites

The area is covered with a topical agent or with occlusive or adherent dressings such as Opsite™, Biobrane™ or Hypafix™. Discomfort is minimal and rapid healing is experienced; the dressing is either left intact until healing has occurred or removed after a few days. Acticoat®, a nanocrystalline silver-containing dressing, may become the donor site dressing of choice. It is applied and left in place for 2 weeks and may be attached with Hypafix™.

FOLLOW-UP

All wounds that have not healed within 3 weeks and all grafted areas will require pressure garments for 6–18 months. All burnt, grafted, and donor areas should be protected from direct sun exposure for at least 6–12 months. Topical application of a bland moisturizing agent may improve skin texture and color.

Early reconstruction principles form an integral part of all surgical procedures, and long-term problems can be circumvented or minimized by proper positioning, supportive splints and pressure devices, judicious use of skeletal traction, and suspension to maintain joint excursions and mobility.

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94

Management of major abdominal trauma

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> If a disease were killing our children in the proportions that accidents are, people would be outraged and demand that this killer be stopped.
>
> *C. Everett Koop, MD, ScD*

HISTORY

Who could have imagined the influence of James Simpson's publication in 1968 on the successful *non-operative* treatment of select children presumed to have splenic injury. Initially suggested in the early 1950s by Tim Warnsborough, then Chief of General Surgery at the Hospital for Sick Children in Toronto, it is remarkable to consider that the era of non-operative management for pediatric spleen injury began with the report of 12 children treated between 1956 and 1965. The diagnosis of splenic injury in this select group was made by clinical findings together with routine laboratory and plain X-ray findings. Keep in mind that the report pre-dated ultrasound, computed tomography (CT), or isotope imaging.

Nearly four decades later, the standard treatment of hemodynamically stable children with splenic injury is non-operative and this concept has now been successfully applied to most blunt injuries of the liver, kidney, and pancreas as well. Surgical restraint has been the theme, based on an increased awareness of the anatomic patterns and physiologic responses characteristic of injured children. Our colleagues in adult trauma care have slowly acknowledged this success and applied many of the principles learned in pediatric trauma to their patients.

Few surgeons have extensive experience with massive abdominal solid organ injury requiring immediate surgery. It is imperative that surgeons familiarize themselves with current treatment algorithms for life-threatening abdominal trauma. Important contributions have been made to the diagnosis and treatment of children with abdominal injury by radiologists and endoscopists. The resolution and speed of CT, the screening capabilities of focused abdominal sonography for trauma (FAST), and the percutaneous, angiographic, and endoscopic interventions of non-surgeon members of the pediatric trauma team have all enhanced patient care and improved outcomes. This chapter focuses on the more common blunt injuries of the spleen, liver, duodenum, pancreas, and kidney.

RESUSCITATION

A multi-trauma patient entering a medical facility should be treated by an organized team of surgeons, physicians, and nurses. The composition of the team will vary, but the senior surgeon should be the team leader. Preparation is mandatory, and includes ensuring the availability of equipment appropriate to children of varying ages and the establishment of a resuscitation protocol. The 'checklist approach' is the surest way to accomplish the essential steps of diagnosis and treatment while individualizing care for each patient. The initial evaluation of the acutely injured child is similar to that of the adult. Plain radiographs of the cervical spine, chest, and pelvis are obtained following the initial survey and evaluation of A (airway), B (breathing), and C (circulation). As imaging modalities have improved, treatment algorithms have changed significantly in children with a suspected intra-abdominal injury. Prompt identification of potentially life-threatening injuries is now possible in the vast majority of children.

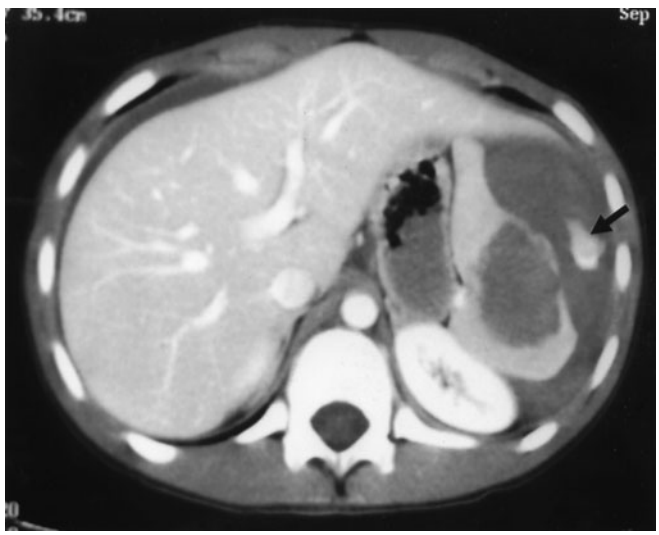
Airway control, vascular access, spinal precautions, and temperature regulation must be assured throughout the initial evaluation and treatment of injured children. The reader is referred to the *Textbook of Pediatric Advanced Life Support* (American Academy of Pediatrics) and the *Textbook of*

Advanced Trauma Life Support (American College of Surgeons) for specific details of airway management, pharmacologic therapy, and central venous access in injured children.

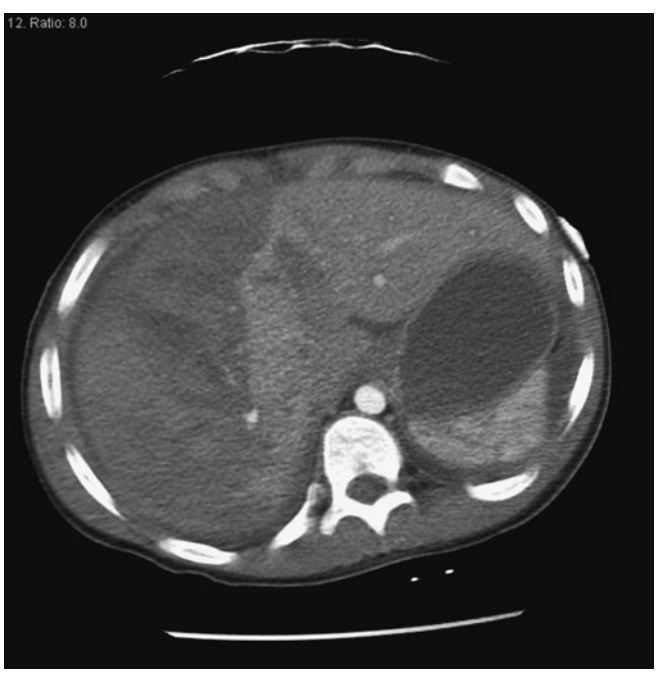
DIAGNOSIS OF BLUNT ABDOMINAL INJURIES

Recognition of significant abdominal injuries in children with blunt multi-system trauma can be difficult. Physical examination is inaccurate in more than 30 percent of cases, particularly in those patients seen soon after injury or with central nervous system injuries and an abnormal neurologic examination. It is no surprise that advances in diagnosis have paralleled the development of new imaging technology. Prompt and accurate recognition of abdominal injuries, now possible with CT, allows for focused treatment plans.

Computerized tomography has become the imaging study of choice for the evaluation of injured children because of several of its advantages. It is now readily accessible in most healthcare facilities, is non-invasive, is a very accurate method of identifying and qualifying the extent of abdominal injury, and has reduced the incidence of non-therapeutic exploratory laparotomy.



1a

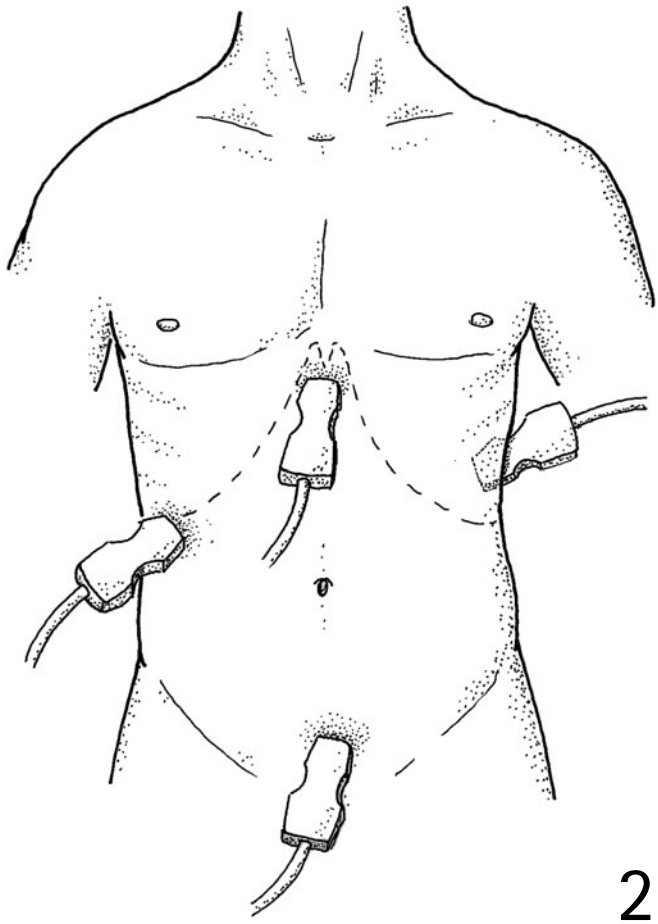


1b

1a,b The use of intravenous contrast is essential, and the utilization of 'dynamic' methods of scanning has optimized vascular and parenchymal enhancement. The impact on resource utilization and outcome of a contrast 'blush' on CT in children with blunt spleen and liver injury continues to be debated. A head CT, if indicated, should be performed first without contrast, to avoid contrast concealing a hemorrhagic brain injury. Enteral contrast for enhancement of the gastrointestinal tract is generally not required in the acute trauma setting and can lead to aspiration.

Focused abdominal sonography for trauma

Clinician-performed sonography for the early evaluation of the injured child is currently being assessed to determine its optimal use. The standard four-view FAST examination includes Morrison's pouch/right upper quadrant, the left flank to include the perisplenic anatomy/left upper quadrant, and a subxiphoid view to visualize the pericardium and the pouch of Douglas/pelvis. This bedside examination may be useful as a rapid screening study, particularly in those patients too unstable to undergo an abdominal CT scan. Early reports have found FAST to be a useful screening tool in children, with a high specificity (95 percent), but a low sensitivity (33 percent) in identifying intestinal injury. A lack of identifiable free fluid does not exclude a significant injury. FAST may be very useful in decreasing the number of CT scans performed for 'low-likelihood' injuries.



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TREATMENT OF SPECIFIC ABDOMINAL INJURIES

Spleen and liver

The spleen and liver are the organs most commonly injured in blunt abdominal trauma, with each accounting for one-third of injuries. Non-operative treatment of isolated splenic and hepatic injuries in stable children is now standard practice. Although non-operative treatment of children with isolated blunt spleen or liver injury has been universally successful, there has been great variation in the management algorithms used by individual pediatric surgeons. Review of the National Pediatric Trauma Registry (NPTR) and recent surveys of the American Pediatric Surgical Association (APSA) membership confirm the wide disparity in practice.

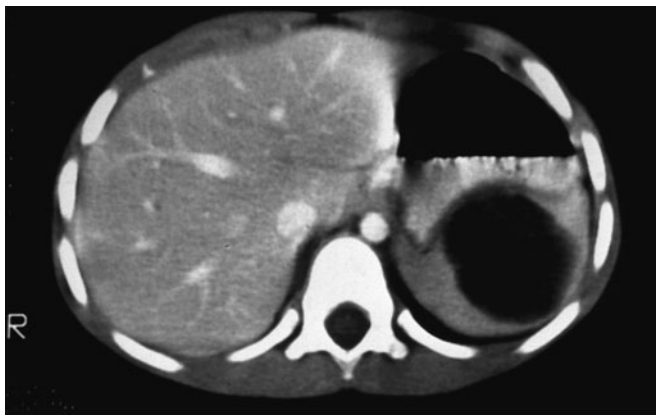
Recently, the APSA Trauma Committee defined consensus guidelines for resource utilization in hemodynamically stable children with isolated liver or spleen injury based on CT grading by analyzing a contemporary, multi-institution database of 832 children treated non-operatively at 32 centers in North America from 1995 to 1997. Consensus guidelines on intensive care unit (ICU) stay, length of hospital stay, use of follow-up imaging, and physical activity restriction for clinically stable children with isolated spleen or liver injuries (Grades I–IV) were defined by analysis of this database (Table 94.1).

The guidelines were then applied prospectively in 312 children with liver or spleen injuries treated non-operatively at 16 centers from 1998 to 2000. **It is imperative to emphasize that these proposed guidelines assume hemodynamic stability.** The extremely low rates of transfusion and operation document the stability of the study patients. Compared with the previously studied 832 patients, the 312 patients managed prospectively by the proposed guidelines had a significant reduction in ICU stay ($p < 0.0001$), hospital stay ($p < 0.0006$), follow-up imaging ($p < 0.0001$), and interval of physical activity restriction ($p < 0.04$) within each grade of injury.

Table 94.1 *Proposed guidelines for resource utilization in children with isolated spleen or liver injury*

CT grade	I	II	III	IV
ICU days	None	None	None	1
Hospital stay (days)	2	3	4	5
Pre-discharge imaging	None	None	None	None
Post-discharge imaging	None	None	None	None
Activity restriction (weeks)[a]	3	4	5	6

[a]Return to *full contact, competitive sports* (i.e. football, wrestling, hockey, lacrosse, mountain climbing, etc.) should be at the discretion of the individual pediatric trauma surgeon. The proposed guidelines for return to unrestricted activity include *'normal' age-appropriate activities.*
CT, computed tomography; ICU, intensive care unit.



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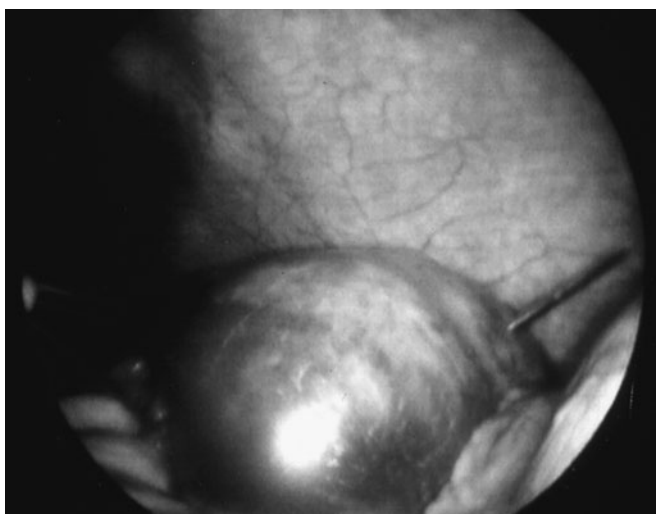

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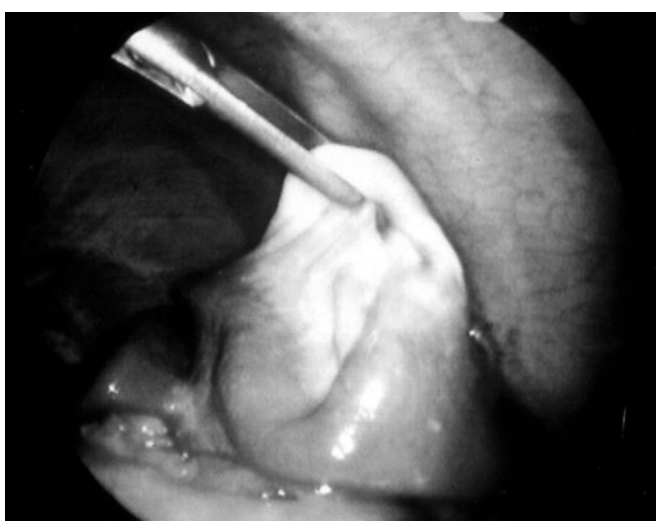
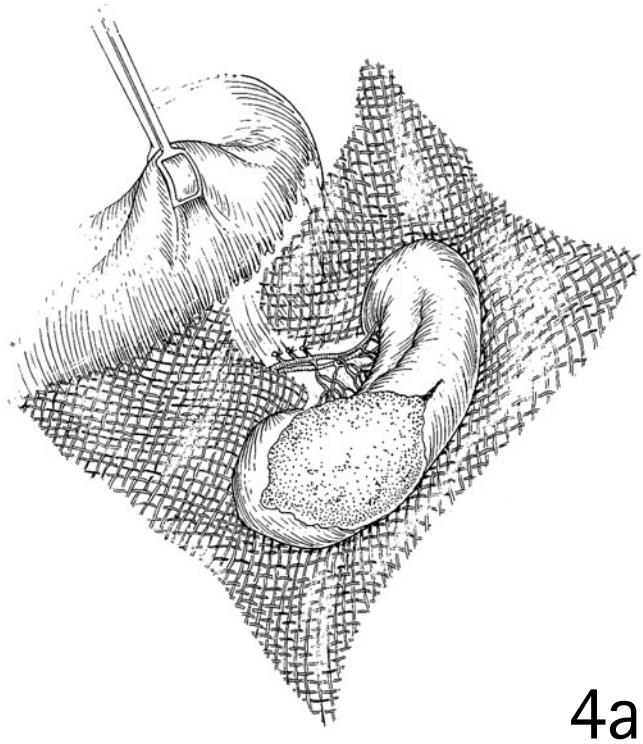


3c

3a–c Routine follow-up imaging studies have identified pseudocysts and pseudoaneurysms following splenic injury. Splenic pseudoaneurysms often cause no symptoms and appear to resolve with time. The true incidence of self-limited, post-traumatic splenic pseudoaneurysms is unknown, as routine follow-up imaging after successful non-operative treatment has been largely abandoned. Once identified, the actual risk of splenic pseudoaneurysm rupture is also unclear. These lesions can be treated successfully with angiographic embolization techniques, obviating the need for open surgery and loss of splenic parenchyma. Splenic pseudocysts can achieve enormous size, leading to pain and gastrointestinal disturbance (a). Simple percutaneous aspiration leads to a high recurrence rate. Laparoscopic excision and marsupialization is highly effective (b and c).



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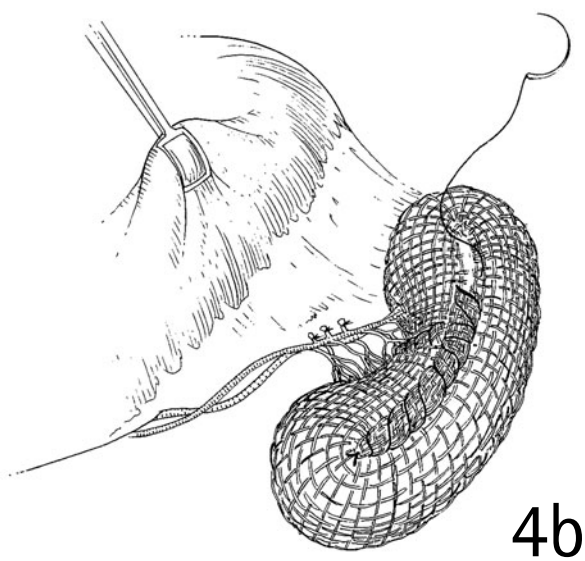
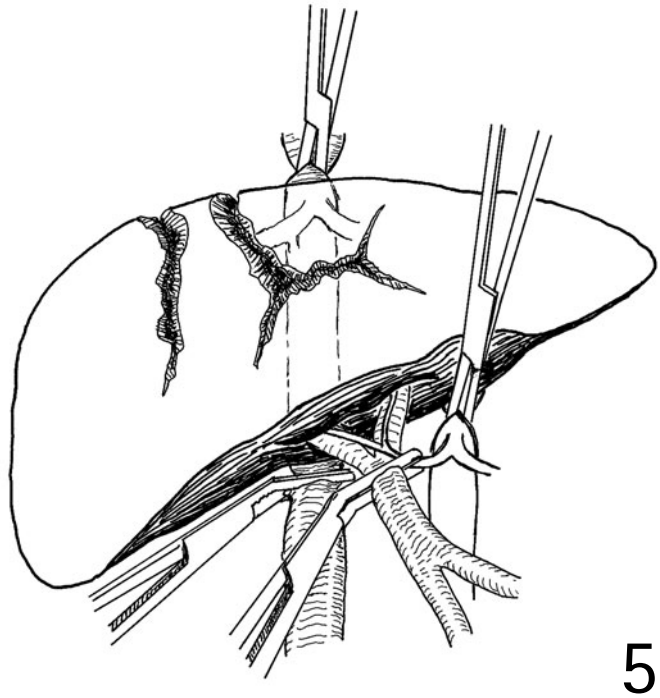


4b

4a,b Even the most severe solid organ injuries can be treated without surgery if there is prompt response to resuscitation. In contrast, emergency laparotomy and/or embolization are indicated in patients who are hemodynamically unstable despite fluid and red blood cell transfusion. Most spleen and liver injuries requiring operation are amenable to simple methods of hemostasis using a combination of manual compression, direct suture, topical hemostatic agents, and woven polyglycolic mesh wrapping.

5 In young children with significant hepatic injury, the sternum can be divided rapidly to expose the suprahepatic or intrapericardial inferior vena cava, allowing for total hepatic vascular isolation with occlusion of the porta hepatis, suprahepatic and infrahepatic inferior vena cava, and supraceliac aorta (optional). Children will tolerate periods of vascular isolation as long as their blood volume is replenished. With this exposure, the liver and major perihepatic veins can be isolated and the bleeding controlled to permit direct suture repair or ligation of the offending vessel. Although the cumbersome and dangerous technique of atriocaval shunting has been largely abandoned, newer endovascular balloon catheters can be useful for temporary vascular occlusion to allow access to the juxtahepatic vena cava.



5

The early morbidity and mortality of severe hepatic injuries are related to the effects of massive blood loss and replacement with large volumes of cold blood products. The consequences of prolonged operations with massive blood product replacement include hypothermia, coagulopathy, and acidosis. Although the surgical team may keep pace with blood loss, life-threatening physiologic and metabolic consequences are inevitable, and many of these critically ill patients are unlikely to survive once their physiologic reserves have been exceeded. Maintenance of physiologic stability during the struggle for surgical control of severe bleeding is a formidable challenge even for the most experienced operative team, particularly when hypothermia, coagulopathy, and acidosis occur. This triad creates a vicious cycle in which each derangement exacerbates the others, and the physiologic and metabolic consequences of the triad often preclude completion of the procedure. Lethal coagulopathy from dilution, hypothermia, and acidosis can occur rapidly. The infusion of activated recombinant Factor VII in patients with massive hemorrhage has shown promising results.

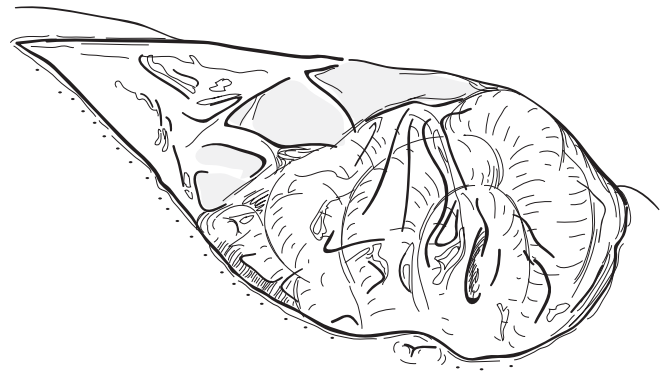
Increased emphasis on physiologic and metabolic stability in emergency abdominal operations has led to the development of staged, multidisciplinary treatment plans including abbreviated laparotomy, perihepatic packing, temporary abdominal closure, angiographic embolization, and endoscopic biliary stenting. Abbreviated laparotomy with packing for hemostasis allowing resuscitation prior to planned re-operation is an alternative in unstable patients where further blood loss would be untenable. This 'damage control' philosophy is a systematic, phased approach to the management of the exsanguinating trauma patient. The three phases of damage control are detailed in Table 94.2. Once patients are re-warmed, coagulation factors replaced, and oxygen delivery optimized, they can be returned to the operating room for pack removal and definitive repair of injuries.

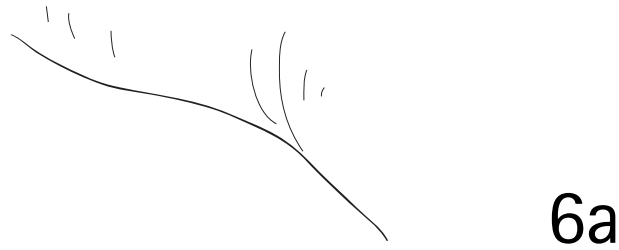
While the success of abdominal packing is encouraging, it may contribute to significant morbidity such as intra-abdominal sepsis, organ failure, and increased intra-abdominal pressure. It is essential to emphasize that the success of the abbreviated laparotomy and planned re-operation depends on an early decision to employ this strategy prior to irreversible shock. Abdominal packing, when employed as a desperate, last-ditch resort after prolonged attempts at hemostasis have failed, has been uniformly unsuccessful.

Table 94.2 *'Damage control' strategy in the exsanguinating trauma patient*

Phase 1	Abbreviated laparotomy for exploration Control of hemorrhage and contamination Packing and temporary abdominal wall closure
Phase 2	Aggressive ICU resuscitation Core re-warming Optimize volume and oxygen delivery Correction of coagulopathy
Phase 3	Planned re-operation(s) for packing change Definitive repair of injuries Abdominal wall closure

ICU, intensive care unit.





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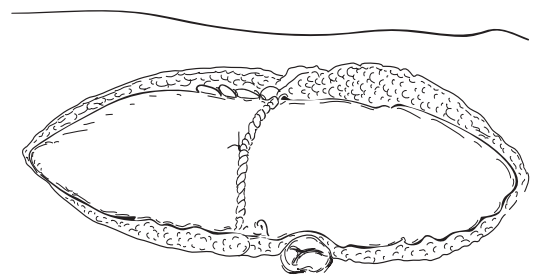


6b

6a,b The obvious benefits of hemostasis provided by packing are also balanced against the potential deleterious effects of increased intra-abdominal pressure on ventilation, cardiac output, renal function, mesenteric circulation, and intracranial pressure. Timely alleviation of the secondary 'abdominal compartment syndrome' may be a critical salvage maneuver for patients. Temporary abdominal wall closure at the time of packing can prevent the occurrence of this syndrome. We recommend temporary abdominal wall expansion in all patients requiring packing until the hemostasis is obtained and visceral edema subsides. Many materials have been suggested for use in temporary patch abdominoplasty, including silastic sheeting (a), Goretex® patches (b), intravenous bags, cystoscopy bags, ostomy appliances, and various mesh materials.

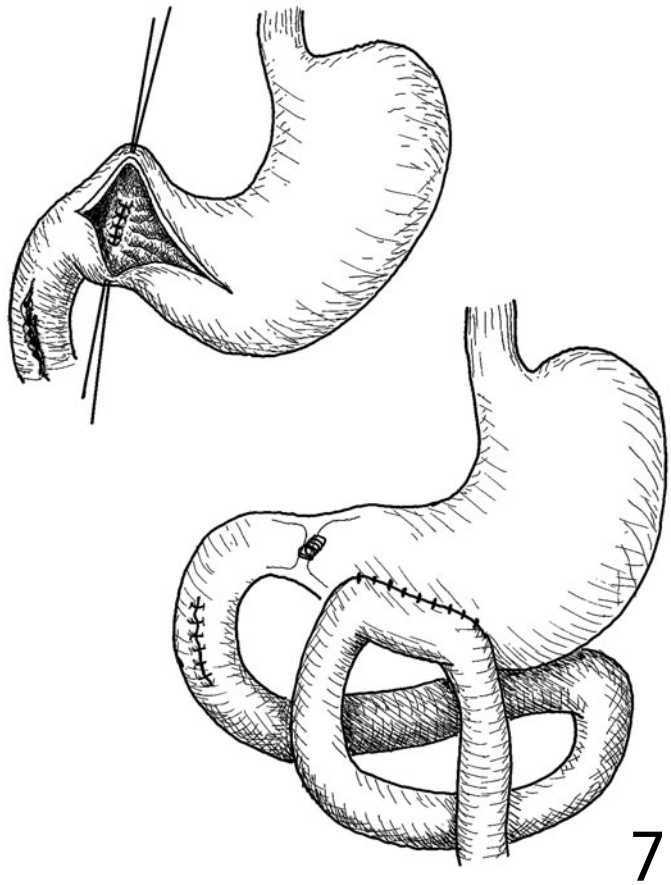
A staged operative strategy for unstable trauma patients represents *advanced* surgical care and requires sound judgment and technical expertise. Intra-abdominal packing for the control of exsanguinating hemorrhage is a life-saving maneuver in highly selected patients in whom coagulopathy, hypothermia, and acidosis render further surgical procedures unduly hazardous. Early identification of patients likely to benefit from abbreviated laparotomy techniques is crucial for success.

Duodenum and pancreas

7 Patients sustaining duodenal perforation are treated operatively in a variety of ways depending on the severity of the injury and the surgeon's preference (Box 94.1). The authors recommend primary closure of the duodenal perforation (whenever possible). Extensive lateral duodenal injury should be treated by primary duodenal repair and 'pyloric exclusion' consisting of temporary closure of the pylorus with an absorbable suture and gastrojejunostomy. Closed suction drainage of the repair is not depicted in this illustration. Feeding jejunostomy is often added to the procedure. When the duodenum is excluded, complete healing of the injury routinely occurs prior to the spontaneous re-opening of the pyloric channel and spontaneous closure of the gastrojejunostomy.

Box 94.1 *Surgical options in duodenal trauma*

- Repair of the duodenum.
- Diversion of the gastrointestinal tract (pyloric exclusion or a duodenal diverticulization).
- Gastric decompression (gastric tube insertion or gastrojejunostomy).
- Gastrointestinal tract access for feeding (jejunostomy tube or gastrojejunal anastomosis).
- Decompression of the duodenum (duodenostomy tube).
- Biliary tube drainage.
- Wide drainage of the repaired area (lateral duodenal drains).



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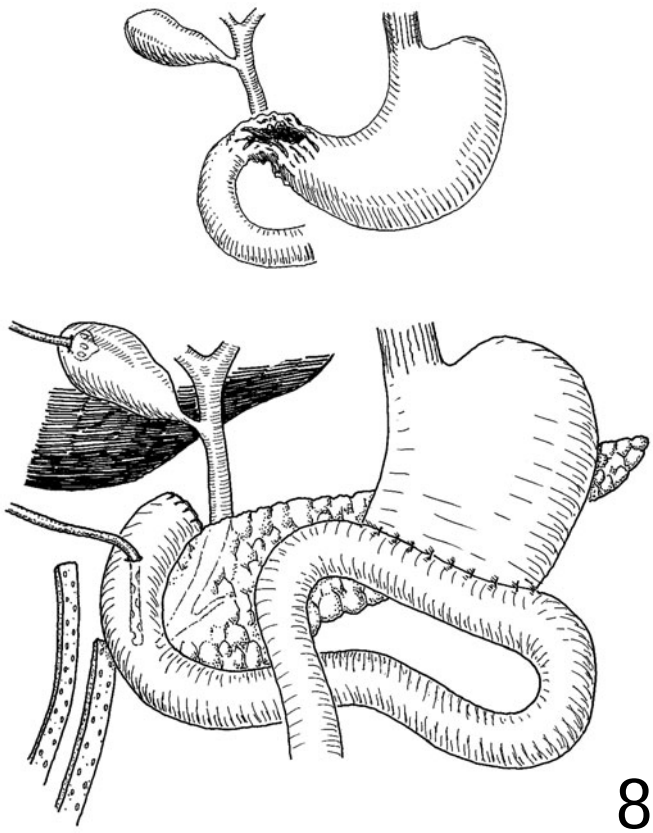


8 Duodenal diverticularization is an effective procedure for combined proximal duodenal and pancreatic injury. Resection and closure of the duodenal stump with decompressive tube duodenostomy, biliary drainage via tube cholecystectomy, gastrojejunostomy, and multiple closed suction drains are depicted. A feeding jejunostomy should be strongly considered (not depicted). No matter what repair the surgeon selects, a summary of the literature demonstrates that protecting the duodenal closure (drain and exclusion) and a route for enteral feeds (gastrojejunostomy ± or feeding jejunostomy) reduces morbidity and length of stay. A pancreaticoduodenectomy (Whipple procedure) should rarely be required. Although occasionally reported in the literature, pancreaticoduodenectomy should be reserved for the most severe injuries to the duodenum and pancreas when the common blood supply is destroyed and any possibility of reconstruction is impossible.

Injuries to the pancreas are slightly more common than duodenal injuries, with estimated ranges from 3 percent to 12 percent in children sustaining blunt abdominal trauma. A summary comparing the San Diego and Toronto protocols is depicted in Figure 94.1. The striking differences in these series are the 100 percent diagnostic sensitivity of CT scanning in Toronto versus 69 percent in San Diego, and the 44 percent operative rate in San Diego versus 0 percent in Toronto. The Toronto group concludes that distal parenchymal atrophy or ductal recanalization occurs uniformly with no long-term morbidity in patients following the non-operative treatment of blunt pancreatic trauma.

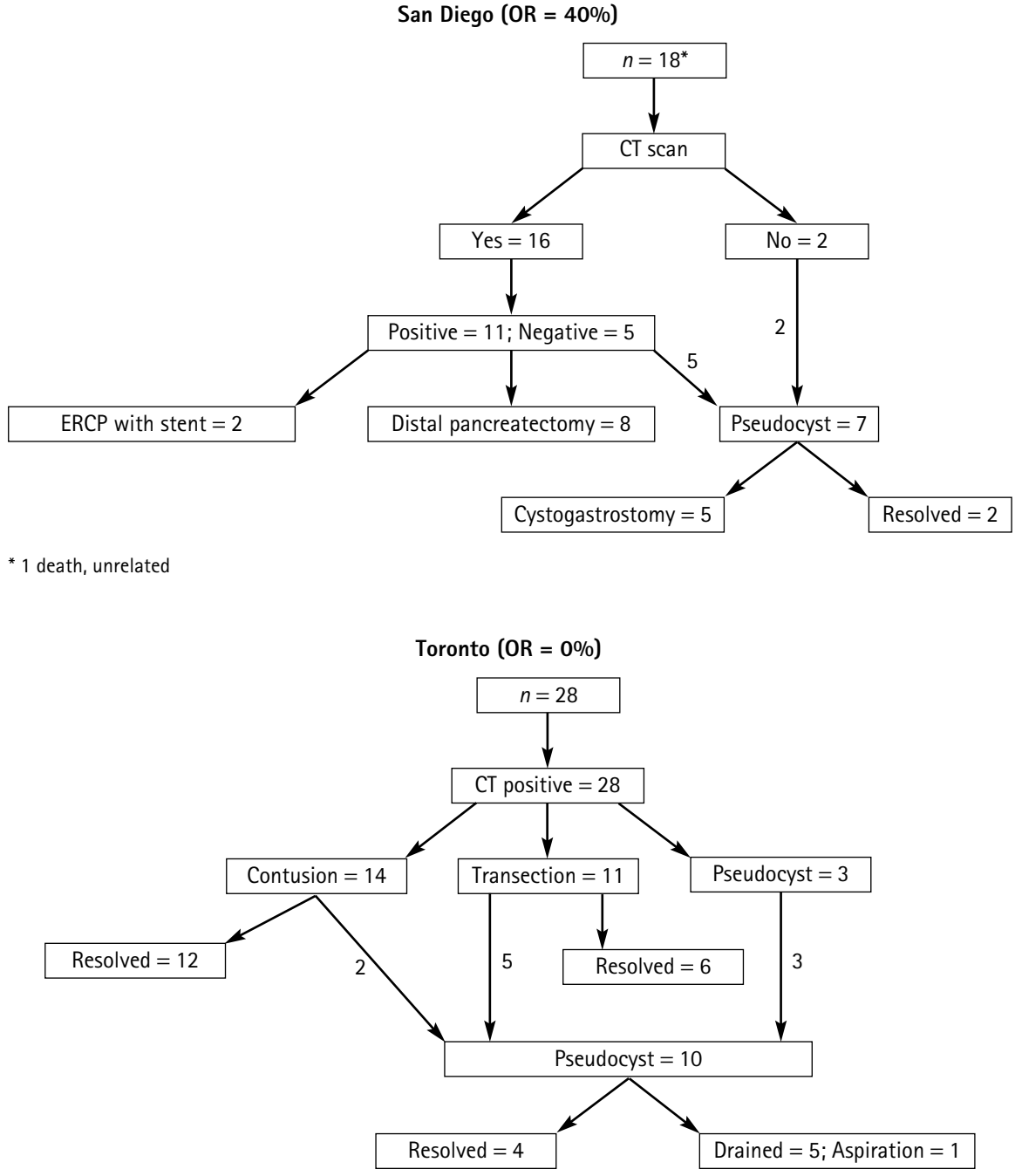
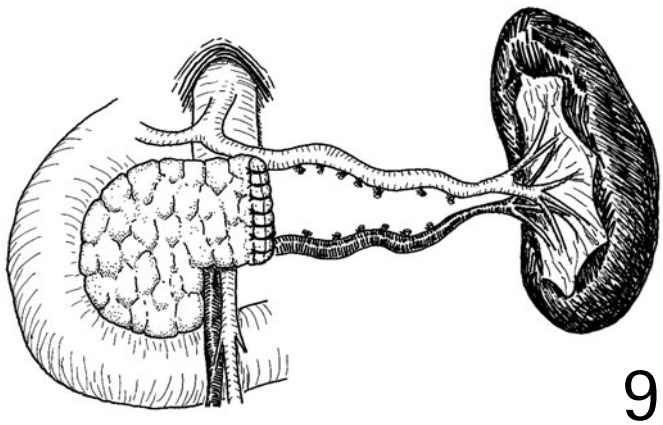


Fig. 94.1 A comparison of protocols in the management of blunt pancreas injury in children. (OR = operation; CT = computed tomography; ERCP = endoscopic retrograde cholangiopancreatography.)

9 Reports from major pediatric trauma centers are clearly in conflict. Some favor and document the efficacy and safety of observational care for virtually all pancreatic traumas including ductal disruption, whereas others favor early distal pancreatectomy for transection to the left of the spine. It is clear that with simple transection of the pancreas at or to the left of the spine, spleen-sparing distal pancreatectomy can accomplish definitive care for this isolated injury with short hospitalization and acceptable morbidity.

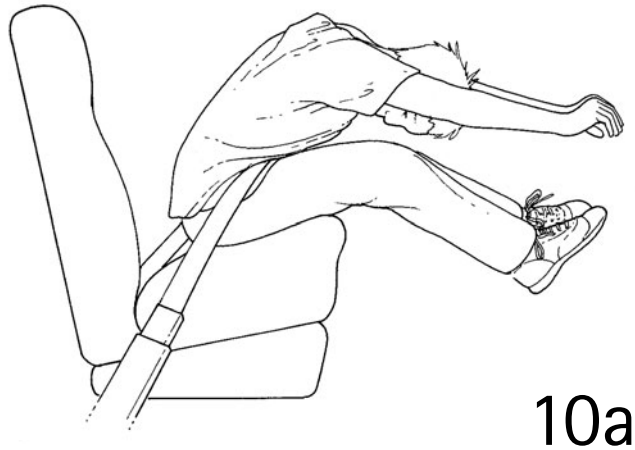


Kidney

Renal injuries in children are often caused by high-energy impact associated with motor vehicle accidents or other serious abdominal trauma. Hematuria is present in 41–68 percent of children following blunt abdominal trauma. Most authors report direct correlation between the amount of hematuria and the severity of genitourinary injury, but renovascular injuries may result in no hematuria and bladder contusion or disruption often causes large amounts of blood in the urine. The magnitude of hematuria requiring diagnostic evaluation following blunt abdominal trauma continues to be a subject of debate, as does the optimal imaging study for the detection of renal injuries. Pre-existing or congenital renal abnormalities, such as hydronephrosis, tumors, or abnormal position, may predispose the kidney to trauma despite relatively mild traumatic forces. Historically, congenital abnormalities in injured kidneys have been reported to vary from 1 percent to 21 percent. More accurate recent reviews have shown that incidence rates are 1–5 percent. Renal abnormalities, particularly hydronephrotic kidneys, may be first diagnosed after minor blunt abdominal trauma. These patients usually present with hematuria following blunt trauma.

It is imperative to acknowledge that major renal injuries such as ureteropelvic junction (UPJ) disruption or segmental arterial thrombosis may occur without the presence of hematuria or hypotension. Therefore, a high index of suspicion is necessary to diagnose these injuries. Non-visualization of the injured kidney on intravenous pyelogram and failure to uptake contrast with a large associated perirenal hematoma on CT are hallmark findings for renal artery thrombosis. Ureteropelvic junction disruption is classically seen as perihilar extravasation of contrast with non-visualization of the distal ureter.

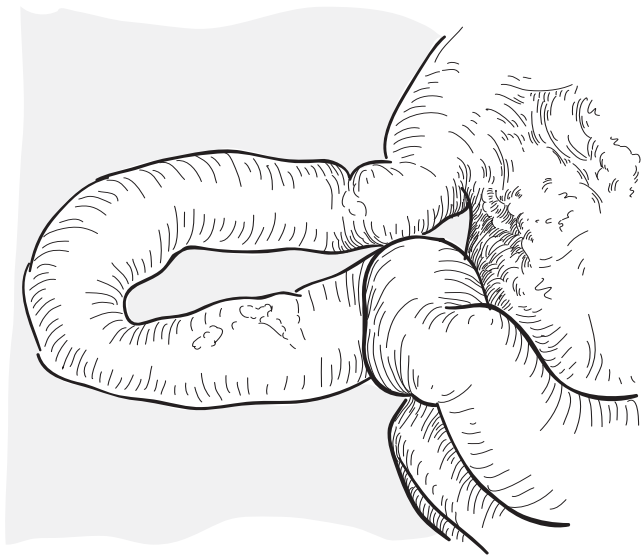
The majority of blunt renal injuries are treated without operation when uncontrolled hemorrhage or other indications for abdominal exploration are absent. This approach is safe and effective in 77–86 percent of children and most have excellent functional outcome without hypertension. Successful renal salvage at operation by partial nephrectomy or nephrorrhaphy depends on the severity of both the renal injury and associated injuries. Collecting system injuries should be repaired with absorbable sutures after evacuation of pelvic clots and debridement of devascularized parenchyma. Intravenous infusion of indigo-carmine (a vital dye excreted in the urine) at operation may help identify sites of extravasation, and proximal control of the renal vessels prior to opening Gerota's fascia may facilitate retroperitoneal exploration. Early control of the vessels increases the rate of renal salvage. When proximal vascular control is performed before any renal exploration, nephrectomy is required in less than 12 percent of cases. When primary vascular control is not achieved and massive bleeding is encountered, the nephrectomy rate increases. Nephrectomy is recommended for major renal injuries in hemodynamically unstable patients with multiple injuries, and in those patients with avulsion injuries. Vascular repair can be attempted within 12 hours of injury and in the absence of multiple injuries.



10a

The 'seatbelt syndrome'

10a–c A decrease in motor vehicle-related fatalities has occurred in association with mandatory seatbelt legislation. Concurrently, a pattern of injuries caused by lap-belt restraints has emerged. This 'seatbelt syndrome' includes abdominal wall contusion, injury to a hollow viscus, and vertebral fracture. The mechanism of injury is hyperflexion of the torso caused by deceleration forces with the lap-belt as a fulcrum (a). Children are at particular risk for these injuries due to their higher center of gravity, thin abdominal wall, non-prominent iliac crest, and immature supporting structures of the vertebral bodies. Bowel injuries associated with lap-belt use can occur by several mechanisms: compression against the vertebral column with a crush injury, shearing of the bowel and mesentery at fixed points in the retroperitoneum, and immediate closed-loop burst injuries on the anti-mesenteric border when fluid-filled loops are subjected to sudden increases in intraluminal pressure. The crush and shearing mechanisms may lead to progressive ischemia with delayed perforation or stricture (b). Children with the 'seatbelt sign' across their lower abdomen should be admitted for serial examinations even if the initial examination and diagnostic tests are normal (c).



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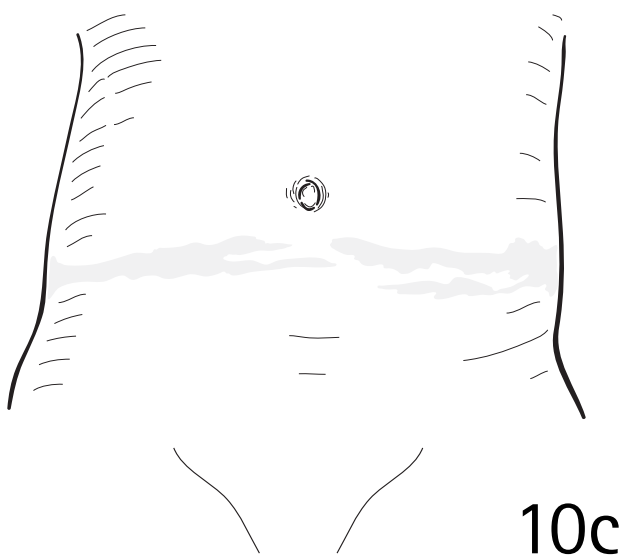

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CONCLUSIONS

The essential ingredient of pediatric trauma care is commitment to the special needs of injured children – personal, institutional, and community commitment. Recent advances in the delivery of trauma and critical care in children have resulted in improved outcome following major injuries. Incorporation of newer minimal access, endoscopy, and interventional radiology techniques is strongly urged. Although an increasing emphasis on non-operative treatment has occurred in the last two decades, the pediatric surgeon should remain the principal physician in the multidisciplinary care of these critically injured children; *the decision not to operate is always a surgical decision*.

Acknowledgment

Original illustrations 2, 5, 7, 8, and 9 by Mark Mazziotti, MD.

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SECTION IX

Miscellaneous

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95

Principles of maternal–fetal surgery

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PRINCIPLES AND JUSTIFICATION

Advanced fetal diagnostic techniques (e.g., chorionic villous sampling, amniocentesis) and serial imaging (e.g., ultrasound, magnetic resonance imaging) have led to an increased understanding of the natural history and outcome of many fetal anomalies. As a result, the majority of fetuses with anomalies amenable to surgery are best served by planned delivery and care after birth. There is, however, a small proportion of fetuses for whom an untreated anomaly will result in death in utero or shortly after birth. Prenatal intervention has, in large part, been predicated on those anomalies that cause either low-output or high-output cardiac failure resulting in hydrops, defined as skin and/or nuchal edema or fluid accumulation in two of three body cavities (pleura, pericardium, peritoneum). Examples include fetuses with hydrops from a space-occupying chest mass (e.g., cystic adenomatoid malformation, pericardial teratoma, tension hydrothorax), sacrococcygeal teratoma (SCT), select heart defects, twin–twin transfusion syndrome, and twin reversed-arterial perfusion (TRAP) sequence. Pulmonary and kidney failure can be predicted in a subset of fetuses with complete and early gestation urinary tract obstruction (e.g., posterior urethral valves) and may benefit from ultrasound-guided vesicoamniotic shunt placement. Fetuses with threatened postnatal airway obstruction from a cervical teratoma or lymphangioma may benefit from the ex utero intrapartum treatment (EXIT) strategy. Controversial areas of maternal–fetal intervention include non-lethal anomalies such as laser therapy for amniotic band syndrome and repair of a myelomeningocele.

Patient selection

The prerequisites for consideration of maternal–fetal intervention include:

- the absence of severe associated anomalies;
- a normal karyotype;
- a well-defined natural history, for which, when considering the maternal–fetal unit, the risk:benefit ratio favors intervention;
- the presence of a correctable lesion, which, if uncorrected, will lead to fetal death or irreversible organ dysfunction before birth;
- no uterine or placental anomalies;
- support of the institutional multidisciplinary fetal review board.

Finally, the maternal social support structure and other maternal comorbidities must be considered.

PREOPERATIVE

Assessment and preparation

The fetus is assessed for a normal karyotype via chorionic villous sampling or amniocentesis. Serial ultrasonography is used to characterize the fetal abnormality, assess its natural history, and examine for placental position, fetal lie, and any uterine abnormalities. A fetal echocardiogram is used to rule out congenital heart disease. The pregnant woman undergoes a complete physical examination, serum screening for hepatitis and human immunodeficiency virus (HIV) status, and a search for signs of pre-eclampsia (hypertension, edema, proteinuria) and for any medical condition that may be a contraindication to surgery or general anesthesia.

Anesthesia and tocolytic therapy

Breeching the uterus, whether by puncture or incision, incites uterine contractions. Empiric tocolysis is instituted prior to

open fetal surgery cases using a combination of preoperative indomethacin followed by intravenous β-mimetics and magnesium sulfate for intraoperative and postoperative care. A high dose of an inhalational halogenated agent as well as an epidural placed preoperatively provides anesthesia for both mother and fetus, with the added benefit of uterine relaxation and prevention of uterine contractions and the initiation of labor.

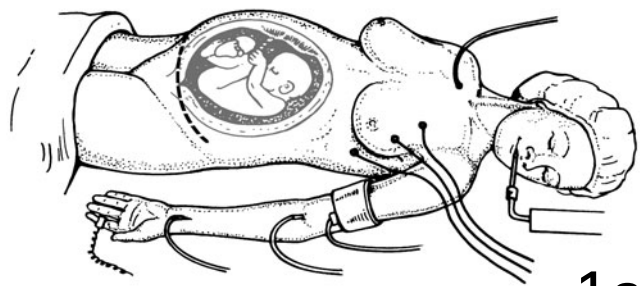
Maternal intraoperative monitoring includes an arterial line, pulse oximetry, bladder drainage catheter, and sequential extremity venous compression device. Cephazolin is administered to minimize the risk of wound infection and chorioamnionitis. The pregnant woman is always positioned with a roll under her right side to avoid inferior vena cava compression by the gravid uterus. Many physicians, support people, and equipment need to be near the patient. For these reasons, along with the uncertainties of the ultimate best position to perform the operation based on fetal lie and placental position, low lithotomy positioning is extremely helpful because it allows access to the uterus from three positions.

FETAL SURGERY TECHNIQUES

Open hysterotomy technique

MATERNAL INCISION

1 A low (suprapubic) transverse skin incision that is slightly curved upward laterally and is on-line with the anterior superior iliac crests is used. A vertical skin incision is performed for obese patients (body mass index > 30) or those with a previous vertical skin scar. Depending on the size of the uterus and placental location, the abdominal muscles can be transected (Mallard incision) or retracted (Pfannensteil incision). A fixed ring retractor facilitates uterine exposure. The fetus and placenta are located by ultrasound and the hysterotomy location chosen. With ultrasound guidance, the edges of the placenta are drawn on the uterine surface with a marking pen.



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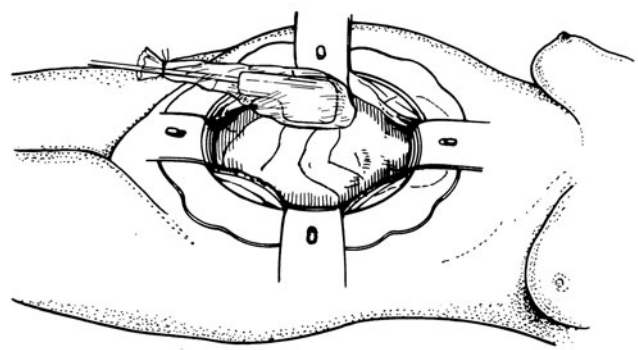
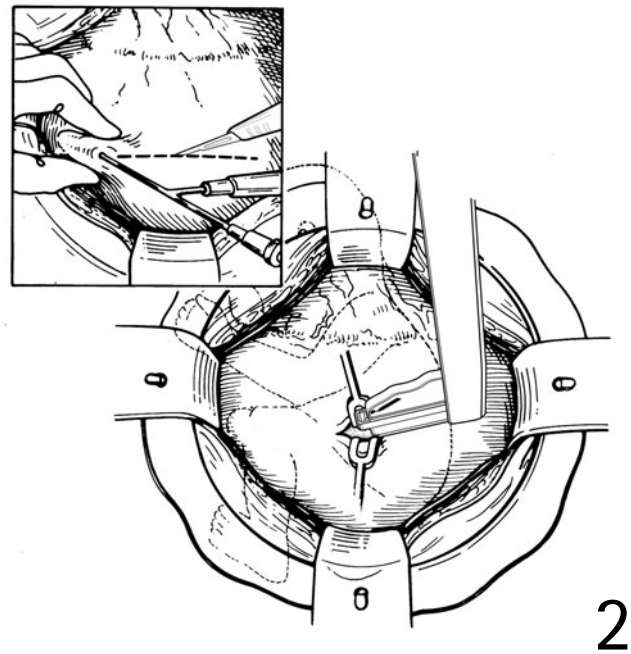


1b

HYSTEROTOMY

2 An anterior placenta requires tipping the uterus forward so that a posterior hysterotomy can be performed, while a posterior or fundic placenta allows for the uterus to remain in situ. At the very minimum, the hysterotomy should be parallel to and not less than 3 cm from the placental edge. If polyhydramnios is present, amnioreduction is performed to avoid underestimating the proximity of the placental edge to the hysterotomy. Initial uterine entry is accomplished through a 1–2-cm hysterotomy using a scalpel. This is performed in the midportion of the proposed hysterotomy. The hysterotomy is completed by placing the foot plate of a specifically designed uterine stapling device into the uterus. The staple line is hemostatic and apposes the membranes to the undersurface of the myometrium using two rows of absorbable Lactomer® staples (United States Surgical Corporation, Norwalk, CT). Palpation and ultrasound are used to exclude the presence of fetal tissue in the stapler jaws before firing. A 6–8-cm uterine incision is created, usually by using two fires of the stapler, in opposite directions. Myometrial bleeding often occurs where the staple lines fail to intersect; this is controlled with a full-thickness, figure-of-eight, absorbable, monofilament 2/0 suture.



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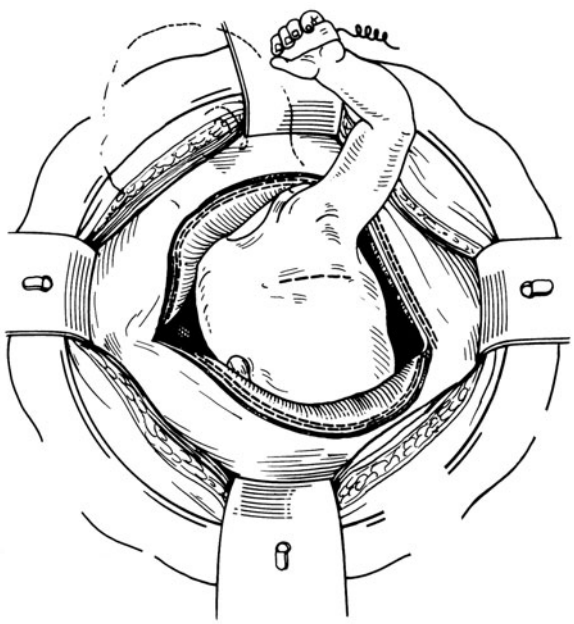


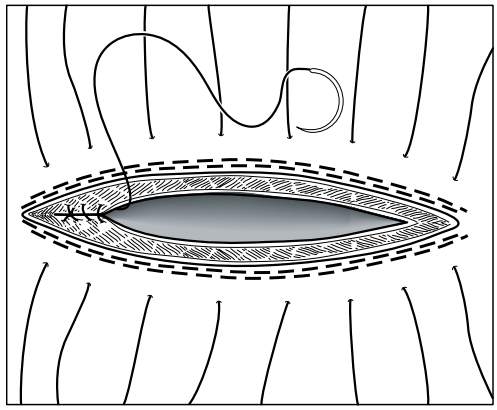
3

FETAL EXPOSURE

3 The fetus is directly visualized and manually positioned within the uterus, with the target body part or cavity exposed while the rest of the fetus remains within the uterus. The fetus is continually bathed by an infusion of warmed lactated Ringer's solution via a soft catheter placed deeply into the uterus. A sterile pulse oximeter is placed on the fetal hand and a sterile extension cord is passed across the field to the anesthesiologist. The fetus is given an intramuscular injection of fentanyl and pancuronium prior to the incision. A standard surgical operation is now performed on the fetus, in this case, a left thoracotomy for a space-occupying lung lesion. Fetal well-being is monitored throughout the procedure with a combination of pulse oximetry and echocardiography.

WOUND CLOSURES

4a–c The fetal wound is closed as for any postnatal operation. A hermetic two-layer uterine closure is performed with long-term absorbable monofilament suture. First, interrupted sutures are placed 2 cm from the hysterotomy edge and left untied (a). Next, the uterine edges are approximated with a running, full-thickness suture (b). The staple line does not need to be excised. Just prior to completing the first layer, the uterine cavity is filled to a low-normal amniotic fluid volume with warmed lactated Ringer's containing 500 mg of nafcillin or vancomycin. The interrupted, imbricating sutures are now tied (c). The maternal abdominal wall and skin are closed in the usual fashion.



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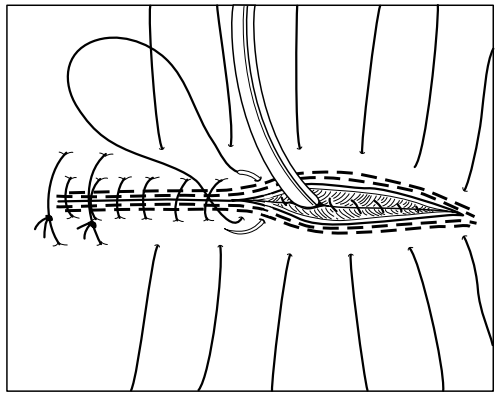

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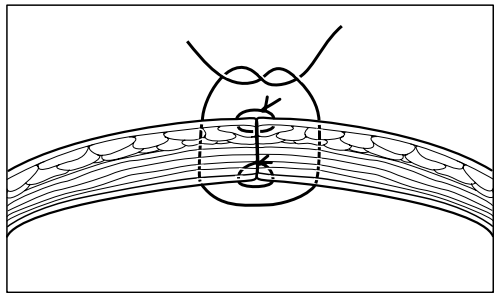

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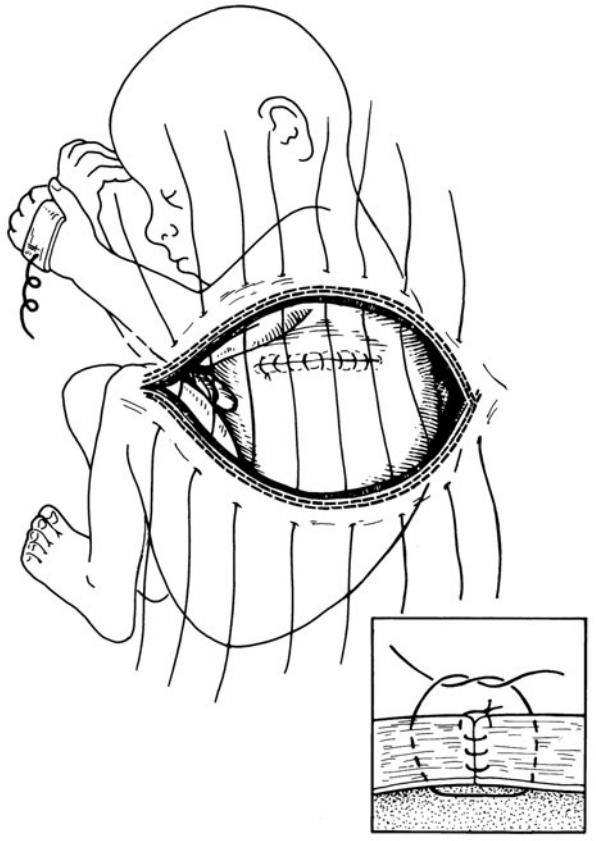
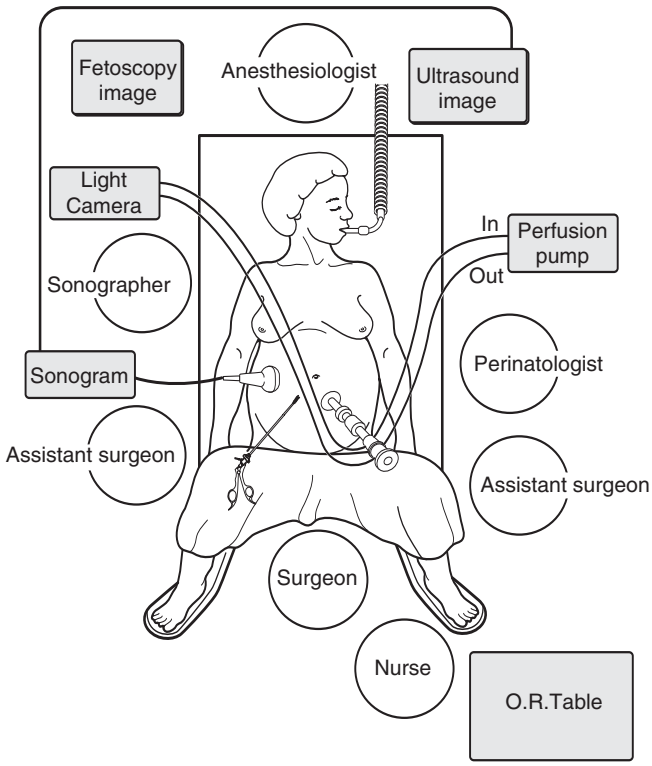


4d

Fetoscopic technique

POSITIONING

5 The procedure is performed using intravenous sedation and an epidural catheter. If necessary, general anesthesia is used. The patient is placed in low lithotomy position and two monitors are used: one for the fetoscopic image, and the other for intraoperative ultrasonography. Intraoperative ultrasound is used to confirm placental position, umbilical cord insertion (if applicable), and the location of the fetus or twins.



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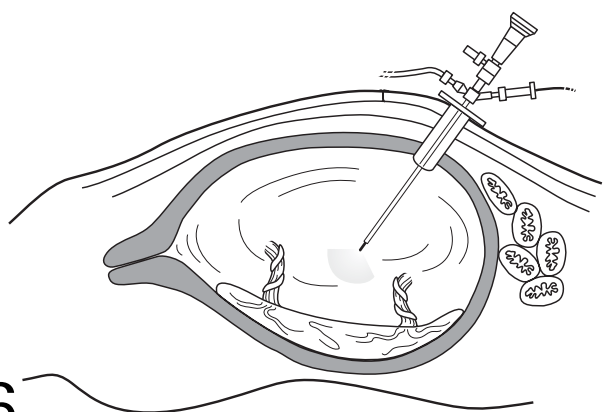


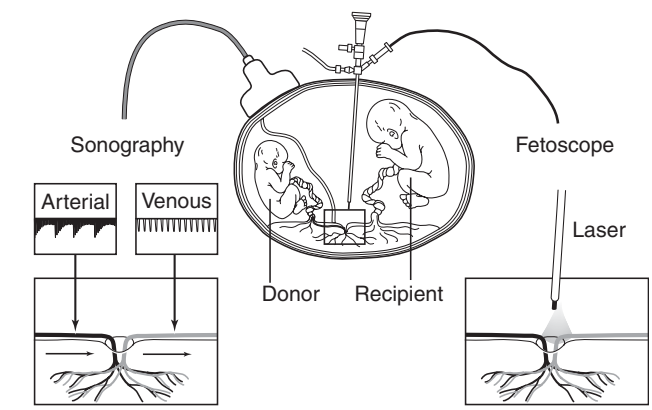
6

TROCAR INSERTION

6 A small (4 mm) incision is made in the skin to allow ultrasound-guided placement of a 3 mm trocar into the amniotic cavity. An irrigating hysteroscope is used to exchange the turbid amniotic fluid with clear lactated Ringer's solution in order to improve visualization. A level I infuser or an extracorporeal membrane oxygenation (ECMO) circuit can be used as the irrigating pump.

PROCEDURE

7 For cases of severe twin–twin transfusion syndrome, the putative unpaired placental vessels need to be coagulated. Through the side port of the hysteroscope, a 600-μ laser fiber is inserted and unpaired vessels along the vascular equator are coagulated. The port is removed and the skin closed. Amniotic fluid leak is rare with ports ≤ 3 mm in diameter.



7

Exit strategy

The EXIT procedure is designed to achieve cardiorespiratory stabilization while maintaining uteroplacental blood flow. It is most commonly used for fetuses with potential airway obstruction due to a neck mass such as a teratoma or lymphangioma. It is similar to a Cesarean delivery except myometrial bleeding is controlled and the umbilical cord is not cut until an airway is obtained – by orotracheal intubation, tracheostomy, or mass resection followed by intubation or tracheostomy.

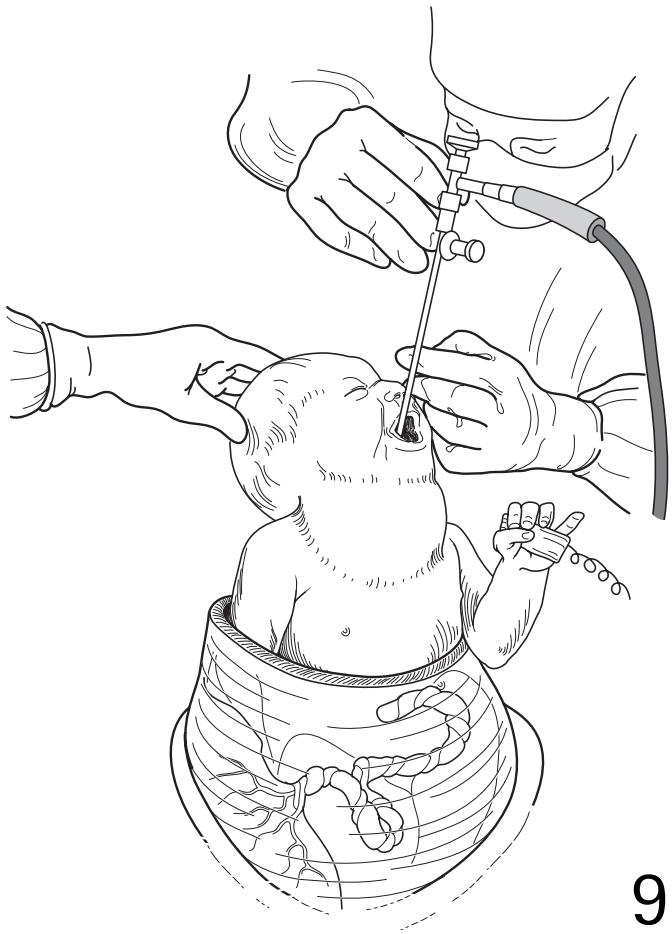
8

INCISION AND FETAL EXPOSURE

8 As described for open hysterotomy surgery, a low transverse skin-crease incision is used. If the operation is performed in the late third trimester and the placenta is posterior or fundic, the lower uterine segment is opened after creating a bladder flap. A hemostatic hysterotomy is created using the uterine stapler. An anterior or previa placenta often necessitates moving the uterus out of the abdomen/pelvis. Intraoperative ultrasonography is critical to map placental position. Two applications of the uterine stapler are usually necessary for an adequate opening. Only the necessary fetal part(s) are removed in order to maintain uterine volume and avoid vigorous contractions and placental separation. In the case of a cervical teratoma, the head, shoulders, and one upper extremity are exteriorized. A sterile pulse oximeter is attached to the palm of the fetal hand. The fetal eyes are covered with a warm, wet laparotomy pad. The fetus is continuously bathed in warm saline. Care is taken to avoid manipulation or unnecessary exposure of the umbilical cord in order to avoid spasm of its vessels.

SECURING THE AIRWAY

9 A variety of sterile instruments need to be available, such as a laryngoscope with at least two different-sized blades, extra bulbs (batteries are not sterilized and are inserted separately), two sizes of a rigid bronchoscope, a light cord, various endotracheal (some with surfactant adapters) and tracheostomy tubes, endotracheal tube stylettes, a hand-bag device with a manometer and sterile tubing that is passed off the field to an oxygen source, a sterile neonatal stethoscope, and a sterile syringe filled with surfactant (if necessary). A mixture of a narcotic (fentanyl) and a paralytic agent (pancuronium) is administered intramuscularly (shoulder) to the fetus immediately after the hysterotomy. During the EXIT procedure, direct endotracheal intubation is attempted in all cases via direct laryngoscopy, and often using rigid bronchoscopy as an adjunct. Failing intubation, further neck manipulation for tracheotomy or ECMO cannulation can be undertaken. There is the possibility that the trachea cannot be intubated transorally or via a tracheostomy due to a huge mass. In this situation, resection of the mass is the only alternative. Operations as long as 3 hours have been performed on placental support. After the airway is secure, fetal vascular access cannulae can be placed prior to cord clamping.



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10

DELIVERY

10 After the airway is obtained and secured, the umbilical cord is clamped and divided and the child is taken to the resuscitation table by the neonatologist. The placenta is delivered and the uterus closed in the standard fashion. Oxytocin is administered immediately prior to clamping the umbilical cord to enhance uterine tone.

POSTOPERATIVE CARE

Successful postoperative management is predicated on good perioperative analgesia. Continued uterine relaxation is facilitated by exquisite postoperative pain control with an epidural catheter. For most open fetal cases, the tocolytic magnesium sulfate is begun via bolus loading dose during the closure of the hysterotomy, and a continuous infusion of 2–4 g/h is continued into the recovery period with the dose titrated to the desired effect while monitoring for signs of magnesium toxicity such as maternal pulmonary edema. Indomethacin, begun preoperatively, is also continued postoperatively for 48 hours. Daily fetal echocardiography is used to monitor the fetus for ductus arteriosus constriction, tricuspid regurgitation with right side heart failure, or oligohydramnios as side effects of indomethacin treatment. Finally, continuous fetal and uterine monitoring is established by external tocodynamometer and daily routine ultrasonographic assessment.

Postoperatively, preterm labor occurs in 100 percent of patients, but to a varying degree. Indomethacin, magnesium sulfate, and terbutaline are the mainstays of postoperative tocolysis. Before discharge home, maintenance tocolysis is instituted in the form of oral nifedipine or a terbutaline pump. After open hysterotomy surgery, patients are on bedrest with bathroom privileges for the remainder of the pregnancy. This is not necessary after fetoscopic surgery and the EXIT procedure.

OUTCOME

For the fetus, the risk of the procedure is weighed against the benefit of correction of a lethal or debilitating defect. However, the risks and benefits for the mother are more difficult to assess. Maternal safety is paramount, since most fetal malformations do not directly threaten the mother's health. However, she must bear significant risk and discomfort from the surgical procedure and the postoperative tocolytic therapy. There have been no reported maternal deaths and few postoperative maternal complications, but there has been considerable morbidity primarily related to preterm labor and its treatment.

Maternal–fetal intervention carries the risk of short-term morbidity in the form of bleeding, wound infection, amniotic fluid leak, oligohydramnios, preterm labor, premature rupture of membranes, amniotic band syndrome, chorioamnionitis, placental abruption, complications of tocolytic therapy (e.g., pulmonary edema from magnesium sulfate), deep venous thrombosis, and pulmonary embolism. Long-term risk has been more difficult to assess. After a hysterotomy that is not in a well-developed lower uterine segment (this includes virtually all of the mid-gestation hysterotomy procedures), the risk of uterine rupture before and during labor is increased. This risk exists in both the current and subsequent pregnancies. Thus, in order to avoid uterine rupture, the index and all future pregnancies cannot include any period of labor and should be delivered by Cesarean section. The risk of infertility related to maternal–fetal intervention appears to be low, but an increased risk cannot be excluded.

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96

Principles of transplantation: kidney

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HISTORY

Successful renal transplantation in human identical twins was first achieved by Joseph Murray and his colleagues at the Peter Bent Brigham Hospital in Boston a half century ago. This accomplishment was recognized by the award of a Nobel Prize to Murray in 1990, by which time living-related and deceased donor renal transplantations in infants and children were routinely performed as the optimum therapy for end-stage renal disease. While advances in immunosuppression have benefited all patients, young pediatric recipients now have the highest success rate of any age subgroup undergoing renal transplantation.

PRINCIPLES AND JUSTIFICATION

Any pediatric patient with end-stage renal disease should be actively considered for renal transplantation, because renal replacement therapy using non-transplant options is associated with significant delay in growth and development. Even infants and neonates, previously reported to have poor outcomes, have been found recently to do extremely well from medical as well as development perspectives. Adequate recipient size is one of the few absolute physiologic prerequisites for operation. In order to permit an adult donor kidney to fit comfortably in the recipient's abdominal cavity, an arbitrary weight of 6–7 kg is sought. Absolute contraindications to renal transplantation include pre-existing malignancy, systemic infection that cannot be eradicated (including human immunodeficiency virus), and absence of patent major abdominal vasculature for anastomosis.

PREOPERATIVE ASSESSMENT AND PREPARATION

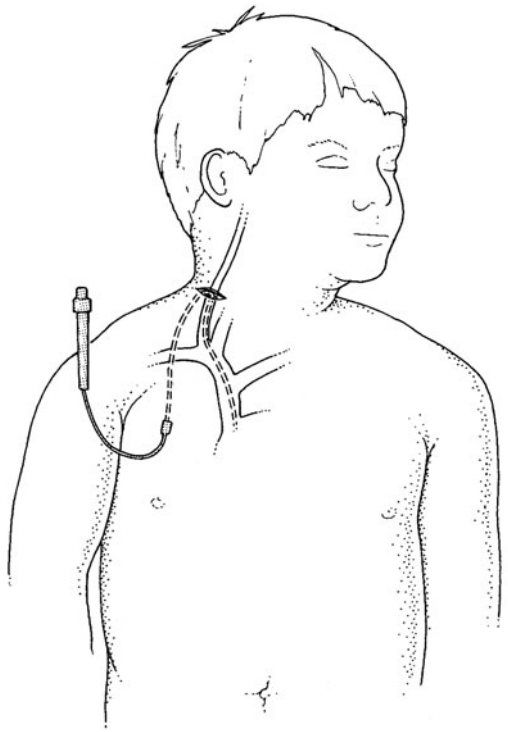
A team approach including the transplant surgeon, pediatric nephrologist, social worker, nurse specialist, and transplant coordinator facilitates a good outcome. Aggressive nutrition and dialysis, usually by a peritoneal method such as nocturnal cycling or intermittent ambulatory peritoneal dialysis, is required to allow growth of small infants with renal failure. If possible, childhood immunizations should be administered before transplantation. The child must be free of acute infectious illness at the time of transplantation.

Unlike adult recipients, children are much more likely to have an inadequate lower urinary tract due to obstructive uropathy or congenital genitourinary abnormalities. An evaluation of the lower urinary system should be complete, and inadequacies must be identified and corrected before transplantation. Strategies for achieving a satisfactory lower urinary tract may include training for intermittent self-catheterization, bilateral native nephroureterectomy, bladder augmentation (usually with colon), or construction of an intestinal conduit.

Adequate social support must be in place. Postoperative outpatient visits and occasional re-admissions to hospital are the rule rather than the exception. Transportation arrangements must be secure. The child's caregivers must have a thorough understanding of the warning signs of post-transplant infection and organ dysfunction.

Anesthesia

General anesthesia for renal transplantation follows the usual principles for major abdominal procedures in children. Attention to intraoperative fluid management and maintenance of core body temperature are particularly critical. The child should be given warmed intravenous fluids, and a ventilator with a temperature-regulated humidifier should be used. Monitoring of arterial blood pressure is mandatory for infants and children weighing less than 15 kg, and central venous pressure should be monitored for all pediatric renal transplant recipients regardless of age or size.



1

OPERATION

Position of patient

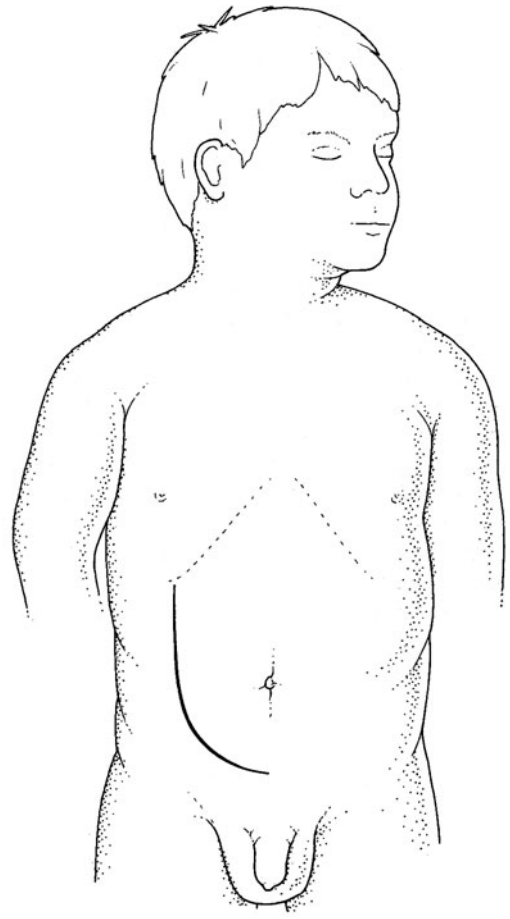
Renal transplantation is performed with the patient in the supine position. A urinary catheter is placed into the bladder or urinary reservoir and connected to a three-way system to allow for instillation of povidone-iodine (Betadine) solution at the time of urinary reconstruction. The head should be extended and turned to one side for placement of a central venous catheter at the beginning of the procedure. If a peritoneal dialysis catheter is in place, any residual fluid should be drained using a sterile technique and the clamped catheter should be secured on the patient's left side. The neck, anterior chest wall, and abdomen are prepared with an iodinated solution.

Central venous catheter placement

1 At the beginning of the procedure, an indwelling multiple-lumen central venous catheter of the Broviac type should be inserted via an incision over the external jugular vein or via the percutaneous approach to the internal jugular or subclavian vein in larger children. The tip of the catheter should be positioned at the junction of the superior vena cava and right atrium and confirmed by fluoroscopy. The catheter should follow a gently curved course and exit on the anterior chest wall. Following insertion, the catheter is passed to the anesthetic team to be connected to an in-line central venous pressure monitoring system.

Incision

2 A curvilinear incision on the right abdomen, which will allow access to the retroperitoneum, major vessels (iliac vessels, vein cava, and aorta), and the bladder, is used. The incision starts at the midline about one finger-breadth superior to the pubic symphysis, extends laterally across the rectus abdominis muscle, and then curves superiorly to about the level of the umbilicus. In small children and infants, the incision must be carried nearly to the costal margin to permit adequate exposure.



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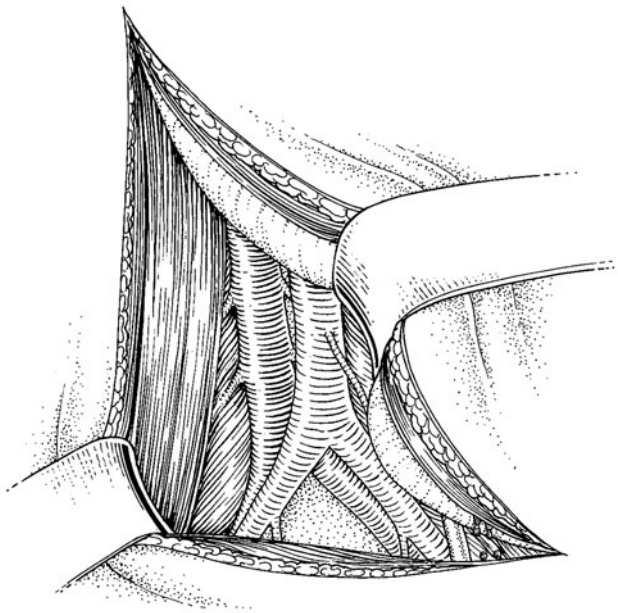


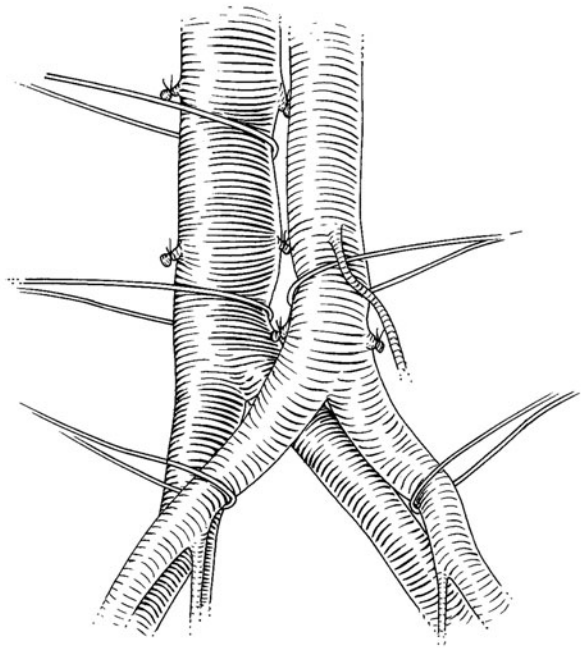
3

Exposure

3 The external oblique, internal oblique, and transversus abdominis muscles are divided, as is the rectus abdominis medially. Extreme care should be taken to avoid inadvertent entry into the peritoneal cavity, which is most likely at the most superior and inferior aspects of the dissection, particularly if there have been previous episodes of peritonitis. Any openings in the peritoneum should be closed with absorbable sutures to prevent leakage of residual dialysis fluid into the operative field. The peritoneum is swept medially and superiorly to expose the iliac vessels, inferior vena cava, and distal aorta. The inferior epigastric vessels are usually ligated and divided medially. The spermatic cord should be conserved by careful dissection and medial retraction. In girls, the round ligament should be divided. At this point, a self-retaining Bookwalter rectractor system should be assembled to establish and maintain exposure for the remainder of the procedure.

Vascular dissection

4 The inferior vena cava and distal aorta should be used for vascular reconstruction in children weighing less than 15 kg. Circumferential dissection of the distal vena cava is accomplished first. Lumbar veins are divided between fine silk ligatures to free a segment of vena cava approximately 2–3 cm in length. The distal aorta and proximal common iliac arteries are controlled with rubber vessel loops. The inferior mesenteric artery does not usually need to be divided. At least one set of lumbar arteries is generally encountered. Care should be exercised during dissection of the aorta to avoid disruption of the abdominal lymphatic trunk.



4

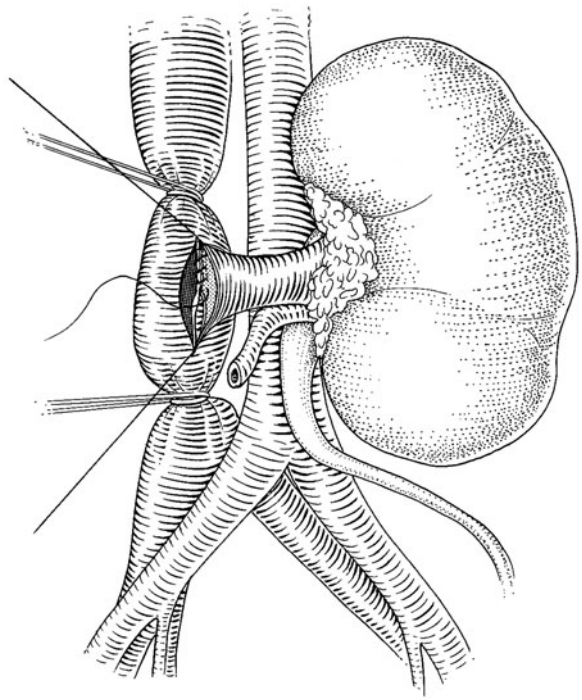
Preparation of donor kidney

Before the administration of heparin and clamping the vessels, the donor kidney is brought to the operative field in a bath of sterile iced saline solution. The renal artery and vein are cleaned of surrounding tissues and side branches are secured. The renal vein must be kept short, usually not longer than 1–1.5 cm, to prevent kinking. If the donor kidney is of deceased donor origin, a decision must be made about whether or not to utilize the entire length of the renal artery with a Carrel patch of donor aorta. Use of the Carrel patch will usually result in a renal artery that is too long unless the recipient weighs more than 30 kg. The renal artery from a living donor kidney should be spatulated. The kidney may be wrapped in a moistened laparotomy pad with a 'keyhole' fashioned for the vessels. This facilitates handling of the kidney in the wound.

Vascular anastomoses

The recipient is given heparin, 50–70 units/kg body weight. Full-dose heparinization is unnecessary, except in cases of known hypercoagulability. Vascular clamps should be carefully chosen to avoid obscuring the field. In infants, tension on double-looped rubber vessel slings is all that is required. This technique has the advantage of elevating the vessels in the field. During construction of the vascular anastomoses, the anesthetic team should volume load the recipient to a central venous pressure of 15–18 cmH_2O and administer mannitol, 1.0 g/kg. These maneuvers will counteract the effects of revascularization and its attendant destabilizing effect due to volume shift. A transplanted adult kidney may account for 20 percent of the circulating blood volume of a 7-kg infant.

5 A longitudinal incision is made in the vena cava and 6/0 or 7/0 polypropylene sutures are placed at the apices. The assistant positions the kidney medially in the wound. The medial side of the anastomosis is performed first from within the lumen of the vein, followed by the lateral side from the outside. Following completion of the venous anastomosis, the kidney is rotated toward the operator's side.



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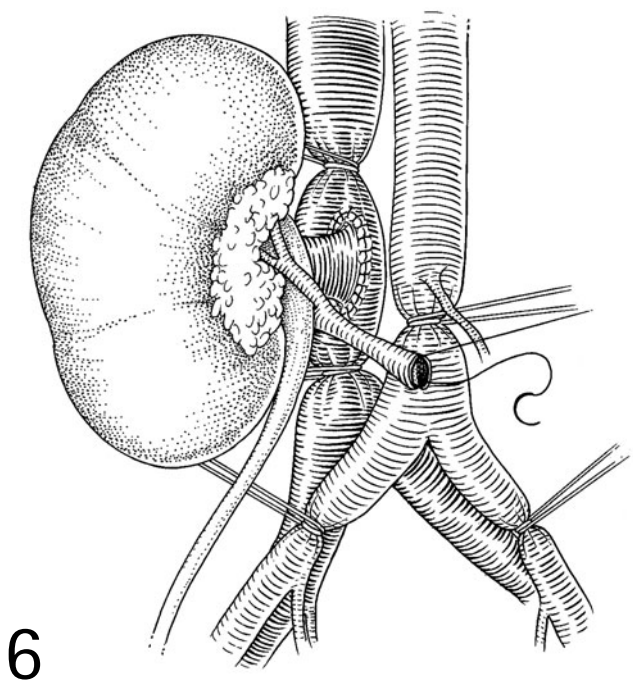


6

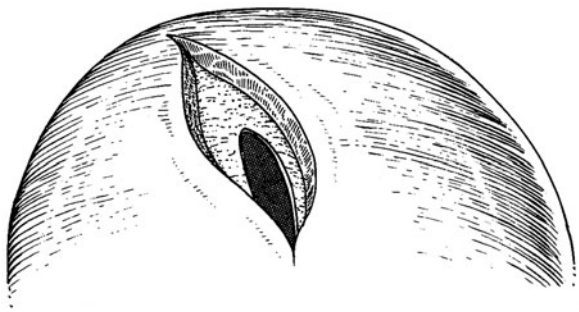
6 The aorta and common iliac arteries are occluded and an aortotomy is made at a site carefully chosen for optimal geometry of the vessels. A 4 mm aortic punch is used to fashion an orifice on the anterior surface of the aorta. This anastomosis is started with a running 6/0 polypropylene suture at the superior aspect. The 'back wall' of the anastomosis is performed from the inside and the 'front wall' from the outside.

Revascularization ensues at the completion of the vascular anastomoses. Heparin need not be reversed with protamine sulfate unless troublesome bleeding occurs. The kidney should be carefully inspected. Optimally, a uniform pink color and normal turgor are followed promptly by the production of urine. Engorgement of the kidney suggests venous obstruction. This may be due to an imperfect anastomosis, but is more commonly attributable to compression of the vena cava by one of the retractor blades. An excellent pulse should be palpable in the renal hilum and a thrill is quite common. Following establishment of hemostasis and assuming satisfactory appearance of the kidney, attention is then turned to the urinary reconstruction.

External ureteroneocystostomy

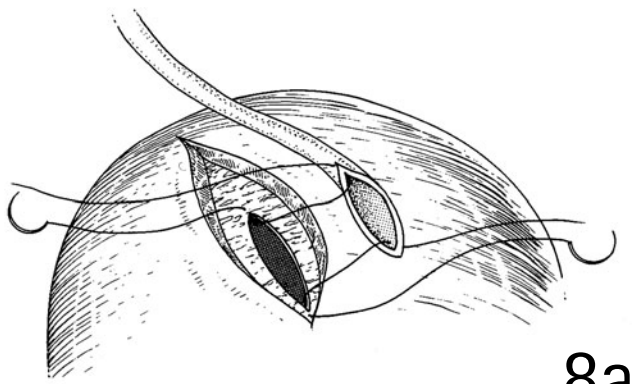
If the bladder is to be used for urinary reconstruction, the urinary catheter is clamped and povidone-iodine solution is instilled to distend the bladder and sterilize its contents. The author's preferred technique is an external ureteroneocystostomy, and the results with this method have been previously reported. The principles of this technique include direct anastomosis of the ureter to bladder mucosa and construction of a submuscular tunnel to prevent reflux into the transplanted kidney.

A site is chosen anteriorly near the dome of the distended bladder. The choice of site should take into account the course of the transplanted ureter. An appropriate balance must be struck between the need for an adequate ureteric length to reach the anastomotic site when the bladder is empty and the requirement of avoiding excessive redundancy with the attendant risk of ureteric obstruction and distal ureteral ischemia. Keeping in mind that the blood supply to the transplanted ureter is completely dependent on small branches from the lower pole renal artery, the tendency should be to err on the side of a shorter ureter.



7

7 The muscular coat of the bladder is divided sharply and small vessels are carefully electrocoagulated. The bladder mucosa is exposed over an area of about 4 cm^2. An ellipse of mucosa is excised at the distal side of the mucosal dissection. The presence of the bladder catheter should be confirmed visually at this time; a thickened peritoneum adjacent to the bladder can fool even the most experienced surgeon.



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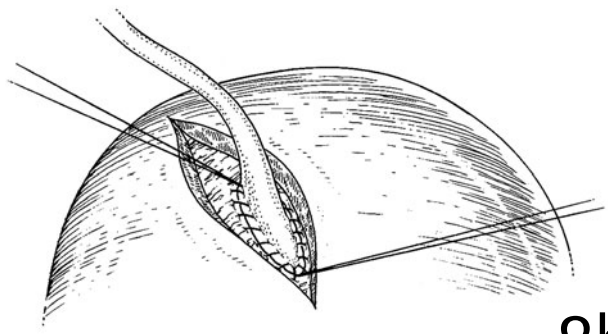
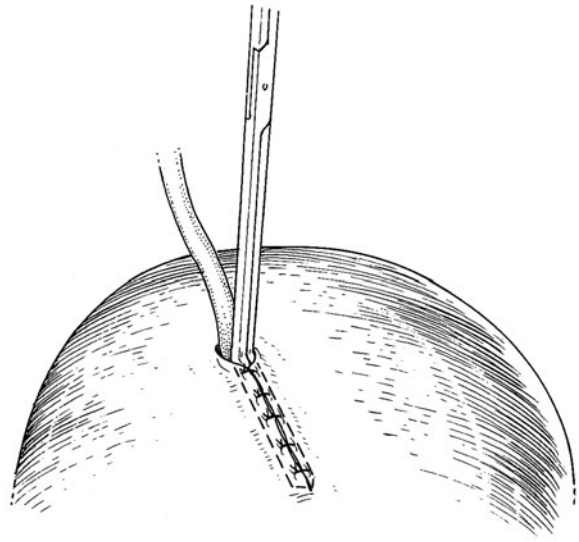


8b

8a,b After sizing and spatulation of the transplant ureter, the anastomosis is constructed with running 5/0 monofilament absorbable sutures. Traction sutures are placed at the 'heel' and 'toe' of the anastomosis. Careful placement of sutures and avoidance of excessive ureteric handling are critically important to avoid stenosis or obstruction. Only the smallest of bites should be taken on the ureter. Construction of the anastomosis should proceed from 'toe' to 'heel' and then back to the 'toe' on the opposite side. A silastic double-J stent may be placed if there is any question about the integrity of the reconstruction. It should be placed after the anastomosis is partially completed, with the upper end positioned in the renal pelvis and the lower end passed into the bladder.

9 On completion of the anastomosis, the ureter is laid in the submuscular space and the bladder muscle is closed over it using interrupted 4/0 monofilament absorbable sutures over a distance of 1–2 cm. When completed, the tunnel should still admit the end of a right-angled clamp, thus ensuring that the ureter will not be obstructed within the tunnel.



9

Wound closure

The transplanted kidney should lie comfortably within the iliac fossa. In extremely small infants (< 7 kg) it may be necessary to open the peritoneum and convert the transplant to an intraperitoneal allograft, but this is required in less than 10 percent of cases. No fixation of the kidney allograft is needed. On removal of the self-retaining retractor, the peritoneal contents will hold the kidney against the iliopsoas and false pelvic side wall. No drains are used. The wound is irrigated and a two-layer fascial closure is completed with interrupted, nonabsorbable sutures. Absorbable suture material should be avoided for the fascial closure because of the deleterious effects of immunosuppressive agents on wound healing. No sutures are placed in the subcutaneous tissues and the skin is closed with a running, absorbable, subcuticular technique.

POSTOPERATIVE CARE

General care and infection prophylaxis

Infants and small children are managed in a pediatric intensive care unit. Although most recipients are extubated in the operating room, a small infant with an adult-sized kidney may require mechanical ventilation for 1–2 days. The urinary catheter is left in place for 3–5 days. Satisfactory bladder emptying may be assured, if necessary, with occasional post-micturitional catheterization. Perioperative prophylaxis against bacterial infection should be given. Prophylaxis is given for 1 month against opportunistic infection. This includes trimethoprim and sulfamethoxazole for *Pneumocystis carinii*, acyclovir for herpes virus, and nystatin for candidiasis. Patients at high risk of *Cytomegalovirus* infection (donor serology positive or recipient serology negative) are given ganciclovir or valganciclovir instead of acyclovir. The mean hospital stay is 7–10 days for uncomplicated cases.

Immunosuppression

Immunosuppressive therapy is commenced immediately before operation with corticosteroids and azathioprine. After operation, induction immunotherapy is given with polyclonal or monoclonal antilymphocyte antibody or monoclonal antibody against the interleukin-2 receptor. A calcineurin inhibitor (either cyclosporine or tacrolimus) is begun when renal allograft function is satisfactorily recovered in order to avoid the nephrotoxic effects of these agents. A deliberate steroid taper is pursued over several months and many pediatric patients can eventually be transferred to alternate-day steroids. Long-term maintenance immunosuppression most commonly consists of a three-drug regimen including calcineurin inhibitor, antimetabolite (usually mycophenolate mofetil), and steroids.

OUTCOME

The overall results of renal transplantation in children have improved steadily in recent years. The most important advances have included aggressive pretransplant dialysis and nutritional management, meticulous surgical technique, and concentration-controlled immunosuppression. Transplant and developmental outcomes have been excellent even among recipients less than 1 year of age with renal failure. Results with living donors in children aged 1–5 years have been particularly good. In the United States, the 5 year patient

survival rate for this group is 96 percent and the 5 year graft survival rate is 84 percent.

FURTHER READING

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97

Principles of transplantation: liver

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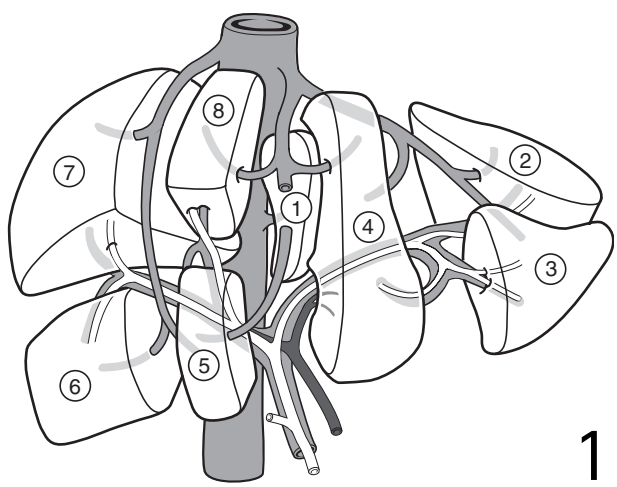
HISTORY

The first successful human liver transplant was performed in 1967 by Thomas Starzl in Denver, Colorado. The introduction of better immunosuppressants such as ciclosporin in 1979 and tacrolimus a decade later, refinements in surgical technique, and advances in organ preservation have each contributed to progressively better outcomes. Although the availability of deceased donor grafts improved as a result of public awareness and national systems of organ sharing and distribution, there was a universal shortage of size-matched donor organs, particularly for small children, who deteriorated or died on the waiting list. To tackle this problem, surgical techniques were devised to enable adult donor livers to be used in small children.

These techniques rely on the segmental anatomy of the liver. *Reduced* liver grafts are prepared by cutting down a deceased donor adult liver to produce a left lateral segment (segments 2 and 3) or left lobe graft (segments 2, 3, and 4) and were introduced in 1984. *Split-liver* transplantation, in which a deceased donor adult liver is divided into two functional grafts, became available in 1988. Pediatric *living-donor* liver transplantation, in which a healthy parent donates part of his or her liver to his or her child, followed in 1989. Living related donation enabled the development of pediatric liver transplant programs in countries without established deceased donor organ systems and has since been extended to adult recipients through the use of right lobe donation.



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PRINCIPLES AND JUSTIFICATION

Indications

The main indications for pediatric liver transplantation are listed in Box 97.1. The main categories are:

- progressive chronic liver disease with incipient liver failure;
- chronic liver disease with secondary major morbidity;
- selected hepatic-based metabolic disorders;
- acute liver failure.

Box 97.1 *Indications for pediatric liver transplantation*

- **Acute liver failure**
  Viral, e.g., hepatitis A, B, and non A-E
  Drugs/toxins
  Metabolic
  Autoimmune
- **Chronic liver disease**
  Cholestatic liver disease
  Biliary atresia
  Progressive familial intrahepatic cholestasis
  Biliary hypoplasia, e.g., Alagille syndrome
  Sclerosing cholangitis
  Metabolic liver disease
  α-1-Antitrypsin deficiency
  Wilson's disease
  Cystic fibrosis
  Tyrosinemia
  Crigler–Najjar (type 1) syndrome
  Urea cycle defects
  Hyperoxaluria
  Neonatal iron storage disorders
  Autoimmune hepatitis
  Liver tumors, e.g., unresectable hepatoblastoma confined to the liver

The commonest indication for liver transplantation in children is biliary atresia, which accounts for at least 50 percent of cases. Those with progressive liver disease suffer from worsening jaundice, recurrent cholangitis, portal hypertension, ascites, and malnutrition. Other, rarer, indications for liver transplantation in children with chronic liver diseases include encephalopathy, metabolic bone disease, intractable pruritus, and hepatopulmonary syndrome.

Inborn errors of metabolism resulting from partial or complete deficiencies of specific liver enzymes or abnormal products of hepatic synthesis constitute the next commonest indication for liver transplantation in children. In many conditions, the disease process damages the liver and transplantation is prompted by hepatic decompensation or the development of a malignant tumor. Alpha-1-antitrypsin deficiency is the commonest of these inborn errors of metabolism; after liver transplantation, the phenotype of the recipient becomes that of the donor, and the α-1-antitrypsin level is restored to normal. Wilson's disease is a genetic disorder of copper metabolism with excessive copper deposition in the liver, basal ganglia, kidney, and other organs. Most cases can be successfully treated medically, but some patients present acutely with liver failure and need an urgent liver transplant. In the future, new specific drug treatments, gene therapy, or hepatocyte transplantation may offer better treatment for some of these hepatic-based metabolic disorders.

The decision to perform a liver transplant in a child with acute liver failure from viral or toxic hepatitis can be very difficult, because a proportion of these cases can recover with supportive therapy alone. A poor prognosis is predicted by progressive encephalopathy, a persistent coagulopathy (International Normalized Ratio (INR) > 4), increasing bilirubin (> 300 μmol/L), cardiovascular instability, and a rapidly shrinking liver. In such patients, liver transplantation provides the best chance of survival, but must be performed before the onset of irreversible neurologic damage. Bioartificial support systems may provide a useful holding measure in some cases.

Contraindications to liver transplant include persistent extrahepatic malignant disease, severe extrahepatic disease that cannot be reversed by liver transplantation, uncontrolled systemic sepsis, severe irreversible neurologic injury, and a severe chronic psychosocial disorder. The ability to adhere to long-term immunosuppression must also be considered. Anatomical constraints such as an interrupted inferior vena cava, isolated portal vein thrombosis, and abdominal situs inversus require planned technical modifications but are *not* contraindications.

Timing

In children with chronic liver disease, transplantation is indicated as soon as progressive deterioration is noted but before the onset of life-threatening complications. A persistently elevated plasma bilirubin (> 150 μmol/L), low albumin (< 35 g/L), and prolonged prothrombin time (INR >1.4) are useful laboratory markers. Clinical features of hepatic decompensation may include growth failure, malnutrition, variceal bleeding, or refractory ascites.

PREOPERATIVE ASSESSMENT AND PREPARATION

Detailed multidisciplinary evaluation is required to:

- assess the severity and effects of the liver disease (including the possibility of occult hepatic malignancy);
- determine the technical feasibility, urgency, and appropriateness of transplantation;
- identify and, if possible, correct metabolic, nutritional, and immunization deficiencies;

- assess the psychosocial impact of transplantation on the family and educate them about the procedure and the long-term sequelae.

A broad range of investigations is needed (Box 97.2). Evaluations are performed by the pediatric hepatologist and transplant surgeon, anesthetist, radiologist, nutritionist, psychologist, social worker, physiotherapist, specialist liver nurse, and transplant coordinator.

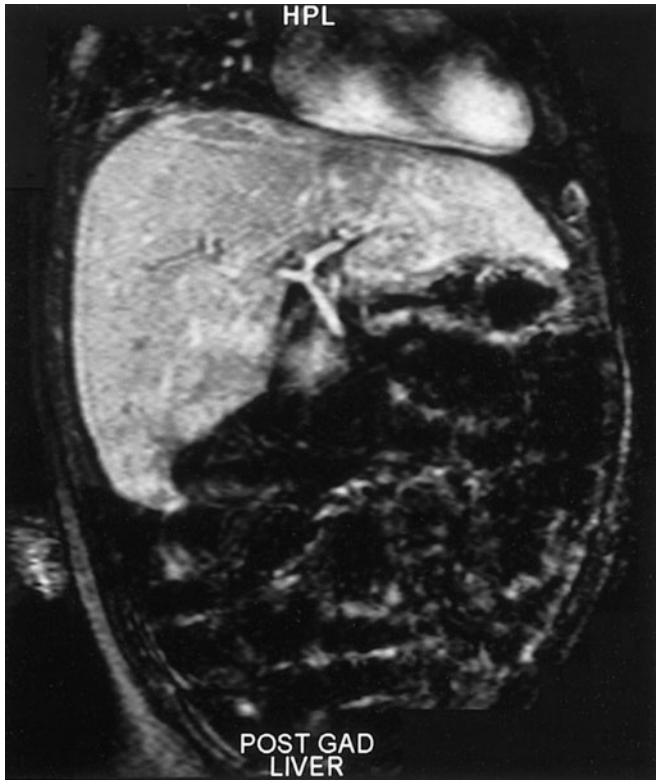
Malnutrition is common because of malabsorption, anorexia, and catabolism. Nutritional status is a major determinant of outcome. Aggressive nutritional support, including nasogastric or even parenteral nutrition, may be needed. Bleeding esophageal varices are treated, preferably by variceal band ligation. Bacterial infection is common in children with end-stage liver disease and must be diagnosed and eradicated prior to transplant. Viral screening should include cytomegalovirus (CMV) and Epstein–Barr virus (EBV) serology, both of which are important potential post-transplant pathogens. Patients should be immunized against *Pneumococcus* and hepatitis A and B in addition to routine vaccines. Live vaccines such as Bacille Calmette–Guérin (BCG) and varicella are best given at least 2 months before transplant, as they are generally contraindicated in the immunosuppressed.

Box 97.2 *Investigation of the pediatric liver transplant recipient*

- Complete history and physical examination, including assessments of growth, nutrition, development
- Hematology – full blood count, clotting, blood group
- Biochemistry – urea, electrolytes, creatinine, liver function tests, glucose, lactate, bone profile, magnesium, lipids, vitamin levels, urine biochemistry
- Immunology – immunoglobulins, autoantibodies
- Microbiology – urine culture, surface swabs
- Viral serology – hepatitis A, B, and C, Cytomegalovirus, Epstein–Barr virus, herpes simplex virus, Varicella zoster virus, human immunodeficiency virus
- Serum α-fetoprotein
- Chest radiograph
- Abdominal ultrasound scan and Doppler studies of hepatic vessels
- Magnetic resonance scan/angiogram (selected cases)
- Echocardiogram and ECG
- Respiratory function tests $\pm$ blood gases
- Glomerular filtration rate
- Dental review

2 Ultrasonography is useful in determining the patency and size of the portal vein. Failure to visualize satisfactory hepatopetal portal blood flow is an indication for magnetic resonance angiography (MRA). This is also recommended in children with the biliary atresia–splenic malformation syndrome and in patients with tumors or the Budd–Chiari syndrome. The pretransplant MRA opposite shows a narrow preduodenal portal vein in an infant with syndromic biliary atresia.

The psychologic assessment and preparation of the child and family are important. Once the child is accepted for transplantation, priority on the waiting list is assigned according to urgency. During the waiting period, regular outpatient monitoring is essential.



2

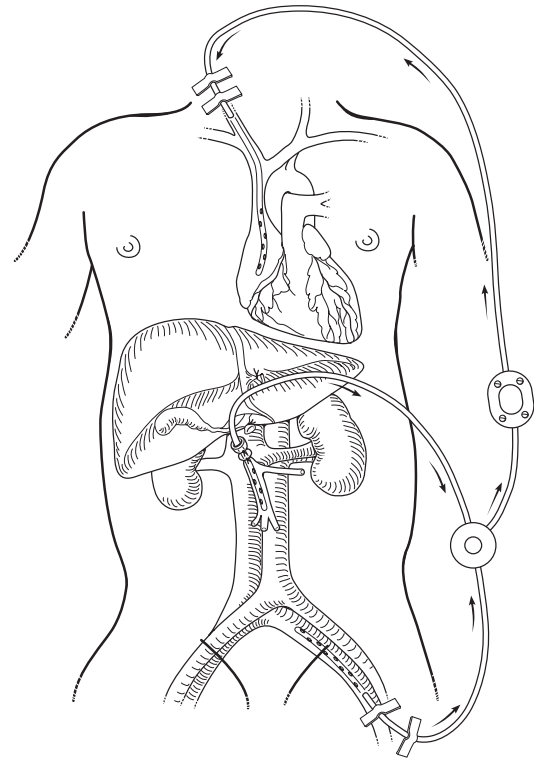
Anesthesia

Children undergoing liver transplantation often have multisystem problems related to their liver disease. Preoperative status can vary from a ventilated child with acute liver failure complicated by severe coagulopathy, cerebral edema, and acute renal failure to that of a small, malnourished infant with biliary atresia, ascites, and portal hypertension. Teamwork to anticipate and prevent complications is essential. A range of blood products including packed red cells, fresh frozen plasma, platelets, and cryoprecipitate must be available. Perioperative broad-spectrum antibiotics and antifungal prophylaxis are given.

Anesthesia is based on an infusion of a short-acting opioid supplemented with a modern volatile anesthetic agent supplied in an oxygen–air mixture. Atracurium or cisatracurium is frequently used for muscle relaxation, as these agents do not rely on hepatic metabolism or renal clearance for excretion. Nitrous oxide is avoided because of the potential for bowel distension during the long operation. A vasopressor should be available, particularly after reperfusion.

Invasive hemodynamic monitoring and central venous access for rapid infusion are supplemented by a central temperature probe, oximetry, capnography, and a urethral catheter. Newer, less invasive methods of measuring cardiac output and systemic vascular resistance are gaining popularity. Blood is analyzed regularly for gases, pH, lactate, hemoglobin and platelet count, prothrombin time, glucose, and electrolytes (including ionized calcium). Thromboelastography is a helpful additional measure of coagulation. Temperature hemostasis is aided by warm ambient temperatures, the use of radiant heaters during induction, an efficient warming mattress and warm air blanket, the warming and humidification of anesthetic gases, and warming of all intravenous fluids. Wrapping the extremities of the child in impervious reflective sheeting is also useful. Pressure areas must be well padded.

3 Venovenous bypass is not routinely used but can be useful in selected older children. Cannulas are inserted into the portal and femoral veins and diverted blood is pumped through an internal jugular vein catheter into the right atrium. This technique helps to maintain venous return and renal perfusion during caval cross-clamping, reduces portal pressure and blood loss during the recipient hepatectomy, and helps to prevent venous congestion of the gut once the portal vein is divided.

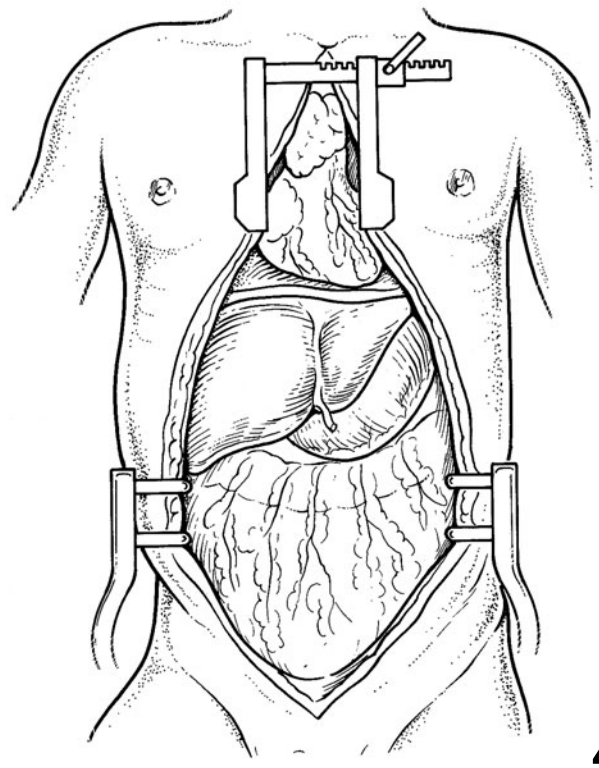


3

DONOR OPERATION

Although livers from controlled, non-heart-beating donors have recently been used in some children, the liver is typically procured from a heart-beating donor in whom brainstem death has been diagnosed by stringent criteria. Most donors for pediatric liver transplantation are aged between 1 and 50 years and have died from a spontaneous intracranial bleed or severe head injury. The donor and recipient should be ABO blood group compatible but tissue typing is not required. The donor should have near-normal biochemical liver function, no history of malignancy, and no evidence of chronic liver disease or past infection with hepatitis B or C, human immunodeficiency virus (HIV), or prions. Donor CMV and EBV serology must be recorded. Most donor livers are removed as part of a multi-organ procurement procedure in which the liver, kidneys, pancreas, heart and/or lungs are removed.

4 A midline incision is made from the suprasternal notch to the pubis. The sternum is opened in the midline using a Gigli saw and sternal retractor. The abdominal viscera are carefully inspected to exclude injury or disease. The liver is mobilized by incising its ligamentous attachments. The gastrohepatic ligament is examined for the presence of an accessory/replaced left hepatic artery arising from the left gastric artery, which, if present, must be preserved in continuity with the celiac axis.



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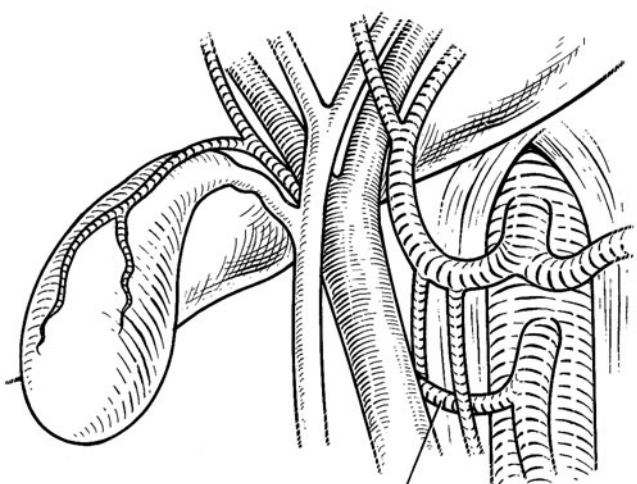
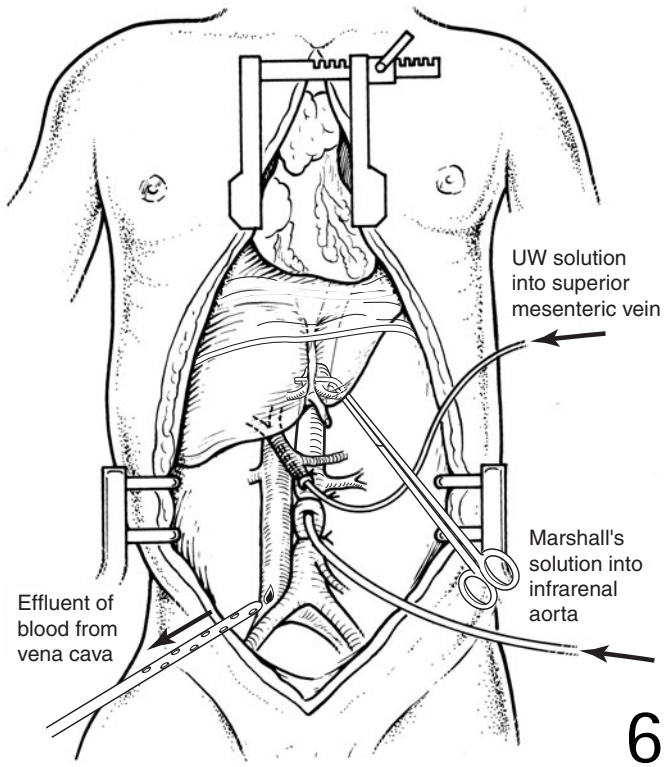


5

5 The porta hepatis is dissected next, checking for an anomalous right hepatic artery arising from the superior mesenteric artery. This usually lies behind the portal vein and must be preserved in continuity with the superior mesenteric artery.

If there is a conventional hepatic arterial supply, the gastroduodenal and right gastric arteries are divided between ligatures followed by ligation of the left gastric and splenic arteries. The common bile duct is divided close to the upper border of the duodenum. The portal vein is carefully dissected.

6 The distal aorta just above the bifurcation is encircled with two strong tapes and the aorta just above or below the diaphragm is next dissected and encircled with a tape. The superior (or inferior) mesenteric vein at the root of the small bowel mesentery is isolated. At this stage, the cardiac team begin preparing the heart and/or lungs for retrieval, after which the donor is heparinized. A large-bore cannula is then inserted into the distal aorta and connected to 1 L of cold perfusate (e.g., Marshall's solution). A smaller cannula is inserted into the superior mesenteric vein and threaded into the portal vein; this is connected to 1 L of cold University of Wisconsin (UW) solution. When all surgical dissection is completed, the diaphragmatic aorta is clamped, the infrarenal vena cava (or intracardiac cava if the heart is not being retrieved) is incised to drain blood from the donor, and cold perfusion begins. Ventilation is stopped.



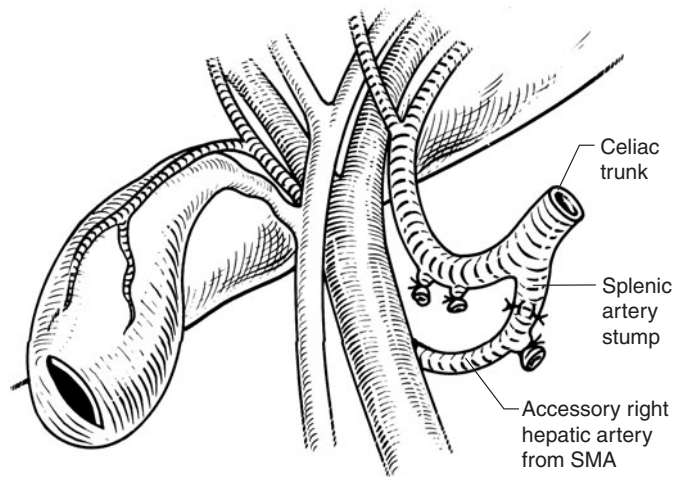
Normally, 1–2 L of Marshall's solution and 1–2 L of UW solution (50 mL/kg) are infused into the aorta and portal vein, respectively. Iced saline slush is poured over the liver and kidneys to provide additional surface cooling. The gallbladder is incised and the bile washed out with cold saline. When the organs are cold and blanched, they are removed. This requires care, as the blanched, non-pulsatile vessels are not so readily identified. The hepatic artery and celiac axis are removed en bloc with a small patch of aorta. The portal vein is retrieved with a stump of the splenic and superior mesenteric veins (assuming the pancreas is not being retrieved). The right lobe of the liver and the vena cava are mobilized, retaining the right adrenal gland with the graft. The infrahepatic vena cava is transected just above the renal veins and the suprahepatic vena cava is divided with a cuff of diaphragm.

On the bench, the liver is further perfused with UW solution via the hepatic artery and portal vein. The bile duct is gently irrigated. The liver is packed in sterile plastic bags containing cold UW solution and stored in ice for transportation. After the kidneys have been removed, the iliac arteries and veins are retrieved for any subsequent vascular reconstruction.

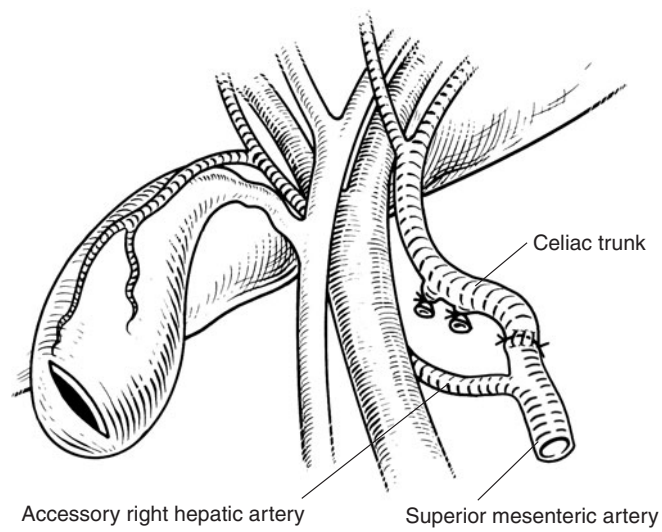
Later, during the recipient hepatectomy, the liver is cleaned and prepared on the back bench; redundant tissue is removed and the phrenic and right adrenal veins are ligated. The hepatic artery and portal vein are prepared for implantation.

Arterial reconstruction

7a,b Arterial reconstruction may be required to simplify implantation of a liver with an anomalous hepatic arterial supply. For example, a right accessory artery arising from the superior mesenteric artery can be anastomosed to the splenic stump of the celiac trunk.



7a



7b

Rapid cooling technique

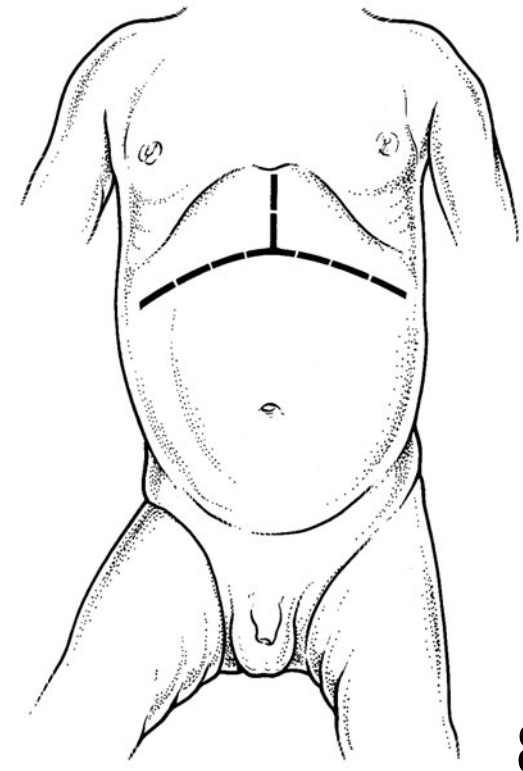
In an unstable donor, a rapid core cooling technique is used. Ice-cold UW solution is infused directly into the abdominal aorta and inferior mesenteric veins immediately after the abdomen has been opened and the donor heparinized.

RECIPIENT OPERATION (ORTHOTOPIC WHOLE GRAFT)

The recipient hepatectomy can be difficult in children with biliary atresia and portal hypertension in whom dense vascular adhesions from previous surgery may make dissection of the portal structures, duodenum, Roux loop, and transverse colon hazardous.

Incision

8 The abdomen is entered through a curved, bilateral subcostal incision. In older children, this may be extended vertically in the midline to the xiphoid process for better exposure. The abdominal muscles are divided with cautery to maintain hemostasis. The liver is exposed using an upper abdominal retractor such as an Omnitract®.



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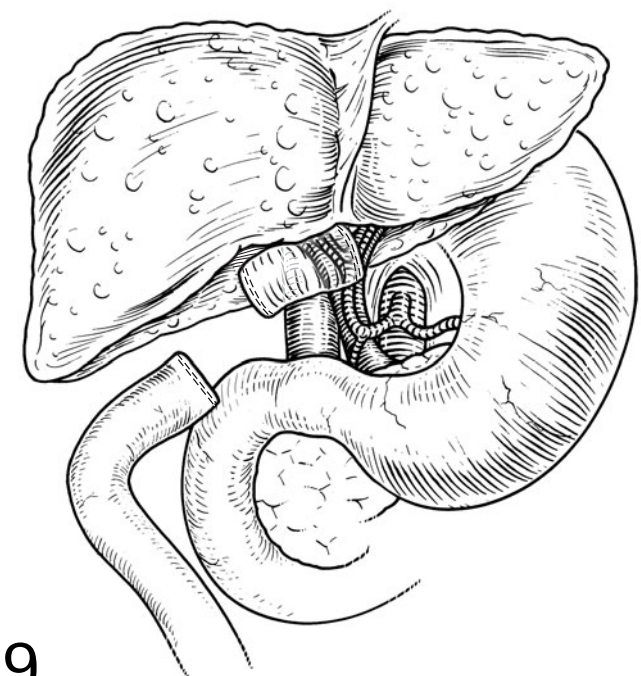


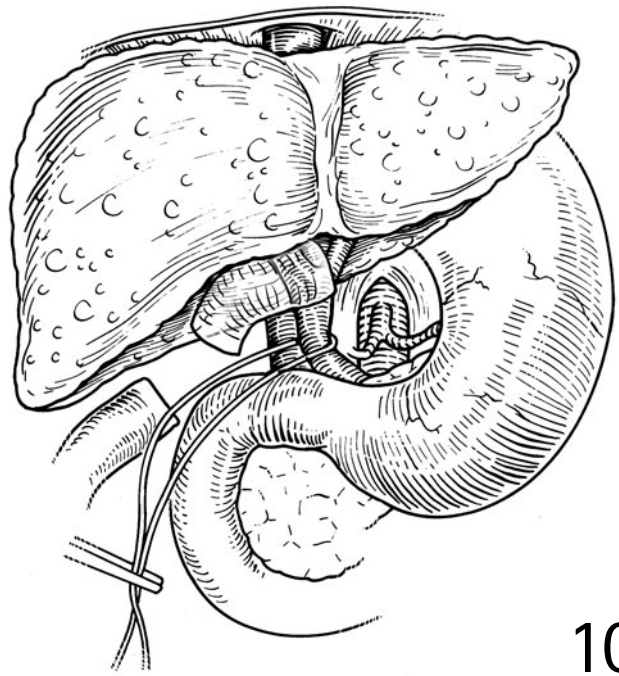
9

Hepatectomy

9 In children who have previously had a portoenterostomy for biliary atresia, the liver is separated from the abdominal wall with cautery, as the normal ligamentous attachments are poorly defined. This dissection becomes easier once freer tissue planes are reached posteriorly. The transverse colon and duodenum are carefully separated from the liver with the aid of precise bipolar cautery. The jejunal Roux loop is identified and divided with a linear stapler close to the liver hilum. The portal structures can then be dissected.

In acute liver failure, the porta hepatis is dissected before mobilizing the liver and the hepatic artery is ligated early to try to minimize hemodynamic instability.

10 In patients in whom the bile duct is present, it is divided between ligatures, leaving a good length of common bile duct. The hepatic artery is similarly divided between ligatures; preserving the bifurcation intact keeps options open for subsequent arterial anastomosis. The portal vein is dissected and isolated; if the portal vein is narrow, either the bifurcation should be preserved or its origin exposed at the confluence of the splenic and superior mesenteric veins.



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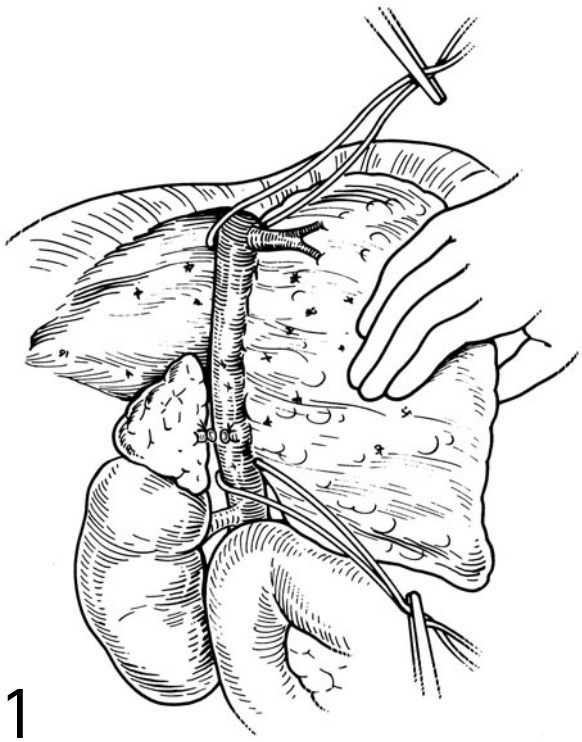
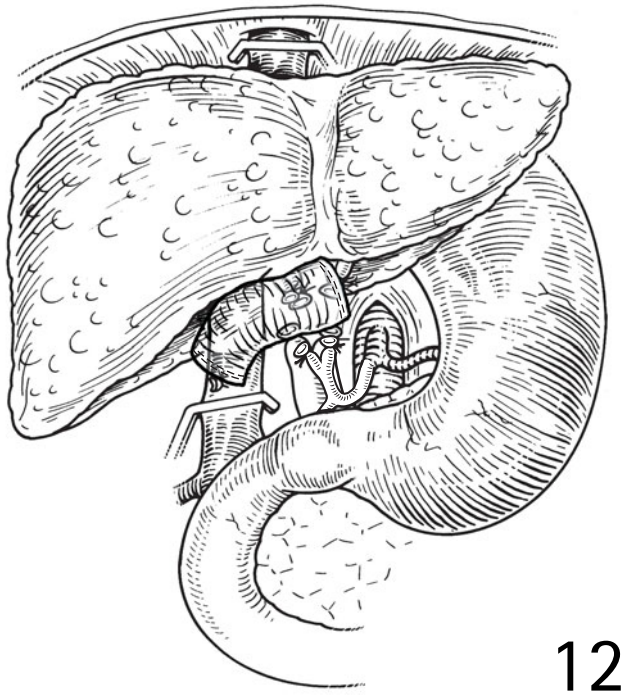


11

11 If not previously divided, the falciform and triangular ligaments are incised. The gastrohepatic ligament is divided between ligatures. The liver is retracted medially and the right lobe is dissected from the diaphragm and retroperitoneum using cautery. The infrahepatic cava is encircled above the renal veins with a tape and, using this plane as a guide, the right adrenal vein is identified and divided between ligatures. The plane behind the retrohepatic cava is carefully dissected from both sides until the suprahepatic vena cava can be encircled ready for subsequent clamping.

12 The portal vein is divided between ligatures or vascular clamps. In some older children, a portal cannula may be inserted for venovenous bypass. Both the suprahepatic and infrahepatic vena cavae are occluded with secure vascular clamps; the former is clamped with a cuff of diaphragm, avoiding the phrenic nerve.



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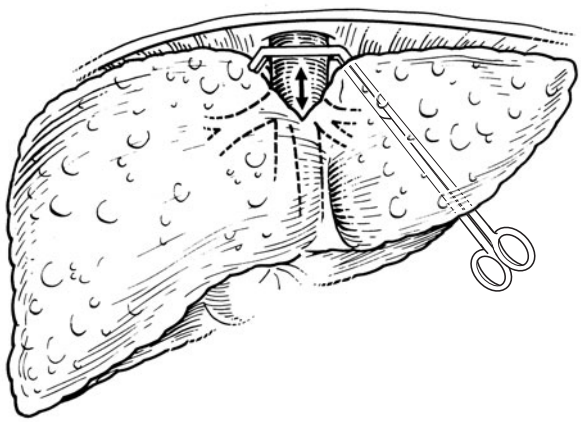
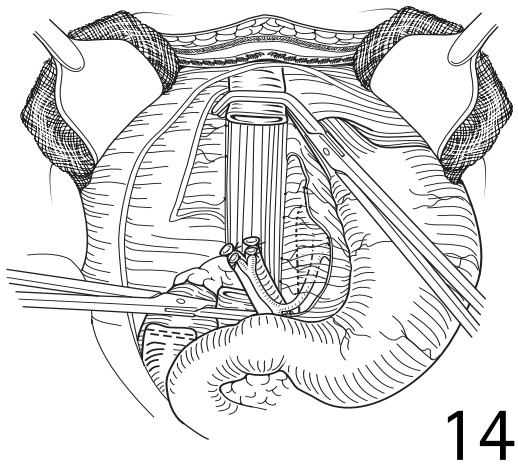


13

13 Provided the patient is hemodynamically stable, the suprahepatic vena cava is divided – extra length can be obtained by incising the liver at the confluence of the hepatic veins.

14 The infrahepatic vena cava is divided and the liver removed. Meticulous hemostasis of the retroperitoneum should be achieved with a combination of cautery, argon beam coagulation, and suture ligation. Access to this area is more difficult once the donor liver has been implanted.



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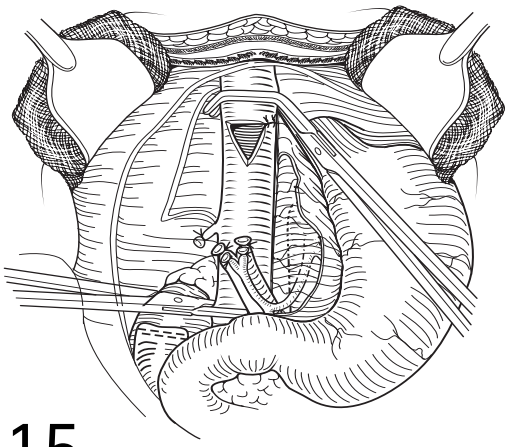
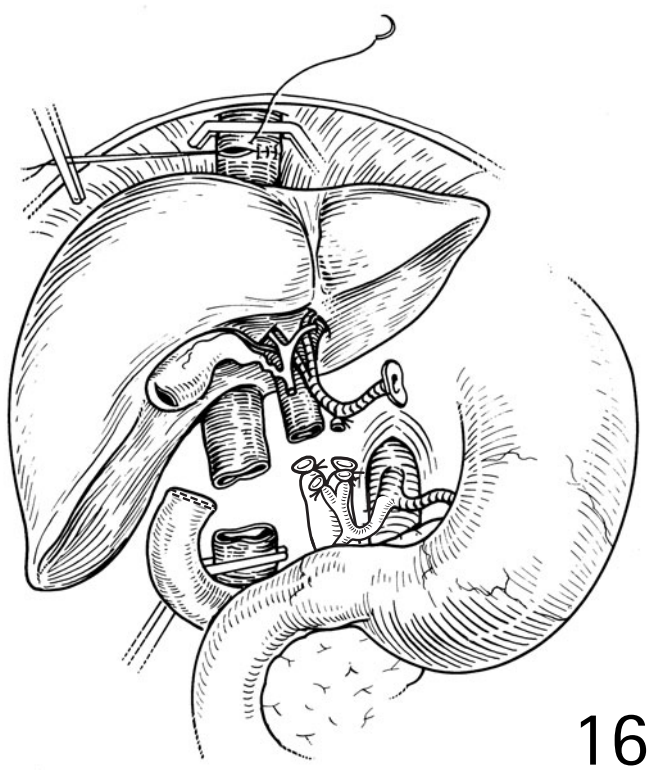


15

15 When a left lateral segment graft is to be transplanted, the recipient vena cava is clamped above and below the liver but the native liver is dissected off the cava, which is left in situ. All venous tributaries on the retrohepatic cava are sutured or ligated. The graft is subsequently 'piggy-backed' onto the recipient cava by anastomosing the donor left hepatic vein to a common orifice of the recipient hepatic veins.

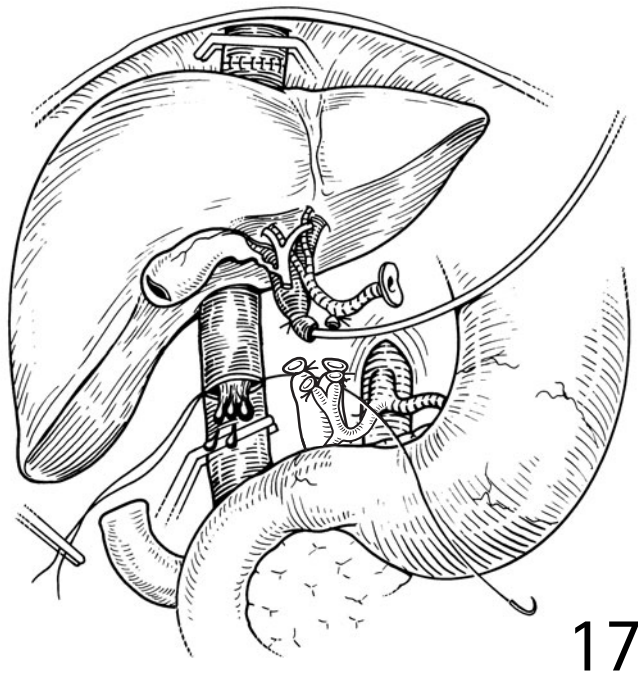
Implantation

16 This must be performed expeditiously to minimize the period of warm ischemia of the graft. Implantation begins with the suprahepatic vena caval anastomosis, which is performed with a continuous 3/0 or 4/0 polypropylene suture. A double-ended suture is placed in each corner of the cava and the left side is tied as the liver is placed in the hepatic fossa. The posterior wall of the cava is anastomosed with a continuous everting suture, which ensures endothelial apposition. If the cava is narrow, the anterior wall is closed with interrupted sutures.



16

17 The infrahepatic vena cava anastomosis is performed next with an everting 4/0 polypropylene suture; the anterior wall is left incomplete. Residual preservative solution rich in potassium is flushed out of the liver with 500 mL of 4.5 percent human albumin solution infused via the portal vein. The effluent flows out of the incompletely anastomosed infrahepatic vena cava, which is then completed.



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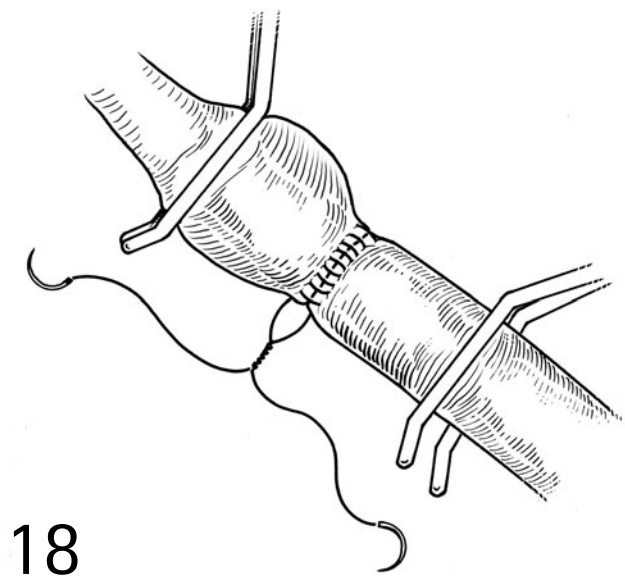
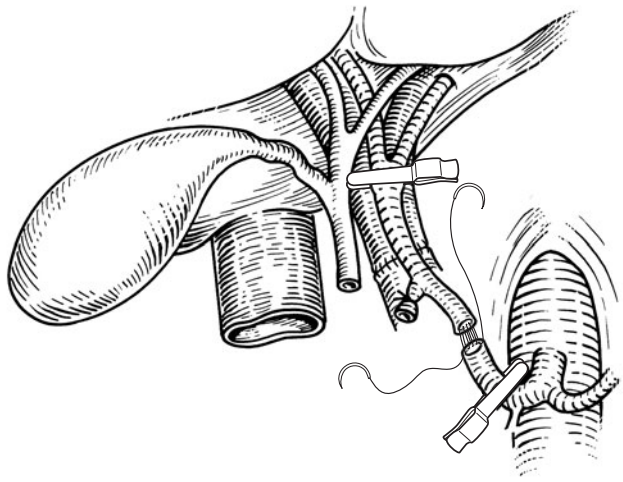


18

18 The portal vein anastomosis is performed next, care being taken to avoid redundancy or rotation, which might cause kinking. The anastomosis is performed with continuous 6/0 or 7/0 polypropylene or Polydioxanone (PDS) sutures. Before tying this suture, the donor portal vein is clamped distal to the anastomosis and the recipient portal vein clamp is released to allow the anastomosis to expand; in addition, a small 'growth factor' is incorporated to help prevent anastomotic stenosis. Interrupted sutures should be used anteriorly in small vessels.

The anesthetist checks the patient for stability and the vascular clamps are removed in sequence. The suprahepatic caval clamp is removed first, followed by the infrahepatic caval clamp. A quick search is made for major bleeding points and the stability of the patient is confirmed before the portal clamp is removed and the liver re-perfused. Any major bleeding points are identified and controlled. The gallbladder is removed using cautery and the cystic duct is transfixed and ligated with an absorbable suture.

19 The donor hepatic artery is gently flushed with heparinized saline and a soft bulldog clamp is applied to prevent backflow of blood from the donor liver, which would interfere with the arterial anastomosis. The hepatic artery is anastomosed end-to-end; a branch patch from the recipient hepatic artery bifurcation or gastroduodenal artery can be used if necessary. The anastomosis can be safely performed with a continuous 7/0 or 8/0 polypropylene suture posteriorly and interrupted sutures anteriorly. Magnification (with loupes or a microscope) is essential with small arteries. If the native artery is small or has poor flow, a donor iliac arterial conduit anastomosed to the recipient's infrarenal aorta may be used to arterialize the graft.



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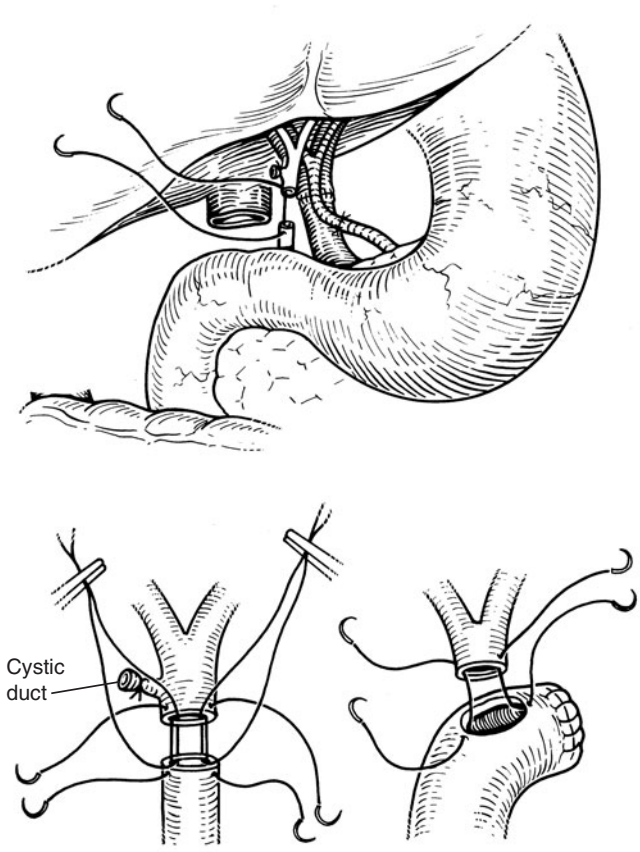


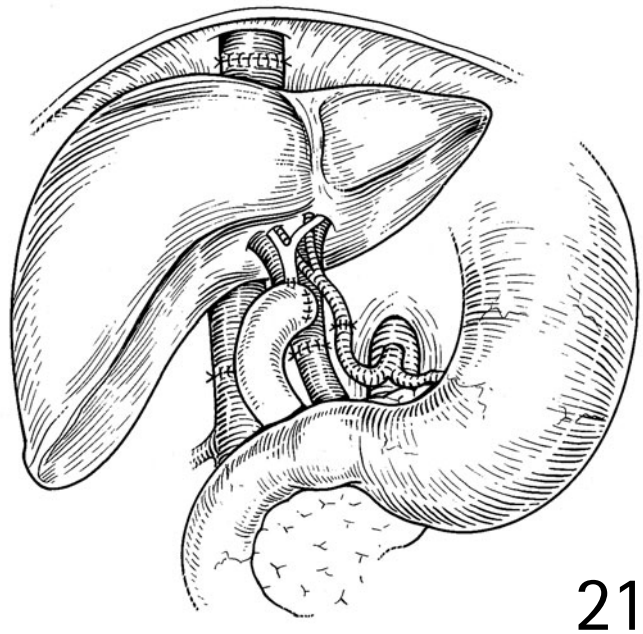
20

20 A duct-to-duct biliary anastomosis is performed using interrupted 6/0 PDS. The donor duct may require trimming to produce a healthy vascular margin. The posterior row of sutures is placed before the ends of the ducts are approximated and the sutures tied.

A Roux-en-Y hepaticojejunostomy is performed in cases of biliary atresia and sclerosing cholangitis or if the recipient common bile duct is very small. A 40 cm Roux loop of jejunum is prepared and brought up to the porta hepatis in a retrocolic position. A small opening is made in the antimesenteric border close to the tip of the Roux loop and the anastomosis is performed with interrupted 6/0 or 7/0 PDS.

Hemostasis and closure

21 Complete hemostasis is secured. All anastomoses are checked. Bleeding at this stage is often secondary to a coagulopathy, and thromboelastography is useful in guiding the use of blood products. The abdomen is closed en masse with a continuous absorbable suture after placing a soft multi-fenestrated large-bore (20–24 Fr) silicone drain posteriorly in the right upper quadrant. The skin is closed with a subcuticular suture. A prosthetic patch (e.g., Vicryl) or skin closure alone (with delayed muscle closure) may occasionally be necessary to avoid an excessively tight abdominal closure.



21

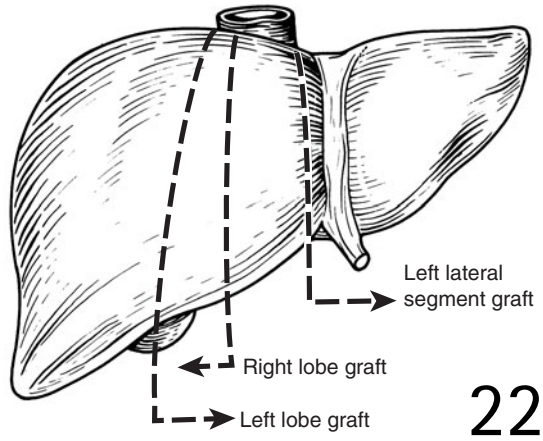
TECHNICAL VARIANT GRAFTS

Segmental liver grafts are prepared from whole deceased donor livers and from living donors. In practice, there are three main types of graft: the left lateral segment (segments 2 and 3), the left lobe (segments 1–4), and the right lobe (segments 5–8 ± 4). The type of graft is determined by the size of the recipient. With cadaveric organs, split-liver grafts are preferable to reduced grafts because this maximizes the donor organ pool. However, when donor anatomy precludes a split-liver transplant, a reduced graft may need to be used. Donor criteria for split or reduced grafts are more stringent than for whole grafts because of the extra insult of the bench procedure; only non-steatotic livers from younger donors are used.

Various size guidelines are helpful in pediatric liver transplantation. As a *minimum*, the graft weight should be at least 0.8 percent of the recipient's body weight in living related liver transplantation; a greater safety margin is advisable in deceased donor transplantation, especially in small infant recipients. The adult liver usually weighs between 1300 g and 1700 g. The left lateral segment comprises approximately 25 percent of the liver weight and the left lobe about 40 percent. The *maximum* graft size that can be safely accommodated in the child's abdomen depends on the recipient's weight, the presence of ascites and hepatomegaly, and other factors, but, as a rough guide, a donor to recipient weight ratio of up to 1.5:1 is often acceptable for whole livers and right lobe grafts, up to 3:1 for a left lobe graft, up to 10:1 for a left lateral segment graft, and up to 25:1 for a monosegment.

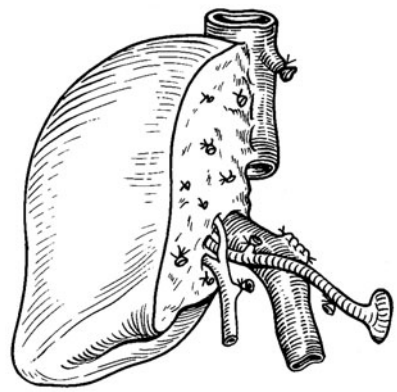
Reduced grafts

Liver reduction is best performed as a bench procedure in the recipient operating suite. The donor liver must be kept totally immersed in cold UW solution at 4°C during the procedure. Care must be taken to avoid excessive traction on the hepatic artery, which could cause intimal injury.



22a

22a–c With right or left lobe grafts, the parenchyma is transected alongside the principal plane slightly to the left or right of the middle hepatic vein, respectively. Biliary and vascular branches on the cut surface of the graft are carefully ligated. The major branches of the portal triad can be divided within the parenchyma. The vena cava is preserved with the graft and the remaining liver is discarded. After division, the cut surface is tested for leaks by gently perfusing UW solution through the portal vein, hepatic artery, and bile duct; these are oversewn. The surface is sprayed with fibrin glue. The lobe is implanted in a similar way to a whole graft.



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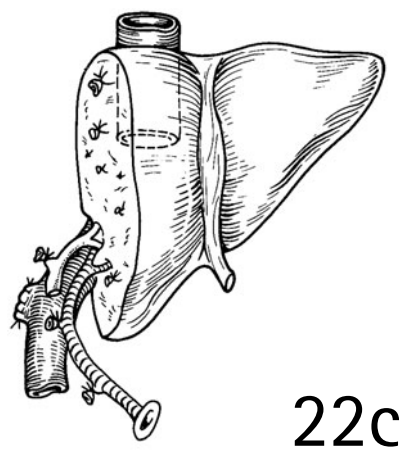
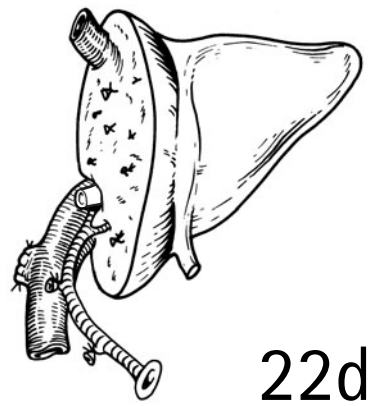


22c

22d In preparing a left lateral segment graft, the plane of parenchymal transection is 0.5–1.0 cm to the right of the falciform ligament. The right-sided branches of the portal triad are dissected and divided outside the liver; the stump of the right hepatic artery is ligated and the right branch of the portal vein is closed with a continuous polypropylene suture. Caudate branches from the origin of the left portal vein are divided. The left hepatic vein is carefully dissected and a cuff is retained for future anastomosis to the recipient vena cava. The common hepatic artery and main portal trunk are retained with the graft. At the level of the cut surface, the bile duct is usually single, but if there are two adjacent segmental ducts, these can often be approximated with fine absorbable sutures.

For liver transplantation in small infants, the left lateral segment can be further reduced, creating a monosegmental graft.



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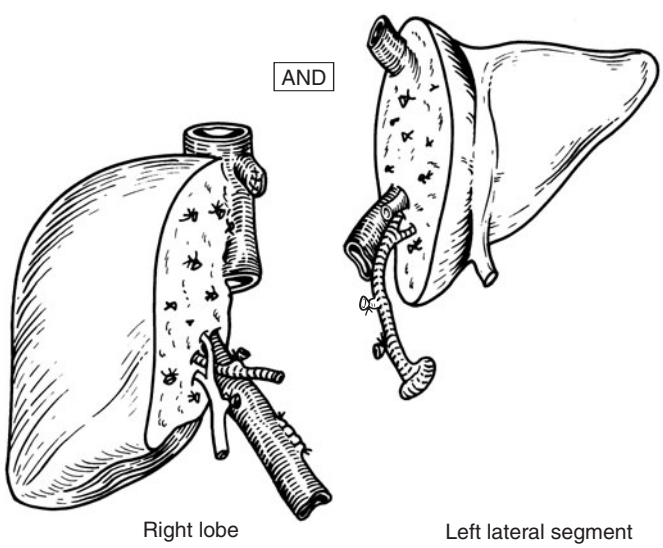

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Split-liver grafts

23 The donor liver is divided into two grafts for transplantation into two recipients. Early assessment of vascular and biliary anatomy is important to ensure that there is no anatomic contraindication to splitting. The gallbladder is removed and the portal vein and hepatic artery carefully dissected to their bifurcations. During right/left lobe splits, a preliminary bench cholangiogram is valuable, but this is not usually necessary for left lateral segment/right lobe splits.

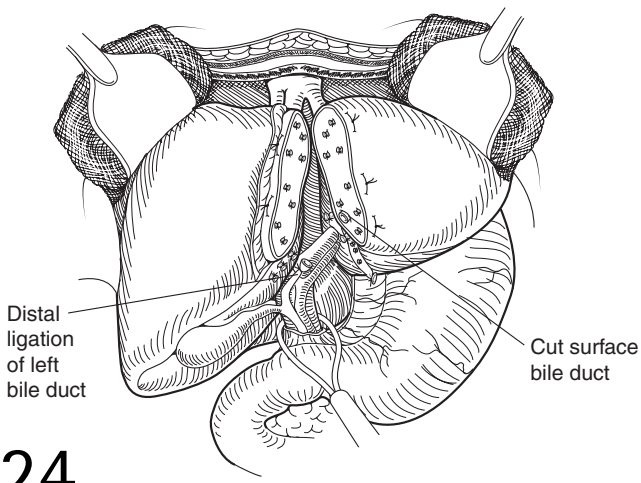
Parenchymal transection along the principal plane will produce a right and a left lobe graft. More commonly, the liver is divided just to the right of the falciform ligament to produce a left lateral segment graft (for a child) and a right lobe graft (for an adult). In most cases, the inferior vena cava remains with the right lobe graft, which also retains the main portal vein, common hepatic duct, and right hepatic artery. Segment 4 is sometimes resected from the right lobe graft since it has been devascularized. The left lateral segment graft retains the left portal vein, a cut surface bile duct, the left hepatic vein, and usually the common hepatic artery. Vessel allocation is modified according to donor and recipient anatomy.

Most liver splits are performed as a bench procedure after deceased donor organ retrieval (ex situ splitting). This takes about 3 hours and the recipient operation must be timed accordingly. Where logistic and organizational aspects per-

mit, in situ splitting in the donor is an alternative; this facilitates cut-surface hemostasis and minimizes the cold ischemic time but requires a more experienced retrieval team.

Living related grafts

The greatest experience is with living related donation of the left lateral segment from parent to child but, in recent years, living related donation of the left lobe and adult-to-adult right lobe donation have become increasingly popular. Living related donation has additional ethical and medical issues. The safety of the donor is paramount. The potential donor should have a compatible blood group and appropriate liver anatomy. He/she requires intensive medical and psychologic evaluation and counseling to determine if he/she is fit for the procedure.



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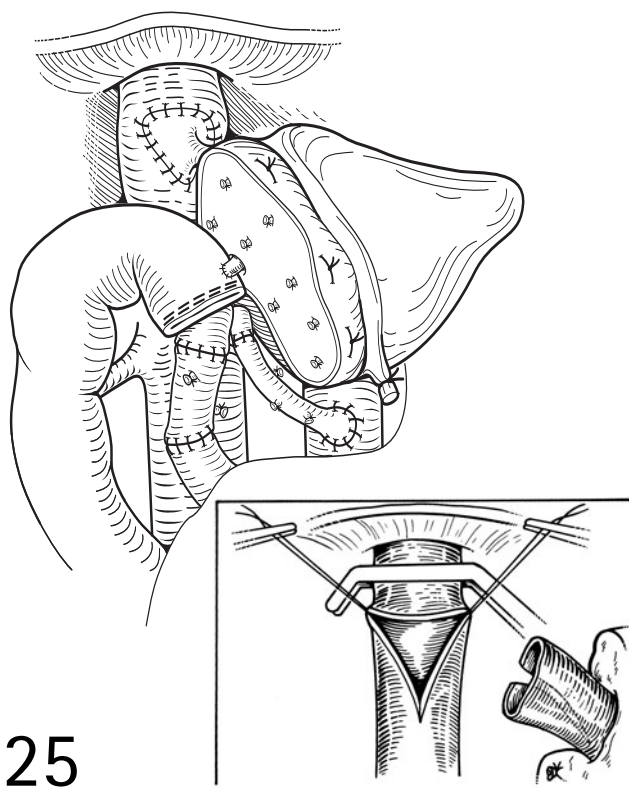
24 Donor and recipient operations are performed electively and in parallel. The abdomen of the donor is opened through a bilateral subcostal incision. In left lateral segment donation, the left side of the liver is mobilized and the left hepatic vein and left portal triad structures are isolated with slings. The parenchyma is transected just to the right of the falciform ligament. Care is taken to ligate vascular and biliary radicles on both sides of the transection plane. When the parenchymal division is almost complete, the left portal vein is mobilized (dividing caudate branches) and the hilar plate and left bile duct divided. After completion of the parenchymal section, the left portal vein, hepatic vein, and hepatic artery are divided and the liver graft is removed and perfused with ice-cold UW solution on the bench. A length of the donor saphenous or jugular vein may need to be retrieved for use as a vascular conduit.

Implantation of the left lateral segment graft

25 The donor left hepatic vein must be widely anastomosed to the recipient cava to prevent venous outflow obstruction. A triangulation technique using a continuous 5/0 polypropylene suture is used. The hepatic vein confluence on the recipient cava may need to be extended inferiorly slightly to the left of the midline to produce a triangle that matches that created by a vertical posterior incision in the donor left hepatic vein. The anastomosis starts with a double-ended suture at the apex of the triangle, moving up each side alternately. Before completing the anterior horizontal part of the anastomosis, the liver is flushed with 4.5 percent albumin.

If the recipient hepatic artery is very small, an arterial conduit from the recipient's abdominal aorta may be needed. This conduit should be prepared and implanted before completing the recipient hepatectomy.

The Roux loop is anastomosed to the bile duct(s) at the cut surface using interrupted 7/0 PDS sutures with the aid of magnifying loupes.



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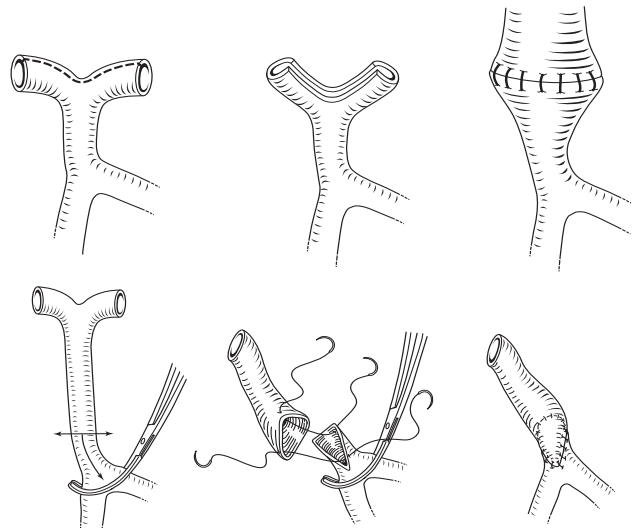

26

26 A narrow portal vein is frequently found in biliary atresia patients. Alternative methods of reconstruction, with or without a donor venous conduit, may be necessary. If a donor conduit is needed, this can be prepared during the anhepatic phase.

Bleeding from the raw cut surface of the segmental graft is controlled by a combination of fibrin glue and other topical hemostatic agents, superficial sutures, and argon beam coagulation. If bleeding is severe or persistent, venous outflow obstruction must be excluded. Suturing the remnants of donor and recipient falciform ligaments helps to stabilize the graft and reduce the risk of torsion of the left hepatic vein.

Orthotopic auxiliary liver grafts

These have been used in children with acute liver failure where subsequent regeneration of the native liver (and removal of the transplanted graft) is anticipated and also in some children with non-cirrhotic liver-based inborn errors of metabolism, e.g., Crigler–Najjar type 1 and urea cycle defects. The native liver is partially resected and a segmental or whole graft inserted in its place. Auxiliary grafts have a greater risk of technical complications, especially related to portal vein flow. In addition, the stability of the recipient with acute liver failure is a concern. In metabolic conditions, there is the same requirement for long-term immunosuppression as with other transplants. Currently, the application of this technique is very limited.

POSTOPERATIVE CARE

The patient is nursed in intensive care. Early extubation is safe after an uncomplicated transplant provided the patient is clinically stable. Arterial blood pressure, temperature, central venous pressure, peripheral perfusion, drain losses, and urine output are closely monitored. The hemoglobin is maintained at 8–10 g/dL. During the first week, daily abdominal Doppler ultrasound scans help to identify any vascular or biliary complications. After an initial postoperative chest radiograph, further radiographs are performed only if clinically indicated. A right-sided pleural effusion is common, but typically resolves spontaneously. The prothrombin time, blood gases, plasma lactate, glucose, and electrolytes are checked regularly and serve as an index of early graft function. Plasma liver function tests, urea and creatinine, calcium, phosphate, magnesium, and immunosuppressant drug levels are recorded daily for the first week. Nasogastric drainage is discontinued as soon as the ileus resolves and enteral feeding is started, usually within 48–72 hours. An acid suppressant is given routinely.

Immunosuppression typically consists of:

- intravenous methylprednisolone given intraoperatively at graft re-perfusion and then at 2 mg/kg per day (maximum 40 mg) for 4 days, followed by oral prednisolone tapered progressively to 0.1 mg/kg per day by 1 month; and
- oral tacrolimus 0.075 mg/kg twice daily or cyclosporin microemulsion 5 mg/kg twice daily (depending on renal function), commencing on day 1 post-transplant and adjusted according to drug levels.

Anti-interleukin-2 receptor monoclonal antibodies such as basiliximab are sometimes used. Steroid-sparing regimens are becoming increasingly popular, but are not appropriate for patients with a history of autoimmune hepatitis.

Early postoperative complications

BLEEDING

This is more likely with a segmental graft or in the presence of graft dysfunction. Continuing hemorrhage despite correction of coagulopathy warrants early re-exploration, especially if

there are signs of a developing abdominal compartment syndrome.

PRIMARY NON-FUNCTION

This occurs in fewer than 5 percent of all transplants and requires emergency re-transplantation. In such cases, signs of satisfactory early graft function (hemodynamic stability, early production of bile, normoglycemia, and resolution of lactic acidosis and coagulopathy) are absent.

VASCULAR THROMBOSIS

Hepatic artery thrombosis after liver transplantation occurs in 4–8 percent of cases and can be devastating. It is commoner in smaller pediatric recipients. Various prophylactic agents such as low-dose heparin and/or acetylsalicylic acid are used and the packed cell volume should initially be maintained below 35 percent. Hepatic artery thrombosis may present in several ways: an acute deterioration in liver function progressing to fatal hepatic necrosis, an insidious onset with biliary complications or sepsis, or an absent arterial signal on routine Doppler ultrasound scan. If hepatic artery thrombosis is suspected within days of the transplant, urgent angiography and/or laparotomy are warranted; if re-vascularization is not possible or unsuccessful, urgent re-transplantation is usually required.

Portal vein thrombosis is uncommon, occurring in approximately 2 percent of cases. The narrow portal vein of children with biliary atresia is a risk factor. Early postoperative occlusion may present with acidosis and deranged clotting, gastrointestinal bleeding, ascites, or deteriorating liver function and demands urgent surgical correction. Late portal vein stenosis may cause portal hypertension (splenomegaly, variceal bleeding), but can often be corrected by percutaneous transhepatic balloon angioplasty.

Vena caval occlusion is very rare. Stenosis of the suprahepatic caval anastomosis leading to ascites and lower body edema is uncommon and can usually be treated by percutaneous angioplasty or stenting.

BILIARY COMPLICATIONS

The incidence of biliary leaks and strictures is about 10 percent, but is related to the type of graft. They may occur in isolation or be secondary to arterial insufficiency. A bile leak may arise from the cut surface of a segmental graft or from a biliary anastomosis. Presentation is with fever and abdominal pain, with or without jaundice. The diagnosis is made by ultrasonography, cholangiography, or radionuclide scan.

Biliary strictures may be anastomotic or non-anastomotic (when they may reflect ischemia, preservation injury, chronic rejection, etc.). They present with jaundice and/or cholangitis. Biliary leaks and strictures can often be managed non-operatively with the aid of interventional radiology and endoscopic stenting. Reconstructive biliary surgery is required for persistent anastomotic strictures, but it is important to assess the hepatic artery beforehand.

ACUTE CELLULAR REJECTION

Acute cellular rejection typically manifests as an increase in the plasma levels of transaminases with or without a low-grade fever and irritability. An urgent Doppler ultrasound scan helps to exclude major surgical problems with the graft before performing a percutaneous needle liver biopsy to confirm the diagnosis. Acute rejection is initially treated with three daily doses of 10 mg/kg intravenous methylprednisolone. Most patients respond within 48 hours. A further pulse of steroids may be given and/or other immunosuppressants introduced in resistant cases.

RENAL COMPLICATIONS

Peritransplant renal dysfunction is common as a result of hepatorenal syndrome, ascites, cardiovascular instability, and nephrotoxic drugs. With good postoperative graft function, most cases recover well, although temporary renal support may be required.

INFECTIONS

Infectious complications are a major cause of mortality and morbidity after liver transplantation because of immunosuppression, indwelling catheters, pre-existing morbidity, malnutrition, and, surgical complications. Early postoperative infections are mostly bacterial or fungal, but viral infections are particularly important later on. Cytomegalovirus infection may cause fever, malaise, diarrhea, gastrointestinal bleeding, abnormal liver function, and pulmonary symptoms. Children are more likely to develop disease if they are primarily infected from a CMV-positive donor than when viral reactivation occurs in a previously immune recipient. Treatment with ganciclovir and reducing the immunosuppression are often effective. Cytomegalovirus prophylaxis is routinely given to naive recipients receiving a CMV-positive graft. Other opportunistic viral infections include herpes simplex, Varicella zoster, adenovirus, and EBV. The last-mentioned is a potential cause of post-transplant lymphoproliferative disease. Both CMV and EBV can be monitored by serial estimations of viral load using the polymerase chain reaction.

OUTCOME

One-year survival after elective pediatric liver transplantation is now consistently above 90 percent, and 5–10-year survival figures are around 80–90 percent. Most survivors enjoy an excellent quality of life, with good growth and complete relief from the symptoms of chronic liver disease. Emergency transplantation for acute liver failure carries a worse prognosis, but even in this situation 70 percent or more patients now

survive. European Liver Transplant Registry data show no difference in the actuarial survival of recipients transplanted when aged less than 2 years compared to older children.

Late complications, including rejection (especially from poor drug compliance), infections, side effects from immunosuppression, and graft dysfunction, continue to pose challenges. It is estimated that about 15 percent of children require re-transplantation at some stage.

Acknowledgment

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FURTHER READING

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Small bowel transplantation

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HISTORY

Intestinal transplantation is a relatively new procedure. As the largest lymphoid organ in the body, transplantation of the intestine presents a significant challenge for immunosuppression. Following animal experiments in the 1960s, notably by Lillehei, and later by Grant, who subsequently performed the first successful liver–intestinal transplant, increasing experience with the procedure has been reported. Deltz reported the first successful long-term small bowel transplant in an adult patient in 1988. In recent years, survival following intestinal transplantation has shown a steadily increasing trend.

PRINCIPLES AND JUSTIFICATION

Parenteral nutrition is life saving in intestinal failure due to short bowel syndrome and a variety of functional causes such as pseudo-obstruction. The potential of a satisfactory and effective life-saving therapy in the form of parenteral nutrition distinguishes intestinal failure from other forms of end-organ failure. Uncomplicated parenteral nutrition in a stable patient population results in a 5-year survival of close to 80 percent. A small subset of patients with refractory parenteral nutrition dependence are at risk of significant morbidity and even mortality due to complications that make it potentially dangerous or even impossible to continue this therapy. The most common of these complications is a syndrome of progressive cholestasis that may lead to end-stage liver disease. Despite satisfactory catheter-care techniques, some patients may also suffer inexplicably from central venous catheter-related complications in the form of life-threatening episodes of bacterial or fungal sepsis or loss of venous access due to venous thrombosis. Thus, the fundamental indication for intestinal transplantation is the combined presence of refractory parenteral nutrition dependence *and evidence of parenteral nutrition failure* with the presence of one of the life-threatening complications of parenteral nutrition.

PREOPERATIVE EVALUATION

A thorough multidisciplinary evaluation is critical prior to a decision to proceed to intestinal transplantation. Key steps in this evaluation are:

- evaluation of residual bowel anatomy;
- evaluation of residual bowel function/motility;
- demonstration of refractory parenteral nutrition dependence and inability to advance enteral feeds;
- evaluation of the presence and extent of liver disease;
- identification of other systemic illnesses/comorbid conditions;
- determination of any significant neurodevelopmental contraindications;
- determination of any psychosocial contraindications.

Following a detailed history and physical examination, it is our practice to carry out abdominal and cervical venous Doppler ultrasound examinations as well as appropriate gastrointestinal contrast radiographs. Based on the clinical examination and biochemical tests of liver function, a liver biopsy may be required for accurate staging of the extent of liver disease.

Such a detailed evaluation has allowed us to avoid the need for intestinal transplantation and even wean some patients off parenteral nutrition altogether. We have also on occasion uncovered previously undiscovered pathology, successful treatment of which has allowed patients to be successfully weaned off parenteral nutrition. On completion of evaluation, the appropriateness of intestinal transplantation for the

patient can be determined with a high degree of confidence, and the choice of the type of graft (i.e., isolated small bowel or combined liver–small bowel) can also be made in most cases. It is important to recognize that there is inevitably a long wait for a suitable cadaver donor in most cases (particularly for recipients weighing less than 10 kg) and an accompanying high mortality rate on the waiting list, especially for patients awaiting combined liver–bowel transplantation. Therefore early referral for patients who appear to be failing parenteral nutrition may be an important step toward improving the outcomes for intestinal transplantation.

The choice of allograft is determined primarily by the presence or absence of advanced liver disease; the presence of cirrhosis with portal hypertension and massive hepatosplenomegaly mandates a combined liver–small bowel transplant. Lesser degrees of liver dysfunction may be reversible with good graft function from an isolated small bowel transplant. The indication for a multi-visceral graft is the presence of disease in other organs (e.g., kidney or pancreas) that requires replacement of the organ, or where abdominal exenteration is required for the removal of diseased viscera.

DONOR OPERATIONS

Choice of donor

Hemodynamically stable donors who are ABO compatible and ideally about 50–75 percent of recipient weight with evidence of good visceral perfusion (normal liver and renal function) and no acidosis constitute satisfactory small bowel donors. We no longer accept donors who are cytomegalovirus (CMV) positive, except for those who are below the age of 1 year and whose CMV positivity may be attributable to passive transfer of maternal antibodies. Donor:recipient size ratios of 0.5–0.6:1 are necessitated by the loss of abdominal domain in the patient with short bowel syndrome who has inevitably had multiple abdominal operations. Patients with functional disorders with few prior operations can accept larger donors. There is growing experience with size-reduction techniques for both isolated bowel and combined liver–bowel grafting that allows donor:recipient ratios of 2:1 or even 3:1.

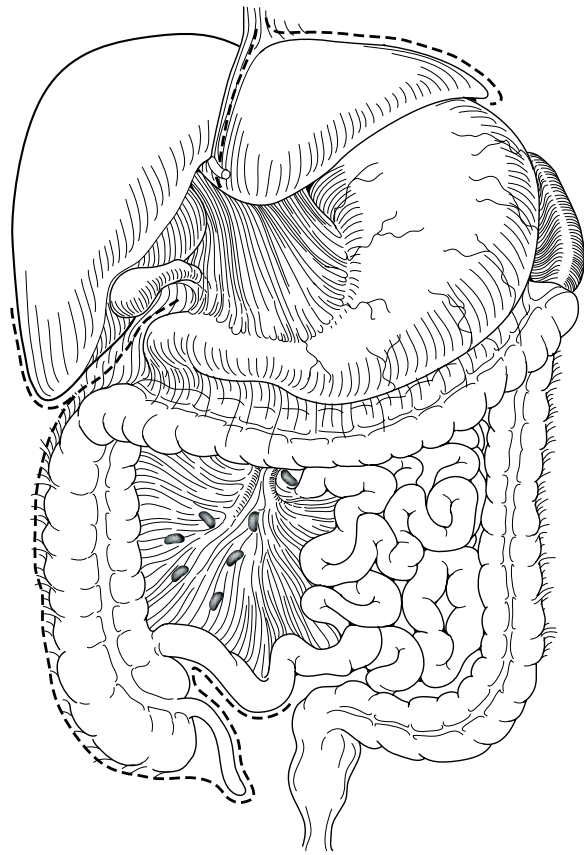
Routine preoperative antibiotics are administered to the donor. Unlike other groups, we no longer practice any specific intestinal decontamination in the donor, with no apparent increase in complications.

Organ procurement

ISOLATED SMALL BOWEL GRAFT

The procedure begins with a long midline incision and a median sternotomy. Superficial mesenteric nodes are procured early for tissue typing. An infusion of antithymocyte globulin, 30 mg/kg, is started at the beginning of the procedure to secure donor lymphocyte depletion *after* the removal of lymph nodes for tissue typing.

1 We section the falciform ligament and the left triangular ligament as well as ligamentous attachments to the right lobe of the liver to avoid inadvertent injury to the liver capsule. Mobilization of the right colon along the lateral avascular plane as well as the root of the mesentery and an extensive Kocherization of the duodenum are carried out. This exposes the aorta to the level of the superior mesenteric artery and the inferior vena cava to the lower aspect of the liver.



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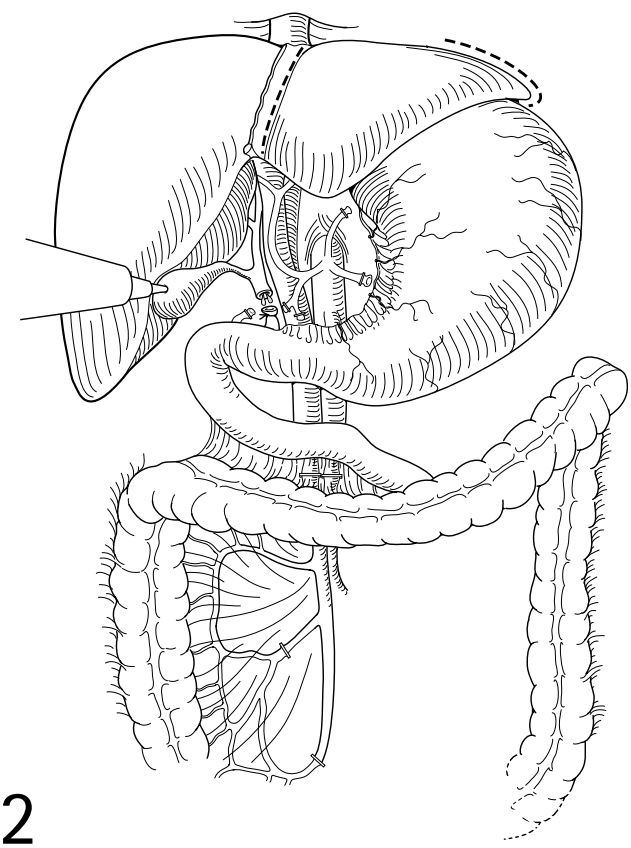
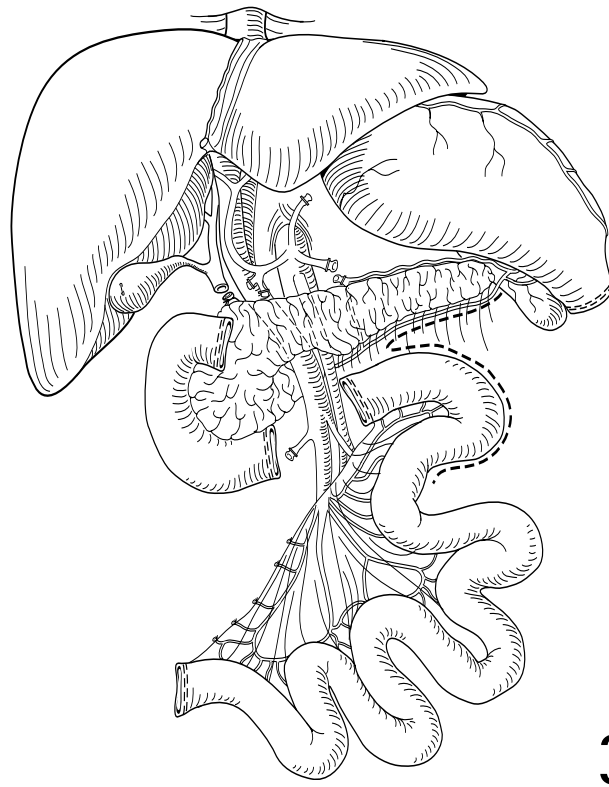


2

2 Attention is turned to hilar dissection. The bile duct is ligated at the superior border of the pancreas, and the duct is divided proximally. Through a small incision in the fundus, the gallbladder is flushed clear of bile with saline. The small venous structures in the hilum as well as lymphatic and adventitial tissues are ligated and divided to visualize the anterior surface of the hepatic artery and portal vein. A superficial position of the portal vein may indicate the presence of a displaced hepatic artery arising from the superior mesenteric artery; if identified posterior to the portal vein, follow the artery to its origin from the superior mesenteric. The hepatogastric ligament is opened and an exploration for a displaced left hepatic artery from the left gastric artery is made. The gastroduodenal artery at the inferior border of the first part of duodenum is identified and divided. After mobilizing the esophagus to the left, the diaphragmatic crura are sectioned longitudinally and an umbilical tape is passed around the aorta at the level of the hiatus. The anterior surface of the pancreas is exposed by dividing the gastrocolic omentum. The superior mesenteric artery and vein are exposed at the lower border of the pancreas by separating the third part of the duodenum from the mesocolon. The middle colic, right colic, and colonic branches of the ileocolic vessels are divided.

3 The small bowel is retracted to the right and the duodenojejunal flexure is fully mobilized. The duodenum just distal to the pylorus is stapled and divided. The stomach is mobilized cephalad and out of the operative field. The small bowel is stapled at the distal ileum. The remaining colonic vessels are divided and the colon is moved out of the field. The retracted colon is omitted and the ostium of the superior mesenteric artery is shown *above* the pancreas for clarity. The portal vein can be visualized deep to the hepatic artery and bile duct in the hilum of the liver. Note the divided middle colic vessels, gastroduodenal, right gastric, and left gastric artery.



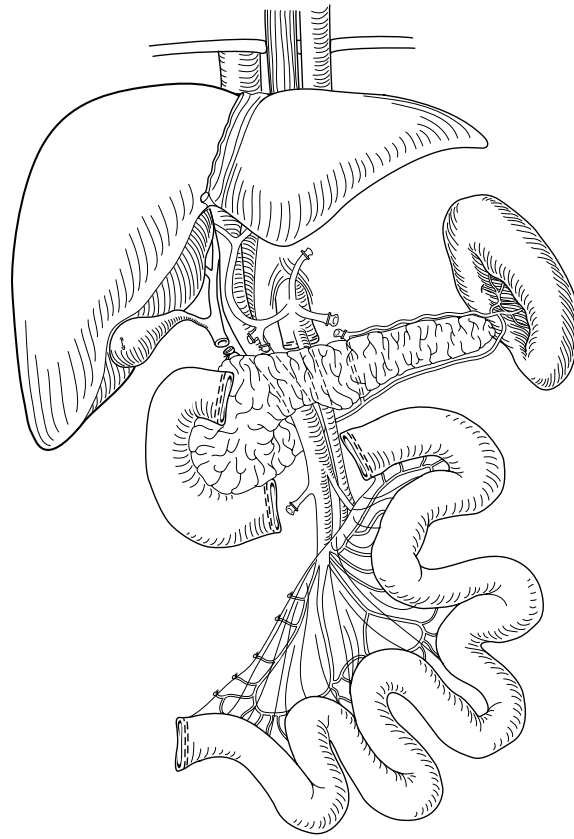
3

COLD FLUSH

Intravenous heparin (300 IU/kg) is administered by the anesthesiology team. An appropriate-size cannula is placed in the abdominal aorta immediately above the bifurcation. A second cannula is placed in the inferior mesenteric vein if the size permits. The heart team (for cardiac procurement) sections the inferior vena cava above the diaphragm to vent the blood and clamps the aorta. We simultaneously clamp the supraceliac aorta immediately below the diaphragm. Cold University of Wisconsin (UW) solution is administered via the aortic cannula and, if present, via the second cannula. The amount of UW solution depends on the size of the donor, and is approximately 50 mL/kg. Cold slush is distributed in the abdominal cavity.

Combined liver–small bowel graft

4 Our technique for combined liver–small bowel grafting avoids any dissection in the porta hepatis. The liver, small bowel, pancreas, and spleen are procured en bloc. A long segment of thoracic aorta is procured in continuity with the celiac trunk and the superior mesenteric artery. The distal aorta is divided a few millimeters below the superior mesenteric artery, leaving enough aortic length to be over-sewn without compromising the flow into the superior mesenteric artery. Aortic (and inferior mesenteric venous) flush with cold UW solution is carried out as before, venting the blood by transecting the inferior vena cava above the diaphragm.



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Cold dissection and removal of organs (isolated small bowel or combined liver–small bowel)

For small-sized donors or in the presence of technical difficulty, we favor removing the liver and intestine en bloc with back-table separation of organs in the case of isolated small bowel grafts. Dissection starts with mobilization of the liver. The diaphragm is divided longitudinally anterior to the aorta; dissection continues around the inferior vena cava and then to the right, dividing the right triangular ligament. The dissection is carried through the right adrenal gland, locating the adrenal vein for back-table ligation. The infrahepatic inferior vena cava is divided above the level of the renal veins. The abdominal aorta is divided at the level of the diaphragm and below the origin of the superior mesenteric artery.

The remaining gastrocolic omentum is sectioned with the short gastric vessels. The stomach is retracted further to the left. The lienocolic and lienophrenic ligaments are divided. The spleen is then held as a handle, retracting medially, continuing to dissect medially posterior to the pancreas in a plane anterior to the left kidney and adrenal gland. The inferior mesenteric vein is divided but marked with a ligature to facilitate later identification of the splenic vein. Transection of the retro-aortic tissues completes the maneuver.

As stated earlier, for the combined liver–small bowel allograft, the supraceliac aorta is not isolated, as the whole thoracic aorta will be harvested in continuity with the abdominal segment bearing the celiac trunk and the superior mesenteric artery. There is no dissection in the porta hepatis except to incise the gallbladder at the fundus and to flush it.

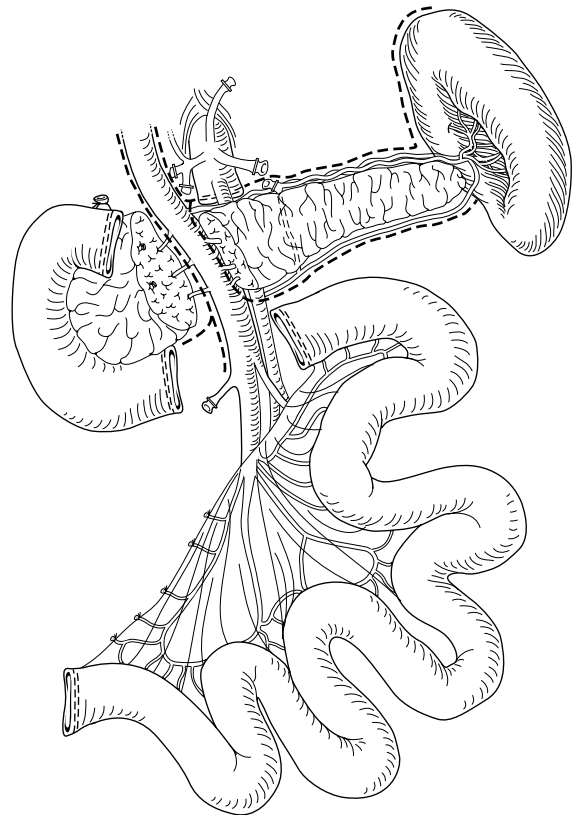
It is routine practice to procure the donor iliac vessels, and in special cases the carotid arteries and jugular veins, in the event that additional vessel length is required in the recipient.

Back-table preparation

If the liver–small bowel–pancreas cluster has been procured for different recipients, the separation of liver from the pancreas–small bowel graft is carried out on the back-table, orientating the viscera in the anatomical position and dividing the portal vein above the superior border of the pancreas. The splenic artery, if not divided earlier, is also divided during this separation. The organs are stored in cold UW solution.

Preparation of the isolated bowel graft

5 The superior mesenteric artery is dissected free from the peri-adventitial tissue, progressing from its aortic take-off distally. The pancreatic branches are ligated and divided, and dissection is continued until the first jejunal branch. The splenic vein is then dissected to its confluence with the portal vein and superior mesenteric vein; the branches to the head and body of the pancreas are ligated and divided. The pancreas is divided at the isthmus, taking due care to avoid injury to the subjacent portal vein. The pancreas and duodenum can now be safely excised, leaving the small bowel graft on its vascular pedicle based on the portal vein and the superior mesenteric artery. We imbricate the staple lines at either end of bowel with 5/0 Prolene suture, and take due care to mark the distal end of the bowel with a long tie to ensure accurate positioning in the recipient. It is our practice also to mark the anterior surface of the portal vein with a marking pen and a stay suture to ensure proper orientation at the time of anastomosis.



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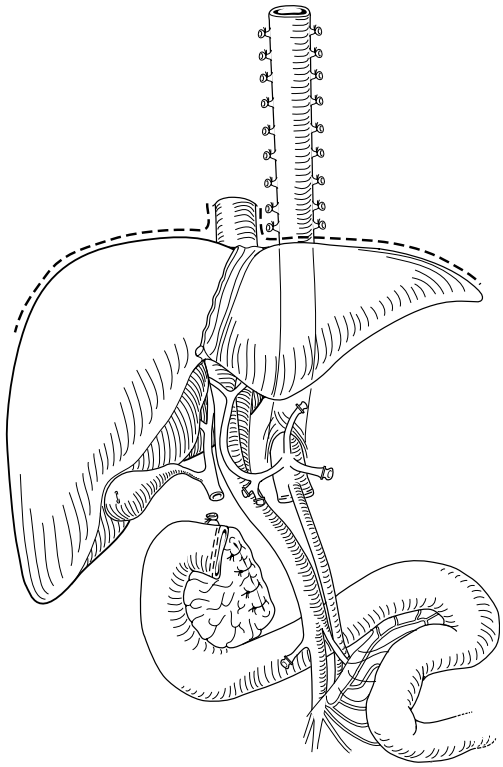


6

Preparation of the liver–small bowel graft

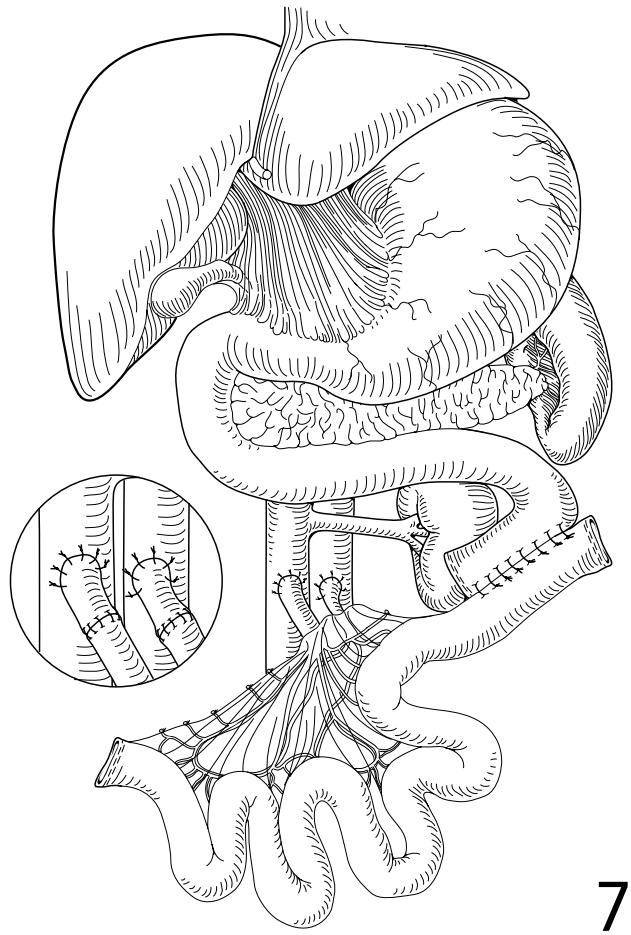
6 The preparation of the liver starts with removal of the diaphragmatic tissue and cleaning around the inferior vena cava. The next step consists of dissection of the thoracic and abdominal aorta with cleaning of the peri-adventitial tissues and individual ligation of the intercostal arteries. The distal aorta is closed using continuous 6/0 or 7/0 polypropylene sutures, just beyond the take-off of the superior mesenteric artery, taking due care to avoid narrowing the ostium. The celiac trunk is dissected with ligation of the splenic and left gastric arteries; the hepatic artery is dissected to the level of the gastroduodenal artery, which is preserved to vascularize the head of the pancreas and duodenum. The splenic vein is dissected up to the confluence with the superior mesenteric vein. The pancreas is divided to the right of the portal vein. If the pancreatic duct can be identified on the cut surface, it is individually ligated with 5/0 polypropylene suture. In any event, the entire cut surface of the pancreas is over-sewn with interrupted 5/0 polypropylene sutures. It must be noted that with forbidding portal hypertension or pancreatic disease, abdominal evisceration including removal of the native pancreas becomes technically simpler or necessary, and in this situation the donor pancreas is left intact with the liver–small bowel graft.

RECIPIENT OPERATION

Isolated small bowel transplant

7 The recipient operation must be timed in concert with the donor operation to keep cold ischemia to the minimum possible.

Abdominal incision can be vertical or horizontal, depending on prior operative scars. A configurable and versatile self-retaining retractor such as the Thomson abdominal retractor facilitates exposure. Extensive adhesiolysis is often required. Prior bowel anastomosis between remnant small bowel and colon is taken down. Obviously non-functional or dilated remnant bowel is excised. The retroperitoneum is exposed, allowing full dissection of the infrarenal abdominal aorta and inferior vena cava up to their pelvic bifurcations. While venous drainage of the graft can be portal or systemic, in the presence of any degree of fibrotic liver disease, systemic venous drainage is preferred. The technical simplicity of the latter has led us to adopt systemic venous drainage of all isolated bowel grafts. Vascular clamps are placed in side-biting fashion when possible, but in the small recipient these are invariably occlusive. The superior mesenteric artery of the graft is anastomosed initially to the infrarenal aorta of the recipient, followed by anastomosis between the superior mesenteric vein of the graft and the inferior vena cava. Due care must be taken to ensure that the venous anastomosis is cephalad to the arterial anastomosis, and that proper orientation is maintained. On occasion, if graft vessels are unduly short, or if lack of recipient abdominal domain precludes a safe vascular anastomosis, we have used interposition grafts for both arterial and venous anastomoses as required, as shown in the inset to the figure. Re-perfusion starts with release of the venous clamps. Bleeding points in the cut edge of the mesentery are controlled, followed by release of the arterial clamps. Once hemostasis is secured, attention is turned to restoring intestinal continuity. We carry out side–side (functional end–end) anastomosis between the proximal end of the graft and the remnant native proximal jejunum or duodenum. Similarly, distal anastomosis is carried out between the graft ileum and the distal native bowel, whether ileum or colon. We perform our anastomoses with a standard two-layered technique due to the theoretically higher risks of poor healing in an immunosuppressed patient. All patients receive a defunctioning distal ileostomy (loop or Bishop–Koop) to allow protocol ileoscopic biopsies for close monitoring of graft histology. It is our practice to leave the skin wound open with dressings to allow healing by secondary intention, in view of the high risk of wound infection.



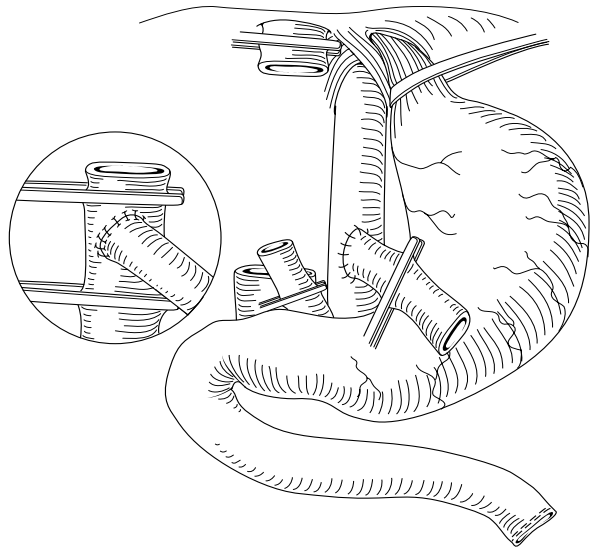
Liver–small bowel transplant

8a,b The liver–small bowel transplant recipient tends to be much sicker, and to require even closer intraoperative and perioperative monitoring, than the recipient of an isolated liver transplant. Close anesthesiological monitoring of coagulation, metabolic disturbances (particularly hyperkalemia), and significant blood loss in the face of extensive adhesions and profound portal hypertension require constant vigilance. We employ horizontal or vertical incisions depending on prior operative scars, but on occasion have had to convert to cruciate extensions. Hepatectomy is performed in the usual manner, leaving the native inferior vena cava clamped above and below the liver. The portal vein is ligated, as is the hepatic artery. At this point, the extent of native bowel is also determined, and prior anastomoses are taken down, excising any dysfunctional or dilated bowel. The supraceliac aorta is dissected free and controlled with a sling. We perform a portocaval shunt between the end of the native portal vein and the infrahepatic inferior vena cava, as shown in the inset; this allows for decompression of all remnant native foregut viscera, such as the stomach, duodenum, and pancreas. The inferior vena cava is then re-clamped above the shunt.

The supraceliac abdominal aorta is then clamped. A segment of donor thoracic aorta is excised and anastomosed in end–side fashion, to be used as an interposition graft, without the composite liver–bowel graft in the way. Aortic flow can be restored immediately after completion of this anastomosis, placing a clamp on the interposition graft. The entire composite graft is then brought into the field.

The anastomoses of the suprahepatic and infrahepatic inferior vena cava are then performed. The donor thoracic aorta (in turn supplying both graft liver and bowel) is then anastomosed to the aortic interposition graft. The use of an interposition graft in this manner allows convenient aortic anastomosis in the depths of the wound, without having the composite allograft in the way, and allowing final arterial anastomosis to the liver–bowel graft at the level of the wound. Re-perfusion is carried out by releasing the venous clamps first, followed by the arterial clamps. Bleeding from the mesenteric edge and from the retroperitoneum is controlled, before proceeding with the proximal and distal intestinal anastomoses.

A loop ileostomy or a Bishop–Koop stoma is brought out in all cases, to allow surveillance endoscopic biopsies of the graft. Following fascial closure with a monofilament non-absorbable suture, the skin wound is left open to heal by secondary intention, in view of the high risk of wound infection. (The native stomach, duodenum, and pancreas are omitted from Illustration b in the interests of clarity, but the site of the proximal intestinal anastomosis to native proximal bowel remnant is shown.)



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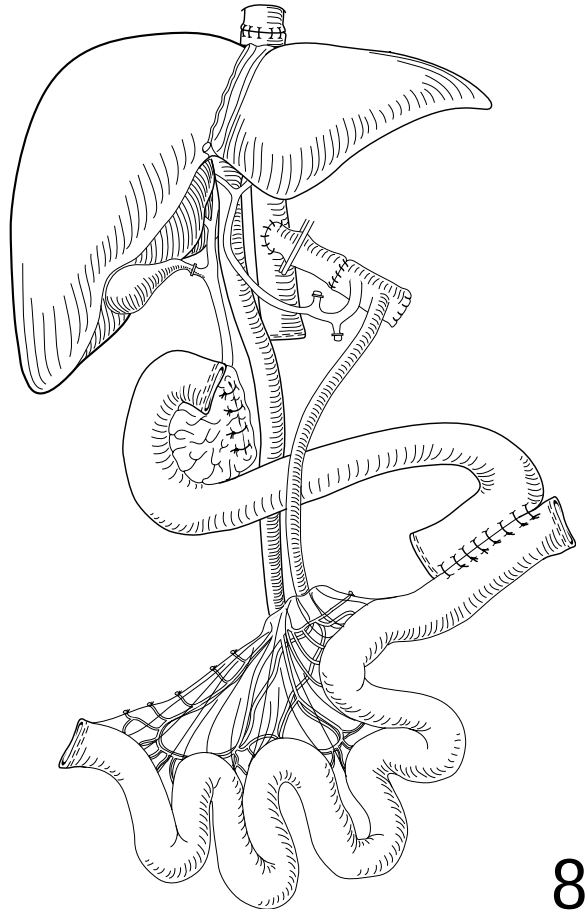

8b

POSTOPERATIVE CARE

Nasogatric decompression is maintained for 5–7 days after transplant, introducing clear liquids, often via a nasojejunal tube, with return of peristaltic activity. Changes in color and appearance of the stoma or its output are a cause for concern and reflect graft vascularity early after transplant. Prophylactic antibiotics are maintained for at least 1 week after transplant. Various prophylactic agents against fungi, viruses such as cytomegalovirus, and opportunist infectious agents such as *Pneumocystis carinii* are routinely prescribed for periods of 3 months to 1 year after transplant.

Most centers use an induction regimen with antibody followed by maintenance immunosuppression with tacrolimus and steroids in the long term. Close monitoring of tacrolimus levels is essential for the first few months after transplant to maintain optimal levels.

With satisfactory stomal activity, fat-free enteral formulas are gradually introduced at 1 week after transplant and advanced gradually based on tolerance and stoma outputs. Transition to complete elemental formulas is delayed until 4–6 weeks postoperatively to allow time for lymphatic reconstitution after transplant. Oral aversion is a common obstacle for transitioning to standard diets in the longer term. The ability to remove the central line after demonstrating full enteral tolerance is a major milestone in these patients. Some patients continue to require supplemental intravenous fluids until stoma takedown between 6 months to 1 year after transplant.

In the absence of any biochemical markers for rejection, we resort to protocol ileoscopies and biopsies of the allograft to diagnose rejection. Histology remains the gold standard for diagnosis. Rejection may be marked by increased ostomy losses and if diagnosed early, may be reversed by steroid boluses, although on occasion it may require antibody treatment or even graft removal.

COMPLICATIONS AND OUTCOMES

Despite the magnitude of the procedures described above, the postoperative management remains the most challenging aspect of the care of these patients.. The high incidence of acute rejection in a lymphoid-rich graft requires very high levels of immunosuppression. This in turn leads to a high incidence of bacterial, viral, and fungal sepsis that requires close monitoring and aggressive early treatment. Bacterial infections, occurring in more than 90 percent of patients, are often due to enteric Gram-negative organisms, which may have to be treated while maintaining significant immunosuppression. In the longer term, complications related to immunosuppression, such as post-transplant lymphoproliferative disease related to Epstein–Barr virus, and chronic rejection continue to pose serious challenges to patient and graft survival. Despite these challenges, the results of intestinal transplantation continue to improve, allowing patient survival at 1 year of the order of 90 percent and graft survival of 70–75 percent. With improving outcomes, survival even at 5 years is now greater than 50 percent. When one recognizes that these outcomes are achieved in patients who have 'failed' parenteral nutrition, it is hoped that earlier patient referral before the onset of advanced liver disease and better patient selection may allow continued refinement and even better outcomes.

FURTHER READING

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Pediatric interventional radiology: an overview

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BACKGROUND

Interventional radiology is a rapidly growing field in pediatrics. Most procedures have adapted techniques originally developed in adult practice to improve on existing pediatric surgical procedures. Radiologists now perform many procedures that were previously exclusively in the domain of pediatric surgery. Continued cooperation between surgeons and radiologists will lead to further improvements.

Most of the following procedures are based on very simple concepts. In one, a structure is first accessed by percutaneous puncture with ultrasound and/or fluoroscopic guidance. A guidewire is inserted, the needle is removed, and a catheter is introduced into the structure over the guidewire. This concept is the basis of angiography, abscess drainage, biliary drainage, nephrostomy, cecostomy, and one method of gastrostomy. Another concept is dilatation by crossing a stenosis with a guidewire followed by a balloon catheter, and inflating the balloon using fluoroscopic guidance. This is the basis of angioplasty (of arteries and veins) and dilatation of tracheobronchial, ureteral, and esophageal and other gastrointestinal strictures.

PREOPERATIVE CARE

In general, the perioperative care required for interventional radiology is the same as for the corresponding operation in pediatric surgery (see Chapter 1). There are exceptions, however. We do not crossmatch blood for a child of any size for central venous access procedures unless we anticipate that transhepatic access will be required or there is an uncorrectable coagulopathy. This is because ultrasound-guided central venous access (see Chapter 3) is almost always easy, and bleeding is extremely rare. On the other hand, percutaneous techniques offer less opportunity for intraoperative recognition and treatment of bleeding complications than the corresponding open procedure. We therefore check coagulation values and platelet levels before most procedures in the chest and abdomen, where postoperative bleeding is more likely to be a significant problem.

Sedation and anesthesia

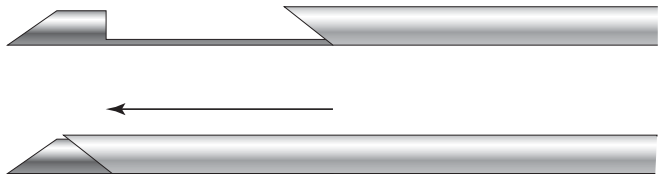
There is a very wide range of anesthetic practice for pediatric intervention. Some centers use mainly intravenous sedation (with various drugs), with good results. At our institution, we prefer general anesthesia, because it is less unpleasant for the child and may make the procedure safer. General anesthesia is essential for procedures that are very painful and/or very long.

When general anesthesia is not used, topical anesthetic cream is applied to the appropriate area before the procedure. Lidocaine buffered with sodium bicarbonate is then injected through this anesthetized area of skin. If the procedure is performed under general anesthetic, levobupivacaine (2.5 mg/mL), a longer-acting local anesthetic, can be used for postoperative pain relief.

For brief but painful procedures, for example renal biopsy or abdominal abscess drainage, older children can breathe an equimolar mixture of oxygen and nitrous oxide (Entonox). This is very effective when used in combination with topical and local anesthetic.

BIOPSY AND DRAINAGE PROCEDURES

1 Image guidance makes percutaneous biopsy and drainage safer and quicker, and blind needle biopsy is now appropriate only for superficial lesions. Percutaneous biopsies in children are usually performed with semi-automatic core needles. The cutting tip is advanced to expose a slot in which tissue is trapped by an outer sleeve, which advances when the needle is fired. Semi-automatic needles are easier to use than manual biopsy needles (e.g., Tru-Cut) and biopsy gun systems.

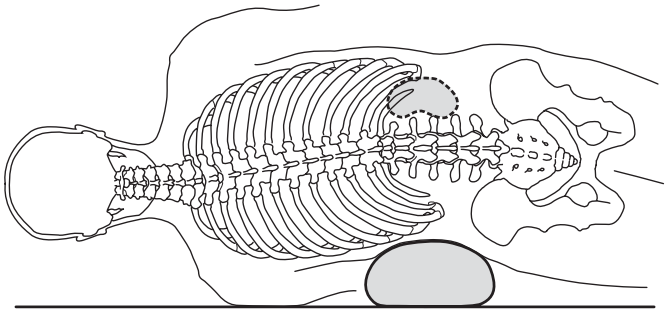


1

Renal biopsy

INDICATIONS

The most common indications for renal biopsy are nephrotic syndrome and impaired function following renal transplantation.



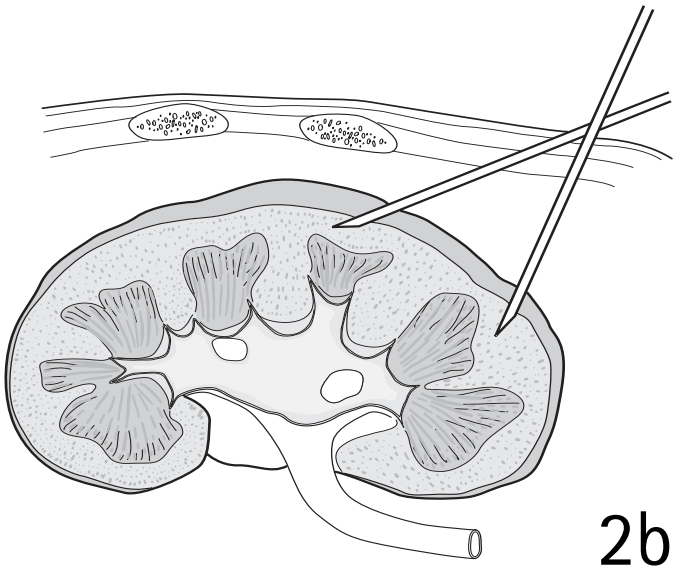
2a

POSITIONING AND ANESTHESIA

2a We perform renal biopsy under general anesthesia in young or uncooperative children, and with sedation or Entonox in older children. Either kidney can be biopsied. The procedure can be performed with the patient prone or seated, but we find it easier to place the patient in a lateral position. This makes it much easier to administer Entonox, and when general anesthesia is used, avoids the need for endotracheal intubation and muscle relaxants. A large pad is placed under the patient's abdomen to increase the convexity of the side to be biopsied. Renal transplant biopsy is performed in the supine position, and does not usually require general anesthesia or sedation.

TECHNIQUE

2b The tract is infiltrated with local anesthetic down to the level of the renal capsule. A 16-gauge semi-automatic core biopsy needle is then inserted through a short stab incision. If the child is able to cooperate, he or she is asked to stop breathing as the needle is advanced into the kidney and the biopsy taken. A shallow or deep trajectory may be appropriate, depending on the size of the kidney and the relationship of the lower pole to the ribs. When the child is under general anesthesia, suspension of breathing is not absolutely necessary, and the anesthetist may prefer to allow the patient to breathe spontaneously. We have a pathology technician present to examine the core and confirm that sufficient glomeruli are present. If not, another core is taken. Using this method, 99 percent of biopsies should have a definitive histological report.



2b

COMPLICATIONS

Significant complications, such as hematuria requiring transfusion, and injury to other organs, should occur in less than 2 percent of biopsies. The most important complication is arteriovenous fistula. These lesions tend to resolve spontaneously, but require embolization (see below) if they are symptomatic.

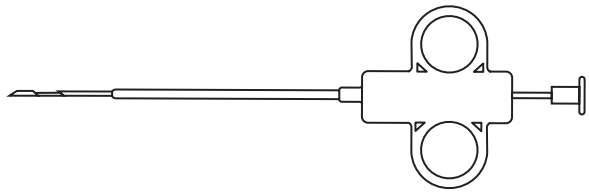
Liver biopsy

INDICATIONS

Percutaneous liver biopsy can be used to evaluate generalized liver disease, liver masses, or small focal lesions. In all these cases we prefer to use ultrasound guidance.

TECHNIQUE

3a-d We use a coaxial technique for almost all liver biopsies. For generalized liver disease we use a coaxial technique with a 17-gauge outer needle and 18-gauge biopsy needle. A subcostal approach is almost always possible, and this avoids the risks of pneumothorax and hemothorax associated with the traditional intercostal approach. The outer needle is advanced into the liver under ultrasound guidance, taking care to avoid the gallbladder and major vessels. Its trocar is then removed and replaced with a semi-automated core biopsy needle. After obtaining three or four cores of tissue, the tract is occluded with gelatin foam plugs or a slurry of collagen in order to prevent bleeding.



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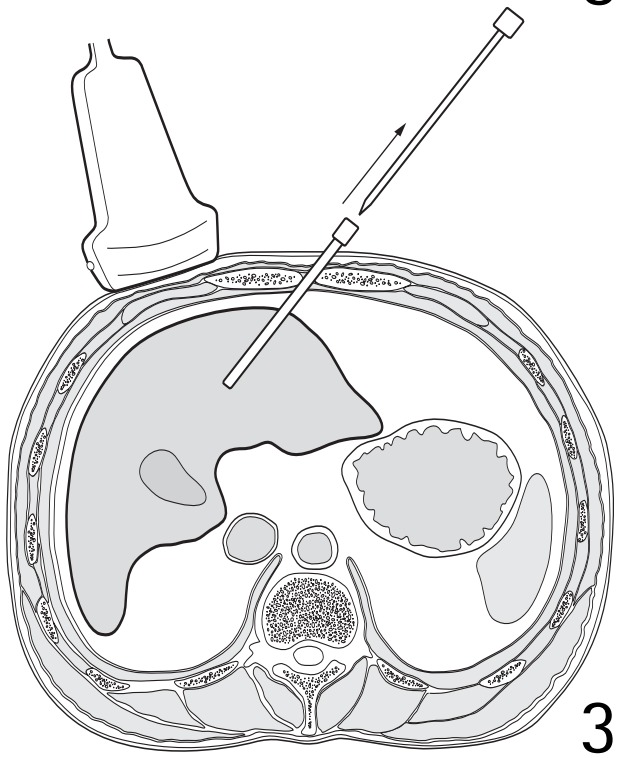

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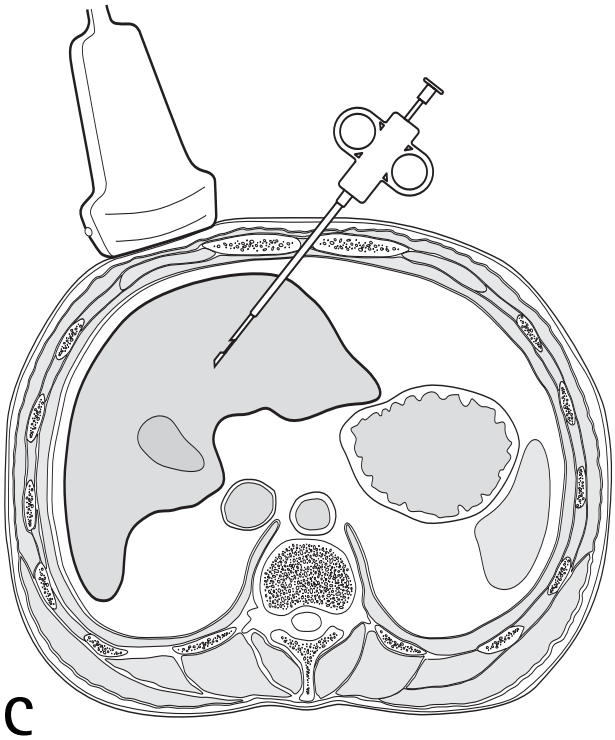

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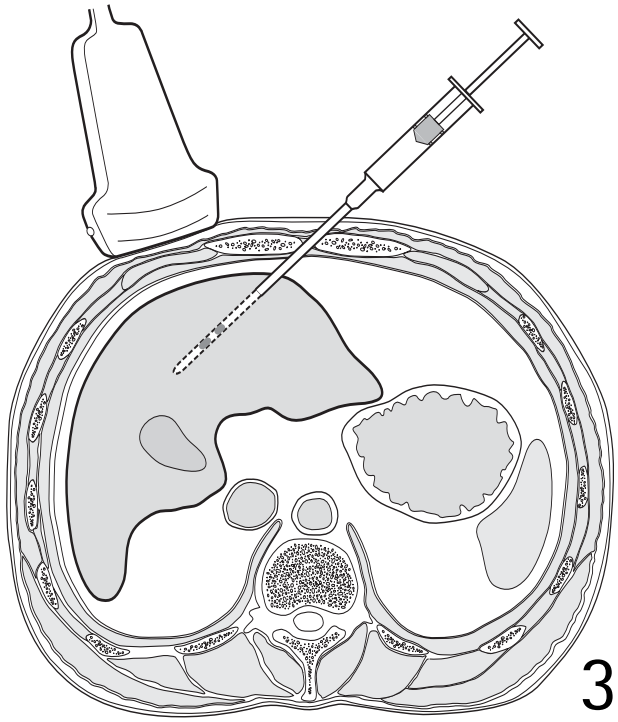


3d

COMPLICATIONS

The most important potential complication of liver biopsy is intraperitoneal bleeding. Frequent meticulous postoperative observations are essential, and should be continued for at least 8 hours. Hemorrhage requiring treatment (by blood transfusion, transarterial embolization, or emergency surgery) is rare in children with normal coagulation parameters. It is more common in children with coagulopathy or thrombocytopenia and in those who have undergone bone marrow transplantation. Transjugular biopsy has been recommended in these patients to try to reduce the risk of post-biopsy bleeding, although it is not clear how effective this is.

Tumor biopsy

INDICATIONS

At our center, the histological diagnosis of most tumors outside the central nervous system is made by ultrasound-guided needle biopsy. Our experience is that needle biopsy is probably safer than, and as accurate as, open (surgical) biopsy. The period of convalescence is shorter, and this may allow earlier commencement of chemotherapy. Laparoscopic biopsy is a useful alternative for certain thoracic and abdominal tumors. Open biopsy is becoming obsolete.

TECHNIQUE

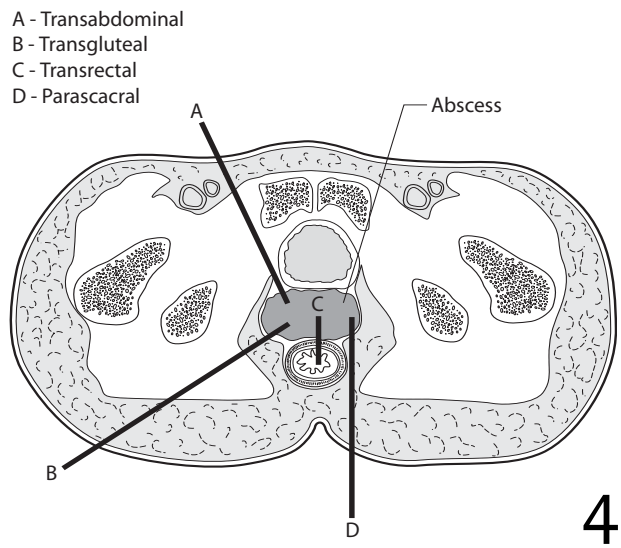
Ultrasound guidance is best for almost all abdominal and soft-tissue biopsies in children. We use computed tomography for very small retroperitoneal lesions, especially in older children, and most lung lesions. Advances in pediatric pathology, including immunohistochemical techniques and the detection of characteristic translocations in tumor tissue, have improved the accuracy of needle biopsy. In most patients, ultrasound-guided tumor biopsy can be performed under the same general anesthetic as insertion of a central venous access device and, where appropriate, bone marrow aspirates and trephines. The coaxial technique (see 'Liver biopsy' above) is particularly useful for tumor biopsy, because numerous cores of tissue can be obtained with a single puncture of the tumor capsule. Plugging the tumor biopsy tract may prevent seeding of tumor cells.

COMPLICATIONS

Complications are very rare. Inadequate biopsies can, in our experience, often be avoided by complete preoperative imaging, and it is usually worth delaying the biopsy for a day or two to make sure that all the relevant imaging is available before the procedure.

Aspiration and drainage of collections

Most fluid collections can be drained by interventional radiology techniques. In the upper abdomen, most collections are approached through the anterior abdominal wall. Certain lesions, such as urinomas or pancreatic collections, are drained retroperitoneally. Pelvic collections can be approached through the anterior abdominal wall, vagina or rectum, or from the buttock.



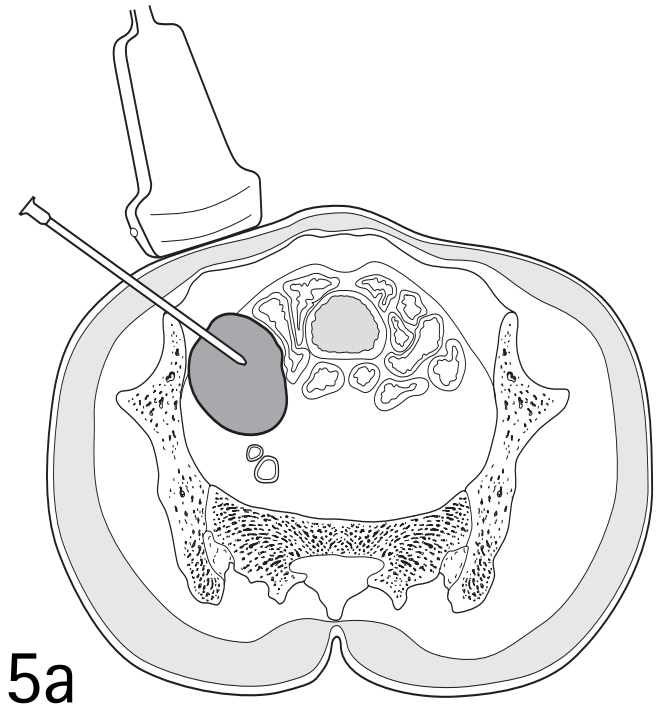
4

TECHNIQUE

5 The usual technique is based on ultrasound-guided puncture of the fluid collection. Guidewire insertion and positioning of the drainage catheter can be monitored with ultrasound or fluoroscopy if necessary. The first step is to select a safe path to the collection by careful ultrasound examination. After injection of local anesthetic, a short stab incision is made in a skin crease and the collection is punctured under real-time ultrasound guidance. We use a trocar needle (usually 18 gauge) for this purpose, because the trocar can be withdrawn slightly into the blunt outer needle, which can then be advanced atraumatically past bowel loops if necessary. When the tip of the needle is in the collection, the trocar is removed and the fluid is aspirated and saved for culture. A guidewire of appropriate diameter is inserted (Table 99.1). The best guidewire has a stiff shaft, and a floppy tip about 30–60 mm long. The floppy tip coils in the collection first, and when increased resistance is felt, the stiff part of the guidewire is in the collection and the needle can be removed.

Table 99.1

Needle	Guidewire diameter gauge
21	0.018 inch (0.46 mm)
20	0.025 inch (0.64 mm)
19	0.035 inch (0.89 mm)
18	0.038 inch (0.97 mm)



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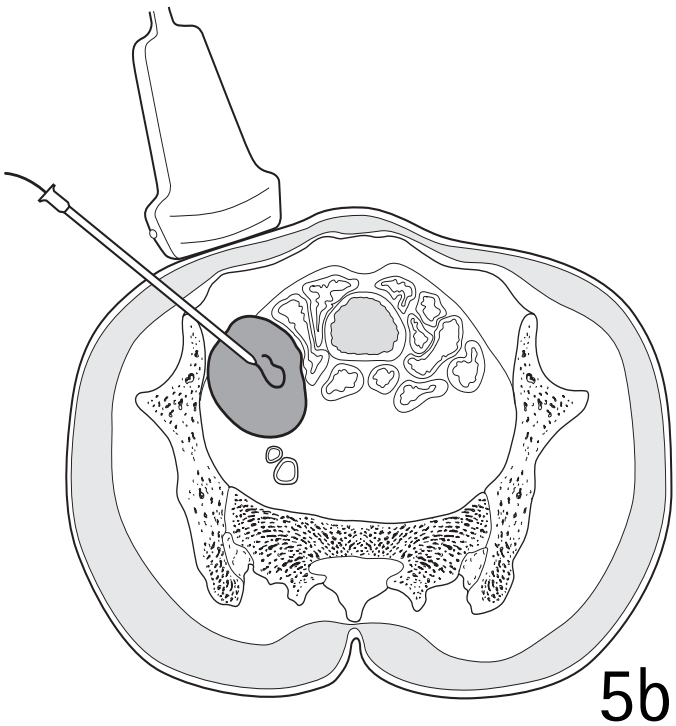

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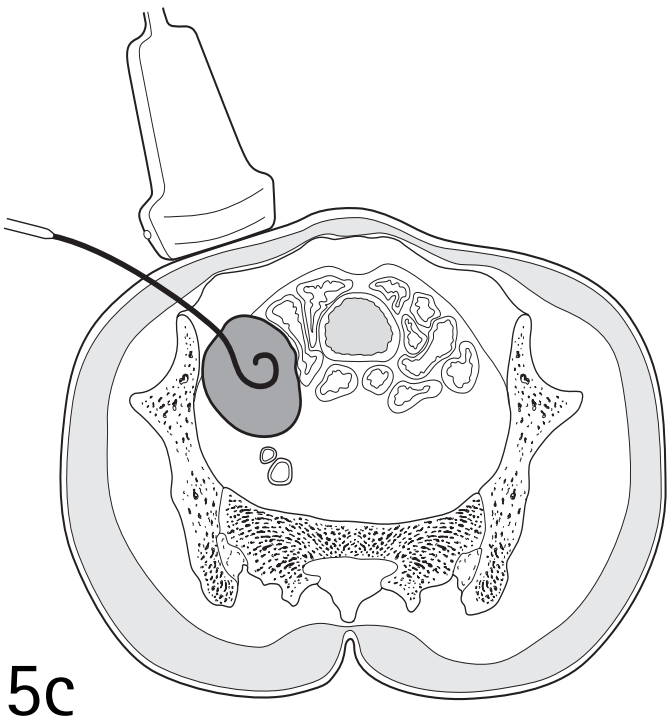
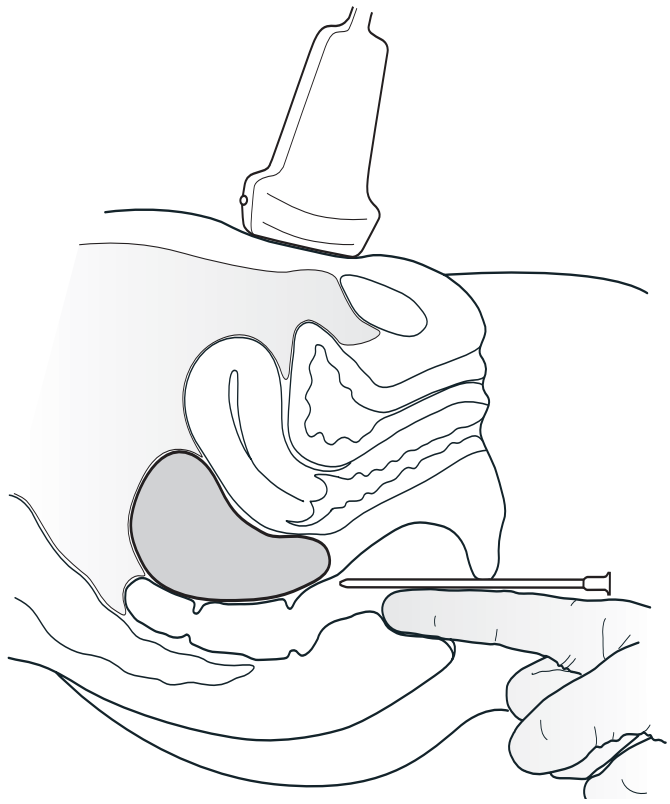


5c

6 We often use a dilator 0.5–1 Fr larger than the intended drainage catheter to establish a tract at this stage. The pigtail catheter (with its internal stiffening catheter) is then advanced over the guidewire and into the collection. The guidewire and stiffener are withdrawn and the pigtail is formed. (There are different methods of doing this, according to the design of the catheter.) Fluid is aspirated to confirm adequate position, and a drainage bag is connected. The catheter can be sutured to the skin.

For transrectal drainage, the needle can be laid along the operator's index finger, with the inner trocar partly withdrawn. When the finger is inserted into the patient's rectum, it is easily seen with transabdominal ultrasound. This enables puncture of a pelvic collection with real-time guidance.



6

GASTROINTESTINAL INTERVENTION

Esophageal dilatation

Fluoroscopically guided balloon dilatation of esophageal strictures is simple and safe, and is replacing bougienage. Endoscopy can be regarded as an additional procedure, with certain advantages over a purely fluoroscopic technique, including the ability to inspect (and possibly biopsy) the esophageal mucosa and to evaluate the stomach and proximal small intestine.

INDICATIONS

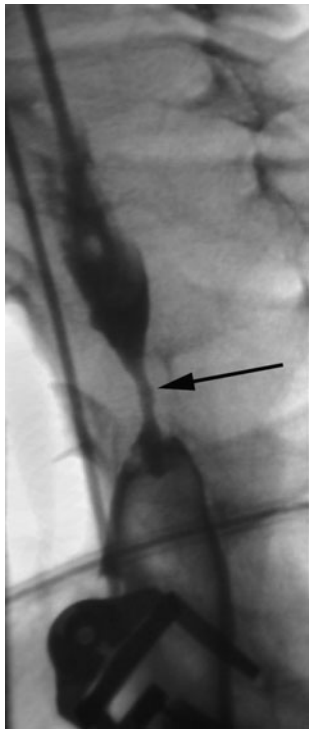
The common causes of esophageal stenosis in children include congenital and surgical strictures, ingestion of corrosive substances, gastroesophageal reflux, achalasia, immunodeficiency, and dystrophic epidermolysis bullosa. The natural history, response to treatment, and probability of complications all depend strongly on the underlying cause of the stricture. Repeated dilatation is often necessary, especially in children with corrosive injury and epidermolysis bullosa. Resection of esophageal strictures (except for congenital stenosis) is rarely necessary.

ANESTHESIA

Although it is possible to dilate the esophagus in a sedated child, we prefer to use general anesthesia, because it is less distressing for the child and seems to minimize the risk of severe vagal response. Endotracheal intubation avoids the severe airway compression that may otherwise occur during balloon dilatation of strictures in small children.

TECHNIQUE

7a–c Most strictures can be treated with balloon dilatation using an antegrade (transoral) approach. A suitably shaped catheter is inserted into the upper esophagus with fluoroscopic guidance, and a contrast study is performed. (Illustration a shows a stricture in the cervical esophagus (arrow) outlined by contrast.) The stricture is then crossed with a guidewire (b), which is advanced to the stomach. The catheter is removed, and replaced with an angioplasty catheter with a balloon of appropriate diameter. The angioplasty catheter is advanced over the guidewire until the radio-opaque markers indicating the position of the balloon straddle the stricture. The balloon is then inflated with dilute radiographic contrast (c). Fluoroscopy is essential at this stage to confirm that the stricture has been successfully dilated, as shown by abolition of the waist on the balloon. The balloon is then deflated, and the catheter and guidewire are removed. Occasionally, it is impossible to cross the stricture from above. When this occurs in children who have a gastrostomy, a retrograde (transgastric) approach is almost always successful. In children whose strictures recur relentlessly after dilatation, mitomycin application and/or temporary stenting can be performed.



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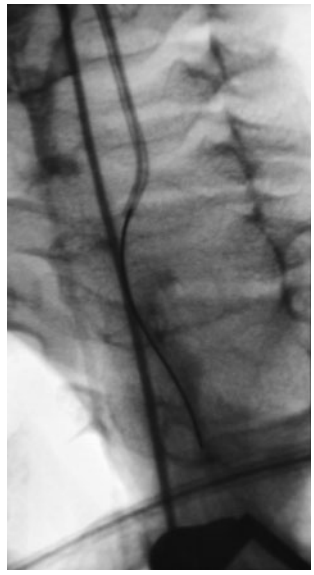

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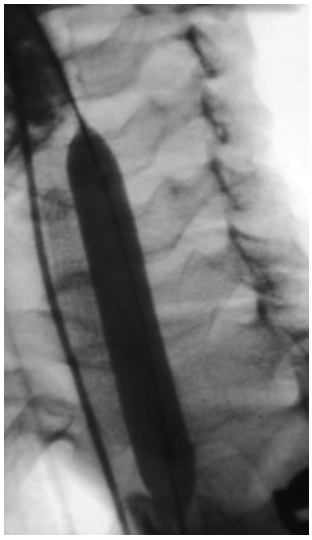


7c

COMPLICATIONS

Complications occur most often during dilatation of corrosive strictures, followed by those associated with gastroesophageal reflux. Three types of esophageal injury occur. Guidewire perforation of the esophagus can be recognized at fluoroscopy and treated conservatively. Submucosal tears of the esophagus are seen as contained extravasation of contrast alongside the inflated balloon. These also seem to do well with conservative treatment. Full-thickness perforation of the esophagus is uncommon, but requires nasogastric or parenteral feeding, broad-spectrum antibiotic therapy, and possibly surgical repair.

Other gastrointestinal dilatation

Balloon dilatation may also be used to treat strictures elsewhere in the gastrointestinal tract, including the pylorus, duodenum, colon, and rectum.

Gastrostomy

Radiologic gastrostomy can be performed by either an antegrade technique, in which the gastrostomy tube is pulled down the esophagus into the stomach, or a retrograde technique, in which it is pushed into the stomach through the

anterior abdominal wall. The antegrade technique is an alternative to the use of endoscopy. The retrograde technique may be useful in children with a narrow or diseased esophagus. The perioperative care is the same as for endoscopic gastrostomy.

TECHNIQUE

The antegrade and retrograde techniques are similar. We usually give the child a small amount (1–2 mL/kg) of barium sulfate suspension orally or by nasogastric tube the evening before the procedure, to opacify the colon. Ultrasound is used to mark the position of the left lobe of the liver. A puncture site is selected, where possible lateral to the rectus abdominis, about 2 cm from the left costal margin. Occasionally, preliminary fluoroscopy shows an anatomical variant that requires the use of a different puncture site. Local anesthetic is infiltrated and a short transverse incision is made in the skin. For antegrade gastrostomy, a snare is passed into the stomach through the mouth at this stage. The stomach is then inflated with air. This can be done via the snare catheter or a separate nasogastric tube. In children with esophageal atresia, the stomach can be inflated after ultrasound-guided puncture with a fine needle. When the stomach is inflated, an 18-gauge trocar needle is inserted through the incision and used to puncture the stomach. This must be done quickly, before the air escapes from the stomach into the small bowel. We find biplane fluoroscopy useful at this point, but it is not essential. Some operators use intravenous glucagon (40 μg/kg, maximum 1 mg) to delay gastric emptying. A small amount of water-soluble contrast may be injected to confirm that the needle has entered the stomach. The snare is opened in the stomach, and a 0.035 inch or 0.038 inch stiff guidewire is advanced through the needle and grasped with the snare. The wire is then carefully pulled out of the mouth, and the tract dilated to permit the retrograde passage of the snare catheter through the abdominal wall to the mouth. The snare catheter is then used to pull the lubricated gastrostomy tube down the esophagus, and out through the anterior abdominal wall. The procedure is otherwise the same as for endoscopic gastrostomy (see Chapter 37).

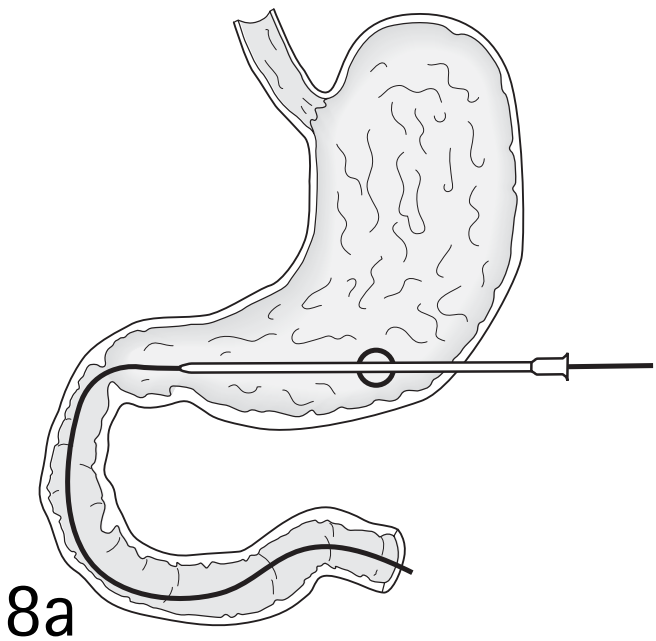
For retrograde gastrostomy, the stomach is punctured in the same manner as for the antegrade technique, but a needle preloaded with a suture anchor device may be used instead of the trocar needle. These temporary retention devices are used to hold the stomach against the anterior abdominal wall as the tract is dilated. A locking pigtail catheter is usually inserted. Although these are more easily dislodged than flanged gastrostomy catheters, they are easy to remove without anesthesia when the tract is mature. An alternative is to insert a transgastric jejunal catheter (with a locking pigtail in the stomach and a long jejunal tip) at the time of creation of the gastrostomy.

COMPLICATIONS

Complications are similar in incidence and nature to those of endoscopic gastrostomy.

Transgastric jejunal intubation

8 This requires the passage of a transpyloric tube through a pre-existing gastrostomy to a position distal to the duodenojejunal flexure, using fluoroscopic guidance. The easiest way to cross the pylorus if there is a mature gastrostomy tract is to remove the gastrostomy catheter and insert a large-bore jejunal tube, dilating the gastrostomy tract if necessary. A stiff dilator (9–15 Fr) is used to engage the pylorus, and a floppy-tipped hydrophilic guidewire is advanced to the jejunum. The dilator is removed, and a long catheter (we use a 7 Fr multipurpose angiographic catheter) is inserted over the guidewire. The floppy-tipped wire is then exchanged for a stiff wire, and the catheter is removed. The transgastric jejunal tube is then lubricated (internally and externally) and inserted over the stiff wire. Injection of contrast through the catheter will confirm its position. Coaxial systems, in which a transpyloric tube can be advanced through a gastrostomy catheter, are also available.



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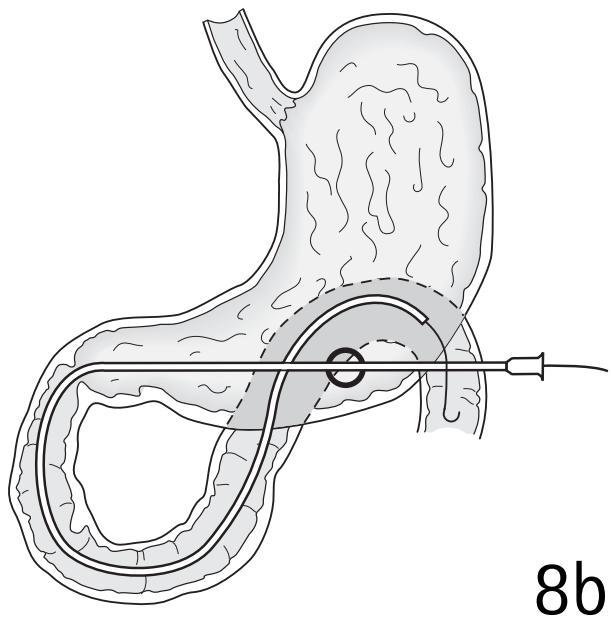


8b

UROLOGIC INTERVENTION

Most urologic interventions are based on nephrostomy. This is usually a simple procedure in children when performed with ultrasound guidance, except when the pelvicalyceal system is not dilated. The nephrostomy tract can also be used for other interventions such as percutaneous stone removal or antegrade ureteral dilatation and stenting.

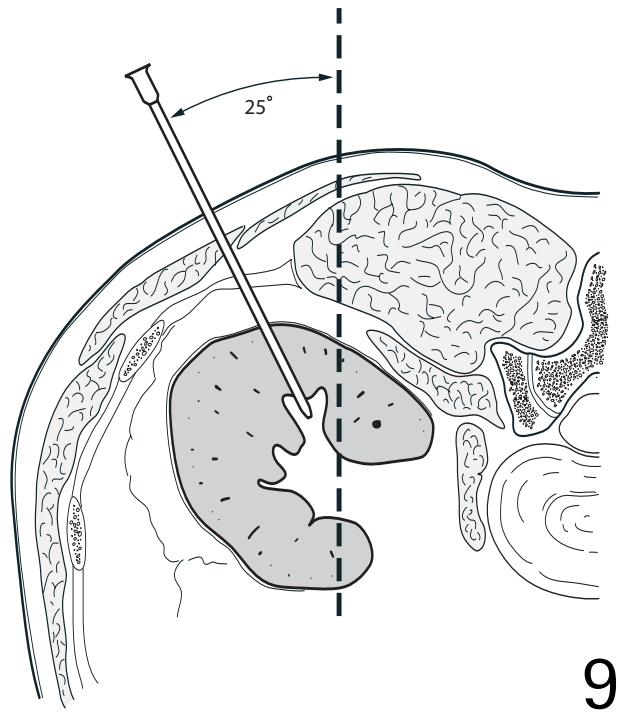
Percutaneous nephrostomy

INDICATIONS

The most common indications for nephrostomy in children are congenital and postoperative obstruction. It is customary to give intravenous antibiotic prophylaxis to reduce the risk of septicemia.

POSITIONING AND ANESTHESIA

9 Unilateral nephrostomy is best performed with the patient in a semi-prone position and the affected side tilted up about 20–30°. This facilitates puncture of a posterior lower pole calyx through the relatively avascular zone of Brödel. Although sedation is an acceptable alternative, we prefer to perform bilateral nephrostomy under general anesthesia with endotracheal intubation, with the patient prone. This requires angling the needle at about 20–30° to the tabletop to achieve the same trajectory. In addition, the tip of the needle is angled cranially, to make it easier to see at ultrasound. This also permits puncture of an interpolar or upper pole calyx, which is sometimes required for ureteral interventions or stone removal.

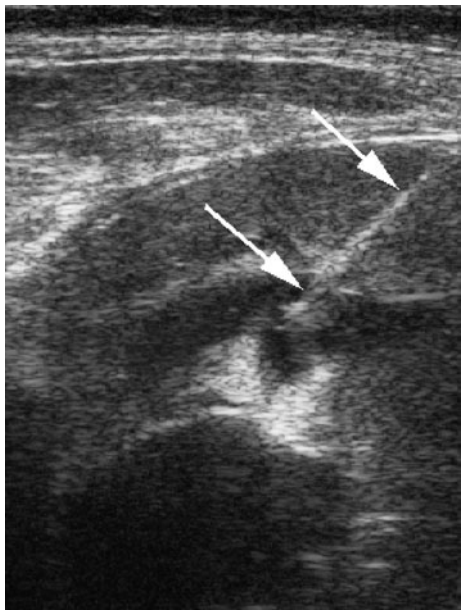


9

TECHNIQUE

10a–c The exit site and proposed needle tract should be injected with local anesthetic. Using real-time ultrasound guidance, a dilated calyx can almost always be punctured with a single needle pass. Various types of needle are available for this purpose. Trocar or sheathed needles have the advantage that when the trocar is removed the outer part of the needle (or sheath) has a blunt tip, which is unlikely to damage the collecting system. Needles with echogenic tips are easier to see at ultrasound, but are not necessary in children. Once the calyx has been punctured, urine is aspirated for culture, and a smaller volume of dilute contrast (to avoid over-distension of the pelvicalyceal system) is then injected to perform antegrade pyelography.

In general, 6 Fr locking pigtail catheters are appropriate for neonates. In older children, and those with suspected pyonephrosis, larger catheters should be used (typically 8.5 Fr). As for abscess drainage, the use of a stiff wire with a floppy tip is recommended. The largest diameter guidewire that can pass through the stiffener of the pigtail catheter is selected, and the smallest needle that accepts the guidewire is used for the puncture. Once the tip of the guidewire is coiled in the renal pelvis, the tract can be over-dilated by a 0.5 to 1 French before inserting the pigtail catheter and stiffener. We usually suture the catheter to the skin. Coaxial access sets, which allow puncture with a 21-gauge or 22-gauge needle, are more difficult to use, but may be helpful when the pelvicalyceal system is not dilated. With these systems, a 0.018 inch guidewire is inserted through the thin needle and then exchanged for a 0.035 inch wire after insertion of a two-part coaxial dilator. The illustration shows nephrostomy in a native kidney. The needle (arrows) can be seen entering a dilated calyx (a). Following tract dilatation, the catheter is inserted over the guidewire using a metal stiffener (arrows, b). In its final position, the pigtail of the catheter is coiled in the renal pelvis, which is outlined with dilute contrast (c).



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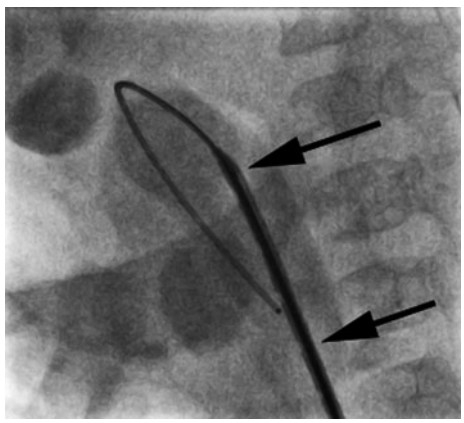

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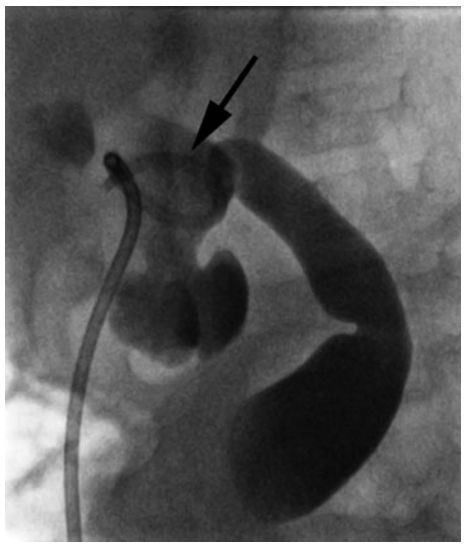


10c

COMPLICATIONS

The most important complication is injury to an intrarenal branch of the renal artery. This may require embolization (see below). Injury to the colon and hemothorax or pneumothorax are unusual.

Ureteral dilatation and antegrade stenting

Ureteral dilatation is most often required in children who have transplant ureteral strictures or postoperative vesicoureteral junction obstruction. Antegrade access is obtained as for nephrostomy, but preferably using a mid-pole (or, if possible, an upper pole) calyx. The renal pelvis and ureter are opacified with dilute contrast. A peel-away sheath is inserted and the stricture is crossed with a hydrophilic guidewire and a catheter of appropriate shape (e.g., Cobra). With the tip of the guidewire in the bladder, a balloon dilatation catheter is positioned across the stricture and inflated with fluoroscopic guidance. A small-diameter balloon (3–5 mm, depending on the size of the child) is usually adequate.

A double-J stent can be placed in the ureter by pushing it over a guidewire with a pusher catheter. A protective nephrostomy tube is probably not necessary unless there is significant bleeding. An alternative to cystoscopic stent removal is to snare the distal end through a Foley catheter under fluoroscopic guidance. Occasionally, transurethral removal is difficult, for example when the lower end of the stent migrates up into the ureter. The stent can then be removed percutaneously by snaring, after puncture of the pelvicalyceal system.

BILIARY INTERVENTION

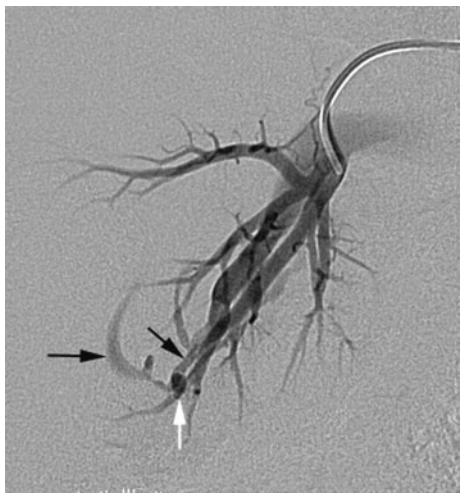
Diagnostic percutaneous transhepatic cholangiography (PTC) is obsolescent. One potential indication is sclerosing cholangitis (at our center, this is seen most often in children with Langerhans' cell histiocytosis), when PTC can be performed at the same time as liver biopsy. When the intrahepatic bile ducts are not dilated, the simplest method is to puncture the gallbladder under ultrasound guidance. Percutaneous transhepatic cholangiography with or without biliary drainage or balloon dilatation of biliary strictures may be required following liver transplantation. Malignant obstructive jaundice is unusual in children; biliary drainage is required in some patients. In these circumstances, ultrasound-guided puncture of a dilated intrahepatic duct allows insertion of an external drainage catheter.

VASCULAR INTERVENTION

11 Diagnostic arteriography and venography are required much less often now than in the past, but interventional angiography is increasing in importance. Arterial embolization is an effective method of controlling iatrogenic or other traumatic hemorrhage. An example is symptomatic arteriovenous fistula following renal biopsy. These lesions may be embolized with coils, which are easy to use and very effective. The aim is to preserve as much renal tissue as possible.

Arterial embolization may also be used for the treatment of arteriovenous malformations and tumors. The management of vascular malformations and arterial interventions such as angioplasty and stenting are beyond the scope of this chapter.

The illustration shows the embolization of an arteriovenous fistula in a renal transplant recipient who developed hypertension after a renal biopsy. In a, there is a complex arteriovenous fistula (white arrow), with two prominent, early-filling veins (black arrows). It was necessary to perform coil occlusion quite proximally in order to control the fistula (b). The hypertension was cured.



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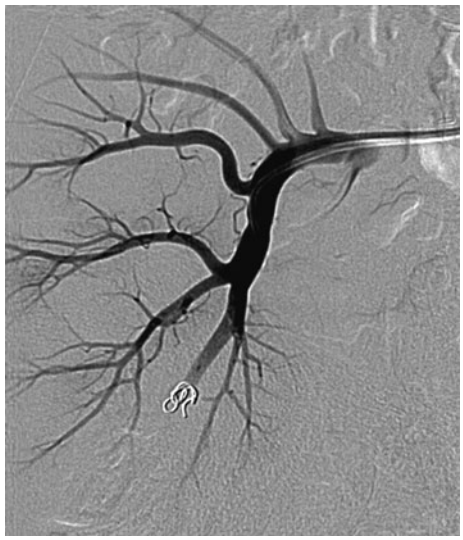

11b

TRACHEOBRONCHIAL INTERVENTION

These techniques are based on bronchography. Balloon dilatation of acquired tracheobronchial stenoses is easy and often effective. The procedure is analogous to esophageal dilatation. The airway can be accessed through an endotracheal or tracheostomy tube, or a laryngeal mask airway. The measurement facilities on modern angiographic equipment are helpful here, because accurate assessment of the diameter and length of the balloon to be used is very important. Fluoroscopic guidance is useful because it allows accurate positioning of the balloon, as well as confirmation that complete inflation has been achieved. A description of stenting and other pediatric airway intervention is beyond the scope of this chapter.

FURTHER READING

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Bariatric surgery – principles

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HISTORY

As the epidemic of pediatric obesity has been increasingly documented, and the efficacy and safety of bariatric surgery for adults have also become evident, more consideration has been given to bariatric procedures for clinically severely obese adolescents. Over the past 30 years, many bariatric procedures with various modifications have been introduced, including intestinal bypass (jejunocolic or jejunoileal), loop gastric bypass, horizontal and vertical gastroplasty, Roux-en-Y gastric bypass (RYGBP), biliopancreatic diversion (with or without duodenal switch), and, most recently, adjustable gastric banding. From the very beginning of the subspecialty of bariatric surgery, the goal of operation was either to restrict the intake of nutrients or to interfere with the absorption of nutrients that are ingested, or both. The long-term goal is to maintain a degree of weight reduction that improves or eliminates obesity-related comorbidities or decreases the risk of future obesity-related medical complications and death.

PRINCIPLES AND JUSTIFICATION

Currently, minimally invasive techniques are being used for all of the modern weight-loss procedures. These include the RYGBP, the vertical banded gastroplasty, the adjustable gastric band, and the biliopancreatic diversion with duodenal switch. The procedure that has been used most commonly in the United States is the RYGBP, as this procedure has been associated with the safest side-effect profile, balanced with excellent long-term efficacy and maintenance of weight loss. This operation effectively creates a 'tool' with which adolescents can lose one-third or more of body mass and concomitantly reduce or eliminate most comorbidities of adolescent obesity.

This chapter focuses specifically on:

- the perioperative preparation of the patient and family once the decision for surgery has been made,
- technical aspects of laparoscopic RYGBP,
- postoperative management.

The National Institutes of Health Bariatric Consensus Development Conference in 1991 concluded that two bariatric procedures, namely vertical banded gastroplasty and RYGBP, should be options for weight management for adults with morbid obesity given the recognized morbidity and mortality of uncorrected severe obesity. This conference concluded that there were insufficient data to make recommendations about bariatric surgery for patients younger than 18 years of age. Significant, durable weight loss and long-term medical benefits have now been documented for adults who have undergone RYGBP. For this reason, current opinion holds that bariatric surgery should also be an option for highly selected adolescents with extreme obesity. Since relatively little is known about the long-term sequelae of bariatric surgery when performed in adolescence, one should use relatively conservative indications for operation. Surgery should be reserved as a last-resort measure after at least 6 months of organized weight-loss attempts have failed. In general, surgery should be considered only for those individuals with comorbidities of obesity.

For those with a body mass index (BMI) ≥ 40 kg/m^2 and severe comorbidities such as obstructive sleep apnea syndrome, type 2 diabetes, or pseudotumor cerebri, bariatric surgery should be considered because clinical experience has demonstrated that (1) these conditions are life threatening, and (2) they are very effectively managed with surgical weight reduction. Bariatric surgery may also be an appropriate option when obesity is more severe (e.g., BMI ≥ 50 kg/m^2) in the presence of somewhat less urgent comorbidities of obesity

such as dyslipidemias, hypertension, degenerative joint disease, asthma, venous stasis, intertriginous infections, gastroesophageal reflux, social discrimination, polycystic ovary syndrome, or metabolic syndrome. Finally, in some individuals with BMI ≥ 50 kg/m^2, activities of daily living (including personal hygiene and school attendance) are difficult, if not impossible. In these cases, surgery should also be considered. The reader is referred to other sources for further discussion of the contraindications and preoperative decision making for adolescent bariatric surgery.

Bariatric programs for adolescents should include expertise in adolescent obesity, nutrition, diet, and behavioral management. It is critical that thorough investigations be conducted to discover unrecognized coexisting obesity-related medical conditions. Minimally invasive surgery has significant advantages over open surgery, including a reduction in length of hospitalization and operative morbidity. Minimally invasive bariatric surgery is one of the most technically difficult operations to perform. Laparoscopic skills utilized in foregut surgery are not directly transferable to bariatric surgery. Expertise in minimally invasive surgery may not confer the same level of expertise in performing minimally invasive bariatric surgery. Surgeons performing bariatric procedures must be well trained, as suggested by the American Society for Bariatric Surgery, American College of Surgeons, and the Society of American Gastrointestinal Endoscopic Surgeons. Prior to performing laparoscopic bariatric operations, surgeons must meet all local credentialing requirements for the performance of open bariatric procedures and advanced laparoscopic operations. Given the controversy centered on adolescent bariatric surgery, the pediatric surgeon aspiring to be a bariatric surgeon should, at a minimum, take a course in bariatric surgery and perform his or her first ten procedures proctored by an experienced laparoscopic bariatric surgeon.

PREOPERATIVE ASSESSMENT AND PREPARATION

The preoperative education of the patient, family, and, to some extent, the referring pediatrician, is of paramount importance. Information about the surgical procedure, alternative operations and the rationale for the operation to be performed, postoperative care, and the considerable lifestyle modifications required afterward must be conveyed verbally and in writing to the patient and caregivers. The adolescent and parents must demonstrate understanding of the details of the procedure, its known and potential risks, and predictable consequences, and that understanding should be reflected in the archived responses to a written test. The referring pediatrician will need to become familiar with the anatomic and physiologic changes that occur after operation and develop an understanding of common postoperative complications to partner effectively with the surgeon in the care of the patient for years following the operation.

On the day prior to surgery, patients are limited to clear liquids. No bowel preparation is required. Preoperative medications include low-molecular-weight heparin (40 mg subcutaneous and continued twice a day postoperatively while in the hospital) and a second-generation cephalosporin (2 g intravenously). Sequential compression boots are also used intraoperatively and postoperatively and should be applied and functioning prior to induction of anesthesia. Patients who have BMI < 50–60 kg/m^2 are appropriate candidates for laparoscopic gastric bypass, depending on the experience of the surgical team. In those with a higher BMI, or in those with prior upper abdominal surgery, an open laparotomy is considered safer than laparoscopic surgery. There are no minimally invasive pediatric surgical procedures more technically challenging than the laparoscopic RYGBP.

Anesthesia

Patients undergo general anesthesia for laparoscopic RYGBP. There is generally no need for epidural analgesia. Routine preoperative consultation with the anesthesiologist and cardiologist is recommended to uncover potentially important information about the patient's airway, the patient's or family's anesthetic history, and to determine if occult cardiovascular disease is present. Preoperative anesthesia consultation also provides an opportunity for unhurried discussion of any concerns or questions the family or patient may have about the technique and risks of anesthesia.

Equipment

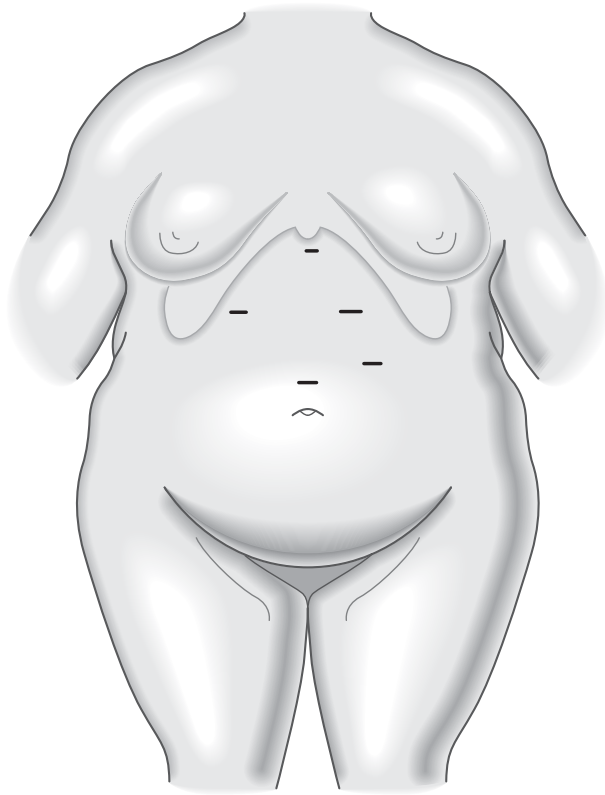
Although extra long (45 cm) laparoscopic instrumentation is available for bariatric procedures, in general, laparoscopic RYGBP can be performed using standard 32 cm adult instrumentation. Various manufacturers supply the equipment required and the choice of one over another is largely a matter of surgeon preference. Specific instrumentation used at Cincinnati Children's Hospital for this procedure includes:

- four 10/12 mm ENDOPATH Optiview trocars (Ethicon Endosurgery),
- 1 percent lidocaine, 10 cc syringe, and a long (spinal) needle,
- one zero-degree, 32 cm laparoscope (for initial abdominal access with ENDOPATH Optiview trocar),
- one 30 degree, 32 cm laparoscope,
- one Nathanson liver retractor (Cook Surgical, Bloomington, IN),
- one 34 Fr orogastric tube (Kimberly Clark, part #15034),
- one esophageal retractor (blunt dissector; Pilling Weck, part #38-1802),
- one locking, atraumatic bowel grasper,
- two non-locking atraumatic bowel graspers,
- one hook scissor for cutting suture,
- one needle driver,

- one Wolfe bipolar electrocautery forceps,
- one ultrasonic dissector,
- one ENDOPATH Endoscopic Linear Cutter 45 mm stapler, 2.5 mm staple (white loads; Ethicon part # ATB45),
- two ENDOPATH Endoscopic Linear Cutter 35 mm staplers, 3.5 mm staple (blue loads; Ethicon part # TSW35),
- multiple 18 cm lengths of 2/0 silk and 3/0 dyed Vicryl on SH needles,
- one Jackson Pratt flat drain.

OPERATION

1 This procedure is primarily a 'one-surgeon' procedure, with the assistant's role largely that of tissue retraction. Thus, the experienced laparoscopic bariatric surgeon can perform this operation without another trained bariatric surgeon as an assistant. During laparoscopic RYGBP, the patient can be flat or in gentle reverse Trendelenberg position with the legs together. The surgeon stands on the right, the scrub nurse to the surgeon's right side, and the assistant on the left side of the patient. Given the patient's extreme girth, it is ergonomically advantageous to stand on a platform 20–40 cm above the floor. For initial abdominal access, the transparent, bladeless, direct viewing (Optiview®, Ethicon Endosurgery [EES], Cincinnati, OH) 12 mm trocar can be used safely and efficiently. This trocar is placed through the midline approximately 10 cm above the umbilicus. The exact site for this trocar is determined by lying a standard 10 mm adult laparoscopic telescope on the patient's abdomen, with the tip of the scope (the end that will be inside) at the nipple level, marking a skin site at the point at the same level as the light cord is inserted on the scope. This skin site will provide optimal visualization of the upper and lower portions of the procedure. The three other 12 mm trocar placement sites and the 5 mm subxiphoid site for liver retraction are shown in the illustration.

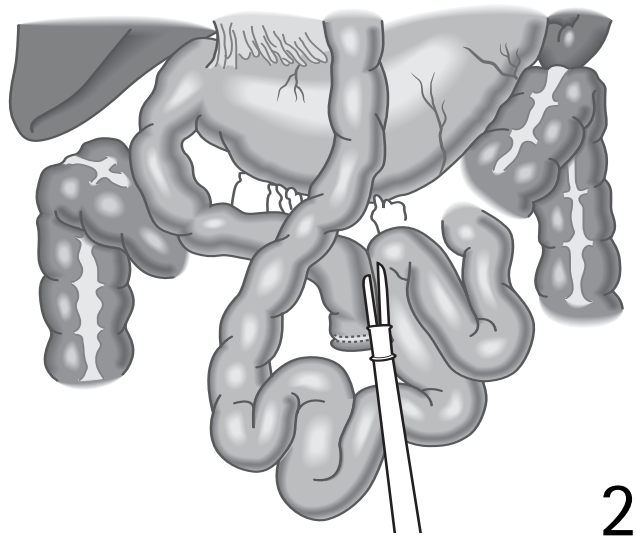


1

Roux limb construction

The omentum is first raised cephalad to expose the transverse mesocolon and origin of the jejunum. Exposure of the small bowel is optimized if the table is placed into slight Trendelenberg position and the omentum is tucked carefully under the liver edge. The transverse mesocolon is grasped with a locking grasper just anterior to the duodenojejunal flexure and elevated anteriorly. The jejunum is divided 25–75 cm beyond the duodenojejunal flexure using the 45 mm endo-GIA (EES, white load). If using a retrocolic construction, the length from the flexure can be 25–30 cm, while an antecolic Roux requires more distance (50–75 cm) to reach comfortably the pouch in the epigastrium. The mesentery is minimally divided at this point using half the length of a second stapler load. Clamping the stapler jaws together for a 10-second count before firing mechanically thins the highly vascular tissue to reduce bleeding from the staple. Bipolar electrocautery is often needed to achieve full hemostasis in this area.

2 There is no unanimity about the exact length of the Roux limb. Current practice employs a 100 cm Roux limb for most patients. With experience, the limb length can be visually estimated by 'walking' hand over hand down the bowel with laparoscopic graspers. Precision in measurement can be increased by using a Penrose drain of known length alongside the bowel. Next, a single stay suture is placed to approximate the antemesenteric borders of the distal end of the biliopancreatic segment (the proximal jejunum) and the point 100 cm distal to the end of the Roux limb. The ultrasonic scalpel is used to make opposing enterotomies, which are spread wide enough with a grasper to accept the stapler jaw. A side-to-side jejunojejunostomy is created using the 45 mm EES stapler with white load, and the resulting enterotomy defect is closed either with a running 3/0 Vicryl (SH needle) or by using another firing of the stapler. There is an increased risk of narrowing the lumen if the stapled closure is not applied precisely. The mesenteric defect is next closed with a running 2/0 silk suture to avoid an internal hernial orifice.



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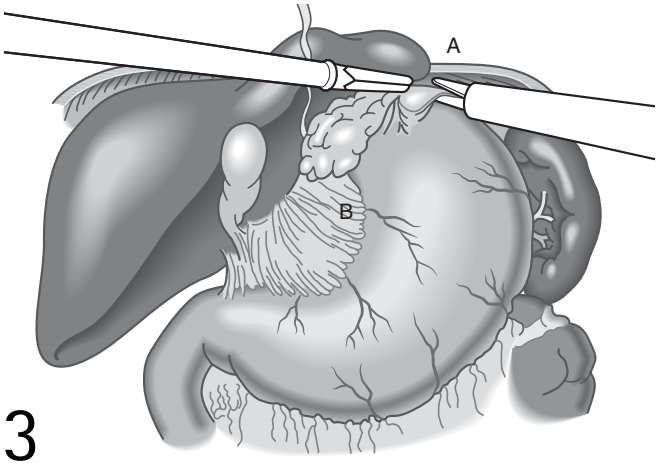
The Roux limb can be tunneled cephalad in a retrocolic fashion, or brought up in an antecolic position. If the retrocolic technique is used, a mesocolic defect will be created through which the Roux limb will pass into the lesser sac. This defect will need to be closed around the Roux limb, and Petersen's defect between the small bowel mesentery and the mesocolon will need to be deliberately closed as well, to prevent internal hernia formation postoperatively. Internal hernias are not more common with the antecolic technique. Since the antecolic technique is technically more straightforward and does not risk troublesome bleeding in the transverse mesocolon, it is a method favored by many high-volume bariatric surgeons. If the antecolic technique is used, the omentum must be draped back inferiorly, and bivalved up to the transverse colon, to reduce the bulk of tissue that the Roux limb will traverse. This omental division also reduces the tension (and thus the risk of stricture) transmitted from the Roux limb to the anastomosis when the patient assumes an erect posture postoperatively. Finally, some surgeons use the bivalved omentum to 'wrap' the gastrojejunal anastomosis at the conclusion of the operation. This technique has theoretic benefit in the event of leakage of the gastrojejunal anastomosis. With an omental wrap, an otherwise uncontrolled leak with resultant diffuse peritonitis may instead become a contained leak that is manageable nonoperatively.



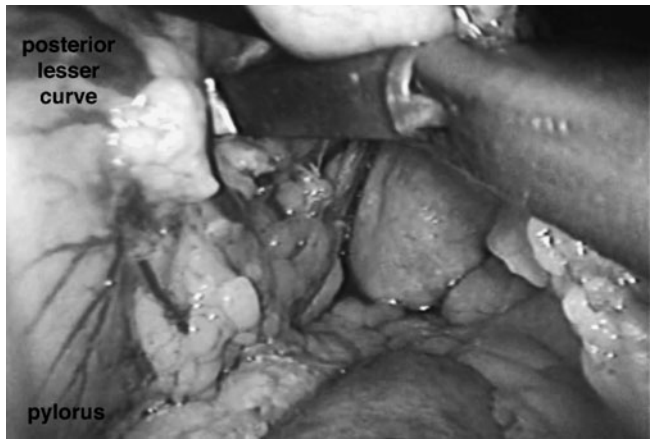
3

Gastric pouch construction

3 The Nathanson retractor is next inserted below the xiphoid and used to retract the left lobe of the liver anteriorly to expose the gastroesophageal junction. The lesser curve gastric pouch will be created around a 34 Fr orogastric tube by beginning with the dissection at the angle of His. Once a sufficient plane has been created between the stomach and diaphragm at the angle of His, the esophageal retractor is used as a blunt dissector to better develop this plane along the length of the left crus.

The creation of the gastric pouch begins with perigastric ultrasonic dissection along the lesser curvature approximately 8–10 cm inferior to the gastroesophageal junction (usually just below the second major lesser curve vessel). The 'flat' side of the blade is used to insure adequate hemostasis during this dissection. Bipolar electrocautery is also commonly needed. This dissection is continued posteriorly in close proximity to the gastric wall until the plane nearly reaches the lesser sac behind the stomach.

4 The greater curvature of the stomach is next elevated and a thin area of the gastrocolic ligament is chosen for entry into the lesser sac, using ultrasonic or hook cautery dissection. Numerous posterior attachments between the lesser curve and retroperitoneum are divided to achieve continuity between the lesser curve dissection that was performed anteriorly. The blunt esophageal dissector is directed from the patient's left to right and placed into the lesser sac behind the stomach and directed through the window created along the lesser curve.



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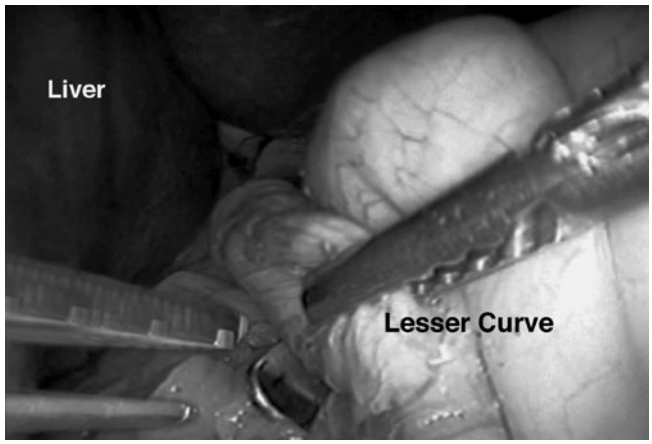

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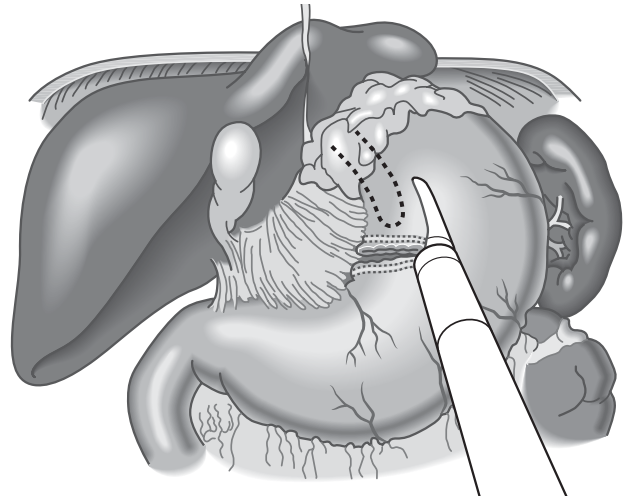
5 The dissector is advanced gently such that the tip traverses the posterior surgical plane to emerge anteriorly when the stomach is released. A tubular lesser curve pouch is next created using the endo-GIA 35 mm stapler with blue loads. The first transverse cut across the lesser curvature is achieved by bringing the stapler into the right upper quadrant port, with the esophageal dissector in place traversing the lesser curve dissection plane created above. The tip of the stapler is brought into contact with the esophageal dissector in the open-jaw orientation and advanced across the lesser curve as the esophageal dissector is backed out. Once in place across the lesser curve, the stapler is fired, creating the inferior-most margin of the pouch. Next, gentle dissection is performed at the apex of the staple line to break completely into the retrogastric lesser sac from above. This simple maneuver is critical as the procedure is far less difficult if the lesser sac is identified and entered from the anterior aspect at this point.

6 The esophageal dissector is now placed through this window into the lesser sac and is used to retract the greater curve of the fundus anteriorly and to the patient's left to facilitate placement of the 34 Fr orogastric tube along the lesser curve of the gastric lumen. The tube is advanced by the anesthesiologist until the tip comes to reside at the level of the transverse cut across the lesser curve. The tube is 'trapped' in this location by application of the endo-GIA in a vertical direction to begin the longer staple line toward the angle of His.



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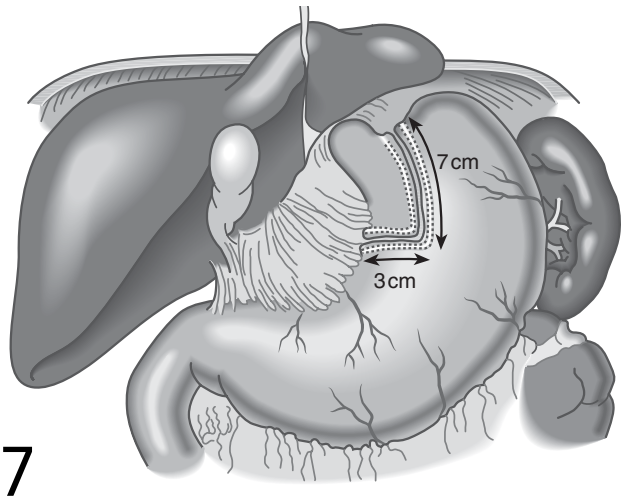

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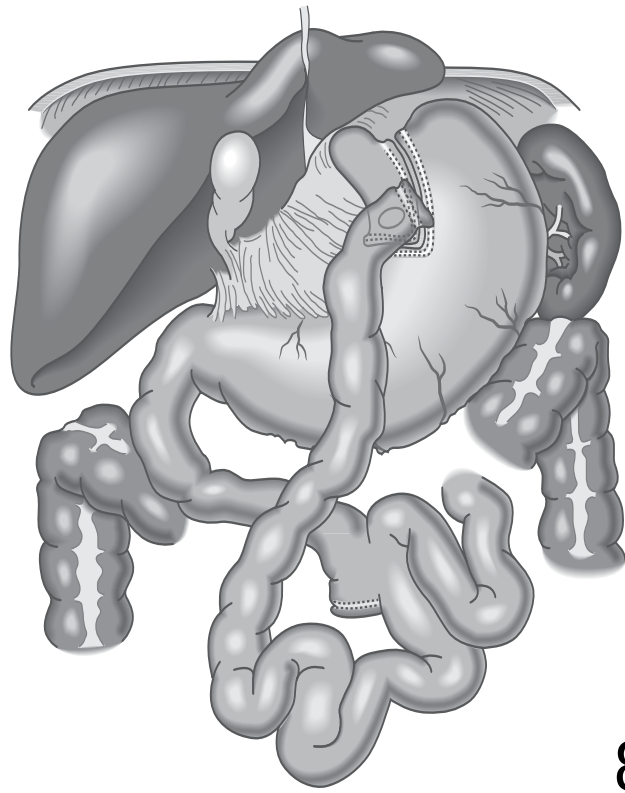
7 This tube sizes the pouch uniformly along its length. The vertical staple line is advanced sequentially by multiple applications of the endo-GIA. Once the angle of His is reached, the pouch is completed.

Gastrojejunal anastomosis

There are numerous techniques for laparoscopic gastrojejunostomy, including hand sewn, the end-to-end stapled technique with anvil inserted into the pouch laparoscopically, the end-to-end stapled technique with the anvil inserted orally (akin to the percutaneous endoscopic method of gastrostomy), and a linearly stapled technique. Some surgeons favor the two-layer, hand-sewn anastomosis, as this method has been found in a large series to have an unequaled 0 percent anastomotic leak rate, is less expensive, and is time efficient. These findings, along with the simplistic elegance of the technique and similarity to the open procedure, make it an attractive option. When considering choices of technique, it is usually best to learn from someone who will be readily available for technical consultation, and to adopt the technique that he or she employs.

The remainder of this chapter focuses on details and pearls of wisdom for the performance of the hand-sewn gastrojejunostomy technique.

8 The anastomosis is created like any other hand-sewn anastomosis done by open laparotomy, by first bringing the Roux limb into proximity with the inferior end of the tubular pouch and running a posterior (outer) suture line using 3/0 Vicryl suture (an 18 cm length of suture on SH needle is commonly used). In this way, the antimesenteric border of the Roux limb is secured to the staple line of the inferiormost aspect of the pouch. The suture is locked and the needle is left attached to discriminate it from the suture used for suturing the anterior (inner) row. Slightly distracted 10 mm enterotomies are next created in the pouch and the jejunum with the ultrasonic scalpel to begin the end-of-pouch to side-of-bowel anastomosis. The orogastric tube should be clamped first to avoid loss of pneumoperitoneum when the pouch is opened. An inner suture is begun at the right corner (at the 3 o'clock position) of the anastomosis and advanced right to left (advanced just beyond the 9 o'clock position of the anastomosis). Once the left corner of the anastomosis has been created, the end is left long and the needle is removed. A second inner suture is advanced from 3 o'clock and tied to the inner suture ending just beyond 9 o'clock. The final outer row is begun at 3 o'clock and tied to the suture left with the needle attached. Thus posterior then anterior full-thickness running layers of the anastomosis are constructed, just as one would do in an open bowel anastomosis. When the anterior running layer is half completed, the anesthesiologist is directed to advance the tube out of the pouch, and the tip is directed into the jejunal lumen, and the remainder of the anterior inner suture line is completed over the tube. Thus, with the tube through the anastomosis, there is assurance of a widely patent anastomosis, with little concern for 'purse-string' narrowing as the anterior and posterior sutures are separate. The anastomosis is completed by placing a final anterior seromuscular layer fully to cover the anastomosis. This layer is tied off to the very first posterior suture line that was originally used to approximate the pouch and jejunum. The integrity and patency of the anastomosis are assessed laparoscopically with intraluminal air insufflation under saline. This pneumatic test is accomplished by temporarily obstructing the Roux limb just beyond the anastomosis and having the anesthesiologist rapidly bolus 60 mL of air into the orogastric tube.



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A drain to prevent fluid collection within the abdomen and to provide for early identification of a leak is left near the anastomosis and exits via the patient's upper left trocar site. A temporary gastrostomy tube is placed in the remnant stomach body if any intraoperative technical challenges are deemed significant enough to warrant deliberate decompression of the bypassed gastric remnant. With the use of the bladeless trocars, the 12 mm port sites do not require fascial closure. In contrast, after a procedure that employs an end-to-end stapling device for the gastrojejunostomy, a 2 cm port is needed and the site usually does require deliberate closure, and these larger port sites are at higher risk of infection and bleeding.

POSTOPERATIVE CARE

Postoperatively, the patients are typically extubated in the operating room after transfer to the hospital bed. They are cared for in a monitored, non-intensive care unit setting, and maintenance fluids are administered based on lean body weight (typically 40–50 percent of actual weight). Early

warning signs of complication include fever, tachycardia, tachypnea, increasing oxygen requirement, oliguria, hiccoughs, regurgitation, left shoulder pain, worsening abdominal pain, a feeling of anxiety, or acute alteration in mental status. These signs warrant aggressive attention and appropriate investigation since they may signal gastrointestinal leak (anastomosis or staple lines), pulmonary embolus, bowel obstruction, or acute dilatation and impending rupture of the bypassed gastric remnant. Routinely, a water-soluble upper gastrointestinal contrast study is obtained on postoperative day 1. After satisfactory passage of contrast is documented, patients are begun on clear liquids and subsequently advanced to a high-protein liquid diet for the first month after operation.

Postoperative monitoring

Gastric bypass essentially results in surgically enforced, very low-calorie, low-carbohydrate dietary intake, thus requiring attention to an adequate intake of important macronutrients and micronutrients postoperatively. Postoperative follow-up after bariatric surgery in adolescence is intensive: weekly for 1 month, then monthly for 6 months, then every third month for the next 17 months. Serum chemistries, complete blood count, urine specific gravity, prothrombin time (evidence of vitamin K adequacy), and representative B complex vitamin levels (e.g., B1, B12, folate) are obtained at 3, 6, 9, and 12 months postoperatively, then yearly. Body composition is assessed with either bioelectrical impedance or dual energy X-ray absorptiometry analysis (DEXA; for patients weighing less than 159 kg) preoperatively and 3, 6, and 12 months postoperatively. The DEXA not only allows for the measurement of rate and relative amounts of fat and lean body mass loss, but also provides a quantitative assessment of bone mineral density changes. This body composition analysis is used to modify dietary plans intended to preserve lean body mass during the period of dramatic weight loss.

Postoperative diet

For the first postoperative month, the diet is essentially a protein-rich liquid diet. Dietary advancement after the first month is a methodical process of introducing new items of gradually increasing complexity, toward a goal of a well-balanced, small-portion (approximately one cup per meal) diet that ensures a daily intake of 1 g of protein per kilogram of ideal weight. Non-steroidal anti-inflammatory medications should be avoided to reduce the risk of intestinal ulceration and bleeding. Ursodiol and ranitidine are prescribed for 6 months, as is the practice for most adult bariatric programs. Postoperative vitamin and mineral supplementation typically consists of two pediatric chewable multivitamins, a calcium supplement, and an iron supplement for menstruating females. B-complex vitamins are supplemented beyond that which is contained in multivitamin preparations, primarily to augment thiamine and folate supplementation, due to severe complications if deficiency develops.

Five basic 'rules' are routinely emphasized with patients and family at each visit.

1. Eat protein first.
2. Drink 2–3 L of water or sugar-free liquids daily.
3. No snacking between meals.
4. Exercise at least 30 minutes per day.
5. Always remember vitamins and minerals.

OUTCOME

Patient satisfaction has generally been very good with this surgical and postoperative approach. Weight loss at a rate of 2.5 kg per week for the first 6 months is not uncommon. An excessive rate of weight loss can lead to liver failure. The operation results in a loss of 60–80 percent of excess weight over the first year postoperatively. When careful attention is given to adequate protein intake, significant improvements in body composition can be expected after operation. At Cincinnati Children's Hospital, adolescents undergoing RYGBP have an average BMI between 50 and 60 kg/m^2. Using detailed body composition analysis, we find that these patients lose nearly 20 percent of both fat mass and lean mass in the first 3 months following RYGBP. Interestingly, absolute fat mass decreases further by 40 percent from 3 to 12 months postoperatively, whereas absolute lean mass does not decrease further after 3 months. Consequently, body composition significantly improves over time, with mean percentage fat mass decreasing from 47 percent preoperatively to 45 percent at 3 months and 35 percent at 12 months. Fat loss appears to plateau by 12 months. This suggests that adolescents undergoing laparoscopic RYGBP dramatically and preferentially reduce body fat mass compared to more modest loss of lean body mass. Additionally, we have found no detrimental effects of RYGBP on bone mineral density after short-term follow-up of 1 year.

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101

Conjoined twins

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HISTORICAL HIGHLIGHTS

The earliest example of conjoined twins is a 17-cm marble statuette portraying parapagus twins, 'the double goddess', dating from the sixth millennium BC. The statue of sisters of Catathoyuk is housed in the Anatolian Civilisation Museum in Ankara, Turkey. Another early example is a stone carving of pygopagus twins dated to 80 BC in the St Marco Museum, Florence, Italy. The earliest attempt at separation of conjoined twins occurred in Kappadokia, Armenia, in AD 970. When one of the male ischiopagus twins died, aged 30 years, an attempt was made to save the surviving twin by separating him from his dead brother, but he died 3 days later.

The first well-documented case is that of the Biddenden maids born in Kent, England, in AD 1100 and joined at the hips and the shoulders. They lived together for 34 years. When Mary fell ill and died, Eliza was advised to be separated but absolutely refused saying, 'as we came together we will also go together'. She died 6 hours later.

The first successful separation of conjoined twins took place in 1689. The surgeon, Johannes Fatio, separated omphalopagus twins in Basel, Switzerland, by 'tracing the umbilical vessels to the navel where he tied them separately. He then transfixed and tied the bridge between the two infants with a silken cord and cut the isthmus.' The ligature fell off on the ninth postoperative day and both children survived.

The most celebrated pair of conjoined twins was Chang and Eng, born on a river boat in Siam in 1911. They were joined at the xiphisternum by a short bridge that stretched so they were eventually able to stand side-by-side. They were taken to the United States where they were exhibited by the showman, Phineas Barnum. They married sisters, lived in North Carolina and had 22 children between them. They lived together for 63 years.

INCIDENCE AND ETIOLOGY

The frequency of conjoined twins has been estimated at 1 in 250 000 live births. Sixty percent of conjoined twins die during gestation or at birth. Females predominate with a ratio of 3 : 1.

Conjoined twins develop from a single fertilized ovum. The most widely held theory of their occurrence is that of failure to undergo complete separation of the embryonic disc at around the fifteenth to seventeenth day of gestation (fissure theory). An alternative postulation, by Spencer, is that secondary fusion occurs between two originally separate monovular embryonic discs (fusion theory).

CLASSIFICATION

The twins are classified according to the most prominent site of fusion, plus 'pagus', the Greek term for fixed (Table 101.1). Thus, thoracopagus, omphalopagus, pygopagus, ischiopagus, and craniopagus signify union at the thorax, umbilicus, perineum, pelvis, and head, respectively (Figs 101.1–101.4). Thoraco-omophalopagus combined accounts for almost three quarters of conjoined twins and is associated with the highest mortality, mainly as a consequence of the frequent

Table 101.1 *Classification of conjoined twins*

Classification	Percent occurrence
Thoracopagus	40%
Omphalopagus	33%
Pygopagus	19%
Ischiopagus	6%
Craniopagus	2%

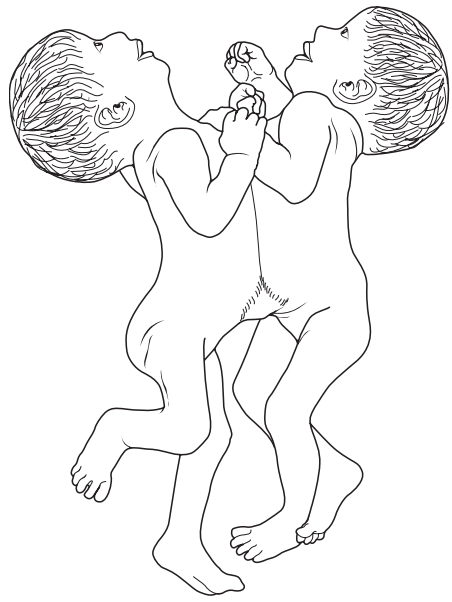

Fig. 101.1 Thoracopagus.

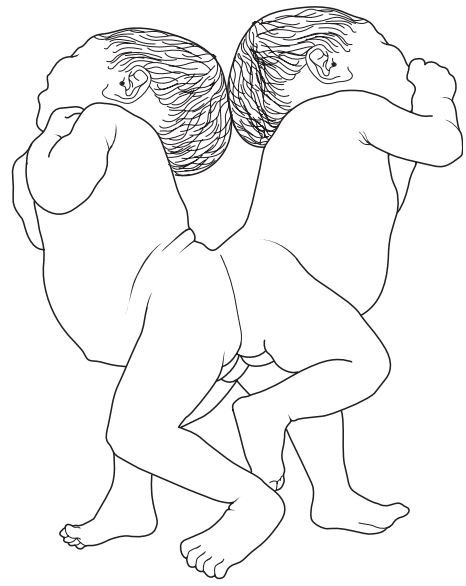

Fig. 101.2 Pygopagus.

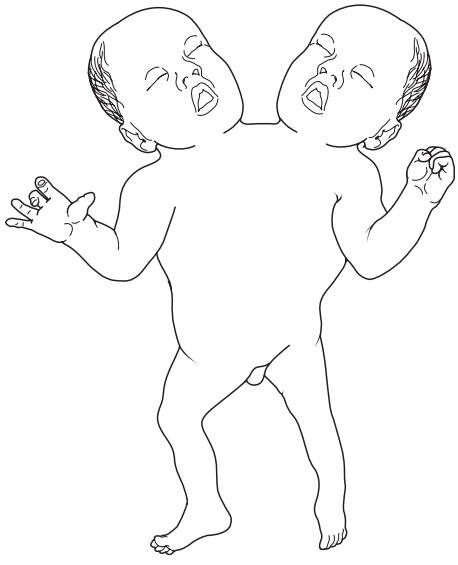

Fig. 101.3 Ischiopagus.

Fig. 101.4 Craniopagus.

association of complex fused cardiac masses. Parapagus refers to extensive ventrolateral conjunction.

PRENATAL DIAGNOSIS

The diagnosis of conjoined twins should be considered in any twin pregnancy that has a single placenta and no visible separating amniotic membrane. Polyhydramnios occurs in as many as 50% of conjoined twin pregnancies compared with 10% of normal twins and 2% of singleton pregnancies.

Prenatal ultrasonography is capable of diagnosing conjoined twin pregnancies as early as 12 weeks' gestation. The sonographic findings include inseparable fetal bodies and skin contours, an unchanged relative position of the fetuses, both fetal heads persistently at the same level, and a single umbilical cord containing more than three vessels. Detailed scanning at around 20 weeks' gestation will accurately define the extent of the conjoined area and provide an assessment of which viscera are shared. Fetal echocardiography is mandatory as twins with a complex shared heart have an extremely poor prognosis and termination of the pregnancy is invari-

ably recommended. Computerized tomography (CT) and magnetic resonance imaging (MRI) may be performed at 32–34 weeks' gestation, but once the decision has been made to proceed with the pregnancy, these investigations can be carried out with greater accuracy after delivery. An additional advantage of prenatal diagnosis is that the time, place, and mode of delivery can be planned. The delivery should take place at or close to the surgical unit where separation will be performed. Delivery must always be by caesarean section at 36–38 weeks' gestation.

MANAGEMENT

The management of conjoined twins can be divided into four separate time frames.

Prenatal

Once the diagnosis of conjoined twins is suspected on prenatal scan at 12 weeks' gestation, it is important to define the anatomy of the union. Termination of the pregnancy is recommended where there is complex cardiac fusion or extensive cerebral fusion. Detailed echocardiography and accurate ultrasonography is essential. The extent of deformity expected following possible subsequent separation must be carefully and accurately explained so that an informed decision can be made either to terminate or to proceed with the pregnancy.

Nonoperative treatment

No attempt at surgical separation should be considered in the presence of complex cardiac or cerebral fusion or where the expected deformity following separation is unacceptable to the parents.

Emergency separation

This procedure is undertaken when one twin is dead or dying and threatening the survival of the remaining twin, or where a life-threatening correctable congenital abnormality (e.g. intestinal atresia, malrotation with or without volvulus, ruptured omphalocele, or anorectal agenesis) is present in one or both twins. Under these circumstances the only chance of saving one or both infants lies in immediate separation. Emergency separation carries a significantly higher mortality rate compared with elective procedures.

Elective separation

This will normally take place between 2 and 4 months of age. It allows the twins to stabilize and thrive and provides time to carry out detailed investigations to define the nature and the extent of union. It also allows the application of methods to be carried out to achieve primary closure of the wound such as tissue expansion. Detailed planning of the operative procedure with all members of the operating team should take place before the separation. The survival rate for elective separation is in excess of 80%.

INVESTIGATIONS

The choice of imaging study will depend on the site of union. For thoraco-omphagopagus twins, essential investigations include echocardiogram, CT. (Fig. 101.5), and MRI scans with particular attention directed to the anatomy of the hearts, livers, and genitourinary systems. Where the livers are fused, it is important to document the presence of separate gallbladders and hepatic veins. Gestational contrast studies are useful in showing separate gastrointestinal systems. MRI angiography has superseded percutaneous angiography to define vascular anatomy. Bony anatomy is best demonstrated on plain X-ray and MRI scan.

ANESTHETIC MANAGEMENT

Two sets of anesthesiologists, one for each infant, are essential, as each has to be separately monitored throughout the procedure. Essential monitoring consists of arterial and central venous catheters, electrocardiogram, pulse oximetry, capnography, and urinary output. Regular blood gas analyses are undertaken throughout the procedure. All drugs and intravenous fluids are calculated on a total weight basis, with half being delivered to each twin. Because of the cross-circulation, drugs given intravenously may have an unpredictable effect. Thus, particular care is essential when administering drugs such as opioids, which should be given incrementally.

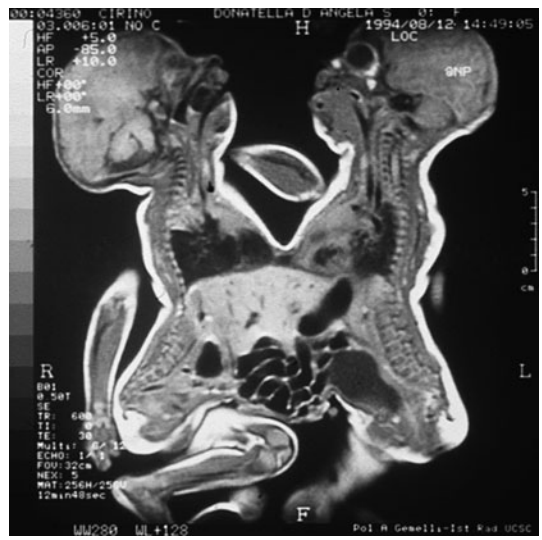

Fig. 101.5 CT scan of omphalo-thoracopagus twins.

THE OPERATIVE PROCEDURE

Technical details of the operative procedure will be dictated by the anatomy of the junction and by the organs and structures shared. In thoracopagus, the liver is invariably shared. In 90% of cases there is a common pericardium which can be separated to provide an individual pericardial sac for each twin. Major myocardial connections are present in 75% of cases and only a few attempts have been made at separation. The upper gastrointestinal tract is common in 50% of cases with a shared biliary system in 25%. In omphalopagus, the liver is shared in 80% of cases and in 33% the intestines join at the level of the Meckel's diverticulum and the common terminal ileum and colon have a dual blood supply. The lower intestinal tract is common in both pygopagus and ischiopagus and the genitourinary tract is shared in 15% of the former and 50% of the latter. It is not uncommon for the ureters in these situations to cross over from one twin and enter the contralateral bladder (Fig. 101.6). The high mortality rate associated with craniopagus is almost entirely due to cerebral fusion, which is also responsible for the neurodevelopmental sequalae in survivors.

Blood loss may be a major intraoperative problem, especially where there is pelvic bony fusion. Blood loss occurring during division of the liver should be minimized by using ultrasonic dissection, meticulously ligating major connecting vessels, and coagulating minor vessels, and by applying fibrin glue (Tisseel®) to the raw surface postoperative ooze of blood and leakage of bile may be prevented. Despite every attempt to define as accurately as possible all anatomical connections prior to surgery, 'unexpected events' are frequently encountered during the operation. Examples in our experience include abnormal vascular communications, and previously unidentified intestinal and genitourinary anomalies. The surgical team should be aware of these variations in anatomy and be prepared to vary the operative procedure accordingly.

When, despite all possible maneuvers, primary closure of the defect proves impossible, it will be necessary to insert prosthetic material (polypropylene mesh, Silastic sheet, Gore-Tex®) as a temporary measure. The insertion of a prosthetic patch in closure of the abdomen is preferable to 'closure under tension' which may embarrass respiration or inhibit venous return.

POSTOPERATIVE MANAGEMENT

Postoperatively, the surviving infant/s are extremely fragile. All intraoperative monitoring must be continued postoperatively in the intensive care unit and because of the prolonged duration of surgery, the infants are electively paralyzed and mechanically ventilated for a variable period of time. Meticulous attention should be directed at monitoring fluid and electrolyte balance, and in particular avoiding overhydration which may precipitate cardiovascular instability. Sepsis is a major cause of mortality and morbidity and strict infectious precautions must be exercised, particularly where large skin defects are present. Late unexpected deaths following separation are unfortunately not uncommon.

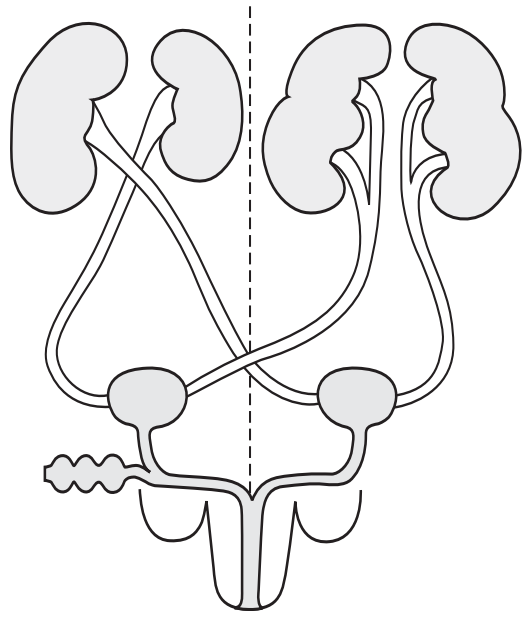

Fig. 101.6 Urological anatomy in one set of conjoined twins.

OUTCOME FOR CONJOINED TWINS TREATED AT GREAT ORMOND STREET HOSPITAL

Since 1985, at Great Ormond Street Hospital, London, we have had experience with 25 pairs of conjoined twins:

- **Nonoperative treatment** was carried out in seven sets all of whom died within a short period of time.
- **Emergency separation** was attempted in seven sets with four survivors (28%).
- **Planned separation** was performed in 11 sets with 19 long-term survivors (87%). Of the three deaths, two occured many months after successful separation.

Comparison with other series in the literature is shown in Table 101.2.

FURTHER READING

Kingston CA, McHugh K, Kumaradevan J, Kiely EM, Spitz L. Imaging in the preoperative assessment of conjoined twins. *RadioGraphics* 2001; **21**: 1187–208.

Spitz L, EM Kiely. Experience in the management of conjoined twins. *The British Journal of Surgery* 2002; **89**: 1188–92.

Spitz L, EM Kiely. Conjoined twins. *The Journal of the American Medical Association* 2002; **289**: 1307–10.

Wilcox DT, Quinn SM, Spitz L, Kiely EM, Ransley PG. Urological problems in conjoined twins. *British Journal of Urology* 1998; **81**: 905–10.

Table 101.2 *Outcome in three major series*

Series	Number	No operation	Emergency procedure*	Planned procedure*
O'Neill	18	5	5 sets 1 (10%)	8 sets 13 (81%)
Cywes	14	4	5 sets 2 (20%)	5 sets 8 (80%)
Spitz/Kiely	25	7	7 sets 4 (29%)	11 sets 17 (87%)

* Results shown are number of survivors, with percent in parentheses.

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