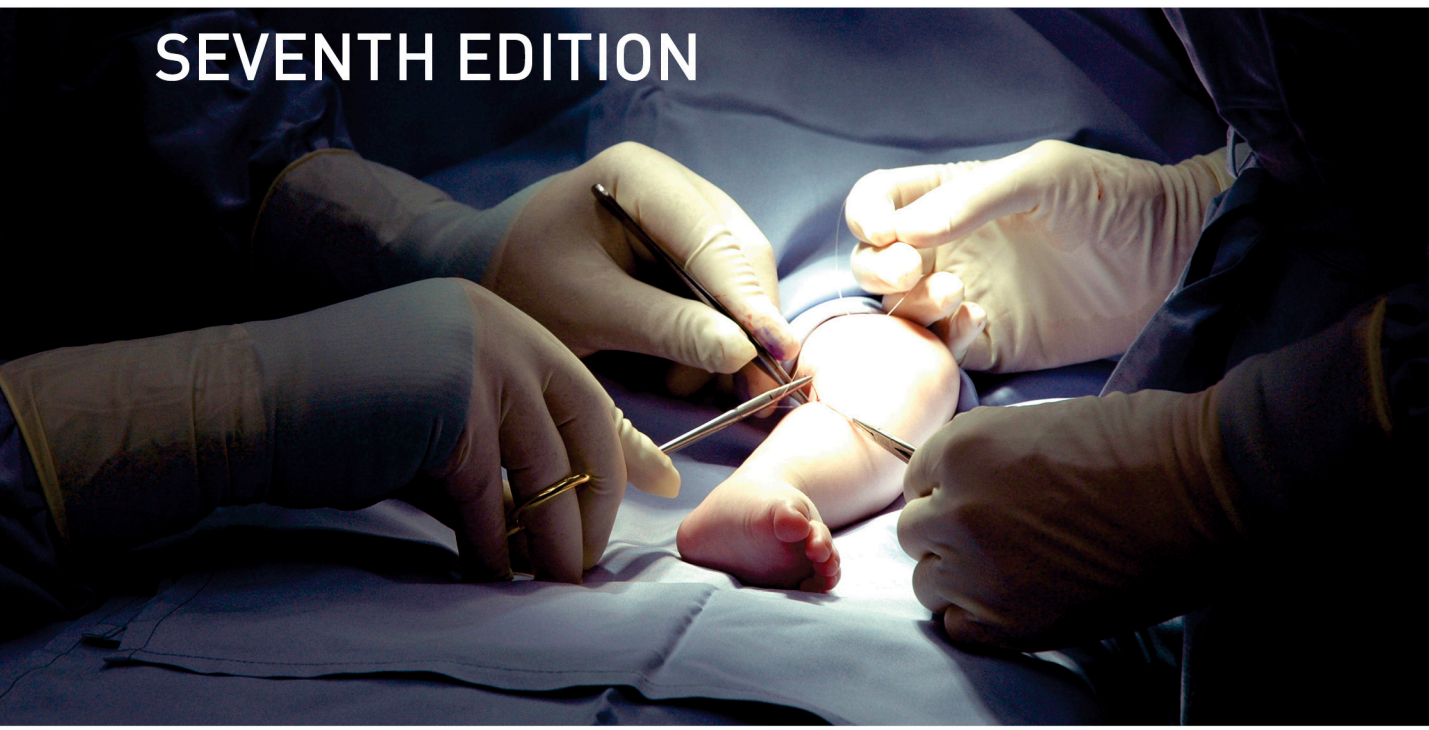
JONES' CLINICAL PAEDIATRIC SURGERY

SEVENTH EDITION



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WILEY Blackwell

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Foreword to the First Edition

The progressive increase in the body of information relative to the surgical specialities has come to present a vexing problem in the instruction of medical students. There is only enough time in the medical curriculum to present an overview to them, and in textbook material, one is reduced either to synoptic sections in textbooks of surgery or to the speciality too detailed for the student or the non-specialist in complete and authoritative textbooks.

There has long been a need for a book of modest size dealing with paediatric surgery in a way suited to the requirements of the medical student, general practitioner and paediatrician. Peter G. Jones and his associates from the distinguished and productive group at the Royal Children's Hospital in Melbourne have succeeded in meeting this need. The book could have been entitled *Surgical Conditions in Infancy and Childhood,* for it deals with children and their afflictions, their symptoms, diagnosis and treatment rather than surgery as such. The reader is told when and how urgently an operation is required, and enough about the nature of the procedure to understand its risks and appreciate its results. This is what students need to know and what paediatricians and general practitioners need to be refreshed on.

Many of the chapters are novel, in that they deal not with categorical diseases but with the conditions that give rise to a specific symptom – Vomiting in the First Month of Life, The Jaundiced Newborn Baby, Surgical Causes of Failure to Thrive. The chapter on genetic counselling is a model of information and good sense.

The book is systematic and thorough. A clean style, logical sequential discussions and avoidance of esoterica allow the presentation of substantial information over the entire field of paediatric surgery in this comfortable-sized volume with well-chosen illustrations and carefully selected bibliography. Many charts and tables, original in conception, enhance the clear presentation.

No other book so satisfactorily meets the need of the student for broad and authoritative coverage in a modest compass. The paediatric house officer (in whose hospital more than 50% of the patients are, after all, surgical) will be serviced equally well. Paediatric surgeons will find between these covers an account of the attitudes, practices and results of one of the world's greatest paediatric surgical centres. The book comes as a fitting tribute to the 100th anniversary of the Royal Children's Hospital.

Mark M. Ravitch
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Tribute to Mr. Peter Jones

Mr Peter Jones (1922–1995) MB, MS, FRCS, FRACS, FACS, FAAP. The first Australian surgeon to obtain the FRACS in paediatric surgery, member of RACS Council (1987–1995), Vice President of the Medical Defence Association of Victoria (1974–1988) and President of the Australian Association of Surgeons (1983–1986). He was legendary as a medical historian and in heraldry, as a great raconteur, but primarily as a great student teacher.

Preface to the Seventh Edition

The objective of the first edition of this book was to bring together information on surgical conditions in infancy and childhood for use by medical students and resident medical officers. It remains a great satisfaction to our contributors that the book has fulfilled this aim successfully and that a seventh edition is now required. Family doctors, paediatricians and many others concerned with the welfare of children have also found this book useful.

A knowledgeable medical publisher once commented to Peter Jones that this book is not about surgery but about paediatrics, and this is what it should be, as we have continued to omit almost all details of operative surgery.

The plan for the sixth edition has been largely retained but with the addition of new coloured photographs. Mr Alan Woodward has retired as an editor, and we have added two new editors, Mr Warwick Teague and Mr Sebastian King. Nearly half of the contributors to this edition are new members of the hospital staff and bring a fresh outlook and state-of-the-art ideas.

It is now about 20 years since Mr Peter Jones died, and this book remains as a dedication to him. Peter was a great teacher and it is a daunting task for those who follow in his footsteps. We hope this new edition will continue to honour the memory of a great paediatric surgeon who understood what students need to know.

Acknowledgements

Many members of the Royal Children's Hospital community have made valuable contributions to this seventh edition. The secretarial staff of the Department, and particularly Mrs Shirley D'Cruz, are thanked sincerely for their untiring support.

PART I
Introduction

CHAPTER 1
Antenatal Diagnosis: Surgical Aspects

CASE 1

At 18 weeks' gestation, right fetal hydronephrosis is diagnosed on ultrasonography.

Q 1.1 Discuss the further management during pregnancy.

Q 1.2 Does antenatal diagnosis improve the postnatal outlook for this condition?

CASE 2

An exomphalos is diagnosed on the 18-week ultrasound scan.

Q 2.1 What further evaluation is required at this stage?

Q 2.2 Does this anomaly influence the timing and mode of delivery?

Antenatal diagnosis is one of the most rapidly developing fields in medical practice. While the genetic and biochemical evaluation of the developing fetus provides the key to many medical diagnoses, the development of accurate ultrasound has provided the impetus to the diagnosis of surgical fetal anomalies. At first, it was expected that antenatal diagnosis of fetal problems would lead to better treatment and an improved outcome. In some cases, this is true. Antenatally diagnosed fetuses with gastroschisis are now routinely delivered in a tertiary-level obstetric hospital with neonatal intensive care in order to prevent hypothermia and delays in surgical treatment, and the results of treatment have improved. In other cases, such as congenital diaphragmatic hernia, these expectations have not been fulfilled because antenatal diagnosis has revealed a number of complex and lethal anomalies which in the past never survived the pregnancy and were recorded in the statistics of fetal death in utero and stillbirth.

Indications and timing for antenatal ultrasound

Most pregnancies are now assessed with a mid-trimester morphology ultrasound scan, which is usually performed at 18–20 weeks' gestation [Fig.1.1]. The main purpose of this examination is to assess the obstetric parameters of the pregnancy, but the increasingly important secondary role of this study is to screen the fetus for anomalies. Most fetal anomalies can be diagnosed at 18 weeks, but some only become apparent later in the pregnancy. Renal anomalies are best seen on a 30-week ultrasound scan, as urine flow is low before 24 weeks. Earlier transvaginal scanning may be performed in special circumstances, such as a previous pregnancy with neural tube defect, and increasingly to detect early signs of aneuploidy. Fetal magnetic resonance imaging is increasingly being used to assess the developing fetus in cases of suspected or confirmed fetal anomalies without exposing the fetus or mother to ionising radiation.

Natural history of fetal anomalies

Before the advent of ultrasonography (as earlier), paediatric surgeons saw only a selected group of infants with congenital anomalies. These babies had survived the pregnancy and lived long enough after birth to reach surgical attention. Thus, the babies coming to surgical treatment were already a selected group, mostly with a good prognosis.

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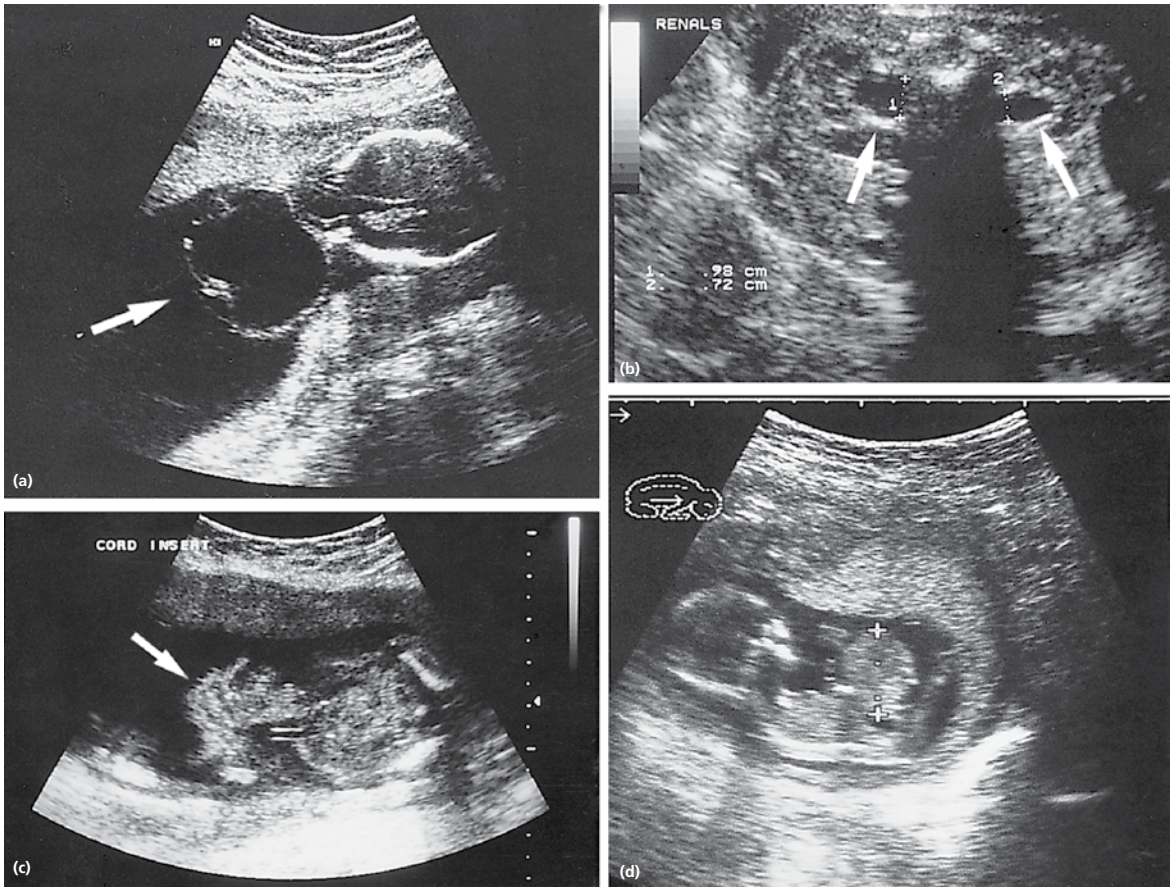

Figure 1.1 (**a**) Encephalocele shown in a cross section of the fetal head. The sac protruding through the posterior skull defect is arrowed. (**b**) Bilateral hydronephrosis shown in an upper abdominal section. The dilated renal pelvis containing clear fluid is marked. (**c**) The irregular outline of the free-floating bowel in the amniotic cavity of a term baby with gastroschisis. (**d**) A longitudinal section through a 14-week fetus showing a large exomphalos. The head is seen to the left of the picture. The large sac (marked) is seen between blurred (moving) images of the arms and legs.

Antenatal diagnosis has exposed surgeons to a new group of conditions with a poor prognosis, and at last, the full spectrum of pathology is coming to surgical attention. For example, posterior urethral valve causing obstruction of the urinary tract was thought to be rare, with an incidence of 1:5000 male births; most cases did well with postnatal valve resection. It is now known that the true incidence of urethral valve is 1:2500 male births, and these additional cases did not come to surgical attention as they developed intrauterine renal failure, with either fetal death or early neonatal death from respiratory failure because of Potter syndrome. It was thought that antenatal diagnosis would improve the outcome of such congenital anomalies, but the overall results have appeared to become worse with the inclusion of these severe *new* cases.

In the same way, antenatal diagnosis has exposed the significant *hidden mortality* of congenital diaphragmatic hernia [Fig. 1.2]. Previously, congenital diaphragmatic hernia diagnosed after birth was not commonly associated with multiple congenital anomalies, but now, antenatal diagnosis has uncovered a more severe subgroup with associated chromosomal anomalies and multiple developmental defects. It is now apparent that the earlier the congenital diaphragmatic hernia is diagnosed in utero, the worse the outcome.

Despite these problems, there are many advantages in antenatal diagnosis. The outcomes of many congenital

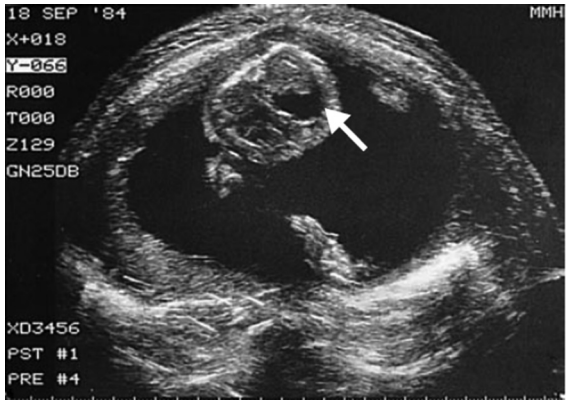

Figure 1.2 Cross section of a uterus with marked polyhydramnios. The fetal chest is seen in cross section within the uterus. The fluid-filled cavity within the left side of the chest is the stomach protruding through a congenital diaphragmatic hernia (arrow).

anomalies are improved by prior knowledge of them before birth.

Management following antenatal diagnosis

Fetal management

Cases diagnosed antenatally may be classified into three groups:

Good prognosis

In some cases, such as a unilateral hydronephrosis, there is no role for active antenatal management, and the main task is to document the progress of the condition through pregnancy with serial ultrasound scans. The detailed diagnosis is made with the more sophisticated range of tests available after birth, and the incidence of urinary tract infections (UTIs) may be reduced with prophylactic antibiotics commenced at birth. Thus, a child with severe vesicoureteric reflux may go through the first year of life without any UTIs. If the parents receive counselling by an experienced surgeon, they have time to understand the condition, its treatment and prognosis. With such preparation, the family may cope better with the birth of a baby with a congenital anomaly.

The paediatric surgeon also has an important role to play in advising the obstetrician on the prognosis of a particular condition. Some cases of exomphalos are easy to repair, whereas in others, the defect may be so large that primary repair will be difficult. In addition, there may be major chromosomal and cardiac anomalies, which may alter the outcome. In other conditions, the outlook for a congenital defect may change as treatment improves. Gastroschisis was a lethal condition before 1970, but now, management has changed and there is a 95% survival rate. In those cases with a good prognosis, fetal intervention is not indicated, and the pregnancy should be allowed to continue to close to term. The mode of delivery will usually be determined on obstetric grounds. Babies with exomphalos may be delivered by vaginal delivery if the birth process is easy. Primary caesarean section may be indicated for major exomphalos to prevent rupture of the exomphalos and damage to the organs such as the liver, as well as for obstetric indications. There is evidence that in fetuses with large neural tube defects, further nerve damage may occur at vaginal delivery, and caesarean section may be preferred in this circumstance. If urgent neonatal surgery is required, for example, in gastroschisis, the baby should be delivered at a tertiary obstetric unit with appropriate neonatal intensive care. In other cases (e.g. cleft lip and palate), where urgent surgery is not required but good family and nursing support is important, delivery closer to the family's home may be more appropriate. Antenatal planning and family counselling give us the opportunity to make the appropriate arrangements for the birth. A baby born with gastroschisis in the middle of winter in a bush nursing hospital in the mountains, many hours away from surgical care, may have a very different prognosis from a baby with the same condition born at a major neonatal centre.

Poor prognosis

Anencephaly, congenital diaphragmatic hernia with major chromosomal anomalies or urethral valve with early intrauterine renal failure are examples of conditions with a poor prognosis. These are lethal conditions, and the outcome is predetermined before the diagnosis is made.

Late deterioration

In most cases, initial assessment of the fetal anomaly will indicate a good prognosis with no reason for interference. However, later in gestation, the fetus may deteriorate, and some action must be undertaken to prevent

a lethal outcome. An example would be posterior urethral valve causing lower urinary tract obstruction. Early in the pregnancy, renal function may be acceptable with good amniotic fluid volumes, but on follow-up ultrasound assessment, there may be loss of amniotic fluid with oligohydramnios as a sign of renal failure. There are several approaches to this problem. If the gestation is at a viable stage, for example, 36 weeks, labour may be induced, and the urethral valve treated at birth. If the risks of premature delivery are higher, for example, at 28 weeks' gestation, temporary relief may be obtained by using percutaneous transuterine techniques to place a shunt catheter from the fetal bladder into the amniotic cavity. These catheters tend to become dislodged by fetal activity. A more definitive approach to drain the urinary tract is intrauterine surgery to perform a vesicostomy and allow the pregnancy to continue. This procedure has been performed with success in a few cases of posterior urethral valve. These patients are highly selected, and only a few special centres are able to perform intrauterine surgery. At present, this surgery is regarded as experimental and reserved for rare situations, but this may not always be the case.

Surgical counselling

When a child is born with unanticipated birth defects, there is inevitably shock and confusion until the diagnosis is clarified, and the family begins to assimilate and accept the information given to them. Important treatment decisions may have to be made urgently while the new parents are still too stunned to play any sensible part in the ongoing care of their baby. Antenatal diagnosis has changed this situation. New parents may now have many weeks to understand and come to terms with their baby's condition. With suitable preparation, they may play an active role in the postnatal treatment choices for their newborn baby.

The paediatric surgeon who treats the particular problem uncovered by antenatal diagnosis is in the best position to advise the parents on the prognosis and further treatment of the baby. Detailed information on the management after birth, with photographs before and after corrective surgery, allows the parents to understand the operative procedures. The opportunity to meet other families with a child treated for the same condition may give time for the pregnant woman and her partner to understand the problem prior to birth. Handling and nurturing the baby immediately after birth is an important part of bonding. Parents and nursing staff suddenly confronted with a newborn baby with an unexpected anomaly, such as sacrococcygeal teratoma, may be afraid to handle the baby prior to the baby being taken away to another hospital for complex surgery. Parents in this situation may take many months to bond with the new baby and to understand fully the nature of the problem. Prepared by antenatal diagnosis, parents realise they may handle and nurture the baby, understand the nature of the surgery and form a bond with the baby. Thus, instead of being stunned by the birth of a baby with a significant malformation, the new parents may play an active part in the postnatal surgical management and provide better informed consent for surgery.

KEY POINTS

- Antenatal diagnosis with ultrasound scanning has revealed the natural history of some anomalies and made the prognosis seem worse (e.g. congenital diaphragmatic hernia, posterior urethra valve).
- Antenatal diagnosis has allowed surgical planning (and occasional fetal intervention), as well as providing time for parents to be informed prior to the birth.

Further reading

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CHAPTER 2
The Care and Transport of the Newborn

CASE 1

A 30-week gestation neonate is born with gastroschisis.

Q 1.1 What advice would you give the referring institution about the management of this infant prior to transport to a tertiary institution?

CASE 2

A 40-week gestation neonate develops respiratory distress shortly after birth. A left congenital diaphragmatic hernia (CDH) is diagnosed.

Q 2.1 List two iatrogenic problems that may occur with positive-pressure ventilation.

Q 2.2 How do you avoid these iatrogenic problems?

The initial care and transport of a sick newborn baby is critically important to the surgical outcome. A detailed preoperative assessment is necessary to detect associated or coexistent developmental anomalies. Vital disturbances should be corrected before operation, and predictable complications of the abnormalities should be anticipated and recognised early.

Respiratory care

The aims of respiratory care are (i) to maintain a clear airway, (ii) to prevent abdominal distension, (iii) to avoid aspiration of gastric contents and (iv) to provide supplementary oxygen if necessary. Various manoeuvres and adjuncts are commonly used in neonatal respiratory care to achieve these aims, including:

1 *Suctioning* of the pharyngeal secretions maintains a clear airway. This is especially important in the premature neonate with poorly developed laryngeal reflexes, and will need to be repeated regularly in neonates with oesophageal atresia.

2 *Prone positioning* improves the airways, assists ventilation and reduces the risk of aspiration of gastric contents with gastro-oesophageal reflux or vomiting. Importantly, this positioning applies to monitored neonates in an intensive care setting and does not contradict the *back to sleep* public health advice pertaining to prevention of sudden infant death syndrome (SIDS).

3 *Nasogastric tube insertion*, size 8 French, will minimise the risk of life-threatening aspiration of vomitus, provided the tube is kept patent and allowed to drain freely with additional aspiration at frequent intervals. It will also reduce abdominal distension and improve ventilation in patients with intestinal obstruction or congenital diaphragmatic hernia (CDH).

4 *Supplementary oxygen therapy* with or without endotracheal intubation and ventilation is provided as required for respiratory distress. Common medical causes of the breathless neonate include transient tachypnoea of the newborn, meconium aspiration, pneumothorax, hyaline membrane disease and apnoea. Surgical causes of respiratory distress include oesophageal atresia and CDH. Ventilation strategies in CDH are complex and require input of specialist and experienced practitioners, who may be neonatologists, intensive care physicians or surgeons. These strategies seek to minimise barotrauma to the poorly developed lungs, which may cause bronchopulmonary damage, pneumothorax and death.

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Blood and fluid loss

Newborn babies do not tolerate blood or fluid loss well. The blood volume of a full-term neonate is 80 mL/kg. Therefore, a loss of only 30 mL blood constitutes a loss of approximately 10% of blood volume, which is equivalent to 500 mL loss in an adult. For this reason, it is routine to crossmatch whole blood prior to neonatal surgery. Blood loss is strictly kept to a minimum and measured by weighing all swabs used. Neonatal blood is relatively concentrated; haemoglobin concentration in the first days of life is about 19 g/dL and the haematocrit 50–70%. In this circumstance, blood loss may be replaced in part with blood and in part with a crystalloid solution, which lowers the viscosity of the blood.

Neonatal bowel obstruction is another common setting resulting in fluid depletion due to vomiting and nasogastric losses. Hypovolaemia is manifest with lethargy, pallor, prolonged capillary return, cool limbs, venoconstriction and cyanosis. Acidosis becomes a complicating factor. In this situation, the baby is fluid resuscitated with an initial *bolus infusion* of 10 mL/kg crystalloid solution of normal saline (0.9% NaCl) over 15 min. Effectiveness of resuscitation is indicated by improved peripheral circulation in response to the bolus. If the response is not adequate or not sustained, further 10 mL/kg bolus infusions of crystalloid may be given and circulatory status monitored.

Control of body temperature

Newborn infants, especially the premature, are at risk of excessive heat loss because of their relatively large surface area-to-volume ratio, lack of subcutaneous insulating fat and immature thermoregulation. The sick neonate with a surgical condition is prone to hypothermia, defined as a core body temperature of less than 36 °C. Neonates counteract hypothermia by increasing metabolic activity and thermogenesis by brown fat metabolism. However, if heat loss exceeds heat production, the body temperature will continue to fall, leading to acidosis and depression of respiratory, cardiac and nervous function.

Heat loss occurs from the body surface by radiation, conduction, convection and the evaporation of water. Excessive heat loss during assessment, procedures, transport and operation must be avoided. Radiant overhead heaters are of particular value during procedures such as intravenous cannulation or the induction of anaesthesia, because they allow unimpeded access to the infant. Neonates with gastroschisis are at superadded risk of heat loss as the eviscerated bowel provides increased surface area for evaporation. Heat loss during transport and assessment is minimised by enclosing the bowel with plastic kitchen wrap or a bowel bag to prevent evaporation. Wet packs should never be applied to a neonate as they will accelerate evaporative and conductive heat losses.

Fluids, electrolytes and nutrition

Many infants with a surgical condition cannot be fed in the perioperative period. Intravenous fluids provide daily maintenance requirements and prevent dehydration. The total volume of fluid given must restore fluid and electrolyte deficits, supply maintenance requirements and replace ongoing losses.

Maintenance fluid requirements are:
60–80 mL/kg on day 1 of life
80–100 mL/kg on day 2 of life
100–150 mL/kg on day 3 of life and thereafter

Maintenance electrolyte requirements are:
Sodium: 3 mmol/kg/day
Chloride: 3 mmol/kg/day
Potassium: 2 mmol/kg per day

Maintenance joule requirements are:
100–140 kJ/kg/day

In the first 2–3 days of life, maintenance requirement for sodium, potassium and chloride is minimal due to a low glomerular filtration rate and low urine output at birth. Therefore, 10% dextrose solution alone is typically sufficient for maintenance needs. Beyond 2–3 days of age, a dextrose–saline solution is required, for example, 10% dextrose in 0.18–0.225% sodium chloride (sodium: 30 mmol/L) with the addition of potassium chloride at 20 mmol/L. However, this solution is inadequate for long-term maintenance of body functions as it has many deficiencies, especially in kilojoules.

In addition to maintenance fluids, many surgical neonates will require replacement of excess fluid and electrolyte losses, especially those with neonatal bowel obstruction. Useful clinical signs of dehydration include prolonged capillary return (>2 seconds), depression of the

fontanelle, dryness of the mucous membranes, reduced tissue turgor and cool peripheries. Reduced urine output and bodyweight loss may precede these findings.

The rule of thumb for estimating fluid loss is that dehydration of 5% or less of body mass has few clinical manifestations: 5–8% shows moderate clinical signs of dehydration; 10% shows severe signs and poor peripheral circulation. Thus, a 3000 g infant who has been vomiting and has a diminished urine output but shows no overt signs of dehydration may have lost approximately 5% of body mass and will require 150 mL (3000 × 5% mL) fluid replacement to correct the deficit. Maintenance fluid requirements must be administered also in addition.

Electrolyte estimations are most useful for identifying a deficiency of electrolytes that are distributed mainly in the extracellular fluid, for example, sodium, but will not be as reliable for electrolytes that are found mainly in the intracellular fluid, for example, potassium. Fluid and electrolyte deficiency due to vomiting needs to be replaced with a crystalloid solution that contains adequate levels of sodium, for example, 0.9% sodium chloride (sodium:150 mmol/L).

Continuing fluid and electrolyte losses need to be measured and replaced. Losses may arise from nasogastric aspirates in bowel obstruction, diarrhoea from an ileostomy or diuresis after the relief of urinary obstruction, for example, after resection of posterior urethral valve. When the losses are high, they are best measured and replaced with an intravenous infusion of electrolytes equivalent to those of the fluid being lost.

Intravenous (parenteral) nutrition will be required when starvation extends beyond 4–5 days. Common indications for parenteral nutrition in the neonate include necrotising enterocolitis, extensive gut resection and gastroschisis. The aim of parenteral nutrition is to provide all substances necessary to sustain normal growth and development. Parenteral nutrition may be maintained for weeks or months as required, although complications include sepsis and jaundice.

Oral nutrition is preferred where possible and breastfeeding is best. Gastrointestinal surgery may make oral feeding impossible for a while: gut enzyme function may be poor, and various substrates in the feeds may not be absorbed. Lactose intolerance is common and leads to diarrhoea with acidic, fluid stools. Other malabsorptive problems relate to sugars, protein, fat and osmolarity of the feeds. These may be managed by changing the formula or, in severe cases, by a period of parenteral nutrition to allow the gastrointestinal tract to recover.

Biochemical abnormalities

Important problems include metabolic acidosis, hypoglycaemia and hypocalcaemia. These must be minimised prior to an operation as they may adversely influence the neonate's response to anaesthetic agents.

Metabolic acidosis

Metabolic acidosis, which may result from hypovolaemia, dehydration, cold stress, renal failure or hypoxia, increases pulmonary vascular resistance and impairs cardiac output. Acidosis is corrected by fixing the underlying cause of the acidosis, and in renal failure, sodium bicarbonate may also be used.

Hypoglycaemia

Hypoglycaemia occurs in the sick newborn, especially if premature. Liver stores of glycogen are small, as are fat stores. Starvation and stress will consume liver glycogen rapidly, resulting in a need for fatty acid metabolism to maintain blood glucose levels, with consequent ketoacidosis. Gluconeogenesis from amino acids or pyruvate is slow to develop in the newborn, due to the relative inactivity of liver enzymes. Eventually, blood glucose levels cannot be maintained, and severe hypoglycaemia results, causing apnoea, convulsions and cerebral damage. These complications of hypoglycaemia may be prevented by intravenous dextrose infusions. Neonates should not be starved for longer than 3 h prior to an operation.

Hypocalcaemia

Hypocalcaemia may occur in neonates with respiratory distress. The ionised calcium level in the blood maintains cell membrane activity. Hypocalcaemia potentially causes twitching and convulsions but may be corrected by slowly infusing calcium gluconate.

Prevention of infection

The poorly developed immune defences of neonates predispose to infection with Gram-positive and Gram-negative organisms. Infection may spread rapidly and

result in septicaemia. Signs of systemic infection in the neonate are often non-specific, but may include hypothermia, pallor and lethargy.

Early recognition and treatment of infection is aided by microbiological cultures from the neonate's nose and umbilicus, and in select cases groin and rectum, both on admission to hospital and while in the hospital. This is important in picking up *marker organisms* such as multiple antibiotic-resistant *Staphylococcus aureus*. When infection is suspected, a septic workup is performed, taking specimens of the cerebrospinal fluid, urine and blood for culture and starting appropriate intravenous antibiotics immediately.

A neonate undergoing an operation is at a significantly increased risk of infection, and care must be taken not to introduce pathogenic organisms: this applies particularly to cross infection in the neonatal ward. Handwashing or antiseptic gel must be applied before and after handling any patient. Prophylactic antibiotics may be used to cover major operations.

Parents

An important part of care for a neonate undergoing an operation is reassurance and support for the neonate's anxious parents. The mother may be confined in a maternity hospital, while her baby is separated from her and undergoing a major operation in another institution. Close communication is important in this situation, and the mother and baby should be brought together as soon as possible. The parents should handle and fondle the baby to facilitate bonding. With goodwill and planning, gentle contact between neonate and mother may be achieved, even in difficult circumstances.

General principles of neonatal transport

Transport of a critically ill neonate is a precarious undertaking, and the following principles should be followed:

1. The neonate's condition should be stabilised before embarkation.
2. The most experienced/qualified personnel available should accompany the patient.
3. Specialised neonatal *retrieval* services should be used.
4. Transport should be as rapid as possible, but without causing further deterioration or incurring unnecessary risks to patient or transporting personnel.
5. Transport should be undertaken early rather than late.
6. All equipment should be checked before setting out.
7. The receiving institution should be notified early so that additional staff and equipment may be prepared for arrival.

Transport of neonatal emergencies

A list of the more common surgical emergencies is given in Table 2.1. Most neonates with these conditions should have transport arranged as soon as the diagnosis is apparent or suspected.

Some developmental anomalies do not require transportation, and specialist consultation at the hospital of birth may suffice (e.g. cleft lip and palate, orthopaedic deformities). Where doubt exists concerning the appropriateness or timing of transportation, specialist advice should be sought.

Table 2.1 Neonatal surgical conditions requiring emergency transport

Obvious malformations	Exomphalos/gastroschisis
	Myelomeningocele/encephalocele
	Anorectal malformation
Respiratory distress	
Upper airway obstruction	Choanal atresia
	Pierre Robin sequence
Lung dysplasia/compression	Congenital diaphragmatic hernia
	Emphysematous lobe
	Pulmonary cyst(s)
	Pneumothorax (insert chest drain first)
Congenital heart disease	
Acute alimentary or abdominal emergencies	Oesophageal atresia
	Intestinal obstruction
	Necrotising enterocolitis
	Haematemesis and/or melaena
Disorders of sex development (DSD)	

Choice of vehicle

The choice between road ambulance, helicopter or fixed-wing aircraft will depend on distance, availability of vehicle, time of day, traffic conditions, airport facilities and weather conditions. In general, fixed-wing aircraft offer no time advantages for transfers of under 160 km (100 miles).

Patients with entrapped gas (e.g. pneumothorax, significant abdominal distension) are better not to travel by air. If air travel is necessary, the aircraft should fly at low levels if it is unpressurised; otherwise, expansion of the trapped gases with decrease in ambient atmospheric pressure may make ventilation difficult.

Communication

Good communication between the referring and receiving institutions is crucial to survival and expedites treatment prior to transportation. Any change in the patient's condition should be reported to the receiving unit in advance of arrival. Detailed documentation of the history and written permission for treatment, including surgery, should be sent with the neonate. In addition, neonates require 10 mL of maternal blood to accompany them, as well as cord blood and the placenta, if available.

Details of stabilisation procedures may be discussed with the transport team, or receiving institution, if difficulties arise while awaiting the transport team's arrival.

Written permission for transport is required. A full explanation of what has been arranged and why, and an accurate prognosis should be given to the parents. They should be allowed as much access as is possible to the neonate prior to transport. The parents may be given a digital photograph of their child, taken before departure or at admission to hospital, if they are to be separated.

Stabilisation of neonates prior to transfer [Table 2.2]

Temperature control

An incubator or radiant warmer is used to keep the neonate warm. Recommended incubator temperatures are shown in Table 2.3. The neonate should remain covered, except for parts required for observation or access. Axillary or rectal temperatures should be taken half-hourly, or quarter-hourly if under a radiant warmer.

Table 2.2 Neonatal medical conditions requiring stabilisation before transport

1. Prematurity
2. Temperature control problems
3. Respiratory distress causing hypoxia and/or respiratory failure
4. Metabolic derangements
   - Hypoglycaemia
   - Metabolic acidosis
   - Hypocalcaemia
5. Shock
6. Convulsions

Table 2.3 Incubator temperature

Neonate's weight (g)	Incubator temperature (°C)
<1000	35–37
1000–1500	34–36
1500–2000	33–35
2000–2500	32–34
>2500	31–33

Respiratory distress

Oxygen requirements

Enough oxygen should be given to abolish cyanosis and ensure adequate saturation. Pulse oximeter oxygen saturation levels >97% indicate adequate oxygenation. If measurements of blood gases are available, an arterial PO_2 of 50–80 mmHg is desirable. Although an excessively high PO_2 is liable to initiate retinopathy of prematurity, a short period of hyperoxia is less likely to be detrimental than a similarly short period of hypoxia.

Respiratory failure

Neonates in severe respiratory failure (on clinical grounds or PCO_2 > 70 mmHg), or those with apnoea, may require endotracheal intubation and intermittent positive-pressure ventilation. Special attention must be paid to those neonates with CDH.

Metabolic derangements

Hypoglycaemia should be corrected by intravenous glucose. Monitoring of neonates at risk should be done with Dextrostix, with intravenous access by the umbilical or a peripheral vein.

An infusion of blood or plasma expander at 10–20 mL/kg over 30–60 min may be required to correct shock.

Acid–base balance should be estimated if facilities are available. Otherwise, a small volume of sodium bicarbonate (3 mmol/kg, slowly IV) may be given to an infant with severe asphyxia, has had recurrent hypoxia or has poor peripheral circulation. The best way, however, to correct acidosis is to correct the underlying abnormality.

Convulsions should be controlled with phenobarbitone (10–15 mg/kg, IV or orally) or diphenylhydantoin (15 mg, IV or orally).

Specialist advice regarding management of specific conditions should be sought from the transport agency. For example, in gastroschisis and exomphalos, the exposed viscera should be wrapped in clean plastic wrap to prevent heat loss; moist packs or gauze should never be used. A nasogastric tube with continuous drainage is required for patients with CDH (Chapter 5), bowel obstruction (Chapter 7) or gastroschisis (Chapter 9). In oesophageal atresia, frequent aspiration of the blind upper oesophageal pouch, at 10–15 min intervals, is essential to minimise the risk of aspiration (Chapter 6).

KEY POINTS

- Sick neonates need stabilisation before transport.
- Early transport is best done by a specialised team.
- Communication with both parents and receiving surgical centre is crucial.

Further reading

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CHAPTER 3
The Child in Hospital

CASE 1

Erin, aged 2 years, is seen in the surgical clinic because of an inguinal hernia. During the explanation prior to filling out the consent form, the surgeon describes the use of 'invisible stitches', a waterproof dressing and local anaesthetic.

Q 1.1 Will the operation be done under local anaesthetic?

Q 1.2 Why are 'invisible stitches' important?

Q 1.3 Why should the dressing be waterproof?

CASE 2

Jacob, aged 6 years, attends the surgical clinic very reluctantly because he is apprehensive about an upcoming epigastric hernia repair.

Q 2.1 What are his major fears likely to be?

Great effort should be made to minimise psychological disturbances in children undergoing surgery. The important factors to consider are the child's age and temperament, the site, nature and extent of the operation, the degree and duration of discomfort afterwards, and the time spent in hospital. Children between 1 and 3 years of age are the most vulnerable and do not like to be separated from their parents. For this reason a parent is encouraged to be with their child during induction of the anaesthetic and to be present in the recovery room as the child awakes from the anaesthetic.

The temperament and ability of children to cope with stress are infinitely variable; the trust which children are prepared to grant those who care for them is a measure of the confidence they have in their own family circle. Major disturbances within the family may affect the patient's equanimity and the ability of parents to give support. Sometimes, elective operations may need to be deferred for stressful family events, such as the following:

- The arrival of a new baby
- A death in the family
- Shifting to a new house

Preparation for admission

Preparation for elective admission is important for children over 4 years of age and, whether assisted by a booklet (see Further Reading) or advice, is largely in the hands of the parents whose acceptance of the situation is its endorsement in the child's eyes. If the parents are calm the child too is usually calm, but if the parents are highly anxious, it is likely their child will be fearful and uncertain – and difficult to manage.

The child needs a brief and simple description of the operation, and if something is to be removed, it should be made clear that it is dispensable. Children should also be told that they will be asleep while the operation is performed, that they will not wake during the operation and that it will be already over when they do wake up. They also will want to know when they will be able to go home, and whether they will be 'stiff' and a little 'sore' for a day or so. It is counter-productive to say that it will not hurt at all, for honesty is essential to preserve trust.

How the child's questions are handled is just as important as the factual content of the answers; possible sources of fear should be dealt with and the pleasant

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aspects suitably emphasised. The amount of information must be adjusted to the child's age and particular needs; more detail will be expected by older children. Many hospitals have 'play specialists' who are expert in addressing children's anxieties and provide distractions for those who are particularly anxious.

Effect of site of operation

Operations on the genitalia or the body's orifices, including circumcision after the age of 2 years, are more likely to cause emotional upset than other operations of the same magnitude. One or both parents should stay with the child and suitable occupational or play therapy can be of considerable value. Most inguinoscrotal operations (e.g. herniotomy or orchidopexy) are well tolerated and the use of local anaesthesia infiltration during surgery means that they have little discomfort afterwards. Many boys who have experienced both operations would prefer, in retrospect, bilateral orchidopexy to tonsillectomy.

Day surgery

Time spent in hospital should be as short as possible. 'Day Surgery' with admission, operation and discharge a few hours later, is cost-effective, convenient and suitable for about 80% of elective paediatric surgery.

The greatest advantage is minimising the psychological impact on the child, which is magnified by sleeping away from home for even one night. There are many other obvious advantages, including minimal disturbances of breast feeding and reduced travelling by parents (i.e. fewer visits to the hospital) and less nosocomial infection, alongside reduced burden on healthcare resources and budget.

Although operative technique is important (haemostasis, secure dressings), day surgery has been made safe and acceptable by special anaesthetic techniques: timing and choice of premedication and general anaesthetic agents, minimal trauma during intubation (particularly the use of the laryngeal mask rather than endotracheal intubation), quick reversal of anaesthesia and long-acting local anaesthetic blocks or caudal analgesia in lieu of the usual post-operative injections of narcotics.

In the most vulnerable 1–3 year old age group, day surgery has reduced the likelihood of behavioural disturbances. Suitable operations for day surgery depend on parental attitudes, logistics and careful selection of individual patients.

Ward atmosphere and procedures

Unlimited visiting by parents, living-in quarters for parents and an understanding and empathetic approach by all staff lead to an informal and friendly atmosphere in hospital. The procedures for investigations or preparation for operation should be scrutinised carefully to see whether they are really necessary. Blood tests or x-rays are rarely required for elective day surgery.

Anaesthesia is an important source of fear and distress. The presence of a parent is very helpful during most anaesthetic inductions. Anaesthetic rooms often have large television screens or electronic games which act as a distraction during induction. Effective premedication, skilful intravenous induction and the prompt administration of hypnotics and analgesics after operation keep discomfort to the absolute minimum. Again, the early presence of a parent in the recovery room may reduce the child's stress as they wake from anaesthesia.

Even after major abdominal operations, some toddlers will be walking within 24 h. They might just as well be playing on the floor or sitting at a table, and today that is where they are, with no subsequent ill effects. A play room is not required for most post-operative patients, since once they can walk to the toilet and play room, they may be discharged home. The child usually sets the pace of convalescence, and as a general rule will show no desire to move when they should rest, for example, during a period of paralytic ileus.

Play materials, a day room, television and bright surroundings, act as constant stimuli to those who are well enough to be 'up and doing'. Play specialists are involved in the management of children who have a longer hospital admission or require frequent dressing changes (e.g. burns patients) and may significantly reduce the amount of analgesia required.

A single, absorbable subcuticular suture may be used to close almost all incisions, which avoids the anxiety and time spent in removing sutures. It also gives an excellent cosmetic result. A waterproof dressing allows normal washing and may be left on until the wound is fully healed.

Parental support

The parents always require consideration, especially when a first-born baby is transferred to a children's hospital on the first day of life. The baby may stay there for several weeks, at precisely the time when the mother's emotions are in turmoil and she would normally be establishing a new and unique relationship. Feelings of guilt at producing a neonate with a congenital abnormality, or inadequacy following removal of the neonate from her care and the lack of close physical contact, may lead her to have difficulty bonding to her baby and produce an exaggeration of the usual puerperal emotional instability. To help overcome this when separation is unavoidable, the mother should be given a photograph of her baby, and should see the baby again as soon as possible, and be involved in the day-to-day care, of her child as much as the illness permits (Chapter 2).

Response of the child

The average child's natural optimism, freedom from unfounded anxiety, remarkable powers of recuperation and apparently short memory for unpleasant experiences may make recovery from even major operations a relatively short and simple matter. Most children are out of bed in 2–3 days and active for much of the day, or already at home by 5 days after many major operations.

Even with minor operations the child may have disturbed behaviour for several months after leaving hospital, and parents should be made aware of this possibility. Signs of insecurity, increased dependency and disturbed sleep are not uncommon but fortunately are of short duration when met with warm affection, reassurance and understanding by the parents.

The undesirable psychological effects of an operation must be put in proper perspective by mentioning the beneficial effects which so often follow operation: the well-being after repair of an uncomfortable hernia; the freely expressed satisfaction at the excision of an unsightly lump or blemish.

Finally, in many older children there is a detectable increase in confidence and poise which comes from facing, and coping adequately with, an operation. This may be the first occasion on which the child has been away from home, and metaphorically at least, standing on his or her own two feet.

The timing of operative procedures

Surgical conditions in infancy and childhood may be classified according to the degree of urgency with which treatment should be carried out. Three categories may be distinguished:

1. The immediate group – conditions where immediate investigation and/or definitive operation is required, for example, torsion of the testis, intussusception, appendicitis.
2. The expedited group – where treatment is not urgent but should be undertaken without undue delay, for example, infant inguinal hernia.
3. The elective group – where operation is performed at an optimum age determined by one or more factors which affect the patient's best interests, for example, undescended testes, hypospadias.

The immediate group

Trauma, acute infections, abdominal emergencies and acute scrotal conditions fall into this category. A particularly important subgroup is neonatal emergencies. Most of these are the result of developmental abnormalities causing functional disorders, some of which may be life-threatening. The best prognosis depends upon early diagnosis and timely transport to a hospital where the appropriate skills and equipment are available. Sometimes this is best done before the neonate is born, as in a congenital diaphragmatic hernia and gastroschisis (see Chapters 4–11); fortunately, most of these conditions are easily diagnosed on antenatal ultrasonography.

The expedited group

Inguinal herniae are prone to strangulation, especially in the first year of life. For this reason, herniotomy should be performed promptly: for those less than 1 year of age, this usually means the operation is performed in the coming days or weeks on the next semi-urgent or elective list (e.g. '6–2 rule': for a baby <6 weeks, herniotomy within 2 days; for infants 6 weeks to 6 months, herniotomy within 2 weeks; for children 6 months to 6 years, herniotomy within 2 months). Investigation of swellings or masses suspected to be malignant should be undertaken within a day or two of their discovery, in close consultation with the regional paediatric oncology service. For many malignancies, several cycles of chemotherapy are given before definitive surgery is undertaken.

The elective group

Factors favouring deferment of operation

Factors which favour deferment of operation, and hence may determine an optimum age, include the following:

1 The possibility of spontaneous correction or cure. In infants, scrotal hydroceles, encysted hydroceles of the cord, true umbilical herniae and sternomastoid tumours all show a strong tendency to spontaneous resolution. An operation is only required for those few that persist well beyond the age of natural resolution.
2 Infantile haemangiomas (Strawberry naevi) progress and enlarge in the first year of life but usually involute and fade spontaneously in the ensuing 2–4 years (Chapter 50). In general, they should be left alone or treated medically. Operative intervention is rarely required and only in specific circumstances, such as a haemangioma which obstructs the visual axis, or has failed to respond to medical management.
3 The difficulties posed by delicate structures may be avoided by postponing operation until they are more robust, although this is seldom the sole reason for deferring operation; for example, an undescended testis may be repaired more easily in a 6–12-month-old boy than shortly after birth.
4 The development of cooperation and comprehension with age. Voluntary exercises are important after some operations and it may be desirable to defer them until the necessary degree of cooperation is forthcoming.
5 The effects of growth are important in some instances. Chest wall deformities are corrected at adolescence, once chest wall growth is almost complete.
6 Coexistent anomalies and intercurrent diseases, for example, infections, will affect the timing of operations. The situation in each patient should be assessed to establish the order of priorities when there are multiple abnormalities and thus determine whether the treatment of non-urgent conditions should be deferred temporarily.

Factors favouring early operation

Factors which favour early operation include capacity for healing and adaptation in the very young. For example, a fracture of a long bone at birth causes such an exuberant growth of callus that clinical union occurs in 7–10 days, and the subsequent moulding will remove any residual bony deformities.

1 Stimulation of development by early treatment occurs in neonates with a developmental dysplasia of the hip. When splinting is commenced in the first week of life, this will prevent the secondary dysplasia of the acetabulum and femur, which once was thought to be the primary cause of the dislocation.
2 Malleability of neonatal tissues is an advantage, for example, talipes, where the best results are obtained when treatment is commenced immediately after birth.
3 Avoidance of undesirable psychological effects. Often these may be prevented by completing treatment, including repetitive painful procedures, before the memory of things past is established (at about 18 months) or before the child goes to school, where obvious deformities or disabilities are likely to attract attention.
4 Effect on the parents. The family as a whole should be considered and when it is not disadvantageous to the child, early operation may resolve parental anxiety and prevent rejection of the child.

KEY POINTS

- All hospital and operative procedures are modified to reduce psychological stress in children.
- As much as possible, all painful procedures are done when children are anaesthetised.
- Invisible stitches, waterproof dressing and local anaesthetic given before waking mean the wound may be left alone post-operatively.
- Day surgery avoids separation anxiety in older children.

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PART II
Neonatal Emergencies

CHAPTER 4
Respiratory Distress in the Newborn

CASE 1

Antenatal ultrasonography has revealed a large solid lesion occupying most of the right chest. At birth respiratory distress develops rapidly: a chest x-ray shows a partly cystic and solid lesion in the right lower zone.

Q 1.1 What is the differential diagnosis?

Q 1.2 What treatment is needed?

CASE 2

After a breech delivery, acute cyanosis and respiratory distress develop in a term neonate. Breath sounds are diminished over the left chest.

Q 2.1 What is the likely problem?

Q 2.2 What emergency treatment may be needed?

When a newborn baby breathes more rapidly than normal, respiratory distress is present. The degree of distress may be slight initially, but progressive deterioration may culminate in irreversible respiratory failure.

Neonatal respiratory distress is not normally the province of the paediatric surgeon, but it may occur in a specific group of neonatal patients in whom the causes are amenable to surgical correction. Respiratory failure may have developed already when the baby presents, and prompt action may save the neonate's life and regain the opportunity for corrective surgery. Those caring for newborns must be able to recognise respiratory distress and the paediatric surgeon must be familiar with its causes and the principles of management.

In only a few cases may a conclusive diagnosis be made clinically, and x-rays of the thorax and abdomen should be obtained as soon as possible.

Recognition of respiratory distress

The key clinical feature is a raised respiratory rate. Tachypnoea is present in the neonate if the respiratory rate exceeds 60 breaths per minute. In addition, tachycardia is almost invariably present, and if the pulse rate exceeds 200 beats per minute, the situation is serious. Bradycardia is also a dangerous sign and often portends imminent respiratory failure.

Other cardiovascular signs, such as apparent 'dextrocardia', and the nature of the peripheral pulses, will provide further clues as to the underlying cause. The abdomen may be scaphoid in babies with a congenital diaphragmatic hernia, but may be distended when there is a pulmonary cause for the respiratory distress. Intestinal obstruction and neonatal peritonitis may cause abdominal distension, thus leading to respiratory embarrassment. Respiration may be laboured or associated with chest wall deformity, or there may be inspiratory (sternal) retraction, indicative of obstruction of the airways.

A surgical cause is present in a minority of babies with respiratory distress, and the surgeon must be familiar with the differential diagnosis, for example, hyaline membrane disease, meconium aspiration and cerebral birth injuries [Table 4.1]. Antenatal ultrasonography, obstetrical details and any abnormal physical signs will help determine the cause of tachypnoea. A baby who is pale and cyanosed but improves with oxygen may have a congenital diaphragmatic hernia (Chapter 5). A scaphoid abdomen and barrel chest, with the heart sounds best heard on the right, are supportive

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Table 4.1 Causes of neonatal respiratory distress

Type of obstruction	Examples
Upper respiratory tract obstruction	
Nasal	Choanal atresia
Pharyngeal	Pierre-Robin syndrome
Laryngeal	Hamartoma of tongue
Tracheal	'Infantile larynx'
	Vocal cord palsy
	Subglottic vascular anomaly
	Laryngeal web or cyst
	Tracheomalacia
	Massive lymphangioma (cystic hygroma)
	Vascular ring
Lower respiratory tract obstruction	Meconium aspiration
	Aspiration of gastric contents
	Lobar emphysema (congenital)
Alveolar disease	Hyaline membrane disease
	Pneumonia
	Congenital heart disease
	Pulmonary oedema
	Congenital diaphragmatic hernia
Pulmonary compression	Pneumothorax
	Congenital diaphragmatic hernia
	Repaired exomphalos or gastroschisis
	Congenital lobar emphysema
	Congenital lung cysts
	Bronchogenic cysts
	Duplication cysts
	Abdominal distension
Neurological disease	Birth asphyxia
	Apnoea of prematurity
	Intracranial haemorrhage
	Convulsions

physical signs of a left congenital diaphragmatic hernia, and a chest x-ray will confirm the diagnosis. By contrast, a baby with cyanosis and respiratory distress which is relieved by crying may have choanal atresia (Chapter 14).

The principles of management

When respiratory failure is present already, urgent treatment is required, regardless of the underlying cause. Accurate diagnosis is based upon the clinical history and signs, and subsequent imaging. The degree of respiratory or metabolic acidosis must be determined to guide the resuscitation required. Where applicable, an operation is undertaken to correct the cause, usually after correction of the physiological disturbances.

Specific conditions

An important aspect of neonatal respiratory distress is that many of the causes have a wide clinical spectrum, for example, a congenital diaphragmatic hernia may produce a direct threat to life within minutes of birth, yet on other occasions may cause no symptoms until well beyond the neonatal period (Chapter 5). Congenital pulmonary airway malformations and pulmonary sequestration are typically diagnosed on antenatal ultrasound, but only infrequently cause respiratory embarrassment in the neonatal period. Choanal atresia is discussed in Chapter 14 and oesophageal atresia in Chapter 6.

Malformations that involve one lung and cause neonatal respiratory distress include congenital lobar emphysema and congenital cystic disease of the lung. The physical signs are not diagnostic and imaging is required to make the diagnosis. There are considerable variations in the clinical picture, and when there is persisting respiratory distress, an operation may be indicated. Resection of the affected lung segment not only removes functionless pulmonary tissue with little or no gaseous exchange but also allows expansion of the normal pulmonary segments that have been compressed by the over-distended segment, lobe or lobes.

Congenital lobar emphysema

The aetiologies of congenital lobar emphysema are variable and include congenital deficiency of the bronchial cartilage and extrinsic compression from an intrathoracic cyst. The end result is expiratory obstruction and air trapping in the affected lobe, leading to massive distension of a pulmonary lobe.

The cardinal symptom is tachypnoea that is most noticeable when the baby feeds. Not infrequently there is a dry cough and stridor. Cyanosis may be an indication for urgent treatment. The mediastinum is displaced and the chest wall over the affected area is prominent and has relatively reduced respiratory excursion; breath sounds are diminished and the percussion note typically is hyper-resonant.

X-rays show an area of increased radiolucency in which there are some bronchovascular markings. There may be downward displacement of the diaphragm on

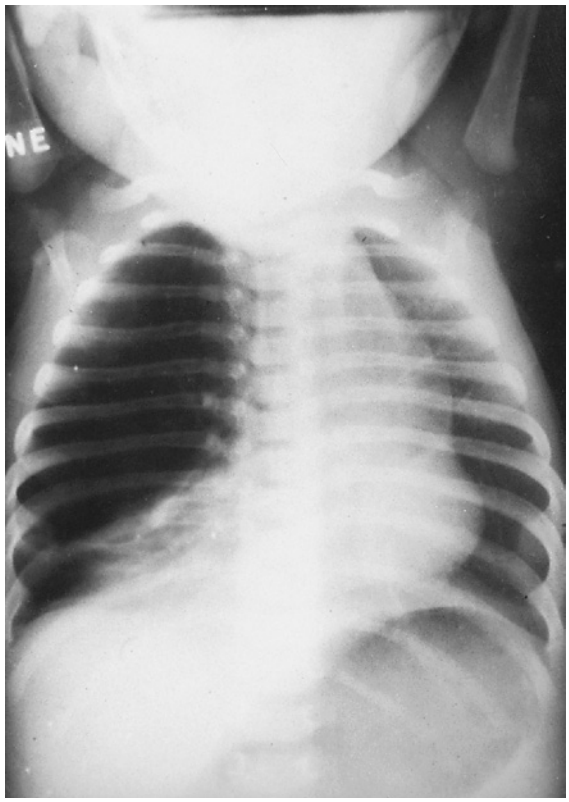

Figure 4.1 Congenital lobar emphysema of the right upper lobe that is overdistended and herniating across the midline.

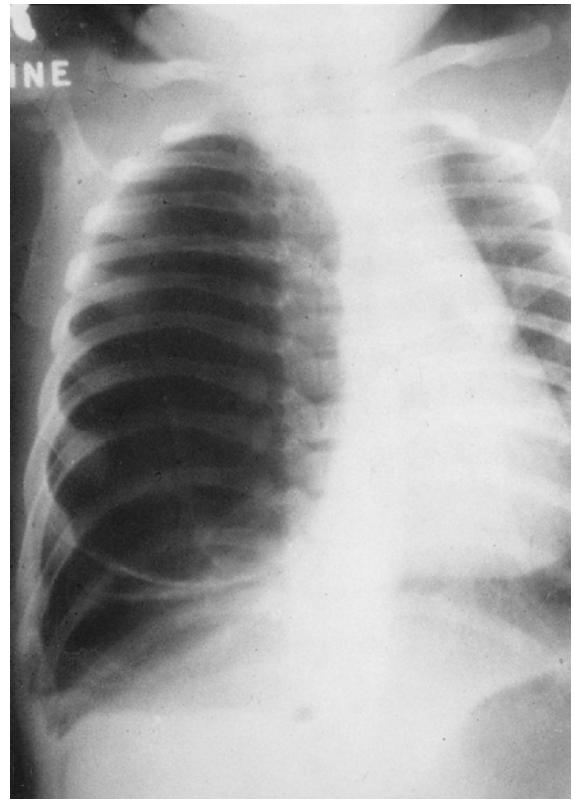

Figure 4.2 Congenital cystic lung. A giant cyst has replaced the right lower lobe, compressing the remaining right lung and herniating across the midline to displace the heart and compress the left lung.

the affected side, and the over-distended lung may herniate across the midline [Fig. 4.1]. The lobes most commonly affected are the left upper lobe or the right middle lobe. An increasing number of patients are now managed non-operatively, but when required, operative management is lobectomy.

Congenital cystic lung

The clinical features are similar to those of congenital lobar emphysema, in that respiratory distress often occurs early, but usually it is more urgent and severe.

X-rays show a large cyst with a sharply defined border [Fig. 4.2] or an extensive multicystic area. There is typically compression and collapse of unaffected areas of the lungs and displacement of the mediastinum.

The operative aim is to remove the portion of the lung that is functionless and interfering with the function of the surrounding normal lung. Depending on the distribution of disease, resection of the affected lobe or even pneumonectomy may be required.

Pulmonary sequestration

Pulmonary sequestration is an uncommon malformation in which there is non-functioning lung tissue which has no connection with the normal bronchial tree, and a blood supply which arises from an anomalous systemic artery, often directly from the aorta [Fig. 4.3] (Chapter 49). It usually occurs on the left side and may be either intralobar or extralobar, depending on whether it shares visceral pleura with the normal lung. It may be diagnosed on antenatal ultrasonography, may present as a pulmonary infection, because of its space-occupying effect, or be found incidentally on chest x-ray. The sequestration is resected by thoracoscopy or by open thoracotomy.

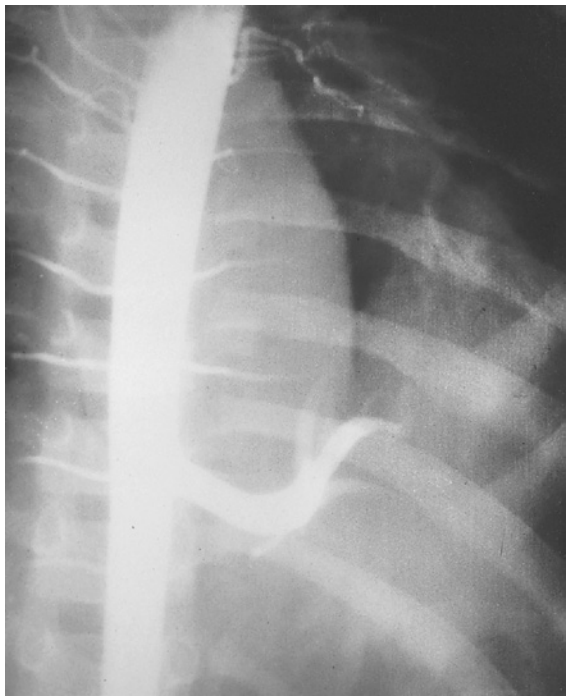

Figure 4.3 Anomalous blood supply from the aorta to a left pulmonary sequestration.

Congenital pulmonary airway malformation

Congenital pulmonary airway malformations (CPAMs) include a range of localised abnormalities in which the bronchiolar tissue is abnormal, with communicating cysts and a relative paucity of cartilage. Previously, these lesions were termed 'congenital cystic adenomatoid malformation' .They may be diagnosed on antenatal ultrasonography as a cystic or solid mass in one part of the lung. Maternal polyhydramnios and mediastinal shift may occur. Many CPAMS observed on antenatal ultrasonography regress and have resolved by term.

The majority of patients born with a CPAM are asymptomatic. However, those CPAMs that present postnatally may do so in three ways:

1 Respiratory distress (60%),
2 Infectious complications, e.g. recurrent pneumonia (20%) and
3 Incidental finding on chest x-ray (20%).

Symptomatic or complicated CPAMs are definitively managed by surgical resection. The management of antenatally diagnosed CPAMs which remain asymptomatic is more controversial, but non-operative management with follow-up is a valid alternative to elective resection in some of these cases.

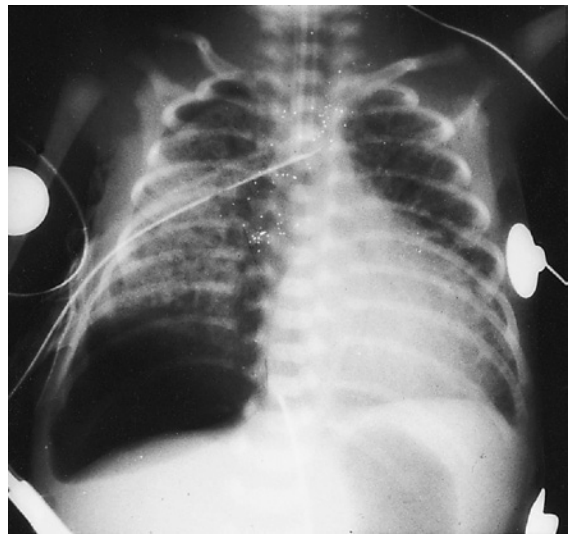

Figure 4.4 Severe pulmonary interstitial emphysema.

Mediastinal conditions

Very rarely, large cystic teratomas and duplication cysts cause respiratory distress and should be removed. In the neonate, oesophageal duplication cysts may present with increasing respiratory distress because of their space-occupying effect compressing the normal airways.

Pulmonary interstitial emphysema

This is an acquired condition of extreme prematurity seen in infants where assisted ventilation is required for severe hyaline membrane disease. High ventilatory pressures force air into the lung interstitium, which tracks along peribronchial spaces, producing interstitial cysts which have a characteristic appearance on x-ray [Fig. 4.4]. Treatment is directed at reducing the ventilatory pressures. In severe and progressive cases, thoracotomy may be required to deflate the cysts. Refinements in neonatology have resulted in a significant decrease in the incidence of this condition, such that it is now seen rarely.

Neonatal Pneumothorax

Pneumothorax may occur as a complication of diffuse pulmonary disease such as meconium aspiration, or of a localised abnormality, for example, subpleural

emphysematous bleb. The pneumothorax may be suspected on clinical grounds by sudden deterioration in condition, displacement of the trachea or apex beat, or a hyper-resonant percussion note, but x-rays are typically required to confirm the diagnosis.

In neonates, the severity of the symptoms frequently is out of proportion to the size of the pneumothorax. Even a small pneumothorax may be associated with severe respiratory distress when there is pre-existing parenchyal lung disease and little respiratory reserve. Intercostal drainage is urgent.

Haemothorax

Haemothorax is an infrequent complication of haemorrhagic disease of the newborn and may produce an alarming clinical picture. This is due to mechanical factors which interfere with respiration and to the reduction of the circulating blood volume. Intercostal drainage and blood transfusion are required.

Acute respiratory failure in the neonate

Acute respiratory failure occurs when oxygenation and/or ventilation are impaired sufficiently to be an immediate threat to life. It is usually the result of asphyxia due to:

1 Birth asphyxia
2 Other injuries sustained during birth
3 Developmental anomalies, including congenital heart disease
4 Hyaline membrane disease in the premature neonate
5 Increased susceptibility to infection

The factors in neonates which predispose to respiratory failure are summarised in Table 4.2. With limited respiratory reserve, respiratory failure may occur rapidly.

Signs of respiratory failure

In the neonate, especially if premature, acute hypoxia causes pallor, apnoea, bradycardia, hypotension and lethargy. The clinical signs of hypercapnia – sweating, tachycardia and hypertension – are seen rarely, but pulmonary haemorrhage, cerebral haemorrhage, severe hyperkalaemia and hypoglycaemia all may occur as the result of hypoxia.

Table 4.2 Factors predisposing neonates to respiratory failure

Factors	Comments
Metabolic rate	Metabolism per kilogram is twice that of adults
Respiratory rate	Lung surface area per kilogram is similar to adult; so neonate has much less respiratory reserve
Compliance	Neonate's chest wall is less able to adjust to reduced lung compliance or increased airway resistance
Airway calibre	Relatively larger total airway resistance than in older children or adults
Airway obstruction	Narrow airways are more prone to obstruction by oedema and secretions
Temperature control	Relatively poor temperature regulation, especially in the premature. In a cold environment, oxygen consumption may increase two- or threefold

General management

A neonate with incipient respiratory failure requires close observation at all times. Neonates should be nursed in an isolette or under a radiant heater so that the temperature is controlled and observation unimpeded. Handling should be kept to a minimum, as it may increase oxygen consumption dramatically. Monitoring of heart rate and oxygen saturation is mandatory. Transcutaneous pO_2 and pCO_2 monitoring and BP monitoring are also preferable.

Oxygen

The method of delivery of oxygen depends upon the neonate's age, oxygen concentration required and the underlying condition. All patients having prolonged oxygen therapy must have continuous oximetry and serial arterial blood gas estimations with adjustment of inspired oxygen concentration to ensure adequate arterial saturation. Premature neonates receiving supplementary oxygen therapy are at risk of retinopathy of prematurity, for which frequent blood gas measurements are required to maintain the arterial pO_2 in the range of 6.6–10.6 kPa (50–80 mmHg). In the newborn, gentle suction is performed at intervals to remove pooled secretions and to stimulate coughing. However, pharyngeal and endotracheal suction may cause a sudden fall in arterial pO_2 that necessitates an increase in the concentration of oxygen in the inspired gases.

Fluids and feeding

Oral feeding should be suspended in children with severe dyspnoea, but enteral nutrition may be continued via nasogastric tube. If abdominal distension occurs, feeding must be discontinued to avoid regurgitation and aspiration, and to prevent splinting of the diaphragm, as these may cause additional respiratory embarrassment. Intravenous infusion may supply fluids and parenteral nutrition, but total fluid intake may need to be restricted in some patients with pulmonary disease.

Sodium bicarbonate may be required to correct metabolic acidosis (Chapter 2). Fluid management requires regular biochemical monitoring and an accurate record of fluid balance.

Temperature control

Seriously ill neonates are particularly vulnerable to cold stress, and consequently maintenance of body temperature is of vital importance (Chapter 2). The preterm neonate has a narrow 'thermoneutral' range in which oxygen consumption is minimised and optimal: abdominal wall skin temperature is optimal between 36 and 36.5 °C. Exposure to an environmental temperature of 20–25 °C increases oxygen consumption threefold and may precipitate cardiorespiratory failure. Critically ill neonates should be nursed in open cots with servocontrolled radiant heat so that access to them is not compromised. Insensible water loss may be increased, particularly in neonates of very low birthweight, but this may be taken into account when planning fluid replacements.

Monitoring

Respiratory and cardiovascular signs should be monitored, along with the oxygen concentration in the inspired air. Blood for gas analysis is obtained by percutaneous puncture or, more accurately, in samples from an indwelling catheter in a peripheral artery, which also may be used for a continuous record of the arterial pressure. Continuous transcutaneous oximetry is routine.

Ventilatory support

In neonates, endotracheal intubation is the preferred type of artificial airway [Table 4.3]. Tracheal tubes of appropriate size and composition may be left *in situ* for long periods with minimal adverse effects or complications.

Humidification of dry inspired gases is necessary to reduce the risk of viscid and retained sputum, atelectasis, blockage of the endotracheal tube with inspissated secretions and to preserve mucociliary function.

Table 4.3 Use of nasotracheal tube in neonates

Advantages	Disadvantages
Provides patent airway	Narrows the upper airways
Overcomes airway obstruction	Bypasses natural humidification, heating and filtering of inspired gases
Allows tracheobronchial toilet and suction	Prevents coughing and expectoration of secretions
Facilitates continuous positive airway pressure	May cause subglottic irritation and stenosis (which may be minimised by a correct-sized tube, allowing a small air leak during positive-pressure ventilation)
Enables mechanical ventilation	

Inspired gases should be delivered to the trachea at 37 °C, fully saturated with water vapour, using a safe, servocontrolled humidifier to help maintain body temperature and reduce insensible fluid losses from the airways.

Regular suctioning of the trachea is necessary to stimulate coughing and to remove accumulated secretions. Suctioning may cause hypoxia and atelectasis and may introduce infection, and techniques are used to avoid these risks. Gentle 'bagging' with an oxygen-rich mixture is used before and after suction to reduce hypoxia and re-expand the lung. In neonates at risk of retinopathy of prematurity, the oxygen concentration in the 'bag' should not be more than 10% higher than the mixture used for ventilation. In older children 100% oxygen may be used.

Continuous positive airways pressure

Continuous positive airways pressure (CPAP) is a technique that employs a distending pressure (5–10 cm H_2O) applied to the airways of a patient who is breathing spontaneously. It is used in pulmonary conditions causing hypoxaemia due to atelectasis, alveolar instability and intrapulmonary shunting. Continuous positive airways pressure increases functional residual capacity and compliance, re-expands areas of atelectasis, decreases intrapulmonary shunting and increases arterial pO_2. In premature neonates, CPAP will often improve the regularity of respiratory movements and decrease apnoeic episodes. The technique requires careful control to avoid reduced cardiac output, retention of fluids, rupture of alveoli and

pneumothorax. Non-invasive CPAP, for example nasal CPAP, should also be used with caution in the neonate with bowel obstruction due to the potential for exacerbation of abdominal distension caused by aerophagia.

Intermittent positive-pressure ventilation

Intermittent positive-pressure ventilation (IPPV) is used to correct hypoventilation and, in some situations (e.g. raised intracranial pressure and pulmonary hypertension), to produce hyperventilation and to lower arterial pCO_2. Mechanical ventilators have been designed specifically for neonatal use. IPPV is often combined with positive end-expiratory pressure (PEEP). PEEP is used for the same reasons as CPAP, that is as a means of improving oxygenation. The hazards of IPPV are greater than those of CPAP and relate directly to the pressure applied. Barotrauma to immature lungs may result in a chronic lung disease in neonates known as bronchopulmonary dysplasia.

Intermittent mandatory ventilation is a technique of mechanical ventilation in which a predetermined minute volume is guaranteed, even when the patient breathes independently from the ventilator. With neonatal ventilators, a constant flow is provided during the expiratory phase from which the neonate may breathe. It is a technique useful for weaning from mechanical ventilation and as a means of minimising barotrauma.

Controlled ventilation involves the use of relaxants and sedatives which paralyse respiratory movements, to completely abolish the work of breathing and improve gas exchange. The technique is useful in critically ill neonates and those with difficult ventilatory problems, but it should only be employed where expert surveillance and sophisticated monitoring are available. Inappropriate pressure settings may cause a pneumothorax with sudden deterioration, and inadvertent disconnection rapidly results in potentially fatal hypoxia.

KEY POINTS

- Neonatal respiratory distress should be diagnosed by tachypnoea, before cyanosis appears.
- A surgical cause is present in the minority but may be identified by physical examination and chest x-ray.

Further reading

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CHAPTER 5
Congenital Diaphragmatic Hernia

CASE 1

Within minutes of birth, a full-term boy develops increasing respiratory distress and becomes cyanosed. He fails to improve with upper airway suctioning. The pregnancy was uneventful. He looks barrel-chested and his abdomen is scaphoid.

Q 1.1 What is the most likely diagnosis?
Q 1.2 What investigation will confirm the diagnosis?
Q 1.3 What factors determine the outcome in these situations?

CASE 2

A newborn with a recently diagnosed left-sided congenital diaphragmatic hernia is about to be transferred to a paediatric surgical centre by air. He is currently being ventilated through an endotracheal tube and just maintaining adequate blood gas levels.

Q 2.1 Should his ventilation be increased during transport?
Q 2.2 Should any other manoeuvre be performed to reduce the likelihood of problems during transport?
Q 2.3 If he suddenly deteriorates, what complication may have happened?

Definitions

The diaphragm develops from four embryonic structures:

1 The septum transversum
2 The left and right pleuro-peritoneal membranes
3 Dorsal oesophageal mesentery
4 Somites at cervical segments 3–5

Congenital diaphragmatic hernia results from failure of formation or fusion of the components of the diaphragm, such that abdominal contents may move through a defect into the thoracic cavity. Sometimes failure of muscularisation may produce a thin, weak diaphragm, referred to as an eventration of the diaphragm.

The Bochdalek type is the most common variety of congenital diaphragmatic hernia (1 in 5000 live births) and results from a defect in the postero-lateral aspect of the diaphragm. During intra-uterine development, the small bowel, stomach, spleen and left lobe of the liver may pass through the defect in the diaphragm into the chest. Lung development is also abnormal in fetuses with congenital diaphragmatic hernia, with hypoplastic lungs and pulmonary vasculature. Recent studies suggest that lung hypoplasia may be a cause rather than consequence of congenital diaphragmatic herniae. ln many neonates the combined ventilation difficulties and pulmonary hypertension are severe enough to produce severe cardiorespiratory distress within minutes of birth and may not be compatible with life.

The Morgagni (retrosternal) type of diaphragmatic hernia is rare and results from a defect in the anterior midline, just behind the sternum [Fig. 5.1]. It usually contains part of the colon or small bowel and, less commonly, part of the liver.

Occasionally, a hernia may occur through the apex of the cupola or at the periphery adjacent to the costal margin. Oesophageal hiatal herniae may also occur and usually produce symptoms of gastro-oesophageal reflux.

Clinical features

Antenatal diagnosis

Most congenital diaphragmatic hernias are now diagnosed on antenatal ultrasonography. Factors that may indicate a worse prognosis on antenatal scanning [Box 5.1] will influence counselling of the parents-to-be.

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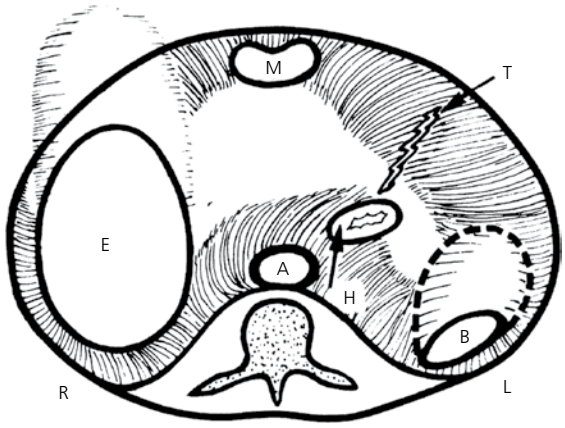

Figure 5.1 Diaphragmatic herniae. Diaphragm as seen from below, showing: B, Bochdalek left posterolateral defect; M, anterior or Morgagni type; H, hiatus for oesophageal and hiatus hernia; E, large eventration in the tendinous portion of the right cupola; T, a tear which causes a post-traumatic hernia; A, aorta; L, left; R, right.

Box 5.1 Possible antenatal markers of severity

- Early gestational age at diagnosis on ultrasonography
- Lung-to-head ratio (LHR) at 24–26 weeks' gestation <1.0
- Small fetal lung volume on 3D ultrasonography and MRI
- >50% liver in chest on right side
- Liver in chest on left side
- Stomach and spleen in chest on left side
- No hernial 'sac' (difficult to confirm on scanning)

Antenatal ultrasonographic diagnosis of a diaphragmatic hernia also allows the mother to be transferred to a tertiary paediatric surgical centre before birth. Successful *in utero* correction of diaphragmatic hernia and fetoscopic tracheal occlusion has been achieved in a research setting, but the techniques are complex and the indications are still being refined. To date, they have not resulted in improved survival, nor reduced morbidity, compared with current postnatal techniques.

Postnatal diagnosis

The majority of neonates born with a Bochdalek (postero-lateral) diaphragmatic hernia become symptomatic at, or shortly after, birth. Where pulmonary hypoplasia is severe, the neonate becomes cyanosed with severe respiratory distress within minutes of birth. In other patients there may be tachypnoea, increased respiratory effort, an hyperinflated chest, scaphoid abdomen and heart sounds are on the right side. This is because 85% of postero-lateral herniae involve the left hemidiaphragm. The remainder are right-sided (13%) or bilateral (2%). Associated anomalies occur in up to 40%, but most are minor and do not affect survival, for example, undescended testes. The most common serious abnormalities are heart defects, which affect 20–25% of patients.

Unlike postero-lateral hernias, anterior (retrosternal) hernias are usually asymptomatic unless strangulation occurs. Very rarely the hernia may protrude into the pericardial cavity rather than into the inferior mediastinum and cause cardiac tamponade, presenting as cardiorespiratory distress in the neonatal period.

Investigation

Diagnosis of a postero-lateral hernia is confirmed by a chest x-ray [Fig. 5.2]. In left-sided defects, loops of bowel may be seen in the left chest. The heart is deviated to the right. The hypoplastic left lung appears small and is further compressed by the mass effect of the herniating liver, gut and spleen. Sometimes the appearance may be difficult to distinguish from basal lung cysts, in

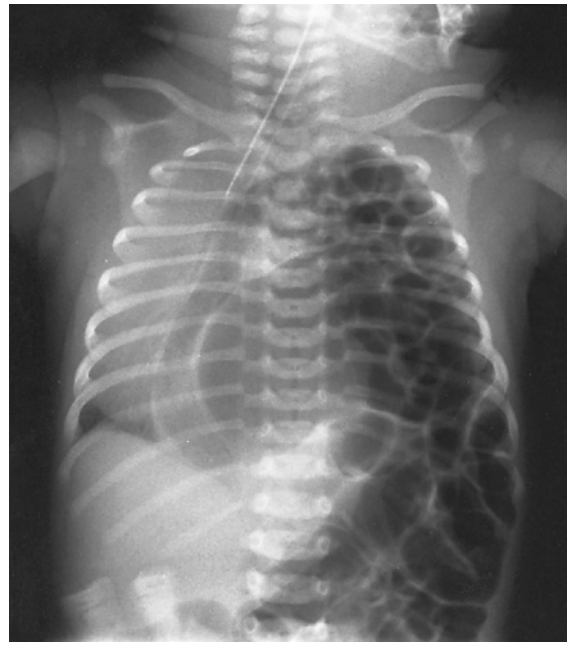

Figure 5.2 X-ray of congenital diaphragmatic hernia (Bochdalek type). Multiple bowel loops fill the left thoracic cavity and the heart is displaced to the right.

which case a repeat chest x-ray is performed after a nasogastric tube has been inserted, the tip of which may be seen in the chest. Alternatively, a barium study will show bowel within the thoracic cavity when there is a diaphragmatic hernia. The presence of a pneumothorax is important to recognise as it may indicate barotrauma and is associated with a poorer outcome.

Treatment

Postero-lateral (Bochdalek) hernia

Where antenatal ultrasonography has identified a diaphragmatic hernia, the best outcomes are achieved if the neonate is transferred to a tertiary paediatric surgical centre prior to birth. This is because these neonates may develop severe pulmonary distress very quickly after birth, making subsequent transfer difficult and potentially dangerous.

Initial treatment involves intensive cardiorespiratory support and insertion of a nasogastric tube to prevent bowel dilatation within the chest. Care must be taken to avoid hyperinflation and barotrauma of the small, hypoplastic lungs. High-frequency oscillatory ventilation in combination with nitric oxide has improved survival rates. Ventilation with a facemask ('bagging') should be avoided as this may force air into the stomach, increasing its volume at the expense of the already compromised lungs. Vigorous endotracheal ventilation should also be avoided because of the risk of causing barotrauma or tension pneumothorax, which may lead to the rapid demise of the neonate. Exogenous surfactant provides no specific benefit in newborns with diaphragmatic hernia. The key to success is careful gentle ventilation that minimises injury to the hypoplastic lungs.

Sudden deterioration of the neonate's condition during initial resuscitation or during transport suggests the development of a tension pneumothorax, and this may necessitate prompt release by needle aspiration or insertion of an intercostal drain. Fortunately, strict avoidance of hyperventilation and limited inflation pressures has made this complication rare.

The primary focus of management of the neonate with congenital diaphragmatic hernia is achieving adequate ventilation with control of pulmonary hypertension. To reduce the risk of injury to hypoplastic lungs in these patients, special ventilatory strategies may be required such as high-frequency oscillation or in very selected cases extracorporeal membrane oxygenation. Surgery to return the herniated contents to the abdominal cavity and to repair the defect in the diaphragm is only performed once the neonate's condition has stabilised. Surgery is typically deferred for at least 24 h after birth, but stabilisation may not be achieved for many days. Surgery generally involves repair through a transverse or subcostal abdominal incision on the same side as the defect. Thoracoscopic surgery has a role in selected cases, usually with smaller diaphragmatic defects.

Survival rates post-surgery of more than 80% are now being achieved, and may be higher in specialist centres. The major cause of death remains pulmonary hypoplasia and pulmonary hypertension, which is due to the small pulmonary vascular bed and to the changing resistance of the pulmonary arterioles: it resolves in most patients with time provided ventilation does not produce additional lung injury (barotrauma).

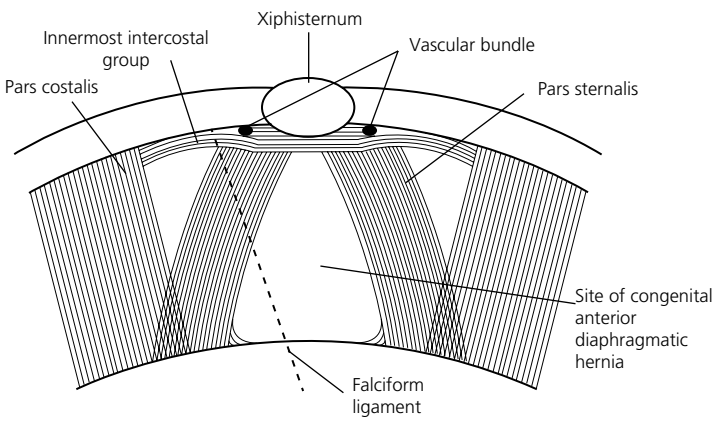

Figure 5.3 Schema of an anterior (Morgagni) diaphragmatic hernia during laparoscopic repair.

Anterior diaphragmatic hernia

Anterior diaphragmatic (Morgagni) herniae are often diagnosed on an incidental x-ray of the chest in a symptomless patient, but repair is still advisable because of the risk of strangulation of the bowel that protrudes through the defect. This is now commonly performed as a laparoscopic procedure [Fig. 5.3], with excellent results.

KEY POINTS

- Congenital diaphragmatic hernia is diagnosed on antenatal ultrasonography or by chest x-ray in a newborn with a barrel chest, scaphoid abdomen and/or respiratory distress.
- Ventilatory support, especially during transport, should be the minimum required to prevent deterioration, as hyperinflation with secondary barotrauma is a significant complication.
- Sudden deterioration is usually the result of a tension pneumothorax.

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CHAPTER 6

Oesophageal Atresia and Tracheo-oesophageal Fistula

CASE 1

A new mother has been admitted to a district hospital where she has delivered a baby boy at 38 weeks' gestation who, despite initial suctioning, appears to be salivating excessively and is very 'mucousy'. He has mild tachypnoea, but is pink.

Q 1.1 What manoeuvre must be undertaken to establish the cause of drooling?

Q 1.2 Are there any other abnormalities likely to be present?

Q 1.3 What needs to be done before and during transfer to a paediatric surgical centre?

CASE 2

Annabel is 38 and had difficult conceiving but now has a newborn girl. The baby looks dysmorphic and is noted to have a deformed forearm and thumb abnormalities and an anorectal abnormality. An orogastric tube could not be passed into the stomach.

Q 2.1 What other abnormalities are likely, and how would they be detected?

Q 2.2 What would you do before proceeding to surgery?

Oesophageal atresia is a congenital anomaly where there is complete interruption of the oesophagus. This produces a blind upper oesophageal pouch and a lower oesophageal segment which usually communicates with the trachea through a distal tracheo-oesophageal fistula. Less common variations of the abnormality also occur [Fig. 6.1].

Pathophysiology

Oesophageal atresia causes saliva (or milk if the neonate has been fed) to accumulate in the blind upper oesophageal pouch and spill over into the trachea, causing choking and cyanosis, and soiling of the lungs. Occasionally, gastric contents may pass through the distal tracheo-oesophageal fistula into the bronchial tree. Pulmonary complications may follow – initially atelectasis and then pneumonia. Abdominal distension from air passing down the fistula into the stomach may elevate and splint the diaphragm, compromising the neonate's ability to ventilate adequately.

Antenatal diagnosis

Oesophageal atresia is being increasingly diagnosed on antenatal ultrasonography by the observation of polyhydramnios, a small stomach or abnormal oesophageal contraction with swallowing. Congenital abnormalities associated with oesophageal atresia may also be evident, thus increasing the diagnostic suspicion.

Early diagnosis

Oesophageal atresia should be recognised as soon after birth as possible, for delay may lead to aspiration and progressive pulmonary complications. The diagnosis is confirmed when an orogastric catheter cannot be passed via the mouth through the oesophagus into the stomach.

Symptoms soon after birth

Oesophageal atresia should be suspected when a neonate appears to be drooling excessively. The neonate may

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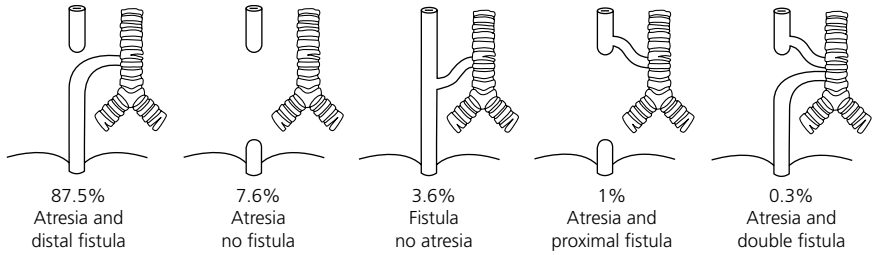

Figure 6.1 The anatomical variants of oesophageal atresia and/or tracheo-oesophageal fistula. The percentage frequency of each variant is shown.

have been resuscitated at birth with suction of mucus using a catheter but then, within minutes, develops rattling respirations, tachypnoea or fine frothy white bubbles of mucus in the nostrils or on the lips. This appearance of excessive salivation or of being 'excessively mucousy' is highly suggestive of oesophageal atresia. Often there is a history of maternal polyhydramnios.

Diagnosis before feeding

A firm 10-French catheter should be introduced via the mouth and passed gently down the oesophagus; if it becomes arrested at 9–11 cm from the lips, the diagnosis of oesophageal atresia has been established. A small catheter may curl up in the upper oesophagus and give a false impression of oesophageal continuity, hence the 10-French catheter [Fig. 6.2]. Ideally, oesophageal atresia should be diagnosed before the neonate is fed, because feeding may cause an acute episode of spluttering, coughing and cyanosis, with aspiration of milk into the lungs.

Pre-operative investigation

X-ray

An x-ray of the thorax and abdomen demonstrates the presence of air in the stomach and small bowel, which indicates that there is a fistula between the trachea and the lower segment of the oesophagus (the most common variant) [Fig. 6.3]. The x-ray also provides information on the condition of the lungs and the presence of vertebral and rib anomalies.

Echocardiography

Nearly 25% of neonates with oesophageal atresia have congenital heart disease. It is important to identify

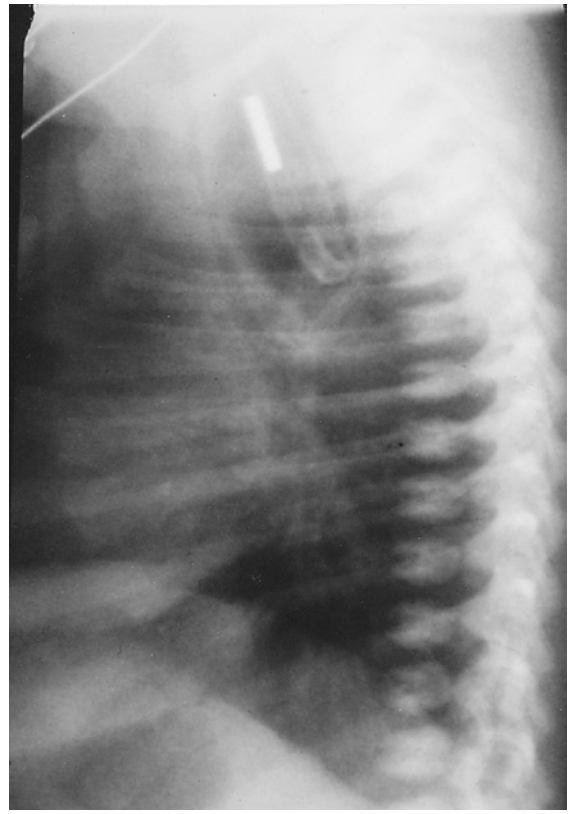

Figure 6.2 A small catheter will curl up in the upper oesophageal pouch and give a false impression of oesophageal continuity. Therefore, a wide-bore catheter, for example, 10-French, should be used.

cardiac lesions pre-operatively because a prostaglandin E_1 infusion must be commenced before repair of the oesophagus if the lesion is 'duct-dependent'. In most babies the cardiac defect does not delay the oesophageal surgery, and oesophageal repair takes precedence over cardiac surgery. An echocardiograph may identify a

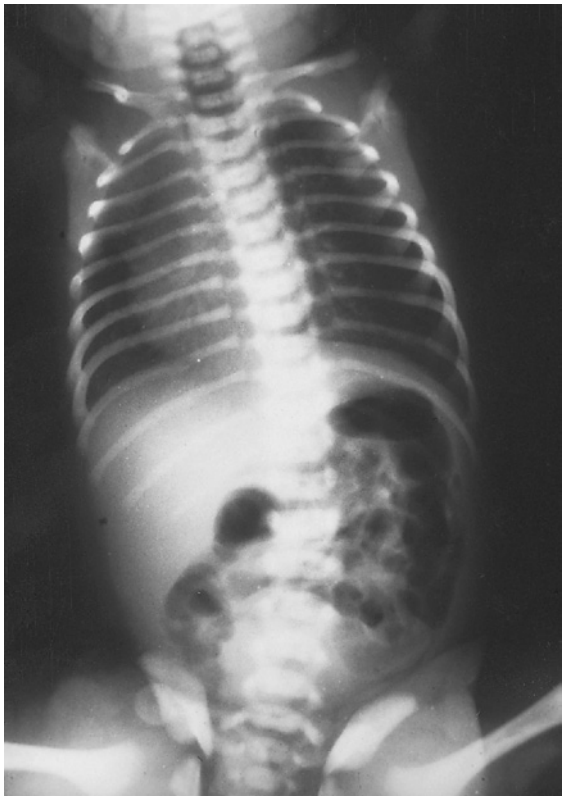

Figure 6.3 Plain x-ray of the chest and abdomen shows air in the oesophageal pouch, stomach and small bowel, indicating that there is a distal tracheo-oesophageal fistula.

Table 6.1 Frequency of associated abnormalities in oesophageal atresia

Abnormalities	Percentage
Cardiac	25
Gastrointestinal (including anorectal)	22
Vertebral or skeletal	20
Renal	15
Chromosomal (usually Trisomy 18 or 21)	7

right aortic arch, though usually this does not influence the operative approach to the oesophagus.

Renal ultrasonography

If the neonate has not passed urine, a renal ultrasound scan must be performed to exclude bilateral renal agenesis. If the neonate has no kidneys or has severely dysplastic kidneys, as occurs in 3%, operative repair of the oesophageal atresia is not justified [Table 6.1].

Table 6.2 Multiple malformation associations seen with oesophageal atresia

VATER (VACTERL)	CHARGE
Vertebral	**C**oloboma
Anorectal	**H**eart
Cardiac	**A**tresia choanae
Tracheo-o**E**sophageal	**R**etarded growth
Renal	**G**enital hypoplasia
Limb (including radial)	**E**ar

Genetic consultation

If the neonate has dysmorphic facies and other features suggestive of a major chromosomal abnormality, early genetic consultation is mandatory. The two most common chromosomal abnormalities are Trisomy 18 and Trisomy 21. In some cases, treatment may be delayed until after the results of chromosomal analysis are known. There are a number of malformation clusters, such as the VATER (or VACTERL) and CHARGE associations, which are common with oesophageal atresia [Table 6.2].

Treatment

The anomaly is treated by early complete correction, with division of the tracheo-oesophageal fistula and construction of an end-to-end oesophageal anastomosis. Before the operation, the upper pouch should be kept empty by frequent suction, and the neonate should be placed in an incubator or under an overhead heater to avoid heat loss. Vitamin K is given intramuscularly and intravenous fluids commenced. Intravenous antibiotics are given pre-operatively.

The operation is usually performed within 12 h of admission to hospital. The fistula is closed through a right posterolateral extrapleural thoracotomy or thoracoscopically, and direct end-to-end anastomosis of the upper and lower segments of the oesophagus establishes oesophageal continuity. Postoperatively, feeds may be commenced after 3 or 4 days.

Anatomical variations

Oesophageal atresia without a fistula

In this variant, the lower oesophagus is not connected to the trachea. On the initial x-ray, these babies show no gas below the diaphragm [Fig. 6.4]. Typically, the gap between

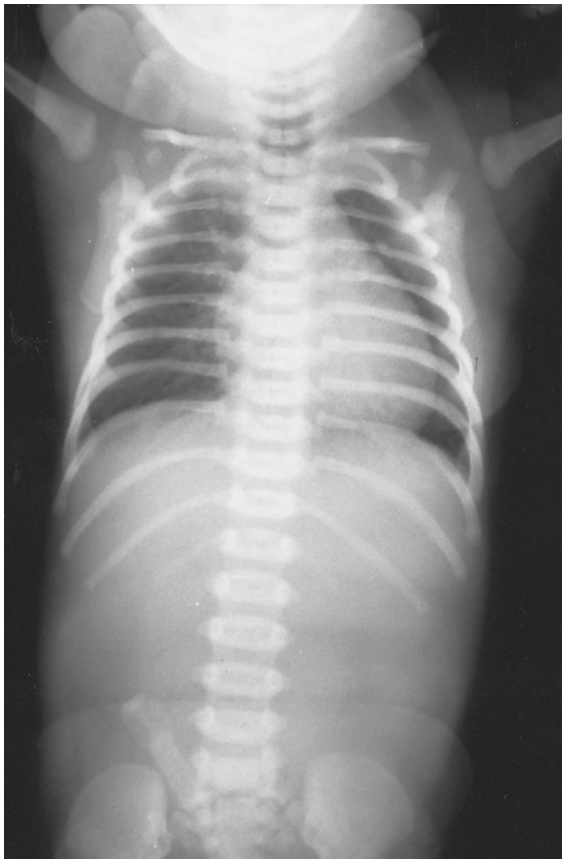

Figure 6.4 In oesophageal atresia without a distal tracheo-oesophageal fistula, there is no air below the diaphragm. Most of these infants have no fistula, while a few have a proximal tracheo-oesophageal fistula.

the two segments of the oesophagus may be so great that a primary anastomosis is impossible at birth. A gastrostomy is fashioned to allow enteral feeding, and the overflow of saliva from the upper pouch into the respiratory tract is controlled by frequent suction. At about 6–12 weeks of age, an oesophageal anastomosis is performed. Occasionally, this fails and oesophageal replacement is required. In a further variant, there may be a fistula between the upper oesophageal pouch and the trachea ('proximal tracheo-oesophageal fistula'): this variant also presents with no gas below the diaphragm on x-ray.

'H' fistula

Sometimes the oesophagus is intact but there is a fistula between the trachea and oesophagus, usually at the level of C7 or T1. These neonates present in the first week or so of life with episodes of coughing, cyanosis during feeding and pulmonary complications. Sometimes they present later with a history of recurrent pulmonary infections or abdominal distension mimicking a bowel obstruction. The diagnosis is made on bronchoscopy or by performing a contrast swallow or mid-oesophageal contrast study, and observing passage of contrast through the fistula into the trachea. Treatment is by operative division of the fistula, usually through a cervical approach.

Special problems

Effect of prematurity

Prematurity is commonly associated with oesophageal atresia. Provided facilities are available, these neonates should have their oesophageal atresia repaired early, before the expected respiratory distress of hyaline membrane disease becomes severe. Failure to divide the tracheo-oesophageal fistula early in these neonates may lead to severe problems with ventilation, because air escapes preferentially through the fistula into the stomach and may even cause gastric perforation and tension pneumoperitoneum.

Gastro-oesophageal reflux

Gastro-oesophageal reflux is common in patients with previously repaired oesophageal atresia and, in conjunction with delayed oesophageal clearance, may contribute to the development of an oesophageal stricture. The child may require treatment with a proton-pump inhibitor or a fundoplication. In its extreme form, aspiration secondary to gastro-oesophageal reflux may lead to acute life-threatening events and is a cause of mortality in oesophageal atresia patients. The longer term effects of gastro-oesophageal reflux on the oesophagus, and the risk of Barrett's oesophagitis and oesophageal cancer, remain unclear.

Tracheomalacia

Tracheomalacia is a structural weakness of the trachea that commonly occurs in association with oesophageal atresia. It is responsible for the 'seal bark' brassy cough characteristic of oesophageal atresia patients. It tends to improve with time but in the neonatal period may cause significant breathing difficulties. Occasionally, splinting of the trachea (tracheopexy, aortopexy) is required to prevent life-threatening collapse of the trachea.

Post-operative complications

There are three main complications:

1 Leak from the oesophageal anastomosis
2 Oesophageal stricture and
3 Recurrent tracheo-oesophageal fistula

The majority of anastomotic leaks are minor and usually treated by withholding oral feeds and commencing antibiotics and total parenteral nutrition. They usually seal spontaneously and re-operation is required only if uncontrolled mediastinitis or empyema develops. Oesophageal stricture may present with dysphagia or choking on feeds. Gastro-oesophageal reflux is frequently a contributing factor. Treatment usually involves radial balloon dilatation of the oesophagus under fluoroscopic control. Sometimes fundoplication is required if there is significant coexisting gastro-oesophageal reflux. A recurrent tracheo-oesophageal fistula rarely closes spontaneously and requires re-exploration and division.

Prognosis

In the absence of associated congenital abnormalities or severe prematurity, survival in oesophageal atresia is now virtually assured. Most patients have a good quality of life into adulthood.

KEY POINTS

- Excessive salivation or drooling in a neonate suggests oesophageal atresia.
- Diagnosis is confirmed by gentle passage of a 10-French catheter through the mouth, which stops at 9–11 cm (distance to stomach is 20–25 cm).
- Babies with oesophageal atresia have a high chance of having other anomalies.
- The prognosis is good in most children following transfer to a tertiary neonatal centre.

Further reading

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De Jong EM, de Haa MAM, Gischler SJ et al. (2010) Pre- and postnatal diagnosis and outcome of fetuses and neonates with esophageal atresia and tracheoesophageal fistula. *Prenat Diagn* **30(3)**: 274–279.

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Sistonen SJ, Pakarinen MP, Rintala RJ (2011) Long-term results of esophageal atresia: Helsinki experience and review of literature. *Ped Surg Int* **27(11)**: 1141–1149.

CHAPTER 7
Bowel Obstruction

CASE 1

A newborn baby develops abdominal distension with bile-stained vomiting and does not pass meconium.

Q 1.1 What are possible diagnoses?
Q 1.2 How would you arrange transport to a neonatal surgical centre?
Q 1.3 What are the principles of resuscitation in this circumstance?

CASE 2

A diagnosis of Hirschsprung disease is made on a newborn baby.

Q 2.1 What is the definitive diagnostic test for Hirschsprung disease?
Q 2.2 What is the operative treatment?
Q 2.3 What is the prognosis?

CASE 3

A 1-day-old baby presents with bile-stained vomiting but no abdominal distension.

Q 3.1 What are the causes of a high neonatal bowel obstruction?
Q 3.2 How do you distinguish between these on investigation?
Q 3.3 How urgent is diagnosis and treatment?

Neonatal bowel obstruction usually presents with the triad of bile-stained vomiting, abdominal distension and failure to pass meconium. A wide range of congenital anomalies of the gut may result in neonatal bowel obstruction. As neonatal physiology and metabolism is significantly different from that in older children, the potential impact upon the neonate must not be underestimated.

Antenatal diagnosis

Dilated fluid-filled loops of gut may be seen on antenatal ultrasound scan and may suggest a bowel obstruction. Sometimes the nature of the obstruction may be characteristic, for example the 'double bubble' of duodenal atresia, but more often the findings do not indicate a specific diagnosis. The normal appearance of the fetal gut is highly variable and an antenatal diagnosis of bowel obstruction should be reserved for those cases with gross gut dilatation. Polyhydramnios may be associated with fetal bowel obstruction, particularly with higher levels of obstruction. Most pregnancies are now checked with an ultrasound scan at 18–20 weeks' gestation, but this may be too early to diagnose many cases of fetal bowel obstruction. Intra-uterine segmental volvulus or intussusception in later pregnancy are the presumed causes of many gut atresias, but ultrasound scans are not routinely performed at this time.

Clinical findings

Bile-stained vomiting in neonates is always significant and must be evaluated carefully as it suggests bowel obstruction.

Abdominal distension is a less specific finding, as gaseous distension may occur without bowel obstruction. Furthermore, some high bowel obstructions, such as malrotation with volvulus or duodenal atresia, often do not have abdominal distension.

The normal-term neonate passes meconium within the first 24 h of life. Neonates with bowel obstruction do not pass meconium, with three notable exceptions: (1) babies with Hirschsprung disease may pass meconium, especially after rectal examination; (2) some

Jones' Clinical Paediatric Surgery, Seventh Edition. Edited by John M. Hutson, Michael O'Brien, Spencer W. Beasley, Warwick J. Teague and Sebastian K. King.

sticky meconium pellets may be passed in babies with meconium ileus and (3) onset of symptoms in malrotation with volvulus may be delayed for some time after birth (after feeding, a stooling pattern will be established).

Imaging

The plain abdominal x-ray is the most important investigation and may show distension of the gut with fluid levels. The number of fluid levels may be related to the level of the obstruction: for example, a double bubble in duodenal atresia; three or four fluid levels in upper jejunal atresia; and many fluid levels in ileal atresia or Hirschsprung disease. An appearance of 'soap bubbles' in the right lower quadrant, in conjunction with a distal bowel obstruction, is suggestive of meconium ileus. Fine calcification indicates antenatal gut perforation with meconium peritonitis. Free gas in the peritoneal cavity is seen when perforation occurs after birth.

Contrast studies are useful in some patients. Incomplete high obstructions are assessed with a upper gastrointestinal contrast study, which may demonstrate a malrotation with volvulus or a duodenal web. A lower gastrointestinal contrast study is a suitable test for low obstructions, such as Hirschsprung disease, ileal atresia or meconium ileus.

Metabolic complications

Neonatal bowel obstruction may lead to rapid and serious metabolic derangement, especially if gut ischaemia occurs. Many of these metabolic problems are related to the neonatal period and must be corrected before undertaking transport. Operative management cannot be contemplated until the neonate has been fully resuscitated. The particular metabolic problems related to bowel obstruction are as follows:

1. Fluid loss from vomiting, lack of fluid intake and sequestration of fluid in the gut and peritoneal cavity leading to diminished circulating fluid volume and poor tissue perfusion. This contributes to hypothermia and acidosis.
2. Tissue glucose stores in neonates are low. If oral intake is impeded and metabolism is stressed by bowel obstruction and poor tissue perfusion, glucose stores will be rapidly exhausted and the neonate will switch to anaerobic metabolism, with consequent hypoglycaemic acidosis. The acidosis has an adverse effect on cardiovascular activity, further exacerbating the problem. Severe hypoglycaemia may cause cerebral damage.
3. The sick neonate is particularly sensitive to hypothermia. Inadequate warming during examination, resuscitation and imaging of the neonate compound the problem. Much of the preparation of a sick neonate for transport is spent in correcting and maintaining body temperature.
4. Respiratory distress is common in many neonates with bowel obstruction as a result of abdominal distension. Inhalation of vomitus may produce pneumonitis and atelectasis.
5. Sepsis from gut organisms (due to transmigration of organisms through an ischaemic or perforated gut wall) causes a rapid deterioration in all metabolic factors. Septicaemia with virulent gut organisms may lead to the rapid demise of the neonate.

General treatment

Emergency transport

The neonatal emergency transport service should come to the sick neonate with a bowel obstruction, as the neonate will not tolerate handling and movement well. Transport is a particularly stressful time and the metabolic problems should be corrected before transfer.

Nasogastric tube

Passing a nasogastric tube to aspirate gut content relieves respiratory distress from abdominal distension and helps to measure the fluid losses. It is mandatory in all cases of bowel obstruction.

Resuscitation

Fluid replacement

Intravenous access is established, while using an overhead heater to prevent heat loss. Rapid resuscitation with 10 ml/kg boluses of Hartmann's solution is given over 15 min for each bolus. The state of hydration of the neonate is assessed in terms of peripheral circulation, urine output and blood parameters. Most neonates will require 10–30 ml/kg of resuscitation fluid prior to operation.

Glucose replacement

Blood glucose levels should be monitored during resuscitation and glucose solution (10% dextrose) should be given as well as Hartmann's solution. Lack of glucose exacerbates acidosis and may cause fitting.

Correction of acidosis

Acid-base measurement is an important part of neonatal resuscitation. Correction of hypothermia, fluid and glucose replacement will help to correct any acidosis.

Sodium bicarbonate given intravenously may sometimes be necessary.

Hypothermia

Hypothermia is a major risk to sick neonates. An overhead heater with monitoring of the neonate's temperature is used during all procedures. X-ray imaging is another time of risk for hypothermia. Much of the expertise involved in neonatal transport services is directed towards preventing hypothermia during transfer to the neonatal surgical centre.

Sepsis

There is a risk of sepsis with neonatal bowel obstruction and intravenous antibiotics are commenced after cultures are taken.

Hirschsprung disease

In 1887, Hirschsprung described two infants who died with gross abdominal distension due to a severely dilated colon containing masses of faeces. Hirschsprung assumed that the disease was caused by the megacolon. However, the disease was later shown to be in the narrow distal bowel where there is a lack of ganglion cells (the intrinsic nerves of the gut) in the submucosal and myenteric plexes. In addition, thickened abnormal (extrinsic) cholinergic nerve fibres are found in the affected segment. The affected gut is in a constant state of spasm and will not relax, causing functional obstruction (and secondary 'megacolon' is not treated).

Hirschsprung disease occurs in 1:5000 births, and there are different sub-types:

1. A larger group (≈80%), in which males are affected five times as often as females, with a relatively short aganglionic segment, usually involving the sigmoid colon and rectum.
2. A smaller group with a long aganglionic segment, equally common in boys and girls, with a higher degree of 'penetrance' and more likely to affect subsequent siblings.
3. Specific genetic abnormalities (e.g. GDNF-ret oncogene; endothelin B receptor system) are being described in Hirschsprung disease and it is likely these genetic markers will become increasingly important in the understanding of the basis of this disease.

The affected segment begins above the anal canal and extends proximally for a variable distance – in most cases as far as the sigmoid colon but sometimes as high as the ascending colon. In a few cases, the affected segment extends into the small bowel, and in rare instances, the entire alimentary canal is devoid of ganglia, excluding hope of survival.

The most common presentation is with complete bowel obstruction in the early neonatal period. In some cases, the infant may present at a few months of age with a history of chronic constipation and failure to thrive. Paradoxically, some of these late-presenting cases have long-segment disease.

The three classic signs are (1) delayed passage of meconium (i.e. beyond 24 h after birth), (2) bile-stained vomitus and (3) abdominal distension.

Clinical features

Abdominal examination will reveal marked gaseous distension. Rectal stimulation with a probe may cause explosive decompression of meconium and faeces through the tight anal sphincters. A nasogastric tube will drain bile-stained fluid.

Investigation

A plain abdominal x-ray usually shows marked gaseous distension of the gut, often with air-fluid levels. A contrast enema will show constriction of the segment of bowel affected by Hirschsprung disease, expanding through a transition zone to a distended megacolon above the functional blockage of the affected segment.

The diagnosis is made by suction rectal biopsy. The biopsy is processed to examine for the absence of ganglion cells by standard microscopy, and histochemical preparations are made for acetylcholinesterase staining of the overgrown extrinsic cholinergic submucosal nerve fibres. The diagnosis of Hirschsprung disease requires sophisticated paediatric pathological services.

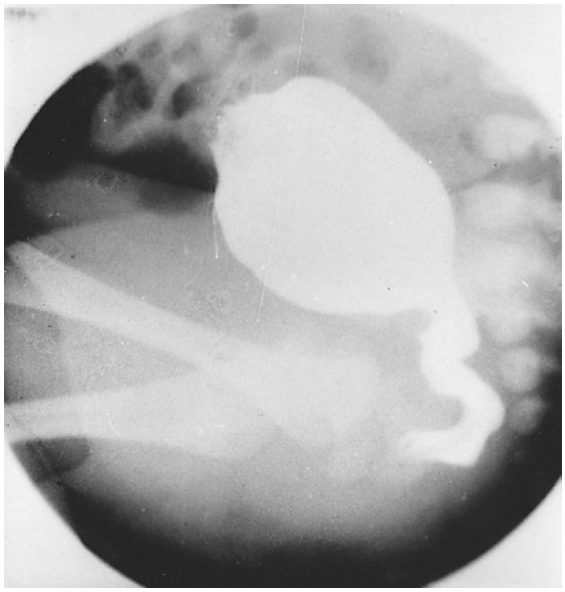

Figure 7.1 Hirschsprung disease transition zone between dilated proximal colon and narrow distal bowel as seen on contrast enema.

Treatment

The bowel is decompressed daily with saline enemas. Although an initial colostomy or ileostomy is sometimes required in more complex cases.

A pull-through operation is performed at a later date when the baby is thriving. The operation delivers the aganglionic rectum and colon out through the anus. The aganglionic segment is excised and normal gut is anastomosed to the anal canal. Laparoscopy for intraoperative biopsy is useful [Fig. 7.1].

Prognosis

The operation for Hirschsprung disease is life-saving, but there may be prolonged morbidity in some cases. Hirschsprung-associated enterocolitis, either before or after the operation, may be life-threatening, with the outpouring of fluid stools causing rapid, severe electrolyte problems along with sepsis. Bowel dysfunction, with diarrhoea and soiling, may be a long-term problem.

Meconium Ileus

Cystic fibrosis causes a change in the physical properties of the meconium that fills the fetal gut. The meconium becomes excessively sticky and tenacious, leading to a mechanical obstruction of the bowel called meconium ileus. In Western countries, cystic fibrosis is the usual underlying cause, and this may be confirmed by genetic tests or by finding high sodium chloride levels in a sweat specimen. Occasionally, meconium ileus occurs as an isolated event and there is no underlying disease.

The sticky meconium impacts as a putty-like substance in the terminal ileum, leading to a small, disused colon distal to the obstruction (known as 'microcolon'). The bowel proximal to the obstructed ileum is distended with sticky meconium.

Clinical findings

The baby presents with abdominal distension, bile-stained vomiting and failure to pass meconium. There may be a family history of cystic fibrosis. The loops of distended gut may be palpable as they are already filled with meconium, rather than the gaseous distension seen in other forms of bowel obstruction. Rectal examination reveals no normal meconium, but pale, mucoid pellets may be passed.

Imaging studies

Plain abdominal x-rays show distended loops of gut filled with a foamy substance. On the erect film (which is done rarely in a neonate) there may be no air-fluid levels as the meconium is too viscous to layer out with gravity. A lower gastrointestinal contrast study will show a microcolon with pellets in the terminal ileum [Fig. 7.2]. Contrast may then pass into the dilated small bowel proximal to the obstructed ileum.

Treatment

Treatment is based on the assumption that the neonate has cystic fibrosis. Antibiotics are commenced at once, and dehydration, which would make secretions even more tenacious, must be prevented.

Sometimes the obstructing meconium may be encouraged to evacuate following a water-soluble contrast enema under fluoroscopy. If this non-invasive method fails, laparotomy is performed and a temporary ileostomy is often performed to allow subsequent bowel washouts to clear the meconium. The prognosis for meconium ileus depends on the underlying cystic fibrosis. Occasionally there is recurrent bowel obstruction due to faecal impaction in the terminal ileum in older children and adolescents. This is known as 'distal intestinal obstruction syndrome'.

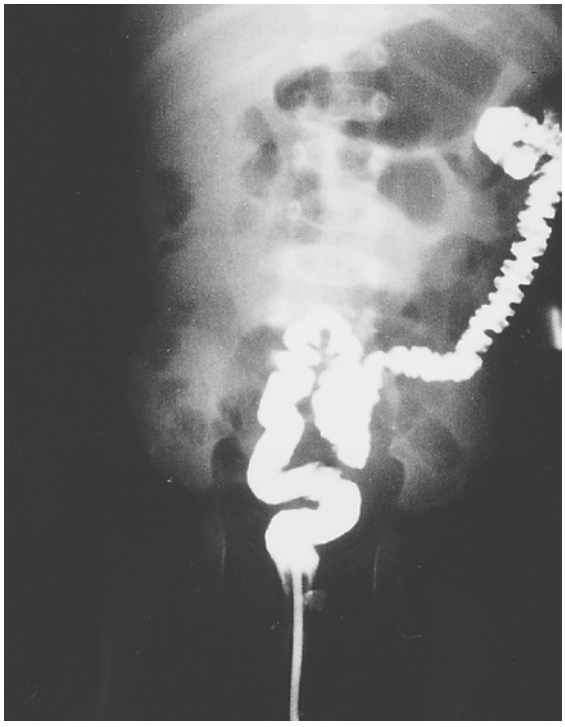

Figure 7.2 'Microcolon'. The unexpanded but otherwise normal bowel distal to any complete intestinal obstruction *in utero*.

Volvulus neonatorum

The normal mesentery of the small bowel has a wide base between the duodeno-jejunal flexure just to the left of the midline in the epigastrium and the ileo-caecal junction in the right iliac fossa. In the case of malrotation of the midgut, the duodeno-jejunal junction and the ileo-caecal junction lie side by side. There is a very narrow base to the small bowel mesentery, allowing the gut to twist around the superior mesenteric vessels (i.e. malrotation with volvulus).

A 360° twist may cause venous and lymphatic engorgement with bile-stained vomiting. An emergency laparotomy at this stage should have a good outcome. A 720° twist will cause arterial ischaemia of the gut from the duodenum to the mid transverse colon, with the potential for a significant increase in morbidity and mortality.

Clinical features

The typical case is a healthy full-term baby who is well for the first few days of life but then develops feeding difficulties with bile-stained vomiting. At this early stage, the abdomen is soft and non-distended. The diagnosis should be suspected at this stage and confirmed with an urgent upper gastrointestinal contrast study. Abdominal distension with tenderness and passage of blood per rectum are late features and indicate major gut ischaemia.

Investigations

An upper gastrointestinal contrast study will demonstrate the abnormal position of the duodeno-jejunal junction and the contrast may spiral through the twisted gut [Fig. 7.3]. Investigation is urgent but must not be allowed to delay the definitive surgical treatment.

Treatment

Laparotomy is urgently required, as ischaemia may lead to gangrene of the midgut (duodenum to the right colon). The volvulus is untwisted, often requiring two or three full rotations to release the caecum. The malrotation of the gut is then corrected by Ladd's operation. The narrow base of the mesentery is broadened by dissection that separates the caecum from the

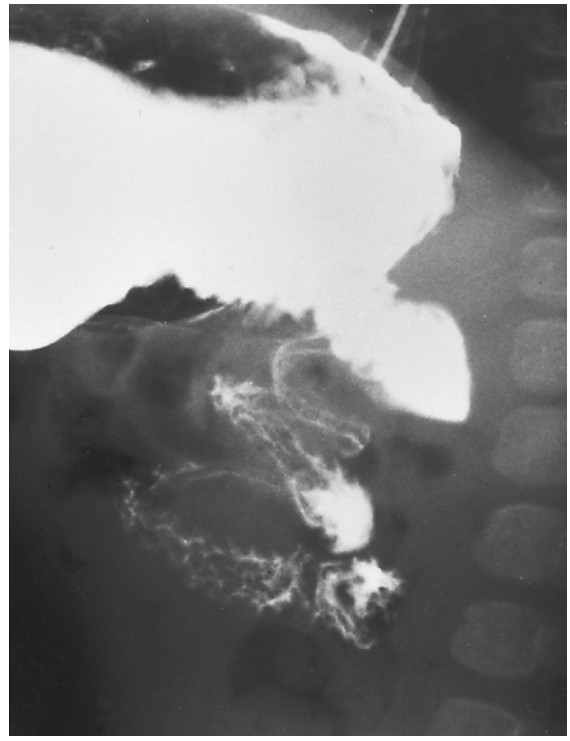

Figure 7.3 Volvulus neonatorum. Upper gastrointestinal contrast study shows a spiral twist of the bowel below the mid-duodenum.

duodenum by dividing abnormal peritoneal adhesions. The small bowel is placed on the right side of the abdomen and the colon is placed on the left side. This leaves the appendix up in the left upper quadrant for which reason appendicectomy may be performed to prevent future confusion. Sometimes the early diagnostic features of volvulus are missed and extensive gut ischaemia occurs. When this happens the surgeon finds nearly all the gut is gangrenous. This situation poses a major problem for future management. The gut is untwisted and a 'second look' laparotomy is performed 24–48 h later to see if any viable gut may be saved.

Duodenal obstruction

Duodenal obstruction is caused by duodenal atresia or by stenosis due to a membrane. Less commonly, an annular pancreas may be wrapped around the duodenum, but usually this is accompanied by severe stenosis or atresia of the duodenum at the site of the envelopment.

Duodenal atresia occurs most commonly in the second part and is associated with Down syndrome in about 30% of cases. An antenatal diagnosis may be suggested by features of polyhydramnios, Down syndrome and/or a double bubble on ultrasonography.

The obstruction develops acutely in the early neonatal period, but signs may be delayed for a day or so while the secretions accumulate in the enlarged stomach and proximal duodenum. In the majority of neonates with duodenal atresia, the obstruction is just distal to the ampulla of Vater (80%), resulting in bile-stained vomiting.

Abdominal x-rays demonstrate a 'double bubble' pattern: two large air bubbles (one in the stomach and the other in the dilated proximal duodenum) each with a fluid level and no gas in the more distal bowel [Fig. 7.4].

A duodenal septum with a hole in the centre may form an incomplete obstruction with episodic vomiting that may be bile-stained. These babies may present in the neonatal period or at a later age if the obstruction is not so severe. The diagnosis is confirmed by a upper gastrointestinal contrast study.

Treatment

In duodenal atresia, with or without an annular pancreas, the obstruction is repaired by duodeno-duodenostomy. The operative treatment for duodenal septum is duodenoplasty, but the bile ducts may pose a special problem as they typically open very close to, or actually into the edge of, the septum.

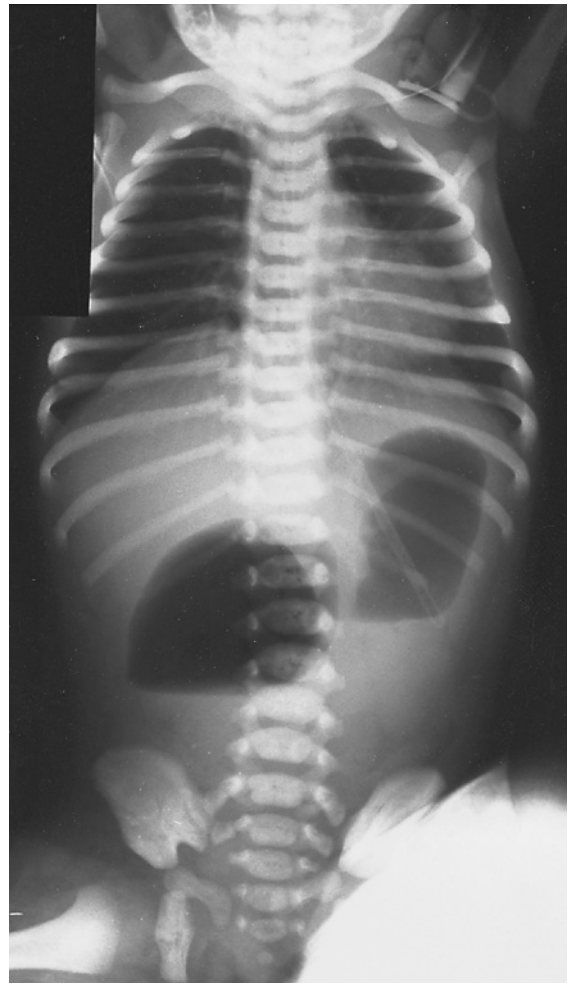

Figure 7.4 Duodenal atresia. Abdominal x-ray shows a 'double bubble', one in the stomach and the other in the dilated proximal duodenum.

Small bowel atresia

Atresia of the bowel beyond the duodenum may occur at any point [Fig. 7.5], most frequently in the distal ileum [Fig. 7.6], and rarely in the colon. There is often only one atresia, although there may be several close together or widely scattered.

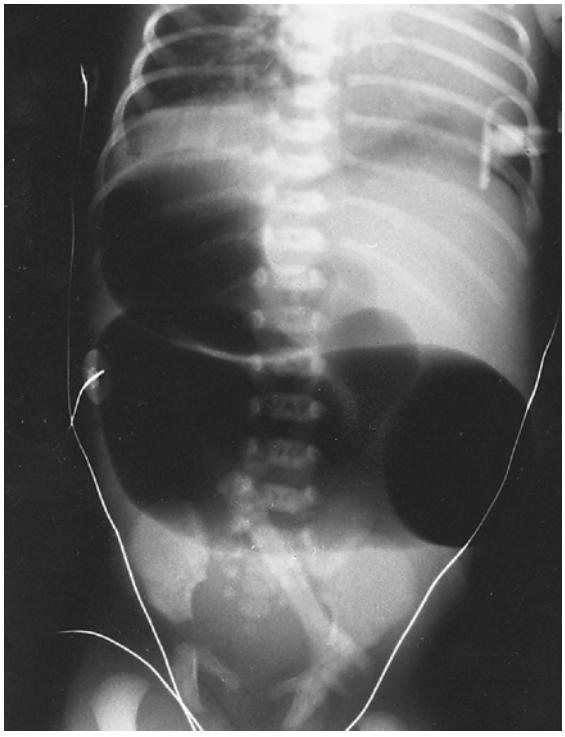

Figure 7.5 Intestinal atresia. Abdominal x-ray shows an obstruction of the jejunum. The neonate also has *situs inversus*.

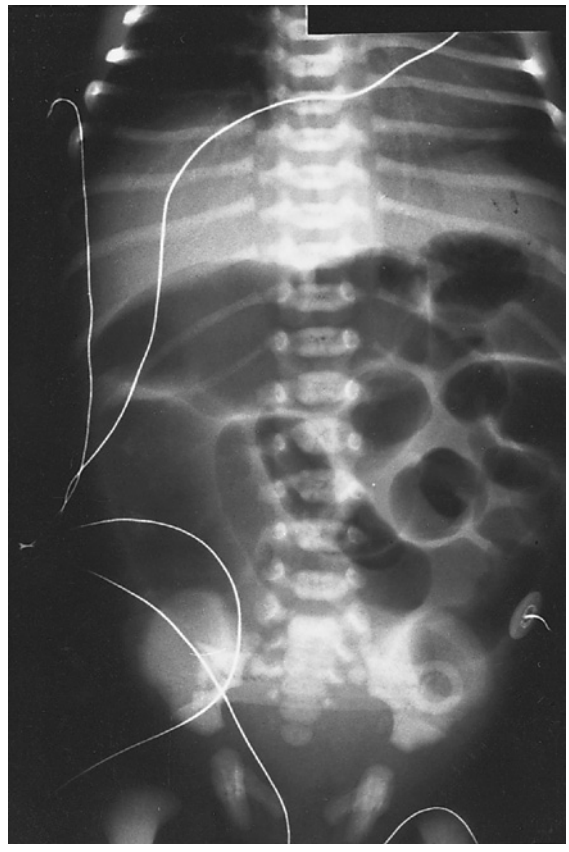

Figure 7.6 Ileal atresia.

The cause may be interruption of the mesenteric arcades by a vascular accident *in utero*, a theory supported by experimental surgery on fetal animals.

The form of the atresia may be a membranous atresia or web with an intact mesentery (type 1), two blind-ending loops with an intact mesentery (type 2), two blind-ending loops with a mesenteric defect (type 3) or bowel affected by multiple atresias (type 4). The adjacent vascular arcades are distorted and their terminal branches may be very small or absent. All the bowel distal to the atresia is collapsed. A lower gastrointestinal contrast study will typically demonstrate a microcolon.

Duplications of the alimentary tract

These are rare developmental anomalies in which a length of bowel is duplicated in such a way that the two segments share the same blood supply and a common wall, while the mucosal linings are separate. They may arise at any point from the mouth to the anus and involve any length from 1 to 2 cm to the entire length of the large bowel. A duplication may or may not communicate with the main alimentary channel.

There are two basic types:

1. Short, closed cystic duplication. The duplicated segment forms a cyst which bulges into the lumen or compresses and angulates the adjoining small bowel, causing obstruction. A small intraluminal cyst may cause obstruction in the neonatal period, but larger ones less intimately connected with the common wall usually cause progressive obstruction later in infancy or in early childhood. Occasionally a large tense cyst is palpable as a very mobile mass in a child without obstructive symptoms.
2. Long, tubular communicating duplication. These cysts are much less common and more likely to be lined by heterotopic (gastric) mucosa, which may cause a peptic ulcer (Chapter 23). If the diagnosis of a duplication containing heterotopic gastric mucosa is

suspected, it may be demonstrable by a ^{99m} Technetium scan. The resection of a long duplication involves the removal of an equivalent length of bowel, but when this is unacceptable because of the inadequate length of bowel remaining, a practical alternative is to remove the lining alone.

Neonatal necrotising enterocolitis

Necrotising enterocolitis is a disease with combined ischaemia and infection of the bowel wall. Although not strictly a 'cause' of intestinal obstruction, it typically presents with abdominal distension and bile-stained vomiting, resembling obstruction. The passage of blood per rectum and a characteristic appearance on abdominal x-ray help to distinguish necrotising enterocolitis from neonatal bowel obstruction. A greater awareness of the entity may have contributed to the increased incidence in recent years, but there has been an absolute increase in the number of cases reported in developed countries.

Predisposing factors

Sick premature neonates may develop necrotising enterocolitis as a further complication of the predisposing factors listed in Table 7.1.

Aetiology

The mechanism has not been elucidated fully. The most widely held theory to explain the intestinal ischaemia is that in a stressed, hypoxic state, blood is preferentially distributed to the heart and brain, at the expense of the splanchnic circulation, skin and muscle.

Local vascular changes have been implicated: for example, a catheter in the umbilical vein, if badly positioned, alters the portal haemodynamics; a catheter in the umbilical artery may have a similar effect on the arterial supply if advanced too far up the aorta and also has the potential to produce emboli.

Certain bacteria appear to be important; *Klebsiella* species resistant to the commonly used antibiotics are found in a significant number of neonates who develop necrotising enterocolitis. Other enteropathogens (e.g. *Escherichia coli*, *Clostridium difficile*, *Streptococcus faecalis* and *Pseudomonas* species) have also been isolated.

The type of enteral feed and when it is commenced may be relevant: for example, the disease appears to occur more frequently in neonates who were fed early with formula. Breast milk may afford some protection against the disease in the 'at risk' neonate.

Table 7.1 Predisposing factors and observed associations in neonatal necrotising enterocolitis

Prematurity
Respiratory distress:
Atelectasis
Hyaline membrane disease
Birth asphyxia
Foetal distress during labour
Prolonged antepartum rupture of membranes
Twins
Caesarean section
Congenital heart disease
Jaundice
Catheterisation of the umbilical vessels
Hyperosmolar feeds
Sepsis

Pathology

Necrotising enterocolitis may be generalised and involve most of the small and large intestine, or may be localised in distribution. The ileum and colon are the two most commonly affected sites. There is histological evidence of impaired perfusion, resulting in tissue anoxia and necrosis. The mucosa is affected most, because of a shunting mechanism, but the process may involve the entire thickness of the bowel wall.

When the mucosa is damaged, production of mucus is impaired and the bacteria normally present in the lumen may invade the intestinal wall (further damaging the bowel) and enter the bloodstream to produce bacteraemia or septicaemia. The damaged mucosa bleeds into the lumen with resultant rectal bleeding, and gas collects in the bowel wall (pneumatosis intestinalis) either due to the activity of gas-forming organisms or by diffusion of intraluminal gas through breaches in the damaged mucosa.

The consequent pathological course is variable:

1. Perforation with generalised peritonitis
2. Perforation with local abscess formation
3. Healing with return of normal function
4. Healing with stricture formation

Clinical picture

A neonate with complications of prematurity is most at risk of necrotising enteterocolitis. The onset of symptoms is between 2 and 14 days after birth. The neonate

becomes ill, lethargic, febrile and not interested in feeds. Abdominal distension and bile-stained vomiting occur, and there may be passage of loose stools containing a variable amount of blood.

When complicated by peritonitis, the anterior abdominal wall becomes oedematous and red, with dilated veins, and palpation causes pain. A mass may be palpable if a localised intraperitoneal abscess has formed or if there is a persistently dilated loop of bowel.

Investigations

The radiological findings are typical. Plain abdominal x-rays demonstrate dilated loops of bowel in which there are intramural bubbles of gas (pneumastosis intestinalis) [Fig. 7.7]. Gas within the portal vein and/or its radicles may be visible. Free gas in the peritoneal cavity, best seen under the diaphragm, is present if the intestine has perforated. Separation of adjacent loops of bowel suggests appreciable amounts of intraperitoneal exudate, an indication of peritonitis, with or without a perforation.

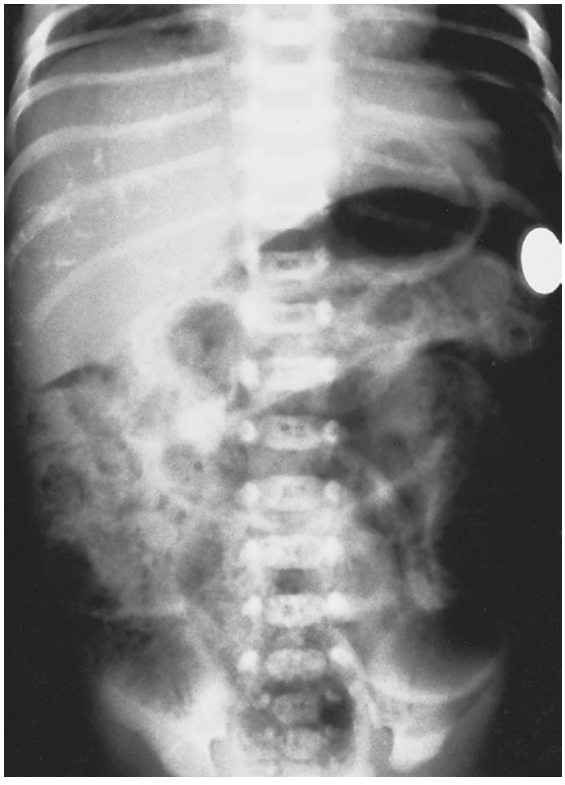

Figure 7.7 Necrotising enterocolitis. Intramural gas.

Bacteriological specimens (e.g. blood culture, nose, throat, umbilicus and rectal swabs) should be taken before antibiotics are commenced or altered.

Biochemistry, haematology, electrolytes, acid–base and bilirubin are monitored. The haemoglobin level may fall progressively as a result of sepsis and haemorrhage, and serial measurements are required. The platelet and white cell counts are depressed in severe disease, and the neonates are frequently acidotic.

Management

Initially this consists of commencing intravenous antibiotics, placing a nasogastric tube to aid intestinal decompression, stopping enteral feeds and ensuring parenteral nutrition.

Intensive measures listed in the section on the pre- and post-operative care of the neonate and adequate respiratory management also may be required.

Frequent clinical and radiological reassessments are essential, for they may show the need for operative intervention.

The indications for operation are as follows:

1 Clinical deterioration despite maximal intensive resuscitation
2 Evidence of bowel necrosis and perforation

Features which are useful in determining the need for operation include free gas on abdominal x-ray, progressive signs of peritonitis (distended, red and tender abdomen), persistent acidosis despite attempted correction and a sudden and profound fall in the platelet count.

Operation is confined to resection of perforated or necrotic bowel, evacuation of intraperitoneal soiling and placement of diverting stomas. The mortality rate until recently was high, but with earlier diagnosis and more effective treatment, the outlook has improved.

KEY POINTS

- Bile-stained vomiting in the neonate is always serious.
- Abdominal distension, bile-stained vomitus and failure to pass meconium indicate bowel obstruction.
- Bile-stained vomiting in a normal baby suggests possible malrotation with volvulus and is an emergency.
- Neonates with bowel obstruction need special transport to the surgical centre.

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CHAPTER 8
Abdominal Wall Defects

CASE 1

Ultrasonography at 18 weeks' gestation showed that the fetus had bowel loops within an expanded umbilical cord. Careful scanning of the remainder of the fetus showed no other abnormality, although the mother was warned of the possibility. Amniocentesis and karyotyping failed to show trisomy 13 or 18 and it was elected to continue the pregnancy. A paediatric surgeon was consulted for advice about management at birth.

Q 1.1 What is the abnormality?
Q 1.2 Why did it occur?
Q 1.3 What 'first aid' treatment is needed before transfer to the surgical centre?
Q 1.4 What is the management and prognosis?

CASE 2

A teenage girl did not want her parents to know she was pregnant. She presented in labour with no prior antenatal visits. A vigorous neonate was delivered, but the midwife noted that most of the small bowel was protruding through a small hole in the abdominal wall, just to the right of the umbilicus.

Q 2.1 Will the baby live?
Q 2.2 Why is the baby's life at risk?
Q 2.3 Is the baby likely to have multiple congenital anomalies?

CASE 3

Frank was born without trouble at term. Antenatally the ultrasonographer was concerned about no urine being visible in the bladder. At birth the attachment of the umbilical cord was low and lay adjacent to an unusual defect with wet, pouting mucosa. The penis was found to be bifid and one testis was undescended.

Q 3.1 What is the embryological defect?
Q 3.2 How may this abnormality be treated?
Q 3.3 Why is the penis split into two halves and what is the likely outcome for sexual function and urinary control?

Exomphalos and gastroschisis

These two developmental abnormalities in the region of the umbilicus are either diagnosed on antenatal ultrasonography or present at birth as neonatal emergencies, and they require urgent treatment.

Currently, the prevalence of exomphalos (omphalocele) and gastroschisis is relatively similar, although the incidence of gastroschisis is increasing in most regions for reasons that are unclear. The relatively high risk of coexisting and signficant abnormalities in a fetus with exomphalos (35%) may be a reason for termination, particularly if amniocentesis identifies a major chromosomal abnormality [Fig. 8.1].

First aid at birth

A baby with an anterior abdominal wall defect is at great risk of heat and water loss from evaporation because of the moist exposed viscera. For this reason, the baby should be placed in a humidicrib, with the entire torso wrapped, including the exposed viscera, in fresh plastic kitchen wrap or aluminium foil. Care must be taken to ensure that the exposed bowel is not twisted at the opening in the abdominal wall [Fig. 8.1]. Do not use hot wet packs as these cool too quickly with evaporative heat loss and chill the neonate. The main objective is to prevent excessive fluid and heat loss during transfer to the receiving neonatal surgical unit.

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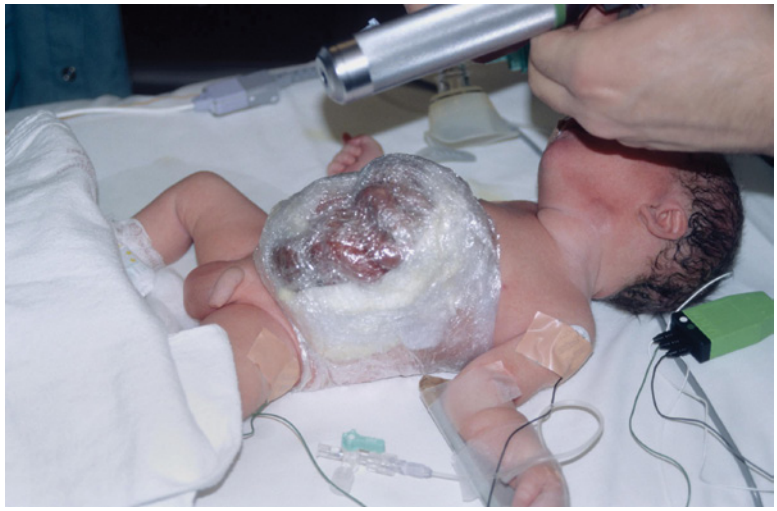

Figure 8.1 First-aid management of gastroschisis. The torso is wrapped in plastic kitchen wrap to reduce heat loss from evaporation. A nasogastric tube keeps the bowel decompressed which facilitates operative reduction of the eviscerated bowel. An intravenous line has been inserted.

Insert a nasogastric tube to decompress the stomach and herniating bowel and do not feed the baby: minimising the gut volume facilitates operative reduction of the herniated bowel.

Commence an intravenous fluid infusion of 10% dextrose to prevent hypogylcaemia during transport, particularly if the baby might have Beckwith–Wiedemann syndrome (organomegaly, exomphalos and hypoglycaemia secondary to dysregulation of insulin-like growth factor 2 expression with hyperinsulinaemia).

Transport

The neonate with an abdominal wall defect should be referred to a fully equipped paediatric surgical centre without delay. Transport should be arranged via a specialised neonatal transport service, if available. If the diagnosis has been made on antenatal ultrasonography, delivery should be undertaken in a tertiary obstetric centre with paediatric surgeons standing by [Box 8.1].

Box 8.1 Immediate 'first aid' for abdominal wall defect

- Wrap the exposed viscera in fresh kitchen wrap (ensuring prolapsed bowel is not twisted)
- Careful examination for other anomalies
- Place infant in Humidicrib
- Nil orally
- IV line with 10% dextrose
- IV Nasogastric tube with aspiration to keep bowel deflated
- If born outside a tertiary paediatric surgical centre:
  - Call Neonatal Emergency Transport Service (NETS)
  - Call Surgical Registrar at tertiary hospital
- Explain management plan to parents
- Get consent for surgery

Exomphalos

This congenital hernia into the base of the umbilical cord is caused by incomplete folding of the embryonic disc and failure of the umbilical ring to form normally. The hernia is covered by fused amniotic membrane and peritoneum.

Very occasionally, the membrane ruptures before birth and the eviscerated bowel becomes matted and indurated with dense adhesions, so that the bowel appears to be shorter than normal. The inflammation is believed to be caused by chemical irritation from meconium and urine in the amniotic fluid. On the uncommon occasion rupture occurs during delivery, the exposed bowel may appear normal.

The size of the defect in the abdominal wall and the volume of the sac are variable: exomphalos *major* has defect greater than 5 cm in diameter and contains gut, liver and/or spleen, whereas exomphalos *minor* has

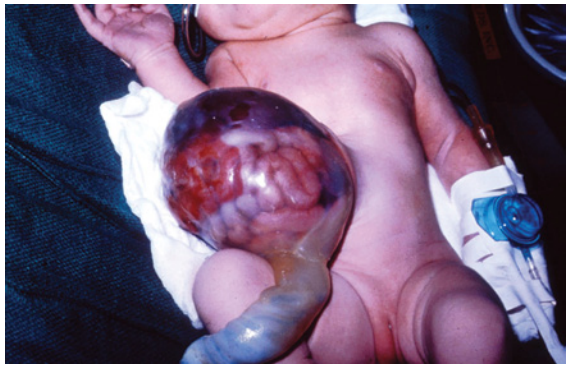

Figure 8.2 Exomphalos.

defect less than 5 cm in diameter and contains only gut within the sac. An intact sac is shiny and translucent but lacks a blood supply and begins to dry out and deteriorate after birth. Within 12 h it becomes opaque and yellowish; later, it becomes black, inelastic and desiccated.

The diagnosis at birth is obvious [Fig. 8.2]; the only difficulty may be in distinguishing a ruptured exomphalos from a gastroschisis. In the latter, there are no sac remnants and the defect is small and separate from the umbilical cord (see below).

Coexisting abnormalities are common in exomphalos, particularly cardiac and renal malformations. Malrotation/non-rotation occurs in 12–20% of cases, but seldom causes volvulus, probably because of adhesions between loops and 'secondary' fixation to the abdominal wall.

Beckwith–Wiedemann syndrome must be recognised early, because of the severe hypoglycaemia which requires immediate correction. These babies produce excess insulin-like growth factor during gestation, which leads to organomegaly, exomphalos minor and excess bodyweight. A large baby (e.g. 4 kg) with exomphalos minor and macroglossia is highly suggestive of the diagnosis. It is suggested that the exomphalos is secondary to the enlarged viscera, which cannot be accommodated inside the abdomen. Postnatal hypoglycaemia is transient but dangerous with risk of brain damage if an immediate infusion of dextrose-containing fluid is not provided.

Investigations

Chest x-ray and echocardiogram are required to identify any cardiac lesion and intercurrent pulmonary conditions, such as atelectasis or meconium inhalation. The kidneys may be examined by ultrasonography. Careful physical examination may suggest other serious anomalies, for example chromosomal anomalies, confirmation of which may modify or even preclude treatment.

Treatment

First aid and the method of transport to the tertiary centre are crucial for optimal outcomes and are discussed at the end of this chapter. The aim of the treatment is to reduce the contents of the exomphalos and repair the defect of the abdominal wall. The method of treatment depends on the following:

1. The general condition of the infant (size, birth weight, maturity, suitability for anaesthesia)
2. Presence of other anomalies
3. Whether the sac is intact or ruptured
4. The size of the umbilical ring and
5. Whether part of the liver has herniated into the sac

Immediate reduction of the bowel and closure of the defect are the best courses when the defect is less than 5 cm in diameter; the neonate is fit for an operation and closure may be obtained. Many units may even attempt non-operative reduction in the ward if the baby is well, with closure of the defect using the umbilical cord.

Excision of the sac (or remnants, if ruptured) and construction of a cylindrical tube (a 'silo') may be used for larger defects. A sheet of silastic or teflon is sewn to the edge of the defect. Alternatively, a post-operative dressing may be attached to the skin around and over the defect, and used to serially reduce the volume of the exomphalos over 7–10 days by imbrication, so that the viscera are returned progressively to the abdomen whilst retaining the integrity of the peritoneum. The prosthesis or dressing is then removed and the defect definitively closed. Use of a silo is indicated when the sac has ruptured with massive evisceration, but it is not without problems, such as infection around the sutures that anchor the prosthesis to the edge of the defect.

Very rarely, non-operative management may be required when anaesthesia is contraindicated because of the poor condition of the neonate (extreme prematurity, cardiac anomaly, meconium inhalation) or when the defect is extremely large (>8 cm in diameter) and contains herniated liver. The sac is painted with an astringent solution or covered with a silver-based dressing to encourage formation of a tough, dry eschar which separates when new skin has covered the area beneath it. This

may take 8–12 weeks or longer. Any substance applied to the exomphalos sac may be absorbed systemically and potentially toxic substances should be used sparingly. Subsequent wound contraction reduces the hernia progressively over 4–8 months and makes the definitive repair easier. Once the eschar has formed, and normal feeding and stools are established, the infant can be managed safely at home, avoiding prolonged, costly and risky (because of cross-infection) hospitalisation. The appearance of the hernia and eschar may be intimidating to the parents, who need support and encouragement to achieve satisfactory bonding.

Gastroschisis

Recent antenatal ultrasound observations suggest that gastroschisis may result from rupture of a physiological hernia in the cord between 6 and 10 weeks' gestation. The fetus usually is normal genetically but has had an 'accident' affecting the umbilical cord. The defect in the abdominal wall is typically small (1–3 cm in diameter) and is nearly always to the right of a normally closed umbilicus.

Evisceration may involve most of the small and large bowel, which become densely matted and adherent with amniotic (chemical) peritonitis and fibrin from vomiting, defaecation and micturition *in utero* [Fig. 8.3], particularly during the last trimester.

It differs from a ruptured exomphalos in that there is:

1 A greater risk of hypothermia
2 A smaller abdominal wall defect and no covering sac
3 Lower incidence of serious coexisting malformations
4 A greater incidence (10%) of a small bowel atresias, but these may be 'occult', that is hidden by the matted fibrin surface (termed 'peel') of the exposed bowel

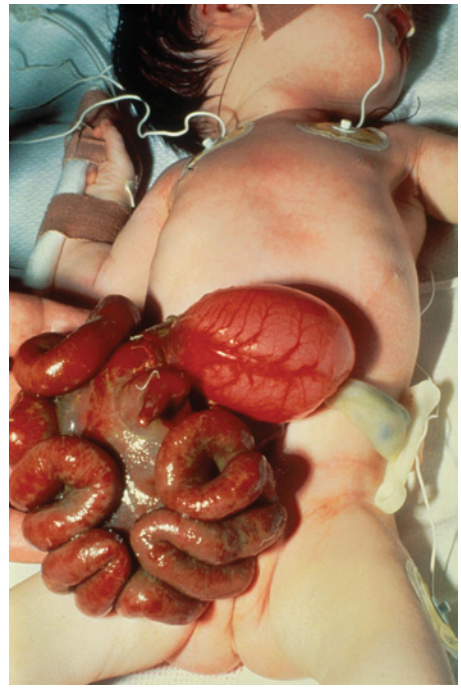

Figure 8.3 Gastroschisis.

Treatment

The goals of management in gastroschisis are as follows:

1 Reduction of the eviscerated organs
2 Closure of the defect
3 Nutritional support
4 Prevention or early treatment of complications

First aid measures are established, and the bowel is carefully inspected for viability and evidence of atresia. If the neonate and bowel are in good condition and the abdominal cavity is of sufficient volume, primary reduction is the preferred method of treatment. This may be done in the operating theatre or at the cot site, with or without general anaesthesia. The defect is then closed, either by sutured repair, or 'plastic' closure in which the umblical cord is used to facilitate closure of the defect. Where primary closure is not possible (or safe), a prosthetic 'silo' can be applied as described earlier. Staged reduction of the viscera is then achieved over 3–10 days, after which delayed closure is achieved as with primary closure.

A dangerous complication of reduction is abdominal compartment syndrome, usually due to overzealous attempts to return the viscera into an abdominal cavity that is too small. Alongside judicious reduction, several active manoeuvres may help reduce the volume of the bowel that has to be reduced, and with this the risk of compartment syndrome. These include the following:

1 Nasogastric suction to minimise bowel contents and gas
2 Anorectal washouts and stimulation to evoke decompression of the colon
3 Enlargement of the defect and intraperitoneal 'milking' of the bowel to evacuate as much meconium as possible through the anus
4 Bladder decompression with urethral catheter

Vigorous stretching of the abdominal cavity walls to increase capacity is deleterious and no longer recommended.

Neonates with gastroschisis typically demonstrate prolonged ileus. Parental nutrition is required to support nutrition whilst gut function and enteral feeding are established, often for many weeks.

High standards of neonatal transport and neonatal surgery lead to a good prognosis for babies born with gastroschisis.

Bladder exstrophy (Ectopia vesicae)

Failure of fusion of the lower abdominal wall during embryonic development leaves the bladder exposed as a flat plaque on the lower abdomen [Fig. 8.4]. There is no covering muscle or skin. The pubic rami do not fuse in the midline and remain widely separated. The ureters protrude from the exposed bladder and dribble urine.

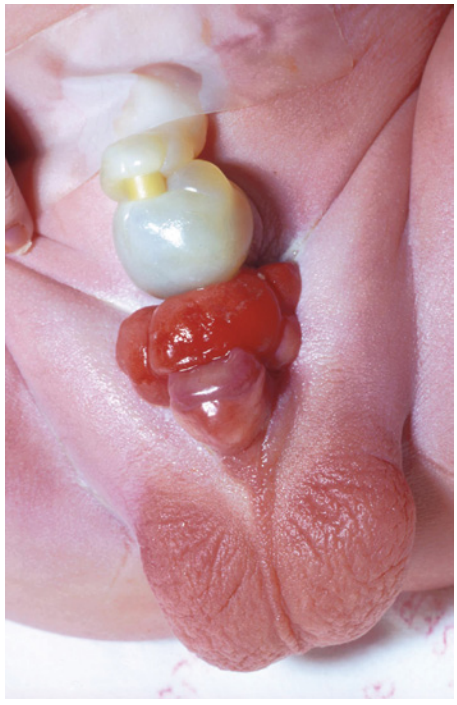

Figure 8.4 Bladder exstrophy.

The urethra is exposed as a flat strip and the bladder outlet sphincters are not functional. Bladder exstrophy, also known as 'ectopia vesicae', is rare, with an incidence of 1:40,000 births. Reconstructive surgery to close the bladder and abdominal wall with restoration of bladder sphincters is one of the greatest challenges of paediatric urology.

An even rarer variant of bladder exstrophy is known as cloacal exstrophy. In this condition, the shortened proximal colon is fused on to the bladder exstrophy and there may be an imperforate anus.

KEY POINTS

- Defects in the anterior abdominal wall are a neonatal emergency because of the risk of heat and water loss, and neonates need immediate first aid.
- Exomphalos is associated with a high risk (35%) of serious anomalies.
- Big babies with exomphalos are at risk of hypoglycaemia and need immediate IV 10% Dextrose.

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CHAPTER 9
Spina Bifida

CASE 1

Jeannie became pregnant at 18, and for various reasons avoided antenatal care and presented in labour. The baby had a red, cystic mass over the lumbar spine and no spontaneous leg movement.

Q 1.1 What physical signs are important to note at birth?
Q 1.2 Will the baby have hydrocephalus?
Q 1.3 What is the prognosis for (a) intellect and (b) walking?

CASE 2

A primigravida woman had both an antenatal alpha-fetoprotein measurement and mid-gestation ultrasonography that did not reveal any fetal anomaly. At birth, the neonate had a small, skin-covered cystic mass over the sacrum. The baby's leg movements were good, but the bladder was palpable.

Q 2.1 What are the urinary tract problems in spina bifida?
Q 2.2 How are urinary and faecal incontinence managed?

Spina bifida is one of the most crippling congenital anomalies. The primary abnormality is incomplete fusion of the neural tube and overlying ectoderm, leading to a defect between the vertebral arches. There is protrusion and dysplasia of the spinal cord and its membranes. The resulting neuronal deficits may cause paraplegia, urinary and faecal incontinence and multiple orthopaedic deformities. Hydrocephalus is a frequent associated anomaly.

The severity of this anomaly has led to widespread antenatal screening with maternal alpha-fetoprotein levels and ultrasonography. In many centres, termination of pregnancy is offered if screening reveals a myelomeningocele. A diet containing folate before conception and during early pregnancy will significantly lower the risk of spina bifida. As a result of these two factors, in western countries the incidence of live-born neonates with spina bifida has decreased dramatically in recent years.

Embryology

Spina bifida and anencephaly are neural tube defects. The fusion of the neural folds should be completed by the fourth week of embryonic development. The mesoderm around the neural tube forms the meninges, vertebral column and muscles. The less severe anomalies involve failure of vertebral arch fusion and protrusion of the meninges to form a meningocele. More severe anomalies involve the neuro-ectoderm, with protrusion of the neural tube itself to form a myelomeningocele. Failure of fusion of the brain causes an encephalocele (see Chapter 12) or anencephaly.

Aetiology and antenatal diagnosis

Spina bifida has been linked to inadequate folate metabolism in the maternal diet. In 6–8% of cases, there is a previous history of hydrocephalus, anencephaly or spina bifida: in these, the risk of spina bifida in subsequent pregnancies is as high as 1:20. With two affected children, the risk is 1:8. Antenatal diagnosis in spina bifida is well established. Second trimester estimation maternal serum alpha-fetoprotein is abnormally elevated due to leakage of fetal cerebrospinal fluid (CSF) into the amniotic fluid. Subsequent antenatal ultrasonography may confirm (or newly detect) the sac and vertebral defect.

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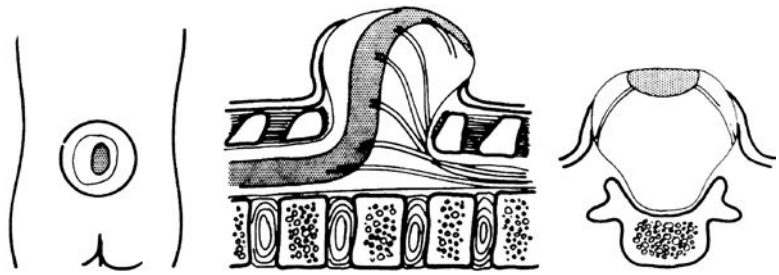

Figure 9.1 Myelomeningocele. Tissue of the spinal cord forms part of the wall of the sac, as well as its contents.

Myelomeningocele

This consists of a bifid spine with protrusion and dysplasia of the meninges and spinal cord. The dysplastic spinal cord is splayed over a meningeal sac filled with CSF and is associated with severe nerve deficits below the level of the lesion. There is a slight predominance of females. The incidence varies widely from one region to another, related to differences in genetics and diet.

Clinical features

The sac is most commonly in the lumbosacral region [Figs. 9.1 and 9.2]. The size of the sac is variable; there is an area of well-developed skin at the periphery but this is thin at the apex, which is covered by the delicate glistening arachnoid membrane with neuronal tissue visible on the surface.

The central area becomes ulcerated and infected, and if left untreated there is a risk of meningitis. Epithelialisation occurs slowly, leaving a puckered scar covered with poor quality skin that is liable to ulceration. The coverings may rupture before, during or after birth, with leakage of CSF. Rachischisis is the most severe form of spina bifida. The neural tube lies open, no sac is present and the spinal cord is a flattened, red, velvet-like ribbon down the centre of back [Fig. 9.3].

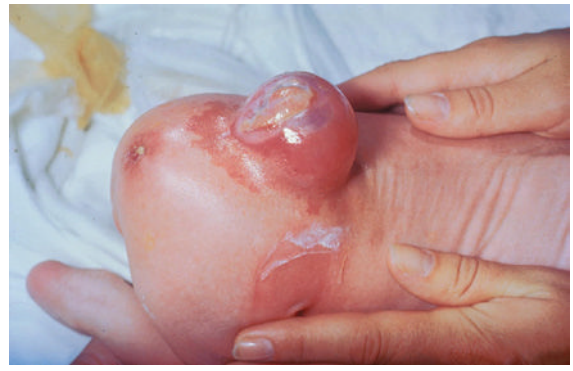

Figure 9.2 Lumbar myelomeningocele: a moderately large sac with well-developed skin at its periphery. The glistening arachnoid membrane and neuronal tissue are exposed on the apex of the sac.

Motor loss

There is a flaccid paralysis of the lower motor neurone type, the extent depending on the level of the neurological lesion. In some patients, upper motor neurone spastic paralysis is also present and is due to either isolated

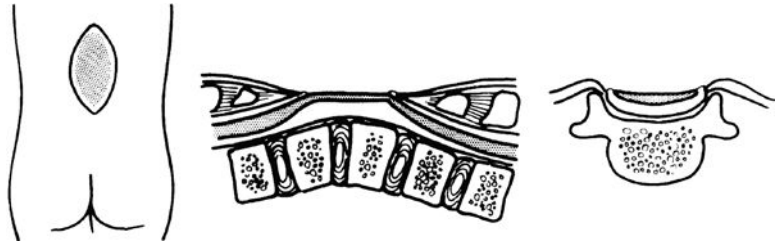

Figure 9.3 Rachischisis: there is no sac and the central canal of the spinal cord lies wide open on the broad deficiency in the posterior vertebral laminae.

normal sections of the cord below the lesion or cerebral or spinal-cord damage from hydrocephalus or meningitis. Children may be grouped according to the level of the lesion. The motor loss may be anticipated by observing the neonate's voluntary (not reflex) movements, and determination of the level is helpful in assessing the probable extent of eventual disability [Table 9.1].

Sensory loss

Sensory loss corresponds closely to the level of motor loss, although the lower level of normal sensation is usually about one segment higher than the lower level of normal motor power. Loss of sensation is most important in the feet, the buttocks and the perineum, because of the risk of pressure sores in these areas.

Meningocele

This is a simple meningeal sac lined by arachnoid membrane and dura, containing CSF and only occasionally neuronal tissue [Fig. 9.4]. It is relatively uncommon (6% of cases of spina bifida cystica) and it may be associated with overlying cutaneous lesions including abnormal pigmentation and hair growth.

The sac may be of any size from a small bulge to an enormous protrusion. It may be tense, but more often is soft and fluctuant; some become tense when the baby cries or when pressure is applied to the fontanelle. The skin over the sac is generally intact, although it may ulcerate occasionally.

Hydrocephalus has been reported but is rare in children with a simple meningocele. The mental state is normal and there are no neurological abnormalities. Deaths are rare and usually are due to meningitis developing before or after operative repair.

Operative management consists of repair of the sac, and when the skin is sound there is no urgency; the repair may be done at any convenient time during infancy. If the sac is ulcerated, it should be repaired immediately after birth, but if this opportunity is missed, epithelium should be allowed to cover the sac, which is then excised at a convenient time later in infancy.

Table 9.1 Assessment of level of paralysis

Level of lesion	Incidence (%)	Motor function
Cervical/upper thoracic	1	Paralysis of legs and trunk
Lower thoracic	27	Complete paraplegia, including psoas
Upper lumbar	23	Hip flexion and adduction present
Lower lumber/ upper sacral	45	Hip movements, knee extension, foot dorsiflexion
Lower sacral	4	All normal movement

Variants of spina bifida

The vertebrae may be bifid without a meningocele or myelomeningocele sac. This may be a normal variant of spinal arch fusion or represent 'spina bifida occulta', which is more serious. Dysplasia of the skin over the spine is a sign of a potentially serious spina bifida: a hairy patch, pigmented naevus, vascular anomaly, lipoma or a sinus may be present. A dermal sinus over the spine may link with an intraspinal dermoid cyst. If this becomes infected, an intraspinal abscess may form with destruction of adjacent spinal cord.

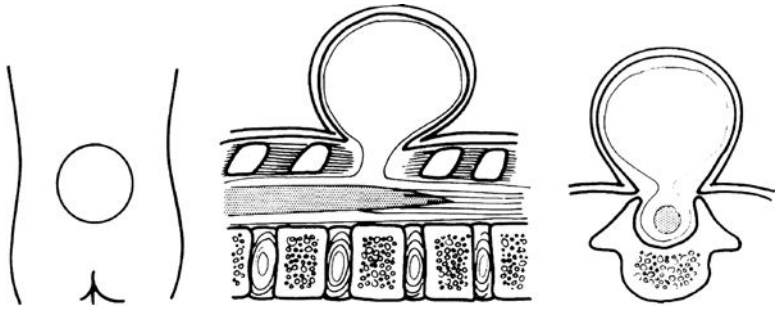

Figure 9.4 Meningocele: the sac contains CSF only and the spinal cord is normal.

A spinal lipoma may be indicated by a bulge over the lumbosacral region. The lower lumbar and sacral nerves run through the fibrolipomatous tissue and progressive nerve damage may occur through spinal cord tethering. These lesions are best demonstrated on magnetic resonance imaging.

Associated anomalies and sequelae of spina bifida

Hydrocephalus

Hydrocephalus is the most important associated anomaly and is present to some degree in almost all cases of myelomeningocele in early infancy. It needs investigation and treatment in about 70% of patients. Hydrocephalus is relatively more common when the myelomeningocele is in the lower thoracic and upper lumbar areas, but it may occur with a myelomeningocele at any level. It is an important factor which influences the child's survival and subsequent mental state. The three most common causes of hydrocephalus are the Arnold–Chiari malformation, stricture of the aqueduct of Sylvius and failure of the subarachnoid space to open up at the level of the tentorium.

Orthopaedic deformities

Orthopaedic deformities are common and greatly complicate management of the paraplegia. They include kyphosis, lordosis, scoliosis, paralytic dislocation of the hips, flexion contractures of the hips and knees, and deformities of the feet. They are caused by inequality of muscular action and bony immobility leading to inadequate growth stimulus to the skeleton.

Urinary tract abnormalities

Ninety-five per cent of children with myelomeningocele have a neuropathic bladder. The kidneys are usually normal at birth, but there is a high incidence of progressive renal damage during the first few years of life if the problems of a high pressure, non-compliant bladder, vesicoureteric reflux and chronic pyelonephritis are not managed effectively.

Other anomalies

Many of the other anomalies that are associated with spina bifida are potentially lethal, for example severe congenital heart disease and visceral malformations.

Complications

Meningitis

Meningitis accounts for approximately one-third of all deaths from spina bifida cystica; it arises either from infection of the ulcerated sac (especially if ruptured) or after operative repair.

Developmental delay

Developmental delay is usually related to hydrocephalus, although separate cerebral deficiencies may also occur. Less than 10% of children without clinical hydrocephalus have developmental delay, and with ventriculo-peritoneal shunting 66% will have normal intelligence. Overall, children with myelomeningocele who survive to school-age have normal intelligence in approximately 75% of cases; 20% have cerebral damage with disability and 2% have severe cerebral damage.

Pressure sores

These may develop on the feet, sacrum and perineum; in the latter two sites, the problem is accentuated by urinary and faecal soiling. Prevention and treatment of pressure sores is of great importance. Full-thickness skin grafts may be helpful to prevent recurrent breakdown for deep, extensive sores in the buttock and sacral area, but it is better to prevent ulcers in the first place.

Special senses

Paralytic squint (i.e. sixth nerve palsy) is a common complication of hydrocephalus, and optic atrophy with blindness occasionally occurs from raised intracranial pressure, usually in older children. Deafness may also occur, apparently unrelated to the hydrocephalus.

Urinary infection

Stasis in the neurogenic bladder predisposes to recurrent urinary tract infections and may lead to chronic pyelonephritis which, if untreated, causes progressive renal scarring. Regular drainage of the bladder by intermittent catheterisation and/or long-term low-dose antibiotics may prevent or limit these problems.

Clean intermittent catheterisation may be started shortly after birth. The parents pass a 'clean' catheter four to five times a day. In 80% of cases, the child wets a little between catheterisation. In 10%, there is marked wetting and the bladder storage capacity may

be increased by using alpha-adrenergic or anticholinergic drugs.

In many children with spina bifida, the neurogenic bladder pattern is that of retention leading to overflow incontinence; the rationale of clean intermittent catheterisation is to empty the bladder frequently, before overflow incontinence occurs. Paradoxically, catheterisation usually lowers the incidence of urinary tract infections by removing stagnant residual urine. Some children on this regimen will also need daily low-dose antibiotics, such as nitrofurantoin or Co-trimoxazole.

As children grow older, wetting becomes less socially acceptable. The main two causes of failure of clean intermittent catheterisation are:

1. poor urine storage due to deficiency in the function of the urethral sphincter and
2. small bladder capacity with a thick-walled, low-compliance bladder

When these children reach school age, the bladder storage may be improved by an operation to tighten the bladder sphincters. The bladder capacity may be increased by endoscopic injection of botulinum toxin into the detrusor muscle or by the more invasive approach of a bladder augmentation using bowel. However, bladder augmentation has many potential complications, including bladder stone formation, bladder rupture and a small incidence of bladder cancer. Consequently, a patient needs careful counselling prior to bladder augmentation and lifelong urological follow-up.

Faecal incontinence

'Accidents' are common during early childhood in patients with spina bifida; normal toilet training should be attempted, and the child is taught to evacuate the stool by contraction of the abdominal muscles. Most children are constipated, and the aim is to produce a firm stool that is not soft enough to leak out and not so hard that it will become impacted. This is typically achieved by a combination of diet, laxatives and bulking agents. After adolescence, most patients are clean and regular and evacuate a firm or hard stool. However, some patients will require a daily suppository or enema. Gross impaction with faecal overflow may be cleared by bowel washouts. The most effective means of washout remains unclear. Some children will have an appendicostomy formed, which allows regular antegrade colonic enemas. This procedure has the potential to significantly improve the quality of life of these children. Biofeedback techniques are useful for some children.

Psychological and social management

Many children show psychological disturbances as a result of their disabilities, especially in adolescence, although only a few are seriously affected. The disturbances are not directly proportional to the degree of disability or to the level of intellectual functioning, nor are they specific to children with spina bifida. Generally, counselling is sufficient, but some children require additional psychiatric treatment. Also, parents may need help to enable them to accept their child's disability. This aspect presents relatively few problems if they have been given a full appraisal of the condition following antenatal diagnosis or soon after birth. However, it may be difficult with parents with previous psychiatric or marital problems.

Assessment in the newborn

Careful assessment by a specialist team (neonatologist, neurologist, neurosurgeon, urologist, orthopaedic surgeon) is essential at birth. A treatment plan should be formulated as early as possible and the following points should be noted:

1. Any other congenital abnormalities, especially those likely to be fatal
2. The type of spina bifida, that is myelomeningocele or meningocele
3. The level, size and state of the sac
4. The presence of hydrocephalus and the degree of cerebral dysplasia
5. The presence of meningitis
6. The severity of orthopaedic disability, recorded by charting muscle activity, especially that of the psoas major, quadriceps and the dorsiflexors and plantar flexors of the ankle
7. The presence of a neurogenic bladder and bowel, as shown by dribbling urine, a patulous anus, perineal anaesthesia and an expressible bladder
8. The presence of upper urinary tract abnormalities or urinary tract infection
9. The family dynamics. Special problems exist with families who are unable to cope with the considerable strains imposed

Most of the aforementioned points may be evaluated in the neonatal period, and the predicted disabilities should be fully discussed with the parents as soon as possible.

Treatment

The aim is to produce an ambulant patient, dry and free of the smell of urine and faeces, who is able to function at optimal intellectual and physical level, is educable and capable of employment and independent living. The severity of the disease in some children precludes the attainment of all these objectives. Adverse factors that carry a poor prognosis include the following:

1 Thoracic lesions associated with complete paraplegia (especially if associated with spinal kyphosis)
2 Hydrocephalus clinically present at birth or rapidly developing after birth
3 Meningitis or ventriculitis
4 Other severe congenital abnormalities
5 Renal impairment

Initial examination and regular supervision in a multidisciplinary specialist clinic are required for accurate assessment and optimal results.

The sac

Early operation prevents meningitis and shortens hospital stay. It encourages acceptance of the child by the parents but does not affect the development of hydrocephalus. In some children, the prognosis is so poor that repair of the sac may be best deferred until the parents have had sufficient time to understand the nature of the problem and give informed consent.

The guidelines for treatment are as follows:

Treatment at birth

Caesarean section and immediate postnatal repair may preserve neurological function in those babies where antenatal ultrasonography has demonstrated good leg movement. There is evidence that trauma during vaginal delivery may exacerbate the neurological deficit. Immediate sac closure is indicated in low lesions without any adverse factors listed above. If the sac is covered with healthy skin there is no urgency to repair it.

Hydrocephalus

Hydrocephalus commonly develops soon after sac closure and is corrected by insertion of a ventriculo-peritoneal (VP) shunt (see also Chapter 12).

Orthopaedic treatment

This is directed at preserving motor development, which should be as near normal as the degree of paralysis will allow. The child with extensive paralysis is given a supportive chair at the age of 3 or 4 months; physiotherapists and occupational therapists encourage activities appropriate to the child's age but which otherwise would be delayed by the paralysis. Standing and walking are encouraged as soon as the child is mature enough to cooperate; for the severely paralysed child, this will be at an age later than normal. Children with low lesions walk well without orthoses. Children with high lesions (above L3) generally walk in long orthoses with extensions to the lower trunk and with elbow crutches. They tend to develop fixed flexion deformity of the hips and knees which may require surgical release. Few children fail to achieve walking, but many of those with high lesions will later cease walking because their mobility is greater in a wheelchair than with extensive orthoses and crutches.

The presence of an active quadriceps muscle enables the child to stand by extending the knees; so long callipers are not required. When the muscles acting on the feet are weak or inactive, plastic ankle-foot orthoses are used to stabilise the feet.

In the common situation in which the dorsiflexors of the ankle are strong but the calf is paralysed, transfer of the *tibialis anterior* tendon posteriorly to the Achilles tendon and calcaneum will prevent progressive deformity.

No orthopaedic treatment is required for children with low sacral lesions, but other deformities which may require treatment are kyphosis and scoliosis. Correction and fusion of paralytic spinal deformities in spina bifida require surgery on the vertebral bodies from in front and on the intact vertebral arches from behind. Internal fixation devices are inserted through both approaches.

Schooling and employment

Most children with spina bifida need assistance at school. Difficulties with access, mobility and continence need to be overcome. Some children with VP shunting for hydrocephalus have specific learning problems, with varying degrees of difficulty with concentration span, attention control, fine motor and perceptual functioning.

Almost all children with spina bifida cystica attend normal school. During the early years in secondary school, vocational guidance is required to direct education and training towards suitable employment. Many professional, commercial, clerical and bench-type jobs are suitable for paraplegic patients.

Prognosis

The antenatal and neonatal mortality for severe spina bifida is high and informed consent for complex and prolonged treatment may be difficult.

Deaths after 1 year of age are usually due to excessive intracranial pressure after failure of a VP shunt for hydrocephalus or due to urinary complications. Most patients, even with severe disabilities, reach adult life with varying degrees of independence. All patients require life-long medical support from doctors and allied staff who understand the nature and implications of their congenital deformities.

KEY POINTS

- Neural tube defects are related to folic acid deficiency in the maternal diet.
- Many neural tube defects are identified antenatally (ultrasonography and alpha-fetoprotein screening).
- Management of spina bifida requires a multidisciplinary team.
- Abnormal skin over the lumbar spine suggests occult spina bifida.

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CHAPTER 10
Disorders of Sex Development

CASE 1

After an uneventful second pregnancy, a woman went into labour quickly and the baby was delivered by the midwife. The midwife thought the baby was a boy, but when the obstetrician arrived she said it was probably a girl! Confusion in the labour ward staff was not resolved until a paediatrician confirmed ambiguous genitalia and arranged transfer to a children's hospital. The parents were very upset and distraught.

Q 1.1 What are the criteria to diagnose a disorder of sex development?

Q 1.2 What should the parents be told?

Q 1.3 Is this an emergency?

Q 1.4 How is the gender of rearing decided?

CASE 2

On routine genital examination, the intern thought the newborn baby had a hypospadias and bifid scrotum, containing one testis.

Q 2.1 Can you be sure this baby is a boy?

Q 2.2 What criteria discriminate babies with a disorder of sex development?

No part of a newborn baby's anatomy arouses as much interest initially as the external genitalia. Throughout the pregnancy, the parents have contemplated whether their child will be a boy or a girl. The announcement of the gender of the child triggers a set of socially predetermined and gender-related responses, gifts, congratulations and celebrations, giving the parents pride and pleasure. It is a crisis, therefore, if the baby's genitalia are abnormal, such that the gender is in doubt [Fig. 10.1]. The urgency of the situation is heightened by the fact that a genital malformation in the newborn may be the outward sign of a potentially life-threatening internal disorder of sex development (DSD), such as congenital adrenal hyperplasia (CAH).

The responsibilities of the attending doctor are to minimise the distress of the parents and family and to arrange for an expedited diagnosis and treatment of the underlying medical disorder which may accompany genital ambiguity.

Definition

Genitalia are described as ambiguous when the phallus is too large for a clitoris and too small for a penis; the urethral opening is near the labioscrotal (genital) folds but there is no normal female introitus; the genital folds remain unfused, with an appearance in between labia and cleft scrotum and the testes may be undescended or impalpable [Fig. 10.1].

The clinical problems

In a newborn baby with ambiguous genitalia, the parents should be told as soon as possible that sex development is incomplete, that the gender cannot be assigned immediately and that consultation with the appropriate specialist will be arranged at once. It will be the specialists' role to outline the steps required to

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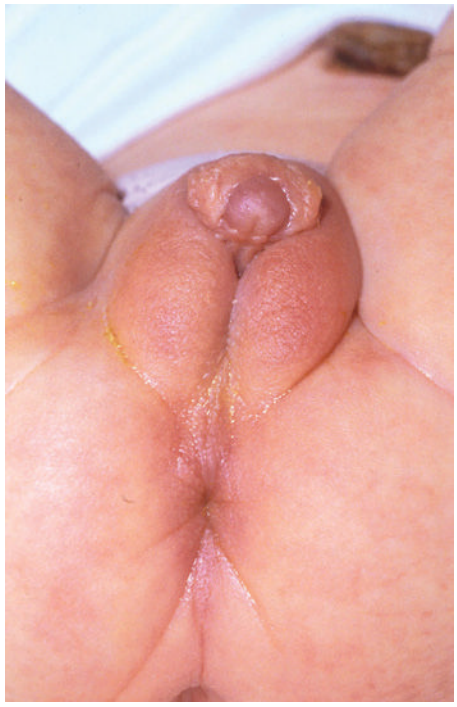

Figure 10.1 Completely ambiguous appearance of external genitalia. Is it a boy or a girl?

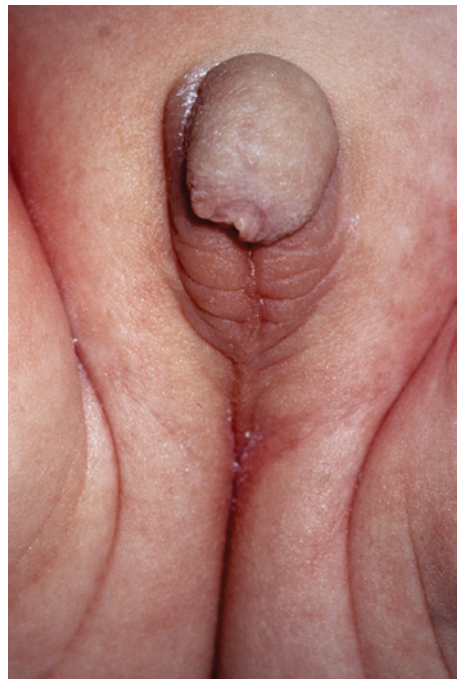

Figure 10.2 Apparent hypospadias and impalpable testes (actually a female with CAH) (Reproduced with permission from Scheffer *et al*. (1988))

obtain the necessary information to determine the cause of the DSD and the sex of rearing.

The investigations of this complex problem are best carried out in the regional referral centre. Two specific types of DSD are the principal source of ambiguity in the newborn: severe hypospadias with a bifid scrotum and/or undescended testes and CAH.

Severe hypospadias with undescended testes

'Hypospadias with undescended testes should be treated initially as a DSD at birth, if either the 'scrotum' is bifid or one or both testes are impalpable. Only a fused scrotum containing two descended testes confirms normal androgenic function, allowing a hypospadiac phallus to be treated as a local anatomical anomaly of penile development in a boy. Once a DSD has been excluded, males with hypospadias and cryptorchidism may be treated by urethroplasty and orchidopexy at 6–12 months.

Congenital adrenal hyperplasia

This life-threatening condition occurs in 1:8000 live births and is the most important condition to be excluded in the management of a DSD causing ambiguous genitalia.

When CAH occurs in females, the appearance of the external genitalia may make the gender difficult to determine [Fig. 10.2]. The ambiguous appearance results from an autosomal recessive defect causing a deficiency of adrenocortical enzymes, especially 21-hydroxylase. This enzyme is necessary for the biosynthesis of both cortisol and aldosterone. Cortisol levels are low, allowing a marked increase in the secretion of pituitary adrenocorticotrophic hormone (ACTH), resulting in adrenal hyperplasia. Only androgens are produced, and in a female, these cause virilisation. Low aldosterone levels allow excessive sodium loss in the urine.

The degree of virilisation is often mild [Fig. 10.3]. If the clitoral enlargement is only minor, the diagnosis may be overlooked – a potentially dangerous situation if the associated biochemical defect produces a 'salt-losing'

situation, with sudden vomiting and collapse (an adrenal crisis) 1–3 weeks after birth.

Investigations

1 Serum electrolytes and blood glucose are obtained urgently, with the expected findings available in 1 h of low sodium, high potassium and hypoglycaemia.
2 Serum 17-hydroxy-progesterone, a progesterone metabolite, is elevated in all but rare forms of CAH due to deficiency of enzymes ordinarily responsible for its further metabolism which takes 12–24 h, for example, 21-hydroxylase.
3 Chromosomal analysis which takes a few days.
4 Twenty-four-hour urine specimen for estimation of pregnanetriol and to obtain a gas–liquid chromatography steroid profile which takes 5 days from collection to get a result.
5 A urogenital sinugram (where X-ray contrast medium is instilled into the external opening) shows a masculinised urethra but connected to a vagina and cervix.
6 A pelvic ultrasound scan confirms the presence of a uterus and Fallopian tubes.

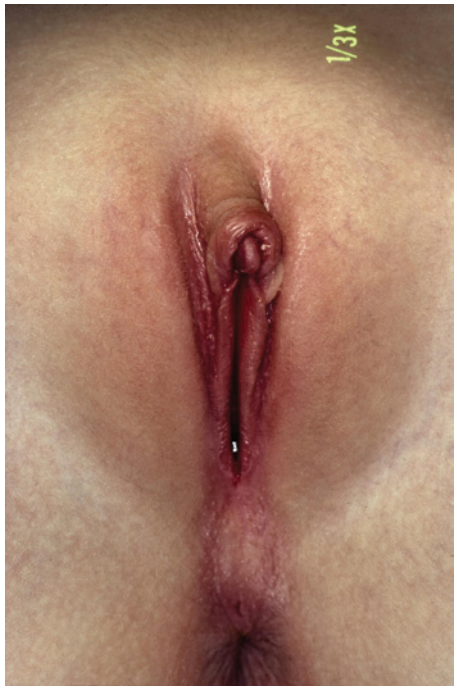

Figure 10.3 An enlarged clitoris in a girl with CAH.

Treatment

1 Intravenous rehydration, initially with normal (0.9%) saline.
2 Hypoglycaemia is monitored and corrected with intravenous dextrose–saline solution.
3 Cortisone acetate or hydrocortisone are given as soon as blood and urine have been collected for examination.

The anatomy should be established as soon as the biochemical status is stabilised, but is less urgent. The anatomical information confirms the diagnosis but is also required to enable planning of the subsequent operative correction which involves clitoroplasty and vaginoplasty. It is important to inform the parents that females with CAH have normal reproductive potential.

Other disorders of sex development

Table 10.1 provides examples of other disorders of sex development, but detailed discussion of this complex subject is beyond the scope of this chapter. Instead, the background is summarised in the following sections.

Table 10.1 Examples of disorders of sex development

Sex chromosome DSD	46,XY DSD	46,XX DSD	Others
45,X (Turner syndrome) 47,XXY (Klinefelter syndrome) 45,X/46,XY (mixed gonadal dysgenesis)	Androgen insensitivity syndromes Persistent Mullerian duct syndrome 5α-reductase deficiency Complete gonadal dysgenesis (Swyer syndrome)	Congenital adrenal hyperplasia	Cloacal exstrophy Vaginal atresia

Internal and external genital development

Gonadal differentiation is determined by the karyotype, with the *SRY* gene on the Y chromosome triggering testicular development of the initially indeterminant gonad. In the absence of *SRY* and presence of two X chromosomes, the gonad undergoes ovarian development. Other internal organs develop as persistent parts of the paired embryonic ducts, the Wolffian (mesonephric) and Műllerian (paramesonephric) ducts which are present initially in both males and females.

The testes *in utero* secrete three hormones: Műllerian inhibitory substance (MIS; also known as anti-Műllerian hormone) from Sertoli cells and insulin-like hormone 3 (Insl3) and testosterone from Leydig cells.

1. MIS promotes regression of the Műllerian ducts. Absence of the Műllerian duct derivatives (uterus, Fallopian tubes and upper vagina) is evidence of MIS secretion, and so a testis containing functional Sertoli cells must be present.
2. Insl3 stimulates growth of the gubernaculum during the first (transabdominal) phase of testicular descent.
3. Testosterone is required to locally stimulate the Wolffian ducts to give rise to the epididymis, vas deferens and seminal vesicle. Without testosterone, the Wolffian ducts atrophy, with failure of development of their derivatives. Testosterone and its metabolite dihydrotestosterone are also responsible for masculinising the external genitalia and controlling the second (inguinoscrotal) phase of testicular descent.

The enzymes required for testosterone synthesis are regulated by autosomal genes, as is the 5-alpha-reductase, which metabolises testosertone to dihydrotestosterone. The gene encoding for the androgen receptor is located on the X chromosome, and so 46-XY individuals who carry mutations of the androgen receptor gene on their X chromosome are not sensitive to androgen during development or after birth.

Practical decisions in management

Three aspects are important in the management of infants with a DSD causing ambiguous genitalia:

1. The specific diagnosis
2. The sex of rearing
3. The explanation and counselling given to the parents

Referral to a regional centre with expertise in managing DSD is essential to ensure optimal anatomical and functional outcomes.

Diagnosis

To reach a specific diagnosis, the advice of a paediatric endocrinologist, and detailed biochemical and anatomical investigations are required because of the following reasons:

1. There may be genetic implications affecting counselling
2. The potential fertility of the neonate should be established
3. Urgent medical treatment may be required, as in CAH
4. A plan for possible operative intervention is required

However, in many children no specific diagnosis is possible.

Sex of rearing

The sex of rearing is determined by the following:

1. The underlying diagnosis (e.g. CAH patients are normally raised as girls, despite virilisation because they have normal female internal genitalia and fertility).
2. The size of the male versus female genitalia (e.g. a child with a micro-phallus and large vagina may be better raised as a girl, despite XY chromosomes).

Correct determination of the appropriate sex of rearing maximises the patient's prospects of fertility and minimises the risk of psychological damage.

Fertility in the male depends upon testes capable of spermatogenesis, a patent pathway, and a penis with sufficient erectile tissue for erection and insemination. A good-sized phallus is rarely present in male infants with a DSD causing ambiguous genitalia, and fertility is unlikely. However, with assistance of reproductive technology (sperm aspiration from epididymis and injection into ova), fertility may be possible in some cases.

In the female, ovulation and a pathway to the uterus are required, but developments in *in vitro* fertilisation, using donor gametes and transplantation of an embryo, now permit pregnancy in a female with a uterus and vagina but no ovaries or Fallopian tubes. There are opportunities, therefore, for active participation in the reproductive process in those raised as females, provided a uterus is present.

Psychological damage is common in both sexes. In a patient raised unsuccessfully as a male, embarrassment due to a micro-phallus, inability to void standing and a phallus inadequate for intercourse, all create major problems.

Counselling parents

Parents should be told frankly, when the gender is unclear, that tests will be carried out urgently, and how long it will take to fully understand the child's pathology.

The gender is the sex of rearing, and once decided, should be reinforced at every opportunity by referring to the infant as 'he' or 'she', and never 'it'. It is important to reassure parents that genital ambiguity and malformations do not lead to homosexuality, a fear many parents experience, but few express. It is helpful to explain to parents that the genitalia go through an undifferentiated stage in both sexes, and that differentiation is extremely complex and not always complete at birth.

KEY POINTS

- Disorders of sex development (DSD) require urgent assessment at birth and always need referral to a regional centre.
- In babies with 'hypospadias' and undescended testes and/or bifid scrotum, full investigation for DSD is required.

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CHAPTER 11
Anorectal Malformations

CASE 1

A baby boy is delivered in a country hospital. He is found to have an absent anal opening and has passed meconium per urethra.

Q 1.1 How would you arrange referral and transport?

Q 1.2 What will you tell the parents about the initial management of this anorectal malformation?

Q 1.3 What are the main long-term issues that this boy will have to face?

CASE 2

A child with a severe anorectal malformation has had an anorectal reconstruction in infancy, but at the age of 5 years he is soiling frequently and is about to start school.

Q 2.1 Which method of imaging would give the best visualisation of the relationship of the bowel to the anorectal sphincters?

Q 2.2 If no fault is found with the reconstructive surgery, how is this problem best managed?

Anorectal malformations are becoming less common. Although *imperforate anus* is the name sometimes given to this condition, in many cases, there is a fistulous opening into the urinary tract in the male or the genital tract in the female. There are many different subtypes of anorectal malformations, and they are often associated with other congenital defects such as the vertebral, anal, cardiac, tracheo-oesophageal, renal and limb (VACTERL) association. Surgical correction of these anomalies is difficult, as the rectum and anus have lost their normal relationship to the sphincter muscles, which themselves may be abnormal in their development and have a poor nerve supply.

Classification

There are a number of variations seen in the anatomy of the perineum in neonates with anorectal malformations [Table 11.1 and Fig. 11.1]. The key difference between the different types of anomaly lies in the relationship of the terminal bowel to the pelvic floor muscles and the *levator ani* muscle in particular. In addition, anorectal malformations are divided into those with or without a fistula to the urogenital tract or the skin. More severe anorectal malformations have arrested development of the bowel above the level of the pelvic floor muscles; these are relatively difficult to treat, and the long-term prognosis for normal continence is not good. In anorectal malformations where the developing bowel passes down through the pelvic floor muscles and anal sphincters, the surgical correction is relatively easy and the long-term prognosis is better, but even in these, continence is not often normal.

Associated anomalies

The mortality and morbidity of anorectal malformations are influenced as much by the associated anomalies as the anorectal malformation itself. Approximately 60% of neonates with an anorectal malformation have a second abnormality. The most common of such abnormalities are in the genitourinary (30%), vertebral (30%), central nervous (20%) and alimentary (10%) systems. A wide range of urinary tract abnormalities, including neuropathic bladder, vesicoureteric reflux, duplication of the ureter and ureterocele, may increase the long-term morbidity. Cardiac and major chromosomal abnormalities may be life-threatening. The vertebral

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Table 11.1 International classification

Major clinical groups	Perineal (cutaneous) fistula
	Rectourethral fistula
	• Bulbar
	• Prostatic
	Rectovesical fistula
	Vestibular fistula
	Cloaca
	No fistula
	Anal stenosis
Rare/Regional variants	Pouch colon
	Rectal atresia/stenosis
	Rectovaginal fistula
	H-type fistula
	Others

From Holschneider and Hutson (2006).

anomalies may be associated with deficiency of the spinal cord and pelvic nerves, contributing to anorectal sphincteric dysfunction and faecal incontinence. In the alimentary tract, oesophageal atresia and duodenal atresia may be seen.

Incidence

Anorectal malformations occur in 1:3000 to 1:5000 births with a slight preponderance in males. Most cases are sporadic and the risk of the same problem occurring in a future pregnancy is small. There are, however, occasional families with a high incidence of anorectal malformations that follow an autosomal dominant inheritance pattern.

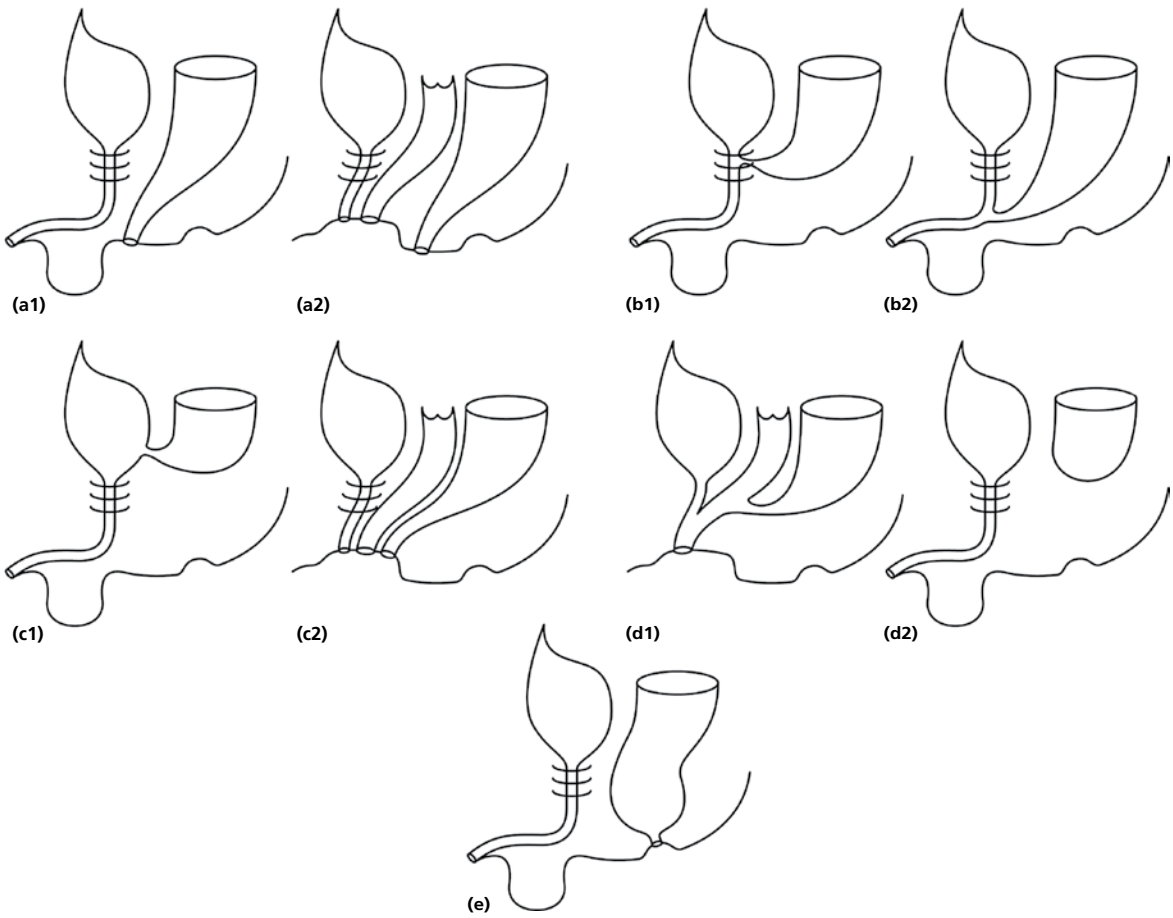

Figure 11.1 A schema of the more common varieties of anorectal malformations. (a1) rectocutaneous fistula (male); (a2) rectocutaneous fistula (female); (b1) rectoprostatic fistula (male); (b2) rectobulbar fistula (male); (c1) rectovesical fistula (male); (c2) rectovestibular fistula (female); (d1) cloacal anomaly (female); (d2) rectal agenesis (often in Down syndrome); (e) anal stenosis.

Clinical features

The newborn baby with a supralevator lesion has no visible anus [Fig. 11.2]. In males, there is a fistula to the urinary tract in approximately 70% of patients: this may lead to the appearance of meconium in the urine, an important diagnostic observation [Table 11.2]. However, as the fistula is too narrow, the neonate will develop features of a distal bowel obstruction. In males with lower and less severe malformations, there is a fistulous opening in the midline of the perineal skin, scrotum or ventral surface of the penis. This is easily visible when it is filled with meconium but may be very minute and requires a careful search with good illumination [Fig. 11.3].

In females, the bowel opening into the genital tract is usually wide enough to decompress the bowel adequately.

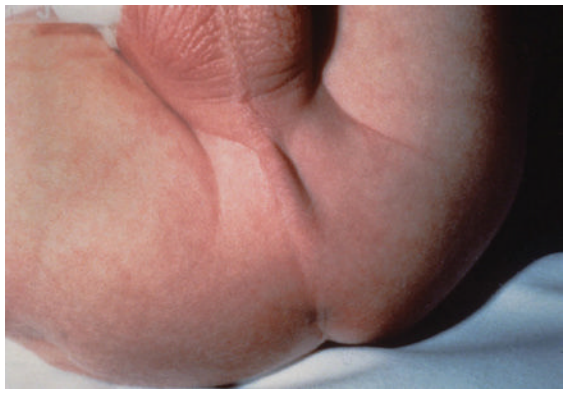

Figure 11.2 The featureless perineum of a male baby with a rectourethral fistula.

Table 11.2 Clinical evaluation of anorectal malformations

Male
A fistulous opening in the skin of the perineum or penis indicates a less severe malformation [Fig. 11.3]
Meconium in the urine indicates a severe malformation with a fistula to the urinary tract (urethra or bladder)
If there is no perineal skin fistula, imaging is required to determine the level at which the bowel stops and confirm whether there is a fistula to the urinary tract
Female
Three openings in the perineum indicate a less severe malformation
Two openings in the perineum (urethra and vagina) with no visible fistula indicate a more severe malformation – the rectum opens directly into the vestibule or vagina
One opening in the perineum (cloaca) indicates a severe complex cloacal anomaly

A detailed search of each perineal orifice is essential, and the internal anatomy may be predicted when the site of the external opening has been located. For example, faeces may be described as coming from the vagina, yet a more careful examination will usually reveal a small orifice tucked into the vestibule just outside the vaginal orifice (i.e. rectovestibular fistula) [Fig. 11.4]. In less severe

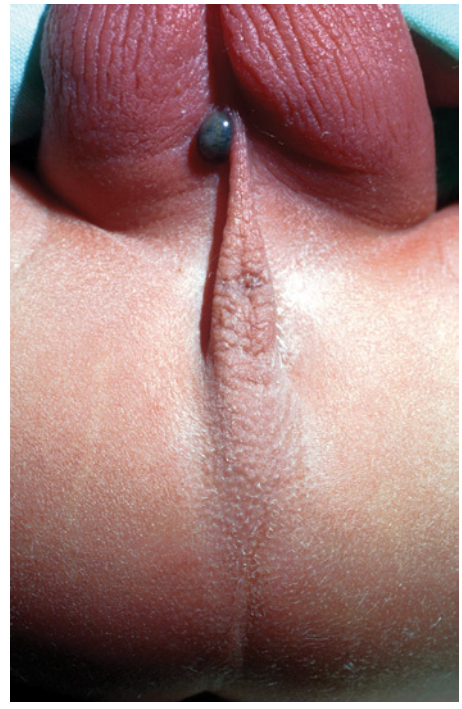

Figure 11.3 A perineal (cutaneous) fistula in a male, with meconium discharging from opening.

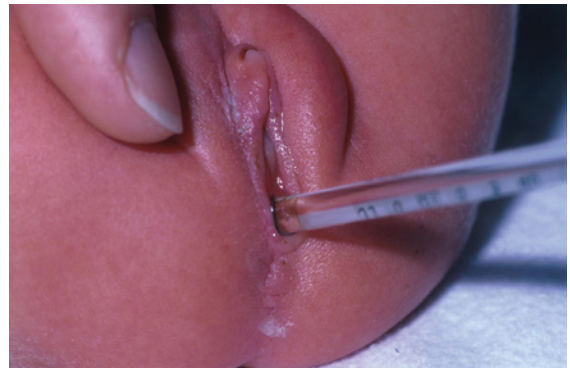

Figure 11.4 A rectovestibular fistula in a female with the thermometer passing cranially up the fistula behind the vagina.

variants, the opening is in the perineum just behind the posterior fourchette as an anocutaneous fistula.

The most severe anomaly seen in the female is the cloaca, where there is only one opening in the perineum: the urethra, vagina and rectum all open into the vault of this common cloacal channel. This is the most difficult of all the anorectal malformations to treat.

A careful and complete physical examination of all babies with an anorectal malformation must be conducted to detect any associated spinal, gastrointestinal and cardiac anomalies. This may include the passage of a stiff 10 French orogastric tube to exclude oesophageal atresia (see Chapter 6), particularly if the neonate has been excessively mucousy after birth. Chromosomal abnormalities such as Down syndrome may also occur.

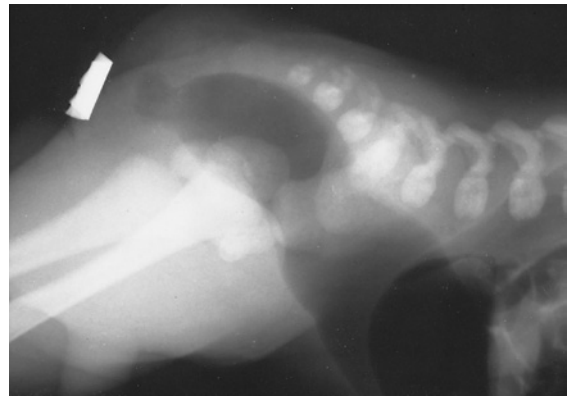

Figure 11.5 Prone, cross-table lateral radiograph in a baby with no fistula, but an imperforate anal membrane, with gas-filled bowel extending nearly to the marker on the skin.

Imaging

The newborn baby with an anorectal malformation that has no perineal opening will need imaging to determine the relationship of the rectum and anus to the anorectal sphincter muscles, to identify a recto-urinary fistula (in the male) and also to demonstrate associated anomalies in the urinary tract, central nervous system, cardiovascular and gastrointestinal systems. The imaging required may include:

1. X-rays of the spine and chest should be examined for associated VACTERL anomalies. Sacral agenesis is of particular importance as loss of the pelvic nerves predicts a poor prognosis for continence.
2. The prone cross-table lateral x-ray uses bowel gas as contrast to establish the position of the terminal bowel against the bony landmarks and determine its relation to the sphincter muscles [Fig. 11.5]. *Invertograms* are no longer performed.
3. Magnetic resonance imaging provides the best evaluation of the state of the sphincter muscles and their relationship to the rectum and anus.
4. Ultrasonography in the first few weeks of life will reveal if there is any deficiency or tethering of the lower spinal cord and will detect any structural abnormalities of the urinary tract. An echocardiograph is also performed.
5. In male neonates with no external opening of the bowel, a micturating cystourethrogram [Fig. 11.6] will often demonstrate the site of any fistula into the urinary tract (e.g. rectourethral fistula or rectoprostatic fistula), as well as coexisting vesicoureteric reflux.

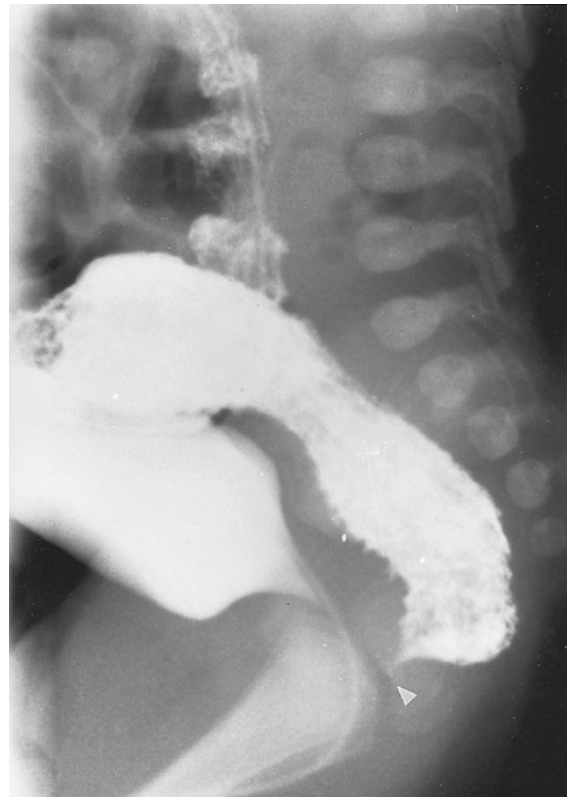

Figure 11.6 A micturating cystourethrogram showing contrast in a rectourethral fistula outlining the distal colon as well as the bladder.

Treatment and prognosis

Certain generalisations may be made:

- The identification of a fistulous opening in the perineum indicates that there is a less severe anomaly and the prognosis is good. In this situation, usually, the only operation required is an anoplasty.
- In males, meconium in the urine indicates a more severe lesion and a fistula between the rectum and the urinary tract. The patient will most likely need a preliminary colostomy.
- In females, a fistula should be expected and a thorough search made for it. In general, three perineal openings indicates a less severe lesion, two a more severe anorectal malformation, and one opening indicates a cloacal abnormality. An expressible bladder with perineal anaesthesia, flat buttocks and sacral agenesis indicate major nerve disruption to the bladder and anorectal sphincters. The prognosis for normal continence is poor.

Malformations with a fistulous opening in the perineum

These malformations are normally managed by an anoplasty shortly after birth. The long-term outlook for continence is good, apart from occasional smearing, staining of underwear and a tendency to constipation with faecal accumulation.

Malformations without a perineal fistula

In most cases, a colostomy is performed at birth. At the age of 3 months or so, definitive corrective surgery (anorectoplasty) is undertaken. This is done by a midline perineal approach or by laparoscopy. A muscle stimulator is used to identify the anal sphincter and pelvic floor muscles forming the *levator ani* sling. The terminal bowel is identified and any recto-urinary fistula is closed. The bowel is brought down through the sphincters to the normal site of the anus. This *pull-through* operation provides anatomical reconstruction of the anorectal malformation. Despite this reconstruction, the results for normal faecal continence are still not good. Although the anorectal sphincter complexes are present in more severe anomalies, the muscles are often poorly developed or the nerve supply is deficient. Recurrent faecal impaction, major soiling or less severe but still distressing minor soiling are common problems. A carefully controlled diet to avoid diarrhoea along with a programme of enemas or home bowel washouts may result in good *assisted continence* in well-organised families. A laparoscopic appendicostomy to enable antegrade colonic irrigation is sometimes needed if faecal incontinence persists into school age. Overall, the morbidity from poor anorectal sphincteric function is determined as much by the long-term support and care of the family in helping with bowel management as by the skill of the surgeon.

KEY POINTS

- Babies with anorectal malformations need careful investigation for other anomalies.
- At birth, immediate transfer to a neonatal surgical centre is optimal.
- Surgical management and prognosis depend on the type of malformation and whether or not a fistula joins the rectum with the skin or urogenital tracts.

Further reading

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PART III
Head and Neck

CHAPTER 12
The Scalp, Skull and Brain

CASE 1

A 3-month-old, ex-premature infant presents with a large head.

Q 1.1 When should you be concerned about enlargement of an infant's head?

CASE 2

A 4-year-old boy presents with early-morning headaches, vomiting and ataxia.

Q 2.1 When does a child with headaches need investigation?

Q 2.2 Can we be optimistic for children with brain tumours?

CASE 3

A 7-year-old girl with a ventriculo-peritoneal shunt is complaining of vomiting and drowsiness.

Q 3.1 Does the absence of ventricular dilatation on CT or MRI scan exclude shunt dysfunction?

CASE 4

You are called to the postnatal ward to see a newborn with a lump at the glabella.

Q 4.1 What do you say to the parents of a baby with an encephalocele?

CASE 5

A 4-month-old infant has a flattened occiput on one side.

Q 5.1 What could the diagnosis be, and what treatment is required?

The infant with a large head

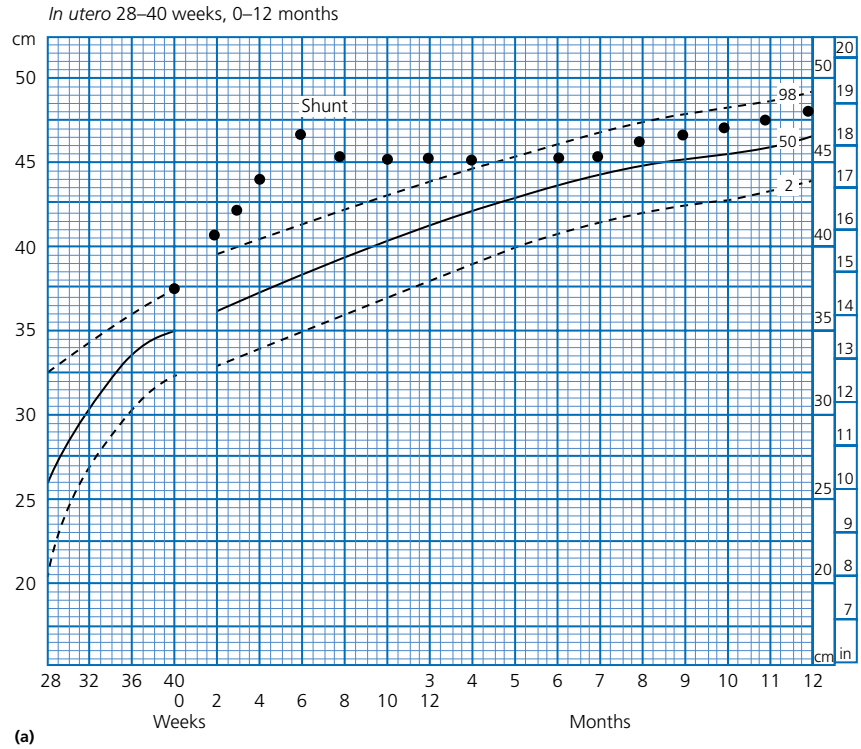
Measurement of head circumference is an essential component of the routine examination of a young child. The growth curve of head circumference must be interpreted, along with the weight and height curves, using standard percentile charts. When suspicion arises that an infant's head is enlarging too rapidly, measurements must be repeated over a period of weeks or months, and compared with the normal curve for this dimension. Deviations from normal [Fig. 12.1] are grouped as follows:

- A steadily increasing divergence from the normal curve, commencing at birth
- A normal curve interrupted by some event, for example, a subdural haemorrhage or an infection, with subsequent increase greater than normal
- An accelerated rate of growth initially, followed by less rapid growth that continues at a high level but parallel to the normal curve, for example, benign enlargement of the subarachnoid space.
- The head circumference commencing at a high level and remaining high but growing at the appropriate rate.

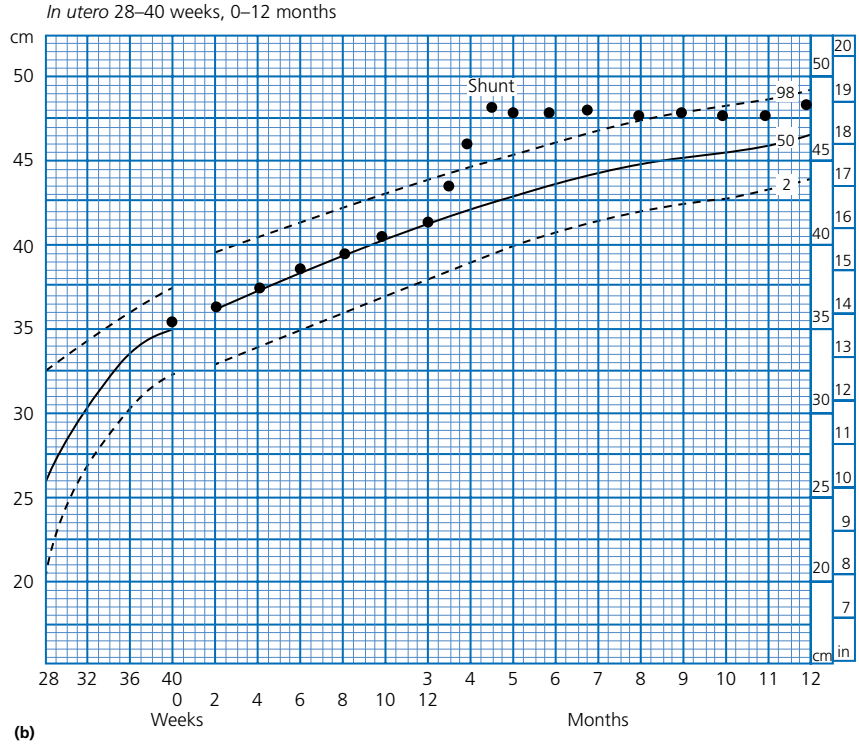
The first two groups need treatment, but surgery may be deferred in the third, unless the accelerated growth in the initial period is very great. Surgery is not usually required for the fourth.

An enlarging head may be the result of factors other than the accumulation of cerebrospinal fluid (CSF), although these are uncommon. The infant's head may enlarge because of thickening of the skull bones, which is readily recognisable in plain x-rays, for example, diffuse fibrous dysplasia. The brain itself may be large, without any increase in the size of the ventricles.

Jones' Clinical Paediatric Surgery, Seventh Edition. Edited by John M. Hutson, Michael O'Brien, Spencer W. Beasley, Warwick J. Teague and Sebastian K. King.



(a)



(b)

Figure 12.1 Variations in the growth curve of the infant head showing mean, 98th and 2nd percentiles. The additional curves represent (**a**) hydrocephalus present from birth and (**b**) acquired hydrocephalus after meningitis at 3 months. In each case, insertion of a shunt is followed by a return towards normal.

Intelligence in these children is often subnormal. One cerebral hemisphere may be larger (hemimegalencephaly), and this may be associated with cortical dysplasia, developmental delay, epilepsy, as well as hemihypertrophy of the body. Localised expanding lesions, for example, subdural haematoma, simple intracerebral or arachnoid cysts or, very occasionally, a cystic neoplasm, may also cause enlargement of the head.

Benign enlargement of the subarachnoid space is also a cause of macrocephaly. It is due to widening of the subarachnoid spaces and must be distinguished from chronic subdural haematoma. It is more common in male infants with a family history of macrocephaly. The head will usually grow at an accelerated rate within the first year of life but settles to grow at a normal rate, although at or above the 98th percentile. The fontanelle may be full in these children. No treatment is required unless the plateau of growth rate is not reached.

Hydrocephalus

Most infants with a large head suffer from excess CSF caused by:

1 Excessive production
2 Obstruction along the pathway of CSF flow
3 Impaired CSF absorption into the veins

Excessive production of CSF causing hydrocephalus is rare and is caused by papilloma, hypertrophy or carcinoma of the choroid plexus.

Obstruction to the flow of CSF is the most common cause of hydrocephalus, which is further subdivided as follows:

1 *Noncommunicating* or obstructive hydrocephalus, in which there is no communication between the ventricles and the subarachnoid space. The ventricles are greatly enlarged without distension of the basal cisterns or cerebral sulci [Fig. 12.2].

   The most frequent causes of obstruction are:

   a Primary developmental anomalies such as aqueduct stenosis or a congenital cyst, for example, supraseller arachnoid cyst or posterior fossa cyst with hypoplasia of the vermis (Dandy–Walker syndrome).
   b Haemorrhage or infection: intracerebral and intraventricular haemorrhage in premature babies is common.
   c Tumours may obstruct the ventricular system in the older child, but only 5% of cases of hydrocephalus in infancy are caused by a neoplasm.

2 *Communicating* hydrocephalus, in which the ventricles do communicate with the basal cisterns, but there is an obstruction in the subarachnoid spaces or in the arachnoid villi/sagittal sinus.

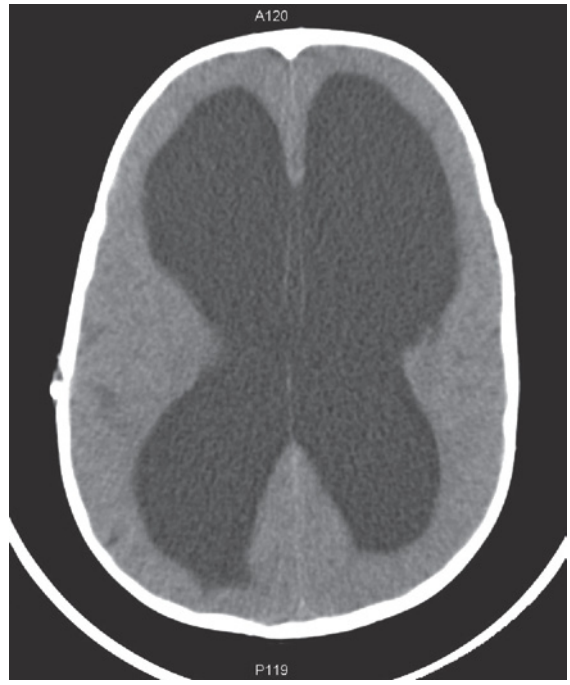


Figure 12.2 Noncommunicating hydrocephalus. CT scan showing massive dilatation of the ventricles.

Failure of absorption of the CSF may occur temporarily as a result of inflammatory exudate around the basal cisterns and arachnoidal villi following meningitis or as a result of haemorrhage in the subarachnoid space. Permanent and severe derangement follows thrombosis of the sagittal or lateral sinuses in the newborn as a result of dehydration; the result is a sudden enlargement of the head. Inadequate absorption of CSF also may occur rarely when the intracranial venous pressure is raised, for example, an arteriovenous malformation (AVM) involving the venous sinuses.

The history, physical signs, a chart of the rate of head growth [Fig. 12.1] and special investigations such as ultrasonography, CT or MRI are all considered to determine the cause [Box 12.1], plan the treatment and estimate the prognosis of the child with a large head.

Clinical signs

A head circumference that is increasing faster than the normal increments for the age of the infant is the main clinical feature and the indication for investigation and treatment. The deviation is depicted by plotting the

Box 12.1 Causes of childhood hydrocephalus

- **1** Communicating hydrocephalus
  - Increased CSF production
    - Choroid plexus papilloma
  - Decreased CSF absorption
    - Haemorrhage
    - Infection
    - Venous hypertension
      - Sinus thrombosis
- **2** Noncommunicating hydrocephalus
  - Congenital
    - Aqueduct of Sylvius stenosis
    - Dandy–Walker malformation
    - Suprasellar arachnoid cysts
  - Acquired
    - Tumours
    - Cysts
    - Haemorrhage

measurements of the circumference obtained at regular intervals on a graph of the normal curve [Fig. 12.1]. Auscultation for a bruit is a useful clinical sign for an underlying vascular malformation.

The shape of the head becomes abnormal. The frontal region is prominent in all types, but with a stricture of the aqueduct, expansion of the lateral ventricles produces an *occipital overhang* above the small posterior fossa as well. The opposite occurs when the fourth ventricle is expanded as a result of occlusion of its foramina; the external occipital protuberance is pushed upwards. Raised intracranial pressure produces a wide anterior fontanelle, palpable separation of the cranial sutures and a raised or drum-like note on percussion of the skull.

Abnormal neurological signs from hydrocephalus alone are unusual. The sixth cranial nerve is vulnerable because of its long course, and a lateral rectus palsy causing internal strabismus may occur. Persistent downward deviation of the eyes (*setting sun* sign) occurs when advanced hydrocephalus causes pressure on the quadrigeminal plate. Fourth or sixth cranial nerve palsies are termed *false localising signs*, as the pathologic process responsible for hydrocephalus does not localise to the affected nerves. When the obstruction is acute and hydrocephalus develops rapidly, there may be brainstem signs with increased extensor tone, rigidly extended lower limbs and clenched hands with the fingers over the infolded thumb. Other possible signs of hydrocephalus are retraction of the head and opisthotonus.

Transillumination of an infant's head by a beam of bright light in a darkened room will often show characteristic patterns. General transillumination indicates a gross and uniform dilatation of the ventricles. Unilateral translucency may indicate a subdural collection of fluid, and other localised bright areas may indicate large cysts or a large dilated fourth ventricle.

Investigations

Ultrasound imaging is a non-invasive means of diagnosing hydrocephalus in infancy and is done by placing the ultrasound probe on the anterior fontanelle. Little or no special preparation is required; the procedure is safe and may be repeated as often as necessary. Once closure of the fontanelle occurs, the technique is no longer applicable.

CT or MRI is used in the older child and in infants if more detail is required. Both modalities provide a clear image of the intracranial anatomy and a precise means of detecting the presence and extent of hydrocephalus, and frequently demonstrate the site and cause of obstruction. MRI, despite the obligatory need for a general anaesthetic in the younger child, is the modality of choice due to the lack of irradiation, the higher resolution of cerebral anatomy and the ability to analyse CSF flow.

The CSF dynamic scan involves the injection of a radionuclide tracer into the CSF pathway. Its passage through the ventricles and subarachnoid space is followed. Obstructions and abnormalities in the passage of CSF, from production to final absorption, may be recorded. With the advent of MRI, radionuclide studies are infrequently used.

In more complex cases, intracranial pressure monitoring may help to determine the need, or otherwise, for treatment.

Plain x-rays are generally of no great value but may be used to confirm a diagnosis of raised intracranial pressure.

Treatment

Not all infants with enlargement of the head require operation, but the child should be investigated if there is evidence of a continued deviation from the normal curve and/or signs of raised intracranial pressure. Operation is indicated if there is sustained deviation

from the normal curve but no obvious evidence of severe brain damage.

Broadly speaking, there are three treatment strategies for controlling an expanding head:

1 Reduction of CSF production: production of CSF may be reduced by a drug that acts directly on the choroid plexus (carbonic anhydrase inhibitor) or by an osmotic agent. Control is frequently incomplete and of short-term benefit only.
2 Reconstitution of CSF pathways within the cranium: removal of a mass may allow CSF to return to a normal flow pattern. Tumours in the posterior fossa frequently cause hydrocephalus, and excision of the tumour leads to a rapid resolution in most cases. Neuro-endoscopy, that is, the placement of an endoscope into the ventricles via a burr hole, is an important technique for inspection, biopsy and therapeutic manoeuvres. Examples of neuro-endoscopic treatments are fenestration of a cyst into the ventricle and creation of an opening in the floor of the third ventricle (third ventriculostomy) to correct an obstructive hydrocephalus.
3 Diversion of CSF to a site outside the cranium: external removal of CSF is the usual method of treating hydrocephalus. In communicating hydrocephalus, particularly in premature neonates, removal may be undertaken intermittently to control the hydrocephalus until normal pathways are re-established, for example, via lumbar puncture or a ventricular reservoir.

A ventriculo-peritoneal shunt is usually the definitive operation of choice in children of all ages. This shunt comprises a ventricular catheter, a valve or flushing device beneath the scalp and a long kink-resistant tube that is tunnelled within the subcutaneous tissues of the chest wall before entering the peritoneal cavity. A long tube is placed within the peritoneal cavity to allow for subsequent growth of the patient. Less frequently, a ventriculo-atrial shunt is performed to divert the CSF into the right atrium.

Complications of shunts

Most children with shunts are *shunt dependent* and will not tolerate malfunction of these devices:

1 Obstruction: most frequently the ventricular catheter becomes occluded with choroid plexus or cerebral tissue. The lower end may be obstructed by the growth of the child, which displaces the lower end into an unsuitable position, by adherence to the greater omentum or by fracture of the tube. Rarely, the valve may malfunction. Revision of the shunt is required.
2 Infection: the shunt system becomes colonised by pathogenic organisms, necessitating externalisation of the shunt for temporary external drainage of the CSF until it is sterilised with antibiotics. The shunt is then revised or replaced.
3 Disconnection
4 Overdrainage. The ventricles become small, and the child may develop chronic headache due to low intracranial pressure. The opening pressure of the valve may need to be raised.

A child with a shunt must be reviewed at regular intervals during the growing years. In general, if the diagnosis is established before hydrocephalus is advanced, if there are no other significant brain anomalies, and if the treatment is appropriate and maintained, then the patient has every chance of developing normally. A child with a shunt is not restricted in activities.

Congenital abnormalities of the cranium

Errors in the development of the scalp, skull and brain are not as common as those of the spinal cord, but they present the same variety of abnormalities. Only the more common or important ones are described here.

Dermoid sinus

This is found most frequently in the mid-occipital region and may communicate with a more deeply situated dermoid cyst containing sebaceous material and hairs. The sinus may have some fine hairs (often a different colour) protruding from it and usually discharges sebaceous material. The deeper component may cause all the signs of an intracranial tumour with cerebellar signs predominating; it may also become infected.

An intracranial dermoid may occur without an external sinus. Infection is uncommon and the cyst presents by causing local pressure or obstruction of the CSF. Rarely, the cyst ruptures, leading to aseptic meningitis.

Dermoid cysts of the scalp are common over the anterior fontanelle and near the orbital margin and are described in Chapter 16.

Craniosynostosis

Cranial sutures act as lines of normal growth. Premature suture closure restricts development of the corresponding region, with compensatory growth occurring at other suture lines. The subsequent distortion in the shape of the skull results in severe cosmetic deformities, but only occasionally does it cause sufficient diminution of the intracranial capacity to limit the growth of the brain. A description of the different deformities is given in Chapter 15.

Treatment

The abnormal appearance and the risk of developmental delay are the two indications for operative intervention. Developmental delay probably occurs in only 10% of these children, and its likelihood is to be suspected when radiographs show signs of increased intracranial pressure, that is, increased cerebral convolutional markings (*copper beating*) and separation of the unfused sutures. Headache, vomiting and papilloedema are rare, but exophthalmos and ophthalmoplegia are not infrequent.

Operative correction ranges from simple linear craniectomy (excision of a strip of bone along the fused suture) to radical removal and repositioning of the vault bones (Chapter 15).

Plagiocephaly

This is a common deformity that skews the entire skull. One frontal region and the opposite occipital region are flat, and the contralateral areas are full and rounded. The effect is that the longest diameter is displaced from the sagittal axis towards the side with the prominent frontal contour.

Congenital plagiocephaly may be caused by contact of the foetal head with the maternal pelvis or with irregularity of the uterine wall, for example, fibroids. Acquired plagiocephaly in the first 3–4 months after birth is more common (Chapter 16). X-rays may show sclerosis along the lambdoid suture line without fusion. The deformity may be minimised by placing babies with deformational plagiocephaly supine but alternating the head position to promote head turning to each side. Providing periods of *tummy time* is most effective.

The deformity tends to improve after the age of 6 months and continues to correct until puberty. A minor degree probably persists indefinitely, though this is not readily detected when hair obscures the contours of the skull.

Premature fusion of the lambdoid suture is an uncommon cause of plagiocephaly. Operative repair is required.

Cranium bifidum (including encephalocele)

Defects at the cephalic end of the embryonic neural tube are much less common than in the thoracolumbar region. The same basic deformities occur, mostly in the occipital region, but in some countries, for example, Thailand, they are more common in the frontal (sincipital) area.

The herniations are in the midline [Fig. 12.3], well covered with skin and lined by meninges, and may contain CSF alone (meningocele) or, more frequently, brain (encephalocele). Occasionally, the herniation occurs into the nasal cavity, and the sac is then covered by mucosa, not skin.

Other intracranial abnormalities also may be present, and imaging is necessary to detect these before operative repair.

Simple excision of the sac, replacement of viable herniated cerebral contents and sound closure of the dura and the bone defect usually may be effected. Occipital encephaloceles may be associated with severe brain dysfunction (developmental delay, visual defects, hydrocephalus), which may preclude treatment. The sincipital encephaloceles are repaired using craniofacial techniques with good cosmetic results and usually a good neurological outcome (Chapter 15).

Intracranial tumours

Tumours of the central nervous system are the largest group of malignancies, excluding leukaemia, in childhood. Radical surgery and adjuvant chemotherapy or radiotherapy in selected cases may produce long-term survival. There are also many benign and slowly growing intracranial tumours that may be cured following operative excision.

Mode of presentation

The mode of presentation in children differs in many ways from that seen in adults:

1 The common types of tumour and their sites of origin are different, for example, the preponderance of tumours in the posterior fossa in childhood [Table 12.1].

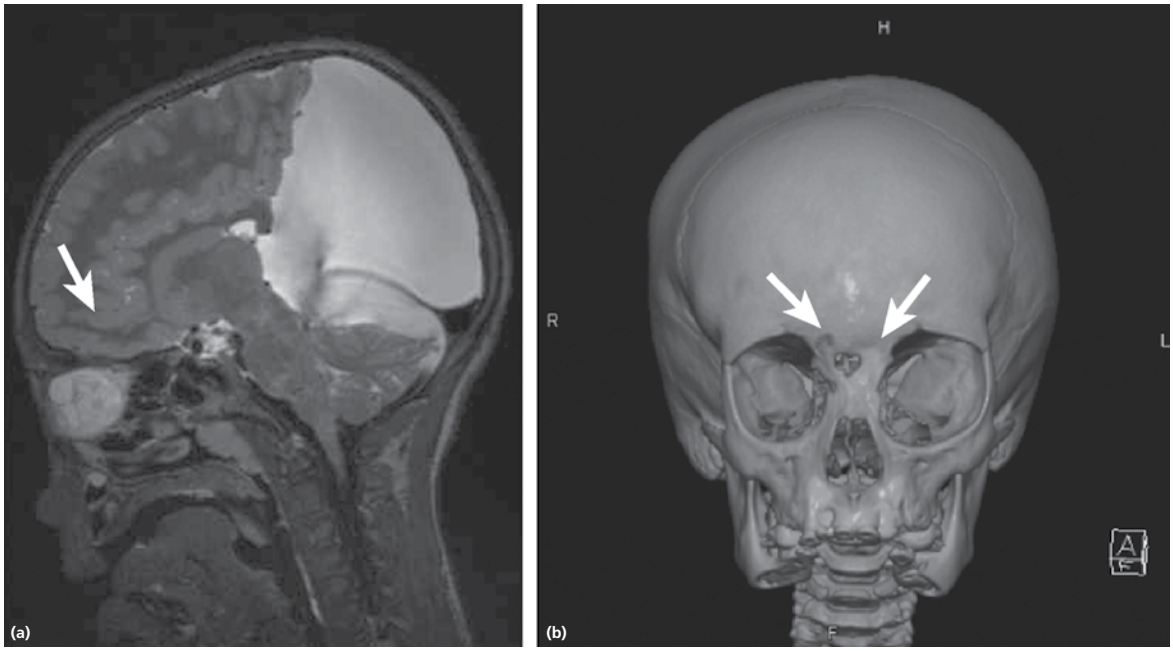


Figure 12.3 Encephalocele. An example of a midline herniation as seen on the (**a**) T2 weighted MRI scans (arrow) with cranium bifidum as seen on the (**b**) CT scan (arrow). This is an example of a neural tube defect.

Table 12.1 Cerebral tumours: Percentage distribution of 300 consecutive tumours at Royal Children's Hospital, Melbourne

Group 1:	Cerebral hemispheres	18%
Group 2:	Third ventricle	17%
	Optic chiasm	5%
	Craniopharyngioma	5%
	Pineal tumour	5%
	Glial tumours	2%
Group 3:	Posterior fossa	53%
	Medulloblastoma	
	Solid/cystic astrocytoma	
	Brainstem glioma	
	Spinal tumours	12%

2 Young children adapt better to an expanding intracranial lesion because of the expansion of the skull; accommodation for weeks or even months is possible, but once this fails, the final decline is often rapid and catastrophic.

3 Many tumours arise close to the CSF pathways in relatively *silent* areas. Neurological signs are few or absent until the flow of CSF is obstructed, when signs of raised intracranial pressure, for example, headache, vomiting and papilloedema, develop with alarming suddenness.

4 Early signs often affect the vision, but loss of acuity or diplopia are not appreciated in early childhood and never arise as symptoms in infants.

5 Neurological signs may present early, while evidence of raised intracranial pressure appears much later. In infants and younger children, the dramatic development of raised intracranial pressure may initiate a search for localising signs that only then are recognised.

Intracranial tumours may be divided into three main groups, each of which produces a more or less typical clinical picture.

Group 1: Glial tumours of the cerebral hemispheres

These are less common than those in the posterior fossa and cover the full spectrum of gliomas. Histology varies from benign to highly malignant but is a much less reliable guide to prognosis than in adult gliomas.

The clinical picture is similar to that of adults, and diagnosis and management follow the same principles. The tumours are excised as far as possible without causing deficit.

Group 2: Tumours near the third ventricle

Tumours near the third ventricle are a very important group in childhood, for example, gliomas of the optic chiasm, craniopharyngioma and pineal region tumours. Their clinical progress is often insidious until signs of ventricular obstruction manifest, but localising neurological signs may be detected early. Those situated anteriorly produce defects in vision and endocrine disturbance and those posteriorly cause hydrocephalus, disturbances in ocular movements and rarely precocious puberty.

Gliomas of the optic chiasm

Gliomas of the optic chiasm and optic nerves are associated with neurofibromatosis in 30–50% of cases. They cause unpredictable field defects, loss of visual acuity, optic atrophy, squint and sometimes proptosis, before obstructing the third ventricle. Infants may present with hydrocephalus or with involvement of the hypothalamus causing wasting and anorexia known as the *diencephalic syndrome*, and a similar lesion in older children may cause precocious puberty.

They usually behave in a very indolent manner, but a large or progressively enlarging tumour may be operatively debulked and many are sensitive to chemotherapy. Shunts to relieve ventricular obstruction are sometimes necessary. Long-term survivals are not uncommon.

Craniopharyngioma

The craniopharyngioma grows insidiously. It arises in, above or behind the sella turcica from a remnant of the primitive Rathke's pouch and compresses the pituitary gland, pituitary stalk or hypothalamus, slowing growth and development and gradually compromising vision. It is variably comprised of solid epithelial components and cysts filled with brown turbid fluid described as *machine oil*. The tumour is usually not suspected until the child has had defective sight for years, growth and development have lagged behind or the child tires easily and is unable to keep up with peers.

Small craniopharyngiomas may be excised totally without damage to the adjacent optic nerve or pituitary gland, but large craniopharyngiomas are one of the most challenging problems in paediatric neurosurgery. There is controversy over whether to attempt a complete excision, with chance of cure but risking serious morbidity, which includes visual loss, persistent pituitary deficiency and hypothalamic dysfunction with morbid obesity. Hormone replacement therapy and DDAVP (arginine vasopressin) have improved the outlook for these patients. Radiotherapy is used for recurrent tumours and as an adjunct for those tumours where excision was deliberately incomplete.

Pineal region tumours

The main types of pineal tumours are:

1. Germ cell origin: germinoma, embryonal carcinoma, yolk sac tumour, choriocarcinoma and teratoma
2. Pineal cell tumours: pineocytoma and pineoblastoma
3. Glial tumours

Pineal tumours often obstruct the aqueduct before local signs develop so that headache, vomiting, papilloedema and impaired consciousness are the presenting features. Later, pressure on the upper brainstem causes a loss of upward gaze, a distinctive localising sign. Precocious puberty is an uncommon feature. These tumours range from highly malignant to benign. Diagnosis is based on imaging and CSF and blood markers. Surgical treatment varies according to pathology, with hydrocephalus often requiring independent treatment. Chemotherapy and radiotherapy are often employed as adjuvant or primary therapy. The prognosis depends on the histology and tumour burden following the primary treatment. Even though germinomas are malignant, they may be cured in the majority with chemotherapy and/or radiotherapy.

Group 3: Posterior fossa tumours

These form about 50% of all intracranial tumours in childhood, but only 25% in adulthood. Those in the cerebellum cause ventricular obstruction early, so that headache and vomiting (characteristically in the early morning) appear before neurological signs such as incoordination, ataxia, hypotonia and tremor. In brainstem gliomas, gross incoordination, ataxia and cranial nerve palsies precede signs of increased intracranial pressure. There are four common tumours in this region.

Medulloblastoma

This is a malignant tumour of the vermis forming a large mass that blocks the fourth ventricle [Fig. 12.4]. It may spread out into the basal cisterns and characteristically disseminates widely throughout the CSF pathways, particularly in the spinal canal.

The tumour occurs more often in males at about 2 years of age, with a typical history of morning headaches

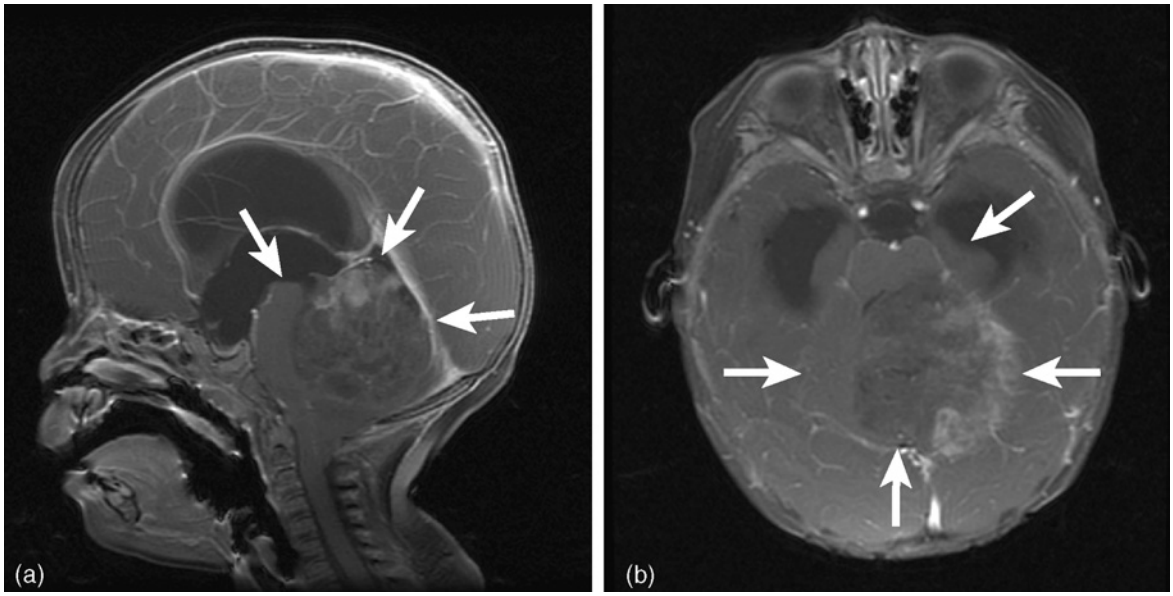


Figure 12.4 Medulloblastoma. An MRI scan showing a massive posterior fossa tumour blocking the fourth ventricle in (**a**) sagittal section and (**b**) transverse section. The tumour is indicated with an arrow.

and vomiting, change in personality and clumsiness of gait. Papilloedema and truncal ataxia are the common neurological signs, and there may be tilting of the head when the child sits up. Following removal, the tumour may recur locally and disseminate, unless it responds to adjuvant chemotherapy and craniospinal radiotherapy in children over the age of 3 years. Children under 3 years of age require aggressive adjuvant chemotherapy. The overall 5 year survival is 60–80% and is dependent on the degree of resection and the molecular biology of the tumour.

Astrocytoma of the cerebellum

An astrocytoma may be a solid tumour or a nodule of tumour in the wall of a cyst or even form the lining of a simple cyst. Children are older (about 5 years) than those with a medulloblastoma; the length of the history is 3–6 months rather than weeks, beginning with morning headaches and vomiting. Much later, usually in the 2–3 weeks before diagnosis, squint, incoordination and ataxia appear. The clinical signs are fairly constant: papilloedema, squint, mild ataxia and incoordination of hand movements.

There is usually a localised tumour, in one lateral lobe, which is often completely excised. Radiotherapy is reserved for tumours with more aggressive histological grades that have recurred. There is a high rate of cure of the benign (pilocytic) astrocytomas following excision alone.

Brainstem glioma

This usually causes diffuse brainstem enlargement without much evidence of a localised tumour. The age incidence is wide, and the signs are caused by cranial nerve palsies and involvement of the long tracts passing through the brainstem: gross strabismus, facial weakness, difficulty in swallowing, hemiparesis and ataxia occur. The child is miserable and pathetic, quite different from those with other types of posterior fossa tumours. Hydrocephalus is uncommon.

The diagnosis is confirmed by MRI, and operative intervention is usually not indicated. Radiotherapy and chemotherapy are used. In most patients, the signs resolve rapidly during treatment but recur within 3–6 months. The prognosis of the diffuse malignant brainstem glioma is very poor despite this therapy. A few patients with less aggressive tumours achieve a longer period of survival. There are some focal and benign tumours of the brainstem that may be largely excised with a good prognosis.

Ependymoma

The mode of presentation is similar to medulloblastoma. At operation, the tumour frequently is attached to the floor of the fourth ventricle and, like the medulloblastoma, the tumour has the same tendency to metastasise in the CSF pathways. The overall prognosis is worse than medulloblastoma, particularly in those with incomplete excision, with 5-year survival rates of 50–65%.

Tumours in the spinal canal

In infancy, neuroblastoma is the most common spinal canal malignancy. Primary vertebral and intrathecal tumours are uncommon. In the first 2–3 years of life, the most common lesion is a metastasis from a medulloblastoma. These lesions may present following a short history of poor limb movement and general malaise.

In later childhood, other intrathecal tumours appear, for example, neurofibromas of the nerve roots and astrocytomas and ependymomas of the spinal cord. The child presents variably with chronic spinal pain that may be severe and unremitting, scoliosis, slowly progressive weakness of the limbs, abnormal reflexes and sphincter disturbance. The diagnosis is established by MRI. Operative decompression, biopsy and excision of the tumour are performed. The child with a spinal tumour may also present acutely with spinal pain and signs of spinal cord compression: paralysis, sensory loss and sphincter disturbance. This is a surgical emergency.

Intracranial vascular disorders

Arteriovenous malformations

AVMs are congenital vascular anomalies of the brain, which comprise abnormal arteriovenous fistulae, which subsequently lead to dilated arteriolised veins, multiple tortuous feeding arteries and a central nidus of capillary-like fistulous vessels. Aneurysms may develop on the feeding vessels due to the high flow rates, and the lesions may vary in size considerably. The presentation is often with rupture, causing intracerebral and sometimes subarachnoid haemorrhage [Box 12.2]. This is the most common cause of spontaneous intracranial haemorrhage in children. Presenting features include sudden severe headache, focal neurological signs, epileptic seizure and rapid obtundation, if the clot enlarges sufficiently. The treatment involves evacuation of clot and excision of the AVM. Elective excision of an unruptured AVM is complex and may involve excision preceded by embolisation. Some small deep-seated AVMs may be treated with focused radiotherapy (stereotactic radiosurgery).

Box 12.2 Presentation of intracranial AVMs

1. Rupture with intracranial (or subarachnoid) haemorrhage
2. Epilepsy
3. Chronic headache
4. Focal signs (adjacent brain ischaemia)

Intracranial aneurysms

Intracranial aneurysms occur at all ages but are generally rare in children. The causes are:

1. Congenital *berry* aneurysms, – these occur at branch points of the major basal cerebral arteries at the circle of Willis and, in children, often reach giant proportions.
2. Related to connective tissue disorders, for example, Ehlers–Danlos syndrome.
3. Post-traumatic – *false* aneurysms either from blunt or penetrating trauma.
4. Mycotic aneurysms, which result from infected emboli, for example, following endocarditis.

Subarachnoid haemorrhage results from aneurysm rupture, and craniotomy and clipping of the aneurysm is the definitive treatment.

Vein of Galen malformations

Vein of Galen malformations are rare. Congenital arteriovenous fistulae arising near the Vein of Galen may result in a large dilatation of this vein, the straight sinus and the posterior venous sinuses. These high-flow fistulae may cause congestive heart failure in newborns or may present later with an enlarging head, hydrocephalus, failure to thrive and epilepsy. Their management is complex requiring neuro-radiological embolisation of the feeding vessels or of the vein of Galen aneurysm itself. Operative division of major feeders may rarely be required. The outlook for the child depends on the completeness of fistula obliteration and how much ischaemic damage is present in the surrounding brain.

KEY POINTS

- A large head requires investigation if the circumference is crossing the percentiles or it is an abnormal shape.
- Frequent, different or early-morning headaches need investigation.
- Ventriculo-peritoneal shunts are life-saving for hydrocephalus and allow normal activity. Recognition of shunt blockage is based on clinical diagnosis.
- Encephalocele is a complex neural tube defect.
- Brain tumours have a variable prognosis, depending on cell type, site of origin, degree of surgical resection and tumour molecular biology.

Further reading

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CHAPTER 13
The Eye

CASE 1

A 3-year-old girl is brought to see you because her parents are concerned that her eyes are misaligned.

Q 1.1 Why should this be taken seriously?

Q 1.2 How can you determine if the parents' observations are correct?

CASE 2

Parents of a 3-month-old boy are worried that he cannot see properly and that at times his eyes seem to wobble uncontrollably.

Q 2.1 Do the parents have anything to be really worried about?

Q 2.2 What are you going to do with this child?

CASE 3

A 6-month-old baby has had a sticky eye since the age of about 1 week. Her mother has to clean the affected eye several times a day.

Q 3.1 How are you going to advise this child's parents and what treatments are available?

Q 3.2 Should this child be prescribed topical antibiotic eye drops?

CASE 4

A 6-year-old boy is brought to see you with a 3 h history of a painful red eye.

Q 4.1 How are you going to assess this child?

Vision loss may have profound effects on a child's development. A systematic clinical approach allows rapid diagnosis of most conditions affecting a child's eye and vision [Box 13.1]. Timely and appropriate management will then reduce the chance of permanent visual loss.

Accurate diagnosis is dependent on obtaining a good history and appropriate clinical examination (see the following) supplemented where necessary by further investigations.

Examination of the child's vision and eye

Measurement of vision in preverbal children is by observation rather than formal testing. Asking the parent 'How well does your child see?' or 'What do you think your child sees?' will provide useful clues to the level of vision.

At birth, an alert infant can fix on a face briefly. By 6 weeks of age most infants smile in response to a face, and also will be able to follow a face or light through an arc of 90°. By 6 months of age, an infant can reach for a small object and actively follow moving objects in the environment. By 12 months, a child can pick up tiny objects such as hundreds and thousands (*sprinkles*) [Box. 13.2].

At 2 years of age, most children can name simple pictures to more accurately document visual acuity. Letter- or shape-matching tests of vision are possible at about 3–4 years of age. Formal Snellen measurement of acuity is managed by most children once they reach school age (5–6 years).

Always document the estimate of vision for each eye. For a preverbal child, this may consist of statements like 'Fixes on a face with left eye, but not with right'. If more formal measurement is possible, document the Snellen fraction, for example, 'right eye – 6/9'. This means that at a distance of 6 m, the child can read the *9* line of letters or symbols. Most tests are performed at 3 or 6 m, but if the child's vision is reduced the chart can be brought closer to the child.

Jones' Clinical Paediatric Surgery, Seventh Edition. Edited by John M. Hutson, Michael O'Brien, Spencer W. Beasley, Warwick J. Teague and Sebastian K. King.

Box 13.1 Common ocular symptoms and signs in children

Suspected poor vision
Misaligned eyes
Wobbly eyes
Inflamed eyes
Droopy eyelids
Watery or sticky eyes
White reflex (leukocoria)
Big or small eye
Injured eye
Headache

Box 13.2 Measuring vision

Parents' assessment	Any age
Fix on face/light through 90°	6 weeks
Fix on moving objects	6 months
Pick up coloured sprinkles	12 months
Name simple pictures	2 years
Letter/shape matching	3–4 years
Snellen eye chart	5–6 years

Reduced visual acuity in a child is often the result of amblyopia. Amblyopia arises during development of the visual cortex when *defective* information is being sent from the eye to the brain. Common causes of amblyopia are strabismus (misaligned eyes or *squint*) and unequal refractive error. To avoid diplopia, the immature brain can suppress the information from one eye when strabismus is present. If the two eyes have an unequal refractive error, one will generally have a more blurry retinal image, and thus, a poor signal will be sent to the visual cortex, resulting in amblyopia. Amblyopia is best treated in young children, and thus, early diagnosis is vital.

Inspection will provide much useful information. Abnormal pupil reactions, significantly misaligned eyes, abnormal eye movements (such as nystagmus), inflamed eyes, droopy eyelids, red, watery or discharging eyes, will all be obvious on simple inspection.

Examination of pupil reactions is important in objectively assessing the integrity of the anterior visual pathways (eyes and optic nerves). Normal pupils are of equal or very nearly equal size. The pupillomotor (efferent) pathway originates in a common area of the third nerve nucleus. Thus, if one eye is blind, the pupils will appear of equal size in ambient light. A difference in pupil reaction and size will become apparent if a bright light source (such as a direct ophthalmoscope) is alternately directed at one eye and then the other (the *swinging light test*). When the light is shone in the normal eye, both pupils will constrict, and when the light is switched to the blind eye, both pupils will dilate. This abnormality is called a *relative afferent pupil defect* and is an important, objective sign of visual impairment in one eye or optic nerve. The afferent pathway of the pupil response leaves the optic nerves at the chiasm to pass to the third nerve nucleus. Thus lesions of the visual system posterior to the chiasm will not cause abnormalities of pupil reactions.

Examining the red reflex with a direct ophthalmoscope will reveal much about the internal structure of the eye. The red reflex is examined with a direct ophthalmoscope set to the zero power lens and observing the child's eyes from a distance of about 1 m. A normal red reflex is red to orange and uniform over the pupil and should be the same in each eye. Children with darker iris pigmentation will tend to have duller red reflexes than those with lightly pigmented eyes.

A white reflex [Fig. 13.1] generally indicates significant pathology within an eye and indicates the need for urgent dilated fundus examination by an ophthalmologist. In children over 1 year of age, the pupils can be safely dilated with cyclopentolate 1% or tropicamide 1%. In children less than 1 year, 0.25 or 0.5% preparations should be used.

Observing the corneal light reflections will indicate strabismus in a child who is not cooperative with cover testing. When a light is positioned directly in front of a child's face, the corneal light reflections should be symmetric. Asymmetry of the corneal light reflections suggests that the eyes are misaligned.

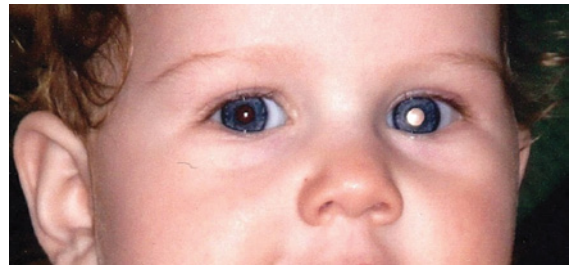

Figure 13.1 The *white* reflex. The normal red reflex is replaced by a white reflection in the left eye in this child with retinoblastoma.

Cover testing is the method of choice to determine if a child has strabismus (misaligned eyes or squint). The cover test is done by first getting the child to fix on an object while the observer determines which eye appears to be misaligned. The eye that appears to be fixing on the object (and not misaligned) is then covered while the apparently misaligned eye is observed. If strabismus is present, a corrective movement of the misaligned eye will be seen as this eye takes up fixation on the object of regard [Fig. 13.2]. If no movement is seen, the eye is uncovered. The cover test is then repeated, but the other eye is covered this time, and the eye that is not covered is again observed for a corrective movement, and if present, strabismus is confirmed. The test can be repeated as many times as necessary. If no movement is seen following repeated covering of either eye, then no strabismus is present. Care must be taken to let the child fix with both eyes open before covering either eye, otherwise normal binocular control may be disrupted and a small latent squint (phoria) may be detected. Latent squints are normal variants and of no significance.

Eversion of the upper eyelid is helpful if a foreign body is suspected to be the cause of a red irritable eye. The upper eyelid is everted by first asking the child to look down. A cotton bud is then applied about 1 cm from the lid margin to act as a fulcrum about which the eyelid will be everted. Finally, to evert the eyelid, the eyelashes are gently pulled initially downwards and then rotated upwards. The subtarsal conjunctiva can then be inspected and any foreign body removed with a second moistened cotton bud.

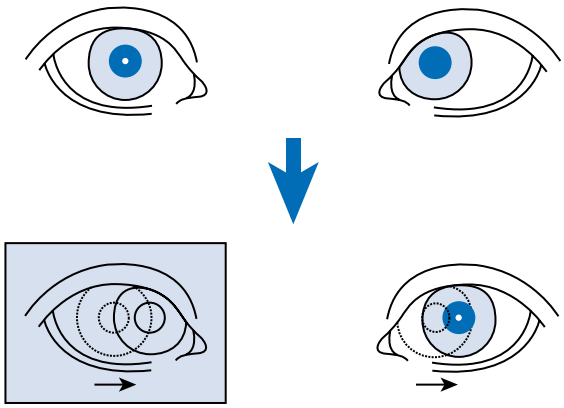

Figure 13.2 Cover test. The child's attention is attracted with a toy or light (top). Once the eye that appears to be looking directly at the toy is covered, the other eye is observed for a refixation movement (bottom). In convergent squint, there is outward movement of uncovered eye (pictured). In divergent squint, the eye moves inwards. If no movement is seen, repeat the cover test covering the other eye.

Refractive errors (focusing problems)

A basic understanding of refractive errors helps a great deal in making sense of ophthalmology. There are three principal refractive errors. These are hypermetropia (long-sightedness), myopia (short-sightedness) and astigmatism. The easiest way to understand refractive errors is to consider the eye in a relaxed state.

When relaxed, a hypermetropic eye focuses light from a distant object behind the retina. To bring such an image into clear focus on the retina, accommodative effort has to be used or a converging (plus) lens placed in front of the eye (i.e. the focal length of the eye has to be changed) [Fig. 13.3]. Conversely, a myopic eye, when relaxed, focuses light from a distant object in front of the retina. No amount of further relaxing will enable the eye to lessen its focal length to bring such an image into clear focus. A diverging (or minus) lens will do this; thus, a myopic eye can only *see* a distant object clearly with the aid of some type of lens [Fig. 13.4]. Astigmatism is more complex but can be thought of as a regular distortion of the image on the retina by an eye that has different focal lengths in different axes.

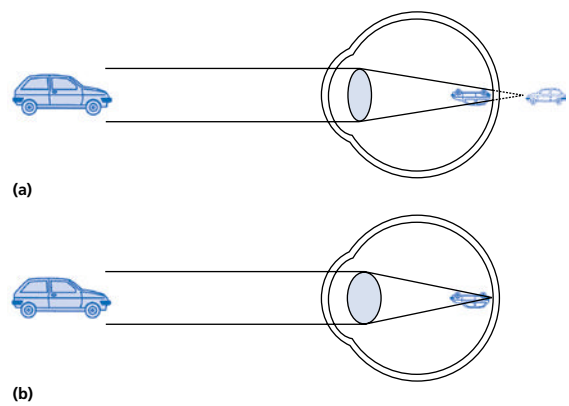

Figure 13.3 Hypermetropia (long-sightedness). (**a**) When the eye is relaxed, the image of a distant object is *focused behind* the retina, producing a blurred retinal image. (**b**) By accommodation (changing focal length), the image is focused on the retina.

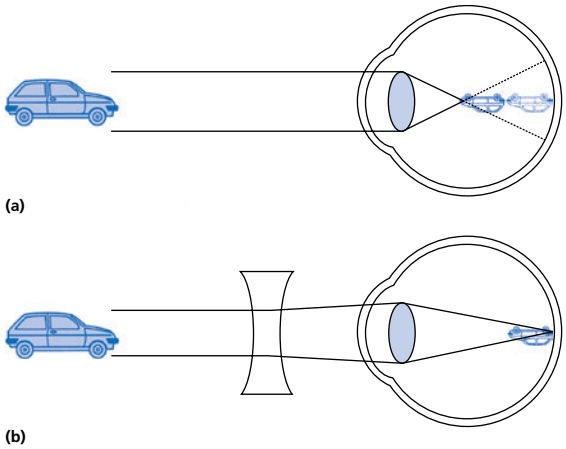

Figure 13.4 Myopia (short-sightedness). (**a**) When the eye is relaxed, the image of a distant object is focused in front of the retina, producing a blurred image. (**b**) Only by placing a diverging lens in front of the eye can a distant object be focused clearly on the retina.

Refractive errors can be compensated for by the use of glasses. In young children, refractive error can be measured accurately with the aid of cycloplegic eye drops and retinoscopy. This method of measuring refractive error is objective and requires minimal cooperation from the child. It can be done very easily on preverbal children to determine need for glasses. Focusing problems of all types are relatively common in childhood. During primary school years, approximately 5% of children wear glasses.

Most children are born a little hypermetropic, and because a child's prodigious accommodative capability can easily overcome this to see clearly, glasses are not necessary. If a child is excessively hypermetropic, large amounts of accommodative effort will be required to focus clearly. Such large amounts of accommodation may result in excessive convergence, and the child will develop a convergent squint (see section Strabismus).

Strabismus (turned eyes or squint)

Strabismus is one of the most common eye problems in childhood. Infrequently, the presence of strabismus indicates a major problem with one eye (e.g. cataract or retinoblastoma). More commonly, it is associated with reduced vision (amblyopia) in one eye. Thus, a turned eye may be secondary to poor vision or may be the cause of reduced vision (amblyopia). Early diagnosis of squint and appropriate intervention increases the chance of restoring or preserving vision.

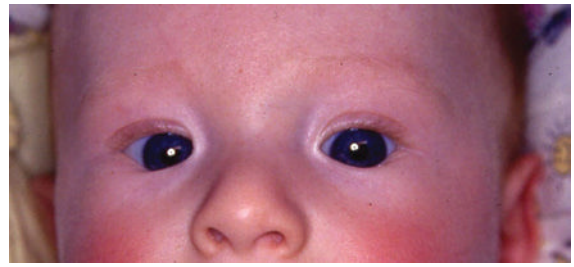

Figure 13.5 Pseudostrabismus. This infant has a prominent epicanthic fold on the right side, giving the appearance of strabismus. Note the sclera on the inner aspect of the right eye is not visible; this contributes to the appearance of misalignment. Cover testing failed to reveal any true strabismus.

The initial assessment of a child suspected of having strabismus involves confirmation of the misalignment (observation, corneal light reflection and cover testing), measurement of vision in each eye, and examination of red reflex to detect major structural defects in either eye. All children with confirmed or suspected strabismus should be referred to an ophthalmologist. If a major structural defect is suspected on the basis of an abnormal red reflex, specialist opinion should be obtained urgently.

Infants have relatively broad and flat nasal bridges, and if this is associated with prominent epicanthic folds, a very strong impression of convergent strabismus can arise. This is known as pseudostrabismus [Fig. 13.5] and is quite common. Careful assessment, as outlined earlier, will enable pseudostrabismus and true strabismus to be distinguished.

Most children with strabismus have a full range of eye movement, and thus, the misaligned eyes are not the result of a muscular abnormality or nerve damage. Childhood strabismus is usually the result of failure of development of normal binocular coordination of the eyes. There may be a primary failure of binocular coordination. This is seen with early onset convergent strabismus (known as infantile esotropia).

Strabismus associated with a full range of eye movement is called concomitant strabismus. If the eyes converge excessively it is known as esotropia, and if the eyes diverge it is called exotropia. Vertical misalignment is called hypertropia if the eye goes up and hypotropia if the eye goes down.

Infantile esotropia is seen before 6 months of age and is generally a large angle convergent strabismus

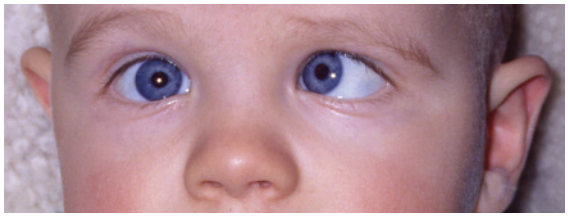

Figure 13.6 Infantile esotropia. This infant has a large angle alternating esotropia. Note the marked asymmetry of the corneal light reflections.

[Fig. 13.6]. These infants seldom have any significant refractive error, and surgery is usually required to realign the eyes successfully. Patching may be necessary for the treatment of amblyopia.

Accommodative esotropia usually has its onset between 18 months and 4 years of age and initially is often intermittent. It occurs in children who are excessively hypermetropic (long-sighted). To overcome hypermetropia and focus a clear image on the retina, accommodative effort is used. The near response consists of the combination of changing focal length of the lens (accommodation) and converging the eyes (so that both are directed at the nearer object of regard). Thus, in children with excessive hypermetropia, there is increased focusing and at times excessive convergence (causing accommodative esotropia). [Fig. 13.7]: this can be corrected by prescribing glasses that compensate for the appropriate amount of hypermetropia. Amblyopia is often seen with accommodative esotropia and requires treatment. If glasses only partly correct the esotropia, surgery may be indicated to obtain optimal alignment.

Intermittent divergent strabismus is unusual before 18 months of age. It is often more noticeable on distance fixation and may be associated with closure of the deviating eye, especially in bright light. Amblyopia is uncommon as the deviation is intermittent, and presumably when the eyes are straight, normal visual development proceeds. In some cases, the divergence becomes more constant, and in such situations, surgery may be undertaken to improve alignment.

All children with strabismus need review periodically until aged 10 years, to detect any amblyopia and to monitor ocular alignment. Even after successful realignment with surgery or glasses, amblyopia may occur and the eyes may also deviate again.

Children require a general anaesthetic (day surgery) for strabismus surgery to be undertaken. The principle

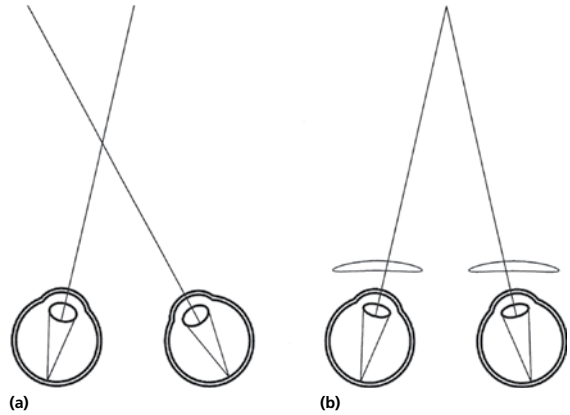

Figure 13.7 Accommodative esotropia. (**a**) Excessive accommodation of hypermetropic eye may result in excessive convergence and esotropia. (**b**) By placing corrective (plus) lens in front of eyes, the amount of accommodation needed to see the object is reduced and so is the convergence; hence, eyes are now correctly aligned.

of such an operation is to align the eyes correctly by weakening and/or strengthening extra ocular muscles so as to rotate one eye relative to the other. Simple analgesia, such as paracetamol, is sufficient for post-operative pain management. Post-operative nausea and vomiting is not unusual in school-age children (over 5 years of age). This is generally self-limited and not a major problem, settling with restriction of oral intake and a little patience.

Nystagmus (wobbly eyes)

Nystagmus is to-and-fro movement of the eyes and is involuntary. In childhood, most nystagmus is a primary defect in the child's ability to keep the eyes still, or the result of early failure of normal development of vision. Nystagmus in childhood is seldom the result of medication, cerebral degeneration, cerebrovascular disease or cerebral tumour as is the case in adults.

Congenital nystagmus is a primary defect in the mechanism that stabilises the eyes and results in mild to moderately reduced vision (6/9–6/24). It is seldom evident before 6–8 weeks of age, and will often reduce or dampen in a particular direction of gaze (called the null point). Thus, to maximise vision, a child may adopt a compensatory head posture to take advantage of the null point. If the compensatory head posture is extreme,

surgery may be necessary to manipulate the null point so that it is closer to the primary position of gaze.

Sensory nystagmus is caused by poor vision and is usually obvious by 3 months of age. Causes include bilateral congenital cataracts, albinism and congenital retinal dystrophy (Leber's congenital amaurosis). These conditions are rare, but prompt recognition will result in a better final visual outcome if treatment is possible (e.g. congenital cataracts). Early intervention will minimise developmental problems associated with lifelong reduced vision (e.g. in albinism and congenital retinal dystrophy).

Watery (epiphora) and sticky eyes

An eye will become watery and sticky because of failure of tear drainage or an inflammatory response (increased production of tears, mucus and cellular response). Congenital nasolacrimal duct obstruction and bacterial conjunctivitis are typical examples of each cause. It is possible that the two problems coexist in the same child. An irritated eye may be painful and erythematous, depending on the cause [Table 13.1].

Congenital nasolacrimal duct obstruction affects about 10% of newborn infants and resolves spontaneously in

Table 13.1 Watery and sticky eyes: Common symptoms, signs and causes

Problem	Symptoms	Signs
Neonatal conjunctivitis (*ophthalmia neonatorum*)	Severe pain	Moderate epiphora Copious discharge Moderate to severe erythema
Congenital nasolacrimal duct obstruction	Painless	Mild to moderate epiphora Mild to copious discharge Minimal erythema
Infantile glaucoma	Photophobia	Moderate epiphora No discharge Minimal erythema Enlarged and cloudy cornea
Viral conjunctivitis	Moderate discomfort	Moderate epiphora Mild discharge Mild to moderate erythema
Bacterial conjunctivitis	Moderate to severe discomfort	Moderate epiphora Copious discharge Moderate to severe erythema
Allergic conjunctivitis	Itch often prominent	Mild to moderate epiphora Stringy discharge Mild erythema
Chemical conjunctivitis	Intense pain	Severe epiphora Mild discharge Moderate to severe erythema
Corneal abrasion	Intense pain	Moderate epiphora No discharge Variable erythema Fluorescein staining
Foreign body	Intense pain	Moderate epiphora No discharge Variable erythema Variable fluoresce in staining
Preseptal cellulitis	Moderate pain	Minimal epiphora Variable discharge Marked erythema and swelling of eyelids – the eye is white
Orbital cellulitis	Severe pain Reduced eye movements	Minimal epiphora Variable discharge Marked erythema and swelling of eyelids – the eye is often inflamed and proptosed

about 95% by 1 year. It presents as a watery and sticky eye in the first weeks of life. Despite the persistent discharge, the eye is generally not red nor inflamed; thus, topical antibiotics are not indicated. The differential diagnosis includes trauma, conjunctivitis and infantile glaucoma (see section Big and small eyes). An inflamed eye suggests infective conjunctivitis.

If the obstruction persists, the lower lid will often become red and sometimes slightly scaly as a result of the skin being constantly moist. This irritation of the skin can be reduced by water-proofing the skin with soft white paraffin ointment. If obstruction persists beyond 12 months of age, probing under a general anaesthetic is indicated and is generally curative.

Ophthalmia neonatorum presents with copious discharge from the eyes in the first few days of life and is the result of infection acquired during birth (e.g. *Neisseria gonorrhoea* and *Chlamydia trachomatis*). Gonococcal conjunctivitis is serious because of the risk of spontaneous perforation of the cornea, loss of vision and generalised sepsis. Chlamydial conjunctivitis is important because of the risk of more generalised chlamydial sepsis. For accurate and prompt diagnosis, conjunctival swabs should be directly inoculated onto culture medium plates and conjunctival scrapings taken for Gram staining and immunofluorescent staining. Systemic, as well as topical, antibiotic therapy is indicated.

Bacterial conjunctivitis occurring beyond the first few days of life is generally the result of relatively innocuous organisms (e.g. *Staphylococcal* spp. and *Haemophilus* spp.). Microbiological investigation is not indicated initially, and a broad-spectrum topical antibiotic should be prescribed (such as soframycin or chloramphenicol).

Viral conjunctivitis is relatively common at all ages and may be very difficult to differentiate from bacterial conjunctivitis. There may be somewhat less discharge with viral conjunctivitis. When there is uncertainty as to aetiology, topical antibiotics as for bacterial conjunctivitis should be used.

Preseptal cellulitis is a bacterial infection of the skin and soft tissue of the eyelids and will present with redness and swelling. Often, there are discharge and watering as well. This infection may respond to oral antibiotics, but frequently, parenteral antibiotics are needed. Less commonly, an infection spreads to the orbital tissues from the surrounding nasal sinuses. This is orbital cellulitis, a more serious infection than preseptal cellulitis and presenting with proptosis (forward protrusion of the eye), redness of eye and eyelids and painful limitation of eye movements. If untreated, orbital cellulitis will frequently result in loss of vision because the raised pressure in the orbit will interfere with the blood supply to the globe, which in turn may lead to infarction of the optic nerve or retina. Treatment involves parenteral antibiotics, urgent CT scan to define the extent of any orbital abscess and drainage of any significant collection of pus.

Big and small eyes

A young child's eye will become bigger if the pressure within it is raised, as in infantile glaucoma. Small eyes in children result mainly from defects in growth of the eye, and there may be other major anomalies of the eye.

Infantile glaucoma (buphthalmos or *ox eye*) is a rare condition with deficient drainage of aqueous fluid from the anterior chamber: the intraocular pressure rises and the infant's sclera and cornea stretch and the eye enlarges. The stretching of the cornea damages the inner corneal layers (Descemet's membrane and associated endothelium).This allows the cornea to become oedematous and opaque. The damaged cornea causes irritation and light sensitivity. Thus, the features of infantile glaucoma are an enlarged, cloudy cornea with watering and photophobia [Fig. 13.8]. There is no significant discharge, which differentiates infantile glaucoma from nasolacrimal obstruction and conjunctivitis.

Treatment for infantile glaucoma is surgery to restore aqueous fluid drainage from the anterior chamber. Such surgery is usually successful, though the stretched

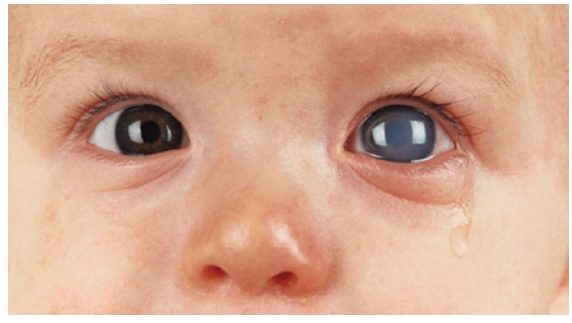

Figure 13.8 Infantile glaucoma presents with photophobia and tearing, and on examination, the affected eye(s) is enlarged and the cornea is cloudy. Compare the left and right eyes in this illustration.

cornea will remain and these eyes are often myopic (short-sighted).

An eye that is small but otherwise normal is termed a nanophthalmic eye. If the eye is associated with an ocular anomaly, the eye is microphthalmic. Microphthalmos is frequently associated with a failure of development of part of the uveal coat of the eye (iris and choroid). Such a defect in the iris or choroid is called a coloboma. Microphthalmic eyes often have poor vision that cannot be improved.

Injured eyes

Trauma to the eye can be physical (blunt or sharp), due to radiation (thermal and electromagnetic) or chemical.

Direct blunt trauma to the eye may disrupt iris blood vessels causing bleeding in the anterior chamber of the eye (hyphaema), tear the iris, dislocate the lens, rupture the choroid and (rarely) rupture the eye wall (sclera) if the force is sufficient. Simple inspection of the eye will reveal most of these injuries, and choroid and globe rupture may be suspected on the basis of the nature of the injury and associated poor vision. Referral to an ophthalmologist is necessary in these cases for confirmation of the injury and further management. The prognosis for vision is poor with severe injuries.

Blunt trauma to the eye may result in a blowout fracture of the bones of the orbital wall rather than rupture of the globe: the orbital floor and medial wall are most often fractured, as they are thin bones. The extraocular muscles and/or their fascial connections may become entrapped in a blowout fracture, leading to restrictive strabismus. Surgery may be needed to free the entrapped tissue and repair the fracture.

Sharp trauma may result from tiny objects, such as a subtarsal foreign body, causing a corneal abrasion, or finger nail scratches, through to penetration of the eye by sharp objects such as a scissors blade or knife. Surface trauma can be easily diagnosed with the help of fluorescein stain and a cobalt blue light. Areas of epithelial abrasion will fluoresce green. If a round ulcer and/or vertical linear abrasions are seen, a subtarsal foreign body should be suspected and the upper lid should be everted and foreign body removed with a moistened cotton bud. Superficial trauma is treated with antibiotic ointment and a patch, and daily review is needed until the ulcer or abrasion is healed.

In penetrating injuries of the eye (cornea or sclera), the intraocular contents may prolapse out through the wound, the iris and pupil may appear distorted, or the anterior chamber may be shallowed. Any suspected penetration of the eye must be referred to an ophthalmologist for further investigation and management. The eye should be protected with a cone that does not exert any pressure on the eye. If vomiting is likely or occurs, an anti-emetic should be given to reduce the chance of further prolapse of intraocular tissue.

Thermal injuries to the eye itself are rare as the eyelids protect the eye. Facial burns may cause scarring, which interferes with lid function, leading to exposure and drying of the eye's surface. If a primary thermal injury to the eye is suspected, fluorescein dye should be used to detect any ulceration. If ulceration is found, treatment is with antibiotic ointment and a patch.

Radiation injuries to the eye are rare in childhood, and most are the result of intentional irradiation as part of medical therapy for facial and ocular neoplasm. Typical injuries are cataract, dry eye syndrome, radiation retinopathy and optic neuropathy. These changes are seen some considerable time after the irradiation.

Chemical burns to the eye are unusual in childhood, but potentially very serious, especially if the chemical is alkaline. Many domestic cleaning agents are alkaline. Strong alkali will denature and dissolve protein and penetrate deeply into the surface of the eye. Acids tend to coagulate surface structures, and this often prevents deeper penetration of the acidic chemical into the eye. Immediate first aid consists of copious irrigation with water for at least 10 min. Local anaesthetic eye drops relieve pain while the eye is irrigated. All chemical burns of the eye should be referred to an ophthalmologist.

White pupil

The pupil is normally black because very little light is reflected back out of the eye. Any abnormal reflecting surface in the eye increases reflected light and causes the pupil to appear coloured rather than black. Cataracts, retinal tumours and chorioretinal colobomas are the most common causes of a white pupil [see Fig. 13.1]. These conditions are all rare but important because of their effect on vision and, in some

instances, the importance of early recognition and treatment.

A cataract is any opacity within the lens. Cataracts will frequently present because a white pupil has been noted. Bilateral congenital cataracts cause poor vision in infancy, while unilateral congenital cataract may go unrecognised as one eye has normal vision. Both bilateral and unilateral congenital cataracts are treatable if diagnosed early. Cataracts are readily detected by inspection of the red reflex with the direct ophthalmoscope.

Most cataracts in childhood are congenital and causes include heredity (dominant, recessive and X-linked), metabolic disorder (e.g. galactosaemia), association with systemic syndrome (e.g. Down syndrome) and congenital infection (e.g. rubella embryopathy). Many, especially unilateral cataracts, are idiopathic.

Management of cataracts in infants involves surgical removal of the cataract and visual rehabilitation with glasses or contact lenses. In older children, an intraocular lens can be implanted in the eye. These children often develop amblyopia and require long-term follow-up.

Retinoblastoma most often presents with a white pupil (the white tumour is seen immediately behind the lens) [Fig. 13.1]. Other presentations are with strabismus, poor vision or a known family history of retinoblastoma. Prompt recognition and treatment is vital to preserve vision and life.

Sporadic and hereditary forms of retinoblastoma are recognised. The sporadic form is the result of two separate mutations that negate the action of the retinoblastoma gene (*Rb gene*) within a single retinoblast cell and thus is always unilateral. The hereditary form arises when the first of these two mutations occurs within a germ cell (most often a sperm). The second mutation occurs within the retinoblast. As all retinoblasts descended from an affected germ cell have the first mutation, more than one retinoblastoma will usually develop, and hence, the hereditary form is often bilateral.

Treatment of retinoblastoma may involve removal of the eye (enucleation), chemotherapy (systemic, intraocular or intra-arterial injection), freezing of the tumour (cryotherapy), laser heating of the tumour (often combined with chemotherapy (thermochemotherapy) or irradiation (both external beam and a local implanted source of irradiation – plaque brachytherapy). Current 5-year survival is about 98%.

Lumpy eyelids

Swellings in the eyelids are common in childhood. Most are the result of minor infections, obstructed oil glands or bruising. Benign tumours occur occasionally and malignant tumours very rarely.

Lid infections are common in children, and most arise in the lash follicles (stye or hordeolum externum) and meibomian glands (hordeolum internum). Unless there is significant secondary erythema of the surrounding lid, topical and systemic antibiotics are not indicated. Occasionally, severe preseptal cellulitis will follow a focal lid infection, and systemic (often intravenous) antibiotics will be needed.

Chronic inflammation of a meibomian gland (chalazion) is generally chemical inflammation rather than infection and occurs when the gland contents escape into the lid following blockage of the duct. A chalazion will appear as a lump in the substance of the lid and is often not particularly inflamed in appearance. Topical antibiotics seldom hasten resolution. Warm compresses may give symptomatic relief and help drainage. Chalazia may persist for many months; some will discharge through the conjunctiva or the skin. On occasions, surgical drainage is indicated for a persistently inflamed and large chalazion.

Angular dermoids occur at inner or outer aspects of the upper lid [Fig. 16.7]. These are benign hamartomas that grow in proportion with the rest of the child. Rarely, direct trauma will cause rupture of a dermoid and significant inflammation will ensue. A deep extension necessitating extensive surgery more often occurs with medial angular dermoids. A CT scan should be undertaken before planned excision if a dermoid is firmly adherent to bone.

Infantile haemangiomas may involve the eyelid, skin or orbit. If the mass causes ptosis or compresses, the eye vision may be affected. Sight-threatening lesions are usually treated with oral propranolol and less frequently with systemic or intralesional corticosteroid.

Malignant tumours of the eyelid and orbit are rare. Rhabdomyosarcoma is the most frequent and presents with rapidly progressive (days to weeks) eyelid swelling and/or proptosis. The overlying skin may appear reddened, but other signs of acute inflammation (pain and fever) are absent. Management includes imaging to delineate site and extent of the lesion, incisional biopsy,

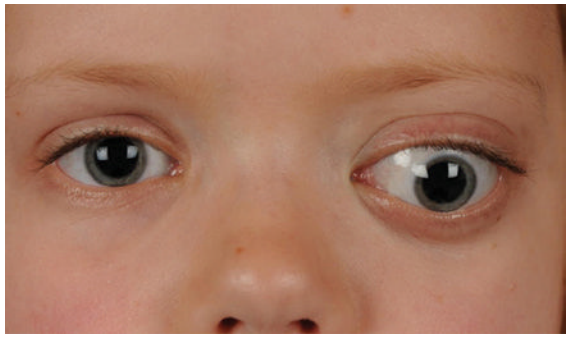

Figure 13.9 Neuroblastoma is a common cause of metastatic tumour of the orbit with proptosis. Note the protrusion and hypotropia (downward direction) of the left eye.

chemotherapy and often external beam irradiation. This management is multidisciplinary. The overall survival rate for this form of rhabdomyosarcoma is excellent. Metastatic orbital tumours occur, with neuroblastoma being the most common [Fig. 13.9].

Droopy lids

Ptosis (or blepharoptosis) is a droopy upper eyelid and results from innervational or muscular defects of the *levator superioris* or Muller's muscles. Innervational defects include third cranial nerve palsy, Horner syndrome (sympathetic nervous system) and myasthenia gravis. Most ptosis in childhood is congenital and has no other systemic associations. Acquired ptosis in childhood requires thorough investigation looking for a cause.

Congenital ptosis is usually an isolated abnormality in the function of one or both levator muscles. The affected muscle is often described as being *dystrophic*, but there is generally no association with more widespread muscular dystrophies. Congenital ptosis will appear worse when the child is tired or unwell; this is not evidence of ocular myasthenia gravis. A child with ptotic eyelids will often adopt a compensatory chin-up head posture to look straight ahead or to look up.

Ptosis will cause visual defects if the lid occludes the visual axis or if it induces astigmatism by altering the corneal curvature. Ptosis is also a cosmetic concern in that it may make an affected child look sleepy or dull. Surgical correction is possible in most cases. Early surgery is indicated when the ptotic eyelid is interfering with the development of vision. If intervention is primarily for reasons of appearance, surgery is usually undertaken just prior to school commencement.

Headache

Headache in children occasionally is caused by an ocular abnormality. Astigmatism and high hypermetropia (long-sightedness) are rare causes of childhood headache. Sustained attention to a near object will often cause some visual discomfort and headache. This is really a fatigue or tension headache and in general does not indicate any significant eye problem. These headaches are often described as a tightening around the head and are not associated with other symptoms.

Migraine headaches are common in childhood and may be associated with visual symptoms. The typical visual aura is blurring of central vision with zigzag bright lines (fortification spectra). These headaches are recurrent; they are often associated with nausea and/or vomiting, photophobia and phonophobia, and the child is almost invariably pale during the episode. Most settle with simple analgesia (paracetamol) and rest. A family history of migraine is common.

Eye examination may help in determining the cause of some headaches. Raised intracranial pressure will usually cause headache that is often worse in the morning and after lying down. The pain is often described as dull and pounding and persistent. It may be associated with nausea and vomiting and sometimes transient blurring of vision (visual obscurations). If the intracranial pressure is raised, fundus examination will reveal papilloedema in most cases. The most common causes of raised intracranial pressure that present with headache are *benign* intracranial pressure (pseudotumour cerebri) and tumours causing obstructive hydrocephalus.

Abnormal head posture

Children will adopt abnormal head postures for many reasons. Structural abnormalities, hearing loss, visual defects, habit and (rarely) central nervous system tumours can all cause abnormal head posture. Sternocleidomastoid muscle *tumour* and hemivertebra are examples of structural abnormalities causing abnormal head posture.

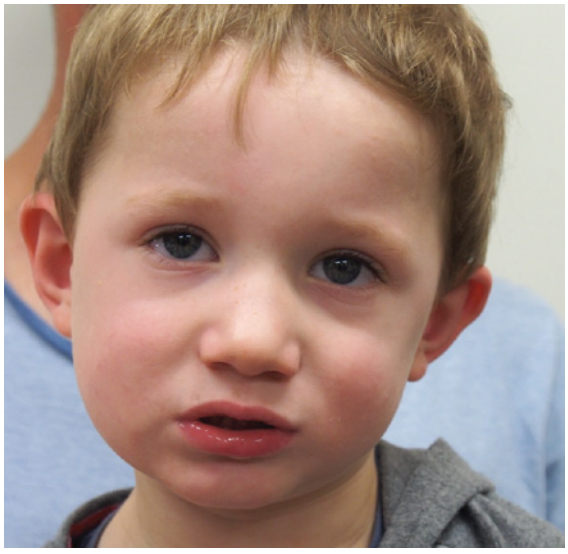

Figure 13.10 This child developed a head-tilt at 5 years of age. Investigation revealed a pineocytoma compressing the quadrigeminal plate.

If vision is improved with the head held in a particular position, a child will characteristically adopt this position [Fig. 13.10]. Visual stimuli for abnormal head posture include strabismus, where binocular depth perception is improved or diplopia is avoided, loss of vision in one eye, dampening of nystagmus, ptosis and severe photophobia.

Post-operative care of the child after eye surgery

The principles of post-operative eye care for children are (i) maintaining general comfort, (ii) protecting the eye when necessary, (iii) minimising specific complications and (iv) returning the child to normal activities as soon as is reasonable.

Pain is usually minimal after paediatric eye surgery. Paracetamol is sufficient for most post-operative pain relief. Immediately after strabismus procedures, children are often distressed and disoriented. This phase may last minutes to an hour or so and, in most instances, will settle with comforting alone. Older children are more likely to require post-operative analgesia than infants.

Nausea and vomiting may occur after strabismus procedures. Avoidance of pre- and intra-operative narcotic analgesia and intra-operative hydration and anti-emetic will minimise these problems. Limited oral intake in the early post-operative period will lessen the occurrence and severity of nausea and vomiting. Rarely, severe vomiting will occur, necessitating readmission and rehydration.

Patches to protect the eye are generally only needed after intraocular surgery (cataract and glaucoma) and eyelid surgery (ptosis repair and drainage of chalazion). Patches will often annoy a child and should be avoided following strabismus surgery.

Post-operative eye drops (antibiotic and steroid preparations) minimise the risk of infection and inflammation, especially following intraocular surgery. Benefit following strabismus procedures is less evident. Lubricating ointment is used for days to weeks after ptosis repair, to protect the ocular surface during healing.

Swimming should probably be avoided for 1–2 weeks after most eye operations because of the eye irritation from chlorinated or salt water. Care must be taken with face- and hair-washing, as soap or shampoo will be more irritating after an eye operation.

KEY POINTS

- Amblyopia may develop if squint and/or poor vision is untreated.
- A white reflex suggests significant pathology and needs urgent referral.
- Conjunctivitis in the first few days of life results from serious infections acquired during birth.

Further reading

Hoyt CS, Taylor D (2012) *Pediatric Ophthalmology and Strabismus*, 4th Edn. Saunders, Philadelphia.

South M, Issacs D (2012) *Practical Paediatrics*, 7th Edn. Churchill Livingstone, London.

Wright KW, Strube YNJ (2012) *Pediatric Ophthalmology and Strabismus*, 3rd Edn. Springer, Berlin.

Useful website

American Association of Pediatric Ophthalmology and Strabismus (for definition and description of eye terms and conditions): www.aapos.org

CHAPTER 14
The Ear, Nose and Throat

CASE 1

An 18-month-old girl with an upper respiratory tract infection for a week has been irritable for the past two days, particularly at night. She has had three ear infections over the past three months and is not talking. She presents with a temperature of 37.8°C and is mildly unwell. Otoscopy reveals opaque tympanic membranes. Her mother thinks she has another ear infection.

Q 1.1 Why is the tympanic membrane opaque?
Q 1.2 What is the management?
Q 1.3 Is hearing likely to be impaired in this child?

CASE 2

A 6-year-old boy presents with recurrent acute tonsillitis and has also been snoring heavily at night. His mother wonders whether or not it is time for him to have his tonsils and adenoids removed.

Q 2.1 What information would you seek regarding the episodes of acute tonsillitis?
Q 2.2 What information would you seek regarding the quality of sleep?

CASE 3

A concerned mother gives a history of her 4-week-old infant having stridor since birth. The child is noisy but appears well and is thriving.

Q 3.1 How would you assess this child?
Q 3.2 What are the possible causes of stridor in this child?

Otitis media

Introduction

The term otitis media describes the presence of a middle ear effusion. Fluid develops because the eustachian tube that normally provides middle ear ventilation, drainage and protection from nasopharyngeal contamination is dysfunctional. For both infective and structural reasons, otitis media is common in the first three years of life, particularly over the winter months.

The tympanic membrane is a window to the middle ear. A normal middle ear is air-containing, and the tympanic membrane therefore appears translucent and freely mobile. An opaque tympanic membrane may be due to fluid in the middle ear or simply thickening of the membrane itself [Fig. 14.1]. The diagnosis of otitis media may be confirmed by demonstrating impaired mobility of the tympanic membrane by either pneumatic otoscopy or tympanometry (type B tympanogram).

Otitis media presents clinically as a spectrum of diseases all characterised by presence of a middle ear effusion, but with varying degrees of bacterial infection. At either end of this spectrum, the child may be labelled clinically as having either

1. Acute suppurative otitis media (ASOM; bacteria cultured in 90% of cases) or
2. Otitis media with effusion (OME; bacteria cultured in 30% of cases)

Acute suppurative otitis media

The diagnosis of ASOM is based on the presence of a middle ear effusion together with features of inflammation that are either local (pain) or systemic (fever, irritability), provided there is no other explanation for the systemic symptoms. Bacteria are cultured in approximately 90%

Jones' Clinical Paediatric Surgery, Seventh Edition. Edited by John M. Hutson, Michael O'Brien, Spencer W. Beasley, Warwick J. Teague and Sebastian K. King.

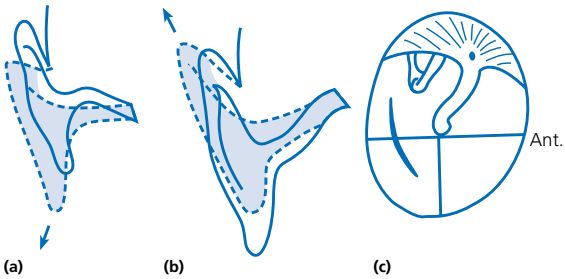

Figure 14.1 The tympanic membrane. (**a**) The direction of the canal in infants and young children: to visualise the membrane, pull the pinna downwards and outwards; (**b**) in older children and adults, the pinna is drawn upwards and backwards; (**c**) the normal eardrum is transparent, with the incus and stapes visible in the background. The site for myringotomy or ventilation tube is shown.

of cases, with the commonest organisms being *Streptococcus pneumoniae*, non-typeable *Haemophilus influenzae* and *Branhamella catarrhalis*. Despite this, most cases of ASOM will settle spontaneously *without* antibiotics, and treatment is not required if symptoms are only mild. Antibiotics should be administered in children under 12 months of age or if significant or prolonged symptoms are present; for example, amoxicillin (40 mg/kg/day in three divided doses for 5 days).

Spontaneous perforation with discharge may occur and is usually associated with relief of pain. The perforation invariably heals within days. Early follow-up of ASOM is generally only required if the child remains unwell after 48 h of antibiotic treatment. Acute drainage of the ear (myringotomy) is occasionally required if significant local or systemic symptoms persist despite adequate antibiotic treatment or if complications occur.

The middle ear effusion associated with ASOM persists for a variable period of time beyond resolution of the infective features [Table 14.1] and, while present, will be associated with some degree of impaired hearing in the involved ear.

Suppurative complications of ASOM are uncommon but need to be considered if systemic symptoms persist. The middle ear space and mastoid air cell system are contiguous. Therefore, any case of ASOM involves inflammation of both the middle ear and the mastoid. Suppurative complications may be confined to the temporal bone or may extend beyond into the central nervous system (CNS). Suppurative complications will generally require surgical drainage of the middle ear by

Table 14.1 Duration of effusion after ASOM

Period after ASOM	Percentage with middle ear effusion
2 weeks	70
1 month	40
2 months	20
3 months	10

insertion of a ventilation tube (grommet) with or without drainage of the mastoid. Intratemporal complications include

- Acute mastoiditis, which presents with postauricular swelling due to a subperiosteal abscess that has spread from the mastoid.
- Sigmoid sinus thrombosis, which presents with features of raised intracranial pressure due to impaired venous drainage. This occurs particularly with involvement of the right side, the usual side of the dominant sinus.
- Facial paralysis, due to inflammation extending to an exposed region of the facial nerve as it traverses the middle ear. This may occur at the time of initial presentation with ASOM.

Extratemporal suppurative complications with CNS involvement are rare, but may accompany intratemporal suppurative infections. These include extradural, subdural and cerebral abscess in the middle or posterior fossa.

Otitis media with effusion

Middle ear effusion is typically associated with mild hearing loss. When bilateral, this may lead to delayed speech development, behavioural problems and educational difficulties. Bacteria may be cultured in 30% of cases and, as a consequence, antibiotic treatment may be of some benefit.

Operative intervention with ventilation tube insertion provides external ventilation of the middle ear but does not correct the underlying eustachian tube dysfunction and is therefore only of benefit while the tubes remain *in situ*. Ventilation tubes should be considered where OME is associated with

- Hearing loss or recurrent ear infections associated with significant morbidity
- A situation that is likely to persist for a significant period of time

Factors that indicate OME will persist include the length of time the effusions have been present, seasonal factors

(OME is associated with upper respiratory tract infections in winter and spring) and anatomical factors such as cleft palate or other craniofacial anomalies.

Standard ventilation tubes usually last for 6–9 months. However, tube designs are available that may remain functional for years. Reinsertion of tubes is necessary in 25% of cases. The longer the tubes remain, the higher the rate of tympanic membrane perforation, which is approximately 1% per year. Tubes may discharge intermittently, particularly with upper respiratory tract infections, and infection is best treated by topical antibiotics.

Cholesteatoma

Cholesteatoma describes the presence of squamous epithelium (skin) in the middle ear. This squamous epithelium may spread throughout the middle ear cleft (middle ear, attic, mastoid) and cause bone destruction due to a combination of enzymatic action and infection. This may result in conductive hearing loss, sensorineural hearing loss, facial paralysis and/or intracranial infection.

Cholesteatoma may be congenital or acquired. Congenital cholesteatoma may be recognised in the early phase by the presence of a white mass deep to the tympanic membrane, usually in the anterosuperior quadrant.

Acquired cholesteatoma usually occurs due to localised tympanic membrane retraction associated with impaired middle ear ventilation as a result of eustachian tube dysfunction. The retraction may involve the tympanic membrane inferior to the lateral process of the malleus (pars tensa) or superior to it (pars flaccida). Development of a retraction pocket may be observed clinically, and cholesteatoma formation may be prevented by insertion of a tympanostomy tube if the retraction pocket is not yet attached to bone. Acquired cholesteatoma may also occur due to traumatic or iatrogenic (e.g. ventilation tube insertion) implantation of squamous epithelium into the middle ear or medial migration of squamous epithelium from the external auditory canal through a tympanic membrane perforation that abuts the external auditory canal. When extensive, removal of cholesteatoma usually requires a combined operative approach to both the mastoid and the middle ear.

Acute tonsillitis

Acute tonsillitis is most commonly viral in origin, especially in children less than 4 years of age. The clinical features include sore throat, fever and systemic upset, inflamed tonsils and cervical lymphadenitis. Bacterial tonsillitis occurs in 15–30% children with a *sore throat*, and is most commonly due to Group-A β-haemolytic streptococcus (GAS) infection. The presence of pharyngotonsillitis and tender tonsillar lymph nodes favours GAS infection and can be considered indications for empirical antibiotic therapy. This diagnosis needs to be differentiated from a viral upper respiratory tract infection or pharyngitis, and specific viral causes of acute tonsillitis, particularly infectious mononucleosis, which do not benefit from antibiotic prescription.

Bacterial infection may be confirmed by obtaining a throat swab. However, a positive culture must be interpreted in the light of streptococci being cultured from the surface of the tonsil in 10–15% of healthy carriers. Demonstrating a rising streptococcal antibody titre is of no value in acute diagnosis, but (anti-streptolysin O titre) ASOT levels may be useful to confirm that bacterial tonsillitis has occurred within the period of between 3 weeks and 3 months beforehand.

Rheumatic carditis and post-streptococcal glomerulonephritis are now rare complications of acute bacterial tonsillitis. Treatment of suspected bacterial tonsillitis with a 10-day course of penicillin aims to minimise morbidity and decrease suppurative complications.

Infection may extend beyond the tonsil as peritonsillar cellulitis or abscess (quinsy) [Box 14.1]. A peritonsillar abscess requires drainage, which may be performed under local anaesthetic in older children.

Tonsillectomy may be considered for recurrent attacks of acute tonsillitis where the history is of frequent infections characterised by significant morbidity, and the age of the child and pattern of infections is such that continuation for a number of years is expected. In short, the

Box 14.1 Signs of quinsy

- Increased systemic symptoms
- Drooling of saliva
- *Hot-potato* speech
- Trismus
- Unilateral bulging tonsil and soft palate
- Fluctuance on palpation

cumulative morbidity of acute tonsillitis is anticipated to be well in excess of that of tonsillectomy. Pain may persist for up to 2 weeks following tonsillectomy. Secondary haemorrhage occurs in 3% of cases, generally between 5 and 10 days after surgery. Most settle spontaneously, unlike immediate postoperative haemorrhage, which is best treated by return to the operating theatre for haemostasis.

Adenotonsillar hypertrophy

Symptomatic enlargement of the adenoids and/or tonsils is common in children; however, the tonsils and adenoids tend to involute by 8–10 years. Obstructive symptoms related to adenotonsillar hypertrophy (ATH) may range from simple snoring to severe obstructive sleep apnoea [Box 14.2]. Where parental observation of sleep pattern and the degree of sleep disturbance is uncertain, objective evidence may be obtained by polysomnography (sleep study).

The integrity of the upper airway during sleep and the severity of obstructive symptoms depend on

- Degree of obstruction by the adenoids and tonsils
- Negative pressure generated during inspiration that will collapse the airway
- Neuromuscular tone that serves to prevent airway collapse

In the presence of ATH, the decision as to whether to proceed to adenotonsillectomy depends on the degree and impact of sleep disturbance and the likelihood that the ATH will continue in the immediate future. Adenoidectomy alone, which has a significantly lower morbidity, should be sufficient when the tonsils are small and only the adenoids are enlarged. The adenoids may be visualised by flexible nasendoscopy performed under local anaesthesia or by a lateral radiograph of the nasopharynx. Adenoidectomy should not be performed in cleft palate because it may cause velopharyngeal incompetence with hypernasal speech.

Box 14.2 Symptoms of obstructive sleep apnoea

- Snoring
- Increased work of breathing
- Sleep disturbance with restlessness and waking
- Episodes of sleep apnoea
- Daytime tiredness
- Failure to thrive
- Cor pulmonale

Sinusitis

The clinical features of acute sinusitis merge with those of viral upper respiratory tract infection, with purulent rhinorrhoea, nasal obstruction, cough and low-grade fever. The diagnosis of acute sinusitis should be reserved for those cases with fever and purulent rhinorrhoea in excess of 4 days. Sinus CT scans tend to be unhelpful as they may be *abnormal* in up to half of otherwise healthy children.

The organisms causing acute sinusitis are the same as those causing ASOM, and a similar antibiotic regime is usually recommended, together with nasal decongestants.

Sinusitis may present with suppurative complications in the orbit and brain. Periorbital and orbital inflammation is usually a complication of acute sinusitis involving the ethmoids. Initially, there is periorbital cellulitis characterised by inflammation of the eyelids. If there is no response to antibiotics, infection may spread to orbital cellulitis with more significant systemic symptoms, chemosis, proptosis, ophthalmoplegia and decreased visual acuity. This is usually associated with a subperiosteal abscess along the medial wall of the orbit adjacent to the ethmoid sinus. Therefore, a CT scan must be performed in all cases of orbital cellulitis. Surgical drainage is required if an abscess is present.

CNS complications most commonly occur following frontal sinus infection and include extradural, subdural and frontal lobe abscesses and cavernous sinus thrombosis.

Congenital stridor

Assessment of the timing of stridor is of paramount importance in the evaluation of stridor. Inspiratory stridor suggests that obstruction is extrathoracic (larynx and cervical trachea), while expiratory stridor suggests an intrathoracic abnormality. Severity of the stridor may be judged by the presence of associated laboured breathing and retractions. Long-standing airway obstruction in young

Box 14.3 Causes of congenital stridor

Nose/nasopharynx
- Mucosal congestion
- Mid-nasal stenosis
- Choanal atresia

Oropharynx
- Glossoptosis (Pierre–Robin sequence)

Supraglottis
- Laryngomalacia

Glottis
- Bilateral vocal cord paralysis

Subglottis
- Acquired subglottic stenosis (post-intubation)
- Congenital subglottic stenosis
- Subglottic haemangioma

Trachea
- Tracheomalacia
  Extrinsic (vascular compression)
  Intrinsic (cartilage softening)
- Tracheal stenosis (complete tracheal rings)

children leads to failure to thrive due to the increased effort of breathing and poor feeding.

The airway may be examined awake by transnasal flexible laryngoscopy, which provides a view of the upper airway from the nose to the larynx. When no diagnosis has been made and the child has significant symptoms, bronchoscopy under general anaesthetic is required.

The causes of stridor may be classified by their location and whether they are structural or functional [Box 14.3]. Obstruction is usually functional in neonates, with the commonest cause being laryngomalacia, characterised by inspiratory collapse of the supraglottis. This presents with low-pitched vibratory inspiratory stridor that increases with increased inspiratory effort (e.g. feeding) and decreases with increased neuromuscular tone (e.g. crying). Intervention is generally not necessary as usually stridor improves by 6 months and resolves between 1 and 2 years.

Neonates are obligate nose-breathers for the first 3 months of life, and nasal obstruction will cause significant upper airway obstruction. A subglottic structural abnormality needs to be considered, particularly with a past history of endotracheal intubation. Stridor in the presence of cutaneous haemangioma may well be due to a subglottic haemangioma, especially if the cutaneous haemangioma is in the *beard* region of the face or neck.

Trauma

Fractured base of skull with temporal bone fracture

A fractured base of skull following a head injury may present with bleeding from the ear. This may also be associated with leakage of cerebrospinal fluid. Alternatively, blood or cerebrospinal fluid may collect behind an intact tympanic membrane. Temporal bone fractures may cause conductive hearing loss due to ossicular injury, sensorineural hearing loss and facial paralysis. Treatment is generally conservative.

Nasal trauma

Nasal trauma, even without a nasal fracture, may cause a septal haematoma recognised as a soft bulge associated with nasal obstruction. If it is not drained, pain and fever will develop as a septal abscess forms, which causes destruction of the septum and collapse of the nose.

The nose should be assessed for a cosmetic deformity due to bone displacement after the oedema has settled, around 5 days. Radiology is of no benefit as the decision to reduce the fracture is based on the cosmetic deformity. Fracture reduction should be performed within 10–14 days of the injury.

Oropharyngeal injury

Oropharyngeal injury typically occurs when a child falls with a stick in the mouth, damaging the palate or posterior pharyngeal wall. Initial assessment involves nasal endoscopy under local anaesthesia to assess the posterior pharyngeal wall, and lateral neck x-ray to detect any air in the retropharyngeal tissues, a retained foreign body and any associated cervical spine injury.

Hospital admission is required when there is
- Significant palatal laceration requiring repair.
- Ongoing bleeding.
- Inability to feed.
- Upper airway obstruction.
- Retropharyngeal injury with risk of retropharyngeal abscess.
- Possible internal carotid artery damage from a lateral injury behind the tonsil. Blunt trauma to this region is particularly dangerous as endothelial disruption may occur, causing carotid thrombosis.

Common conditions of the mouth

Mucus retention cysts

Goblet cells in the buccal mucosa may become blocked and form a pale, pedunculated retention cyst on the inner aspect of the lip, the gingivo-labial sulcus or inside the cheek. They sometimes discharge spontaneously, but usually annoy the patient, worry the parents and occasionally interfere with feeding. They are best removed if troublesome.

Ranula

A ranula is a larger sessile cyst in the floor of the mouth under the tongue, arising as an extravasation cyst of the sublingual salivary gland. It may become large enough to interfere with speech, swallowing and breathing, and may warrant an operation. This involves intraoral sublingual gland excision.

Tongue-tie

In tongue-tie, the lingual frenulum is short and may be attached to the very tip of the tongue. In a minority of cases, the tongue-tie may sufficiently impair neonatal breast feeding to cause significant maternal discomfort as well as poor neonatal weight gain. In this highly select neonatal group, tongue-tie division is likely to improve breast feeding. Interference with speech development is a common reason for parents to seek tongue-tie division. However, early or presumptive tongue-tie division does not protect against future speech difficulties. Children with tongue-tie and speech difficulties should be assessed first by a speech therapist as this may inform whether tongue-tie division can be expected to improve this child's specific difficulties. Other perceived or social indications for tongue-tie division include ability to *lick an ice-cream* or kiss, but these remain controversial.

In the neonate, tongue-ties can be safely divided with or without general anaesthesia. In infants or older children, operative division under general anaesthetic is recommended.

KEY POINTS

- The key to diagnosis of otitis media is identifying the presence of a middle ear effusion, which is best determined by assessment of tympanic membrane mobility.
- Otitis media is largely seasonal, and a conservative approach to ventilation tube insertion should be taken as summer approaches.
- The decision to proceed with adenotonsillectomy for recurrent tonsillitis should be based on knowledge of the natural history of tonsillitis and the pattern of tonsillitis in the child.
- Children who snore and have disturbed sleep should be evaluated for possible obstructive sleep apnoea.
- Inspiratory stridor suggests an extrathoracic cause, expiratory stridor an intrathoracic cause.

Further Reading

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CHAPTER 15
Cleft Lip, Palate and Craniofacial Anomalies

CASE 1

A term neonate has a small jaw, wide cleft palate and airway obstruction when supine.

Q 1.1 What is the diagnosis?

Q 1.2 How should this be managed in the next few hours, few days and in the long term?

CASE 2

A child is born with a unilateral cleft lip and palate.

Q 2.1 What is the risk of a sibling being born with a similar problem?

Q 2.2 What is the risk if a parent is also affected?

CASE 3

Ultrasonography at 16 weeks of gestation shows syndactyly of all digits of all limbs and significantly decreased anterior–posterior cranial dimensions.

Q 3.1 What is the most likely cause?

Q 3.2 What are the principles of management?

Q 3.3 Will the I.Q. be normal?

Cleft lip and palate

A cleft lip can involve not just the lip, but also the alveolus between the lateral incisor and the canine, and the anterior portion of the hard palate as far back as the incisive foramen. The cleft results from failure of mesoderm to merge between the frontonasal process and the maxillary process of the first branchial arch between 4 and 7 weeks of gestation.

The *secondary* palate forms the hard and soft palate behind the incisive foramen. Palatal clefts are caused by failure of fusion of the two hemipalatal shelves between 7 and 10 weeks of gestation. A cleft can involve just the lip, just the palate or a combination of lip and palate.

Incidence, aetiology and risk

Congenital clefts of the lip and palate are common malformations, occurring in approximately 1 in 600 live births. A cleft lip, with or without cleft palate (CL±P), makes up about 70% of patients and is seen more commonly in boys. CL±P is more commonly found in Asians and least frequently in Africans. Cleft palate (CP) alone accounts for 30% of cases, is more common in girls, has no racial differences and is a different clinical group from CL±P. Clefts are inherited in a multifactorial, polygenic way. There is a positive family history in 25% of cases, particularly CL±P. Where one child already has a cleft, the risk of the second being affected is 2% for cleft palate and 4% for cleft lip and palate. Also, if the parent has a cleft, the risk for their first child is 4%. The risk increases to 16% if the parent has another affected child.

Other congenital anomalies should be looked for as they occur in up to 30% of patients and might be life-threatening. One of these is the Pierre–Robin sequence which may be present in those with clefts of the secondary palate. It consists of a small lower jaw (micrognathia), a cleft palate which is usually wide and U-shaped, and the tendency for the base of the tongue to be positioned posteriorly and fall backwards causing obstruction (glossoptosis). The sequence is thought to

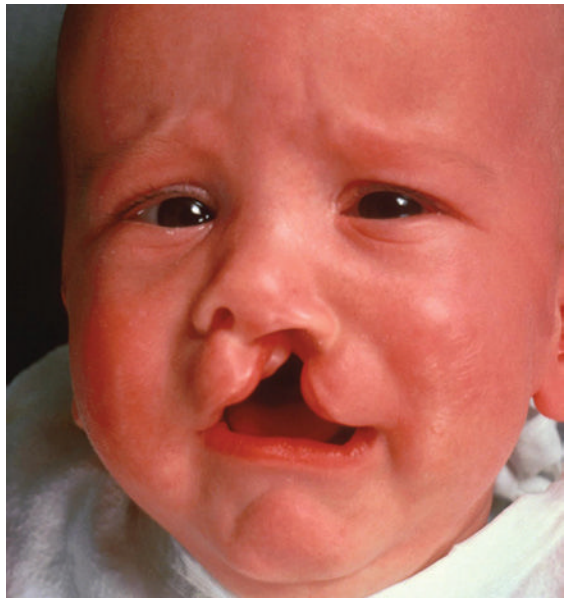
Jones' Clinical Paediatric Surgery, Seventh Edition. Edited by John M. Hutson, Michael O'Brien, Spencer W. Beasley, Warwick J. Teague and Sebastian K. King.

be caused by early underdevelopment of the mandible causing elevation of the tongue that prevents fusion of the palatal shelves around 10 weeks of gestation. These patients may have early feeding difficulties, failure to thrive and apnoeic episodes.

A submucous cleft palate is often overlooked. In these patients, the uvula is bifid and the soft palate is grooved in the midline where there is a cleft in the muscle. There is also a palpable notch in the posterior margin of the hard palate. These patients require careful assessment and usually need surgical repair similar to an overt cleft palate. Isolated cleft of the uvula occurs in 1 in 80 Caucasians and 1 in 10 Asians and is asymptomatic.

Classification

Clefts of the lip may be incomplete, complete, unilateral or bilateral [Fig. 15.1], and the latter may be asymmetrical. Two-thirds also have a cleft of the secondary palate involving both the hard and the soft palate posterior to the incisor foramen.



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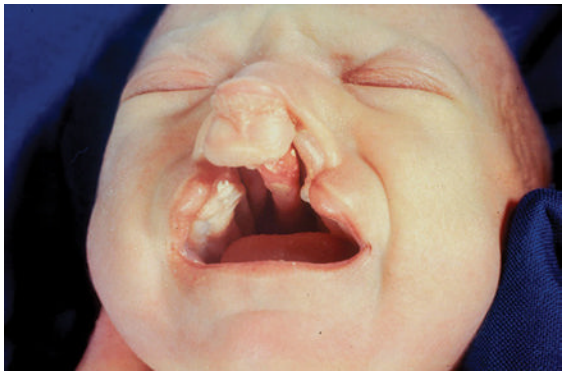

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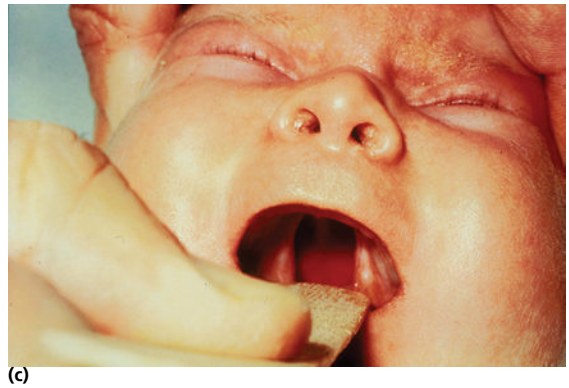

(c)

Figure 15.1 Classification of cleft lip and palate: (**a**) Left unilateral incomplete cleft lip involving the nose, lip, alveolus and primary palate; (**b**) bilateral complete cleft lip and palate; (**c**) isolated complete cleft of secondary palate.

Figure 15.2 Infant feeding bottle for patients with cleft palate (the infant has had cleft lip repair also).

Management

Major clefts of the lip and palate are usually diagnosed prenatally by ultrasonography, so parents can be counselled by a plastic surgeon and geneticist prior to birth. Prenatal explanation and support has made the management far easier than when the diagnosis was only made at birth.

After birth, there is an early referral to the plastic surgical team who coordinate the patient's cleft care. The deformity affects not only the patient's appearance, but also early feeding, hearing, speech, dental and maxillofacial development. The cleft-lip-and-palate team includes specialists from many disciplines and the care requires coordination.

Children with the Pierre–Robin sequence need early careful observation. Difficulty feeding, failure to thrive or apnoeic episodes require admission to neonatal intensive care. Babies who are mildly affected can often be managed conservatively with positional nursing alone until they are more mature. Others may require a nasopharyngeal airway and even nasogastric feeding. More severe cases may need infant mandibular lengthening with distraction osteogenesis. Operations to pull the tongue forward are now rare. Tracheostomies are avoided if at all possible.

Feeding

Sucking is often not difficult for the baby with a small incomplete cleft lip alone. Even though the *Orbicularis Oris* sphincter is incomplete, babies can often make a lip seal around the nipple. However, babies with cleft palate are unable to close the nasal cavity off from the pharyngeal cavity and cannot generate enough negative intraoral pressure to suck. These children need to be fed with a squeeze (or gravity-feed) bottle [Fig. 15.2] which delivers the milk to the back of the tongue. Once this is done, babies can swallow normally. There are a number of specially designed teats which deliver milk as they are compressed in the mouth. Parents are educated and helped to find the best feeding regime that suits their particular child.

Cleft lip repair

Repair of the lip is usually performed when the baby is about 2 months old (or 5 kg weight) and the distorted nose repaired at the same time. In patients with severe bilateral or wide unilateral clefts, presurgical manipulation of the maxillary segments is often attempted by the orthodontist. This makes the surgery easier and often improves the final result. The aim of lip repair is to definitively close all components of the lip, in particular the *Orbicularis Oris* muscle sphincter. The anterior nasal floor is closed and, if conditions are favourable, the anterior palate may be closed at the initial surgery as well [Fig. 15.3]. With the nose, the distorted lower alar cartilages are properly repositioned.

Cleft palate repair

Clefts of the secondary palate are usually repaired between 6 and 9 months of age (about 8 kg weight) prior to acquisition of speech. Unnecessary delay may affect the prognosis for normal speech as the child develops compensatory speech patterns that are difficult to correct later with speech therapy. The aim of the procedure is to create a palatal mechanism which separates the oral and nasal cavities during speech and swallowing. This usually occurs by elevating the soft palate against a

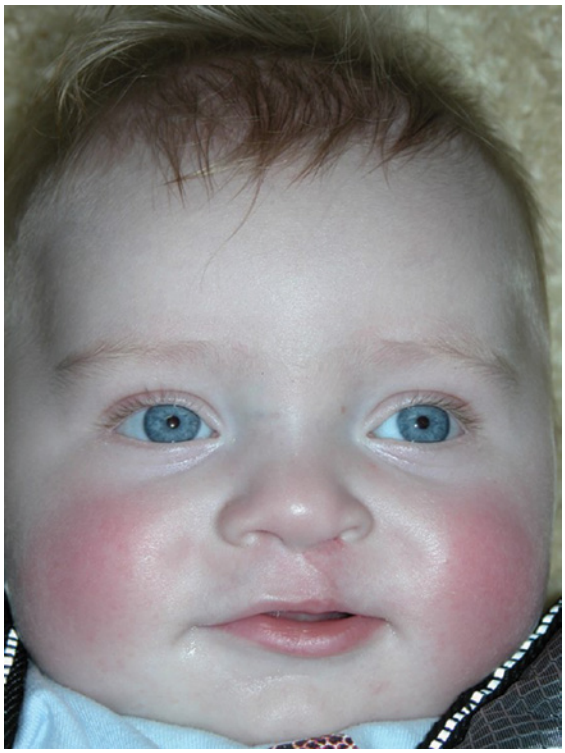

Figure 15.3 A repair of cleft lip.

constricting posterior pharyngeal wall. The key to the repair is accurate dissection and realignment of the cleft *Levator Palatini* muscles of the soft palate, usually involving an operating microscope.

Speech

Approximately 90% of patients with cleft palate will achieve normal or acceptable, intelligible speech. Speech should be assessed periodically following surgery. Speech therapy will benefit patients with articulation problems, or those who use compensatory mechanisms to produce certain sounds. Failed closure of the soft palate to the pharynx (velopharyngeal incompetence) allows nasal escape of air during speech. Significant velopharyngeal incompetence may require further surgery with lengthening of the palate, posterior repositioning of the Levator muscles and possibly adding local tissue flaps to the posterior palate or the pharyngeal wall (pharyngoplasty). Pharyngoplasty is more likely in patients whose cleft palate repairs were delayed and those who had submucous cleft palates where the diagnosis was initially missed. It is also more likely for children with velopharyngeal incompetence without a cleft but neuromuscular weakness of the palatopharyngeal sphincter.

Problems in the ear, nose and throat

Children with a cleft palate often have abnormal eustachian tube drainage leading to a high incidence of middle ear mucus build-up and possible otitis media. Some patients will require tympanostomy tubes (grommets) for middle ear ventilation. The mucus build-up decreases with age and with palate repair. However, hearing may be impaired early on, and cleft patients require frequent otoscopic and audiological evaluation.

Tonsils and adenoids occupy a significant space in the pharynx. Their removal is discouraged in cleft patients, except when there are severe recurrent infections.

Dental and orthodontic treatment

Virtually all children with cleft lip and/or palate will require orthodontic treatment in the long term. Early dental care and hygiene is most important. Adult teeth cannot erupt without the presence of bone. The cleft alveolus usually is bone-grafted between 8 and 12 years to allow the proper eruption of the adult canine tooth. Supernumerary or abnormal teeth are seen quite often in cleft patients and may need removal.

When growth is complete, some patients require orthognathic surgery to re-position the hypoplastic maxilla and to obtain a functional and aesthetic dental occlusion.

Secondary surgery

Secondary surgery may be required to repair any bad scars or asymmetries prior to school. However, with good primary repairs, this is seldom necessary. The nasal deformity has often been dealt with at primary surgery and does not need adjustment for many years.

However, with pubertal growth, the peri-cleft structures often do not have the same potential for development as the normal opposite side. In addition, with puberty, inherited familial characteristics develop, which may accentuate cleft deformities (such as a big nose). At this stage, secondary lip and nose revisions are common. If the upper lip philtrum appears too short or hypoplastic, it can be rebuilt utilising a tissue flap from the central lower lip (Abbé flap). Orthognathic surgery for final jaw aesthetics and dental occlusion is usually completed after facial growth (about 17 years of age for girls and 19 years for boys).

Table 15.1 Craniofacial deformity – a classification

Congenital
Clefting disorders
Cleft lip and palate
Major craniofacial clefts (Tessier classification including Treacher Collins syndrome)
Bizarre facial clefts (amniotic bands)
Craniosynostosis (premature fusion of craniofacial bone sutures)
Non-syndromic (usually single suture)
Syndromic (usually autosomal dominant fibroblast growth factor receptor gene mutations (e.g. Apert, Crouzon)
Encephaloceles (herniation of the CNS beyond the cranial cavity)
Failure of anterior neural tube closure
Some associated with severe major clefting disorders
Microsomias (dysplastic underdevelopment of first and second branchial arch and related structures)
Usually unilateral; 15% bilateral
Disorders of bone growth
Fibrous dysplasia; craniometaphyseal dysplasia; neurofibromatosis etc.
Acquired
Tumour (sarcomas etc.)
Trauma
Disorders of growth (e.g. prognathism; condylar hyperplasia)

Craniofacial anomalies

Facial appearance is to a large extent determined by the underlying bony skeleton [Table 15.1]. It is now possible to elevate the soft tissues of the face and orbits in a subperiosteal plane. Similarly, intracranially the dura can be separated from the skull. This provides a safe method to osteotomise, reshape and reconstruct the craniofacial skeleton. The same techniques can be utilised for congenital deformities, resection of tumours with reconstruction or craniofacial trauma. The principles of exposure and correction are the same, but the timing may differ.

Congenital craniofacial deformities

Principles of dysmorphology

Congenital anomalies may result from malformations, deformations or disruptions. Craniofacial *malformations* are the result of abnormal development, for example, craniosynostosis and clefts of the lip and face. *Deformations* result from extrinsic compression *in utero* or in infancy, for example, deformational plagiocephaly. *Disruptions* result from an extrinsic disruptive intrauterine mechanical force, for example, amniotic bands producing bizarre facial clefts (fitting no particular pattern) and constriction ring syndrome.

Cranial growth

The cranial sutures are not centres of growth, but rather *gaps* that allow the cranial bones to be pushed out by the growing brain. Bony growth occurs secondarily by deposition of bone at the sutures (sutural growth) and at the pericranial surface (appositional growth), as well as absorption of bone from the dural surface. Pathological fusion of a suture restricts growth perpendicular to the suture, with compensatory growth occurring in other non-restricted areas of open sutures.

Growth of the brain and its surrounding cranium is rapid in the first two years of life, reaching half adult size by 9 months and three-quarters by 2 years. In cranial suture synostosis, the ensuing deformity will worsen with ongoing cranial growth in the first two years. If more than one suture is involved, the volume of the cranium or orbital cavities may be restricted with secondary effects on the brain or eyes. In contrast, in deformational plagiocephaly with an otherwise normal skull and normal sutures, once the deforming force is removed, growth of the brain may be expected to improve the deformity without surgical correction. Disruptions, such as bizarre facial clefts, are not likely to be modified by further growth.

Craniosynostosis

Craniosynostosis is premature fusion of one or more cranial sutures [Table 15.2]. The commonest form is fusion of a single suture and no associated syndrome. The suture involved may be unicoronal or bicoronal, metopic, sagittal or, rarely, lambdoid. Each one gives a different characteristic shape to the skull due to lack of growth at right angles to the fused suture; for example, a fused sagittal suture produces a long, narrow *scaphoid* cranium. With non-syndromic sutural synostosis, it is uncommon to have generalised raised intracranial pressure (<15%). In general, the more sutures that are fused in the skull, the greater the chance of raised pressure. Rare cases can occur following rapid decompression of hydrocephalus, or with maternal Epilim ingestion during pregnancy (foetal valproate syndrome), thyrotoxicosis and rickets. If the fused suture is on one side of the head, it grows asymmetrically, leading to plagiocephaly (crooked head). Plagiocephaly can also occur due to uterine deformational processes without synostosis. Deformational plagiocephaly tends to improve with ongoing cranial

Table 15.2 Craniofacial deformity: a classification by head shape

Descriptive terminology	Observations	Possible causes
Plagiocephaly	Crooked or twisted head	• Unilateral coronal synostosis • Lambdoid synostosis (rare) • Intrauterine deformational forces • Torticollis
Scaphocephaly	Long *boat-shaped* head	Sagittal suture synostosis
Trigonocephaly	Triangular, *bow-sprit* forehead	Metopic (frontal) suture synostosis
Turricephaly	Tower-like forehead	• Fusion of sutures around anterior fontanelle (metopic, coronal and sagittal) • Some syndromes: Crouzon, Apert, Pfeiffer
Brachycephaly	Short head (A–P length)	Bicoronal synostosis (syndromic and non-syndromic)
Kleeblatschadel	Cloverleaf skull	Multiple cranial suture synostoses (some severe: Pfeiffer, Crouzon, Apert)
Orbital hypertelorism (Teleorbitism)	Orbits too far apart	• Encephaloceles, median and paramedian • Craniofacial clefts • Some craniosynostoses

growth once the deforming forces are removed, but also can be perpetuated by the baby habitually sleeping in a particular position [Figs. 15.4 and 15.5].

Syndromic craniosynostoses are less common but are usually autosomal-dominant genetic inheritance. The majority of the syndromes have been shown to involve mutations of fibroblast growth factor receptor genes (FGFR 1, 2 and 3). Crouzon, Apert and Muenke syndromes fall into this pattern. Bicoronal synostosis with midface hypoplasia occurs in Crouzon. In Apert syndrome, the synostosis is more severe and associated with complex syndactyly of the hands and feet. Mental delays in development and the chances of raised intracranial pressure are high. There are many craniosynostosis syndromes, and, as the knowledge of genetic mutations is increasing, more correlation with the specific phenotypic clinical expression is expected. Most syndromic synostoses have associated peripheral limb abnormalities. The incidence of raised intracranial pressure, primary brain anomalies and cervical spine anomalies is also increased.

Craniofacial microsomia

Craniofacial microsomia is the most common encompassing term to describe a spectrum of conditions such as hemifacial microsomia, first and second branchial arch syndrome, Goldenhar syndrome, oral-mandibular-auricular syndrome, oculoauriculovertebral syndrome, etc.

Common to all these conditions, there is variable hypoplasia or dysplasia of structures derived from the first and second branchial arches. Usually, there is underdevelopment of the maxilla, mandible and temporomandibular joint. In more severe cases, this can be associated with cleft lip and palate and even lateral macrostomia. Epibulbar dermoids may occur with Goldenhar syndrome, and there may be cranial nerve weaknesses, especially the facial nerve. Preauricular skin tags, sinuses and external ear deformities are a common manifestation, and severe cases may have plagiocephaly and orbital dystopia. Craniofacial microsomias are usually unilateral, but 15% are bilateral.

Encephaloceles

Encephaloceles are congenital herniations of the central nervous system beyond the cranial cavity (see Chapter 12). Sincipital (anterior) encephaloceles usually exit the skull between cranial bone junctions (e.g. fronto-ethmoidal) and are subclassified by where they exit onto the face. A second group can occur in association with severe major facial clefting. Encephaloceles may cause secondary craniofacial deformities such as hypertelorism of the orbits, trigonocephaly and orbital dystopia. About 25% of children with encephalocele have other central nerve system abnormalities.

Facial clefts

Facial clefting is basically classified into three categories: cleft lip and palate (the major common group), major Tessier clefts (relatively rare) and bizarre facial clefting (extremely rare).

The clefting process may range from some mild notching to a complete absence of tissue with an associated

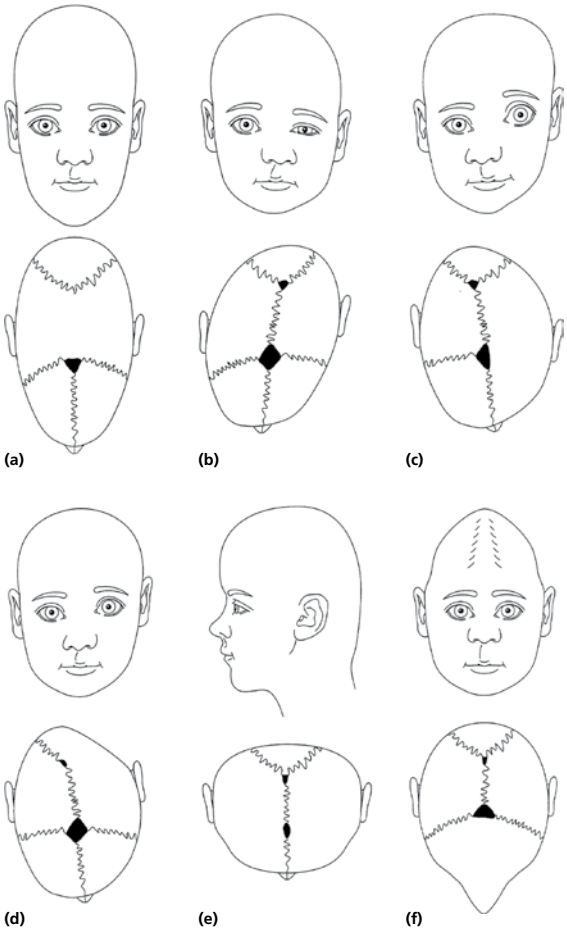

Figure 15.4 Descriptive names are applied to various head shapes which do not necessarily reflect the aetiology. (**a**) Scaphocephaly (*boat-shaped*) with fusion of the sagittal suture. (**b–d**) Plagiocephaly (*crooked head*) may be due to deformation (b), unilateral coronal synostosis (c) or unilateral lambdoid synostosis (d). (**e**) Brachycephaly (*short head*) is usually due to bicoronal synostosis, and often seen with Crouzon or Apert syndrome. (**f**) Trigonocephaly (*triangular forehead*) is seen with isolated metopic suture synostosis.

encephalocele. Very mild clefts may show subtle signs such as defects in the eyebrows or a widow's peak in the hairline. Tessier's classification [Fig. 15.6] is very useful, and the clefts are numbered anticlockwise around the orbit and clockwise around the mouth. The commonest Tessier cleft is midline (0–14 type). This can vary from a mild bifidity of the nose to a complete facial cleft with an encephalocele.

Bizarre facial clefts fit no particular pattern and are thought to be due to amniotic bands in the uterus.

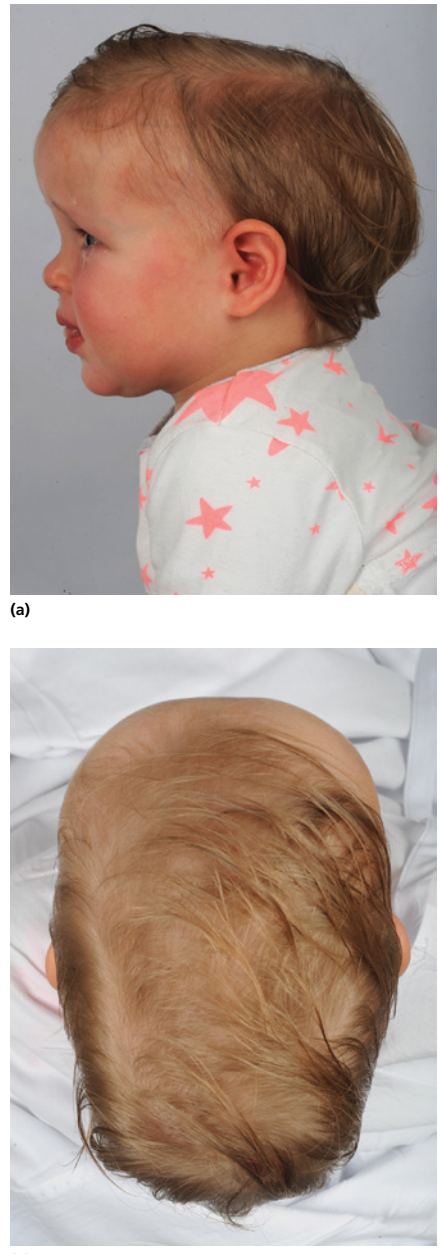

Figure 15.5 Scaphocephaly, shown from the side and above, showing the elongation of the cranium caused by synostosis of the sagittal suture.

Craniofacial neoplasia

Neoplasia is rare in the craniofacial region in infants. Benign lesions include fibro-osseous lesions and dermoids. Neoplasms are best treated with a wide resection

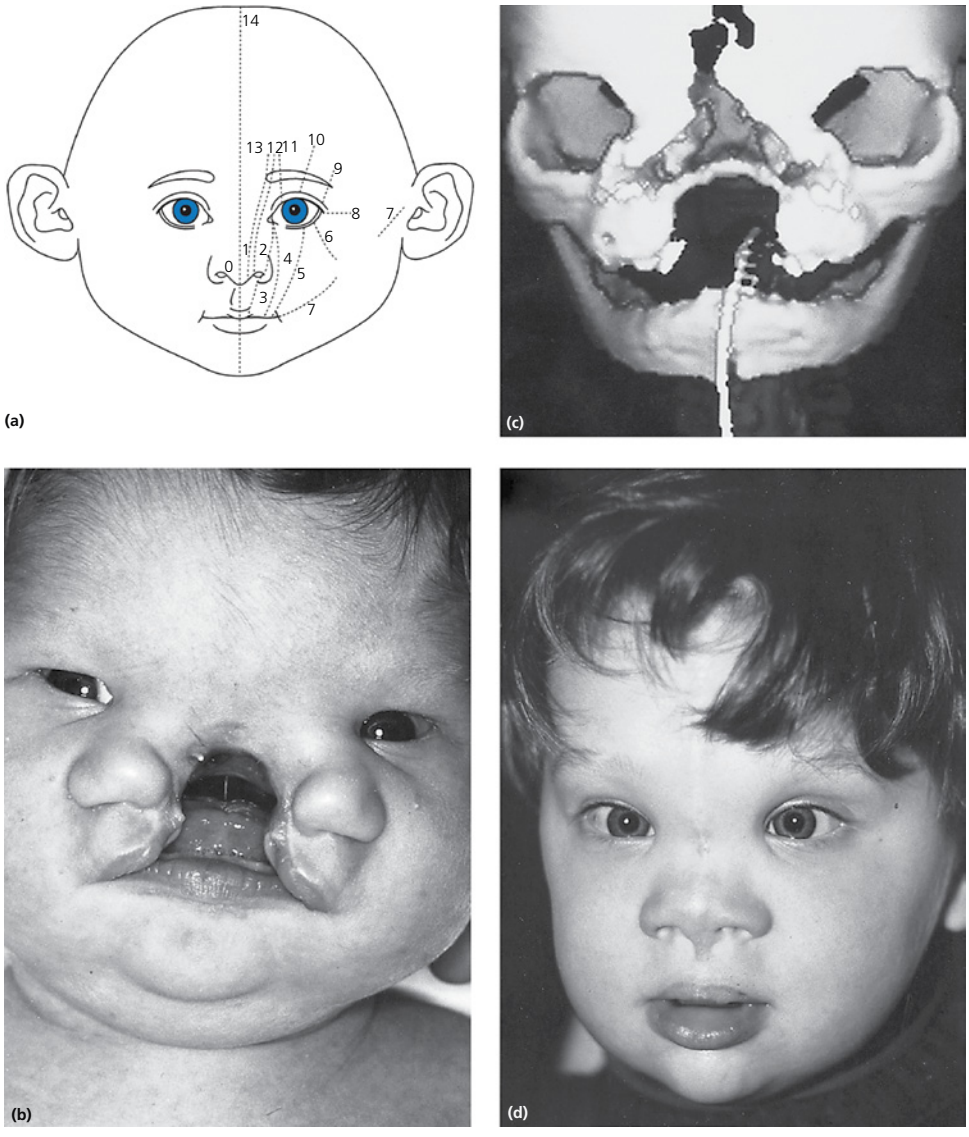

Figure 15.6 (**a**) The Tessier classification of craniofacial clefts. (**b**) A patient with a severe 0–14 midline Tessier cleft. (**c**) 3-D CT scan of the patient (with endotracheal tube *in situ*). The bony cleft continues in the 14 position and hypertelorism of the orbits is demonstrated. (**d**) Postoperative appearance after complete mobilisation of the orbits and maxillae with midline closure.

and reconstruction to prevent local recurrence and secondary growth deforming effects. Utilising craniofacial principles, this can be achieved with little deformity to the patient. Ewing's sarcoma, for example, is treated with induction chemotherapy until maximum reduction in tumour size is achieved. This is followed by craniofacial resection of the remaining tumour mass and reconstruction, prior to maintenance chemotherapy, usually with excellent results. Radiotherapy is avoided in children where possible; it is frequently associated with deforming growth restriction, as well as possible late induction of new tumours.

Trauma

Extensive craniofacial trauma can occur in children. It often results from horse-kick injuries, falls from balconies, bungy-jumping injuries and motor vehicle accidents. Complex fractures are best treated primarily

utilising craniofacial techniques, with accurate reduction of fractures, rigid fixation and primary bone grafts as required; the soft tissues are then meticulously repaired and re-suspended to the repaired framework. Secondary deformities, such as enophthalmos, orbital dystopia and subsequent deformities relating to growth (e.g. failure to develop a frontal sinus unilaterally due to a fracture of the fronto-orbital region in childhood), are also best addressed later with craniofacial techniques.

Management of craniofacial anomalies

Conservative management

Children with deformational craniofacial malformations usually improve slowly after birth, when the deforming stimuli cease (e.g. deformational plagiocephaly and torticollis). However, the spontaneous improvement is often slow, and diagnostic difficulties may arise. Often x-rays and an expert opinion are necessary to exclude synostosis. With deformational plagiocephaly, mild cases will usually return to normal during childhood. More severe cases will take until puberty to improve. Helmet moulding therapy can speed up the process in early infancy (before 9 months). Rarely, extremely severe cases of deformational plagiocephaly may require surgery.

KEY POINTS

- Clefts of lip and palate are usually diagnosed antenatally, but if not, need immediate neonatal referral to the plastic surgical team who coordinate management.
- Babies with cleft lip alone can usually be breastfed.
- Cleft lip is repaired at about 2 months of age (5 kg).
- Cleft palate is repaired at about 6–9 months of age (8 kg), before speech develops.
- Cleft lip and palate require team management throughout childhood for ENT, dental, orthodontic, speech and cosmetic management.
- Craniofacial anomalies need early tertiary referral for multidisciplinary team management and often early surgery in the first year.

Operative management

Patients with craniosynostosis require surgery. This is usually undertaken in the first year of life so that the continuing rapid growth of the brain can be utilised to help attain a normal long-term head shape. The principle of surgery is to remove the fused suture(s) and to rebuild the affected area of the cranial vault to where it should have been had the synostosis not occurred. If the craniosynostosis is severe and associated with raised intracranial pressure, then surgery is urgent. Children with tumours and encephaloceles are often operated on early so as to remove the expanding force of the lesions and return the craniofacial skeleton to a normal configuration.

Most children with craniofacial deformities require monitoring and treatment until their growth ceases as late teenagers. Periodic adjustment surgery may be required during that time and not finalised until adulthood.

Further Reading

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CHAPTER 16
Abnormalities of the Neck and Face

CASE 1

A 2-year-old was well until 2 days ago when he developed fever and a tender lump under the right side of his jaw.
Q 1.1 What is the differential diagnosis?
Q 1.2 Is an operation required, and if so, when?

CASE 2

Anita was first noticed to have a small, hard lump on her eyebrow at 6 months of age. Since then, it has grown gradually larger.
Q 2.1 What is the lesion and how is it treated?

CASE 3

A toddler is noticed to have a lump under his chin in the midline. It varies in size a little and recently became red and sore.
Q 3.1 What is the differential diagnosis of midline neck lumps?
Q 3.2 What treatment does he need?

The neck is a common site for cystic and solid swellings during childhood. There are developmental anomalies (arising from remnants of the branchial arches, the thyroglossal tract, the jugular lymphatics or the skin) or acquired diseases of the lymph nodes, salivary glands or the thyroid gland [Table 16.1].

Branchial cysts, sinuses, fistulae and other remnants

These arise from the branchial arch system. Persisting branchial clefts between the arches may give rise, in order of likelihood, to a branchial sinus (blind-ending tract), branchial fistula (communication between two epithelial-lined surfaces) or branchial cyst. The branchial arch itself may give rise to aberrant mesodermal remnants, usually cartilaginous, lying along the line of development of the arch.

Sinuses and fistulae most commonly arise from the second branchial cleft, occasionally from the first and rarely from the third branchial cleft. Each cleft fistula tract relates in its path to the nerve and artery derived from the next cleft in number ascending order. Thus, *second* branchial cleft fistulae commence in the tonsillar fossa and pass with the *third* cleft-derived glossopharyngeal nerve between the internal and external carotid arteries to end in the skin at the anterior border of the lower third of the sternomastoid muscle. First cleft fistulae run from the external auditory canal to the skin below the lower border of the mandible. The rare third branchial cleft fistula opens internally into the piriform sinus and externally on the skin overlying the anterior border of the lower end of the sternomastoid muscle.

Branchial fistulae usually present in early childhood when a drop of mucus is observed leaking from the external orifice or a persisting damp patch is noticed on the clothing. Sinuses may present at any time during childhood and sometimes may be complicated by infection. The treatment for both is surgical excision.

Branchial cysts are uncommon in childhood. They usually arise from the second cleft and emerge from beneath the anterior border of the sternomastoid muscle in the upper third of the neck. They may extend upwards behind the angle of the jaw to the base of the skull or antero-inferiorly towards the midline, lying on the carotid sheath. The fluid content is milky and

Jones' Clinical Paediatric Surgery, Seventh Edition. Edited by John M. Hutson, Michael O'Brien, Spencer W. Beasley, Warwick J. Teague and Sebastian K. King.

Table 16.1 Swellings in the neck

Developmental anomalies
Branchial cleft: sinus, fistula or cyst
Branchial arch: cartilage
Thyroglossal cyst
Ectopic thyroid
Lymphatic malformation
Epidermal cyst
Acquired lesions
Inflammation of cervical lymphatics:
Acute lymphadenitis
Atypical mycobacterial infection
Acute lymph node abscess
Lymph node tumours:
Primary neoplasia
Secondary
Submandibular gland: calculus
Parotid gland: sialectasis
Thyroid gland: goitre

contains cholesterol crystals. The cyst may become infected and should be excised.

Branchial arch remnants usually arise from the second branchial arch and present as a skin tag (containing cartilage) at the anterior border of the lower third of the sternomastoid. They are excised for cosmetic reasons.

Thyroglossal cyst

The embryological descent of the thyroid anlage from the floor of the mouth leaves a track from the foramen caecum of the tongue to the thyroid isthmus. A cyst (lined by respiratory epithelium) may arise anywhere along the track, but is usually close to and adherent to the hyoid bone (75%), and is one of the common causes of midline neck lump [Table 16.2]. Recognising the attachment of this midline swelling to the underlying hyoid bone is the key to both clinical diagnosis and surgical excision. Typically, there is a tense rounded cyst in the midline or just to one side, which moves on swallowing as well as with protrusion of the tongue. The cyst may also be submental (15%), suprasternal (8%) or lingual (2%) in position. Infection may supervene [Fig. 16.1] and an infected thyroglossal cyst may be mistaken for acute bacterial lymphadenitis in the submental lymph nodes. The thyroglossal cyst and the entire thyroglossal track should be excised, preferably before infection occurs. The resection must include the middle third of the hyoid bone to minimise the risk of recurrence (Sistrunk operation).

Table 16.2 Midline neck swellings

Submental lymphadenitis
Thyroglossal cyst
Ectopic thyroid
Epidermal cyst
Goitre

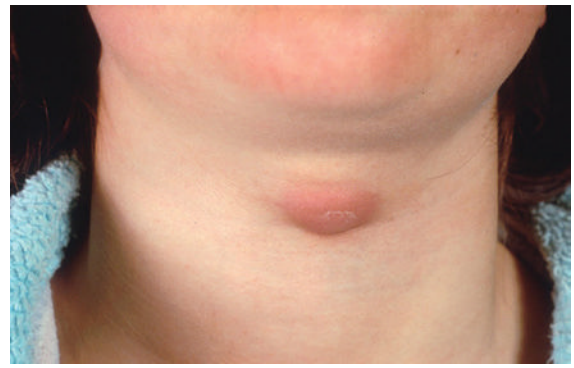

Figure 16.1 Thyroglossal cyst that has become infected.

Ectopic thyroid

Ectopic thyroid is now a rare cause of midline neck swelling, as it presents as *low thyroid function* on neonatal screening. The swelling tends to be softer than that of a thyroglossal cyst but the diagnosis may not be apparent until at operation, when the lesion is found to be solid and vascular. If this lesion is suspected preoperatively, a thyroid isotope scan should be performed to determine the distribution of all functioning thyroid, because the ectopic thyroid may be the only functioning thyroid tissue present. In this situation, it is not excised: the mass is divided in the midline and rotated on its vascular pedicle laterally to lie behind the strap muscles. Other thyroid swellings in children are rare. Neonatal goitre may result from excessive maternal iodine ingestion. Thyrotoxicosis is rare in young children. Adenoma, papillary carcinoma and medullary thyroid carcinoma are seen occasionally in older children, the latter most often in association with a multiple endocrine neoplasia (MEN) syndrome.

Lymphatic malformations

Lymphatic malformations in the region of the head and neck are relatively common. In the past, these malformations were termed cystic hygromas, but this nomenclature is now discouraged as the suffix *oma* wrongly implies these lesions to be *tumours*. Lymphatic malformations are present at birth but may not become noticeable until infancy. They are more common in boys than girls [Fig. 16.2]. Lymphatic malformations in the head and neck region are of broad two types: (1) a *simple* or multicystic lesion compressing adjacent structures or (2) a *complex* lesion infiltrating other structures including the mouth, pharynx, larynx or mediastinum. This second type resembles lymphatic malformations found elsewhere in the body and may contain cavernous haemangiomatous elements.

Simple lymphatic malformations are more common and are usually found as unilateral fluctuant, transilluminable swellings in the anterior triangle. The cysts are of varying sizes and contain clear fluid (lymph). They may enlarge suddenly and rapidly, due to viral or bacterial infection or haemorrhage. The effect of this will depend on the site and size of the cysts. A clinical emergency may arise if the increased swelling compromises the airway. In the absence of these complications, injection of sclerosant is undertaken (and uncommonly an operation) for cosmetic reasons and the prognosis is good.

Complex lymphatic malformations are less common, and complications arise because of extensive soft tissue involvement. These malformations may involve the oropharynx (leading to difficulty with speech and swallowing) or the larynx and trachea (leading to a life-threatening respiratory obstruction). Involvement of the mediastinum and pleural cavity likewise may lead to respiratory embarrassment. They may present on the first day of life and emergency care may necessitate insertion of an endotracheal tube and sometimes a tracheostomy. The baby should be referred for assessment by a multidisciplinary team (e.g. at a Vascular Anomalies Clinic). Imaging, including MRI and ultrasound, is used to delineate the anatomical extent and injection of sclerosants or operative excision may be undertaken relatively early.

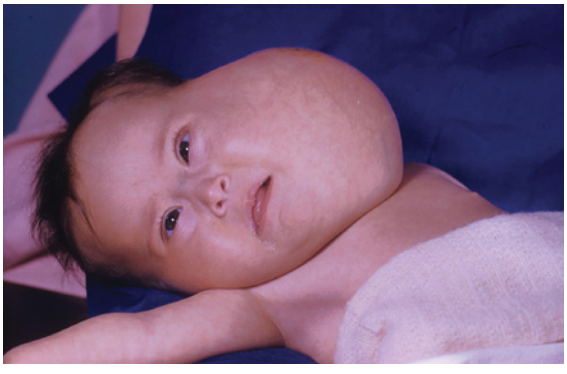

Figure 16.2 Lymphatic malformation in a baby with Down syndrome.

Epidermoid cysts

Inclusion dermoids arise from entrapment of ectodermal cells within the mesodermal layer during fetal growth. They are often in the midline or along lines of fusion, for example, at the external angle of the eye, or in the midline of the neck at which location an epidermoid cyst may be mistaken for a thyroglossal cyst. They contain sebaceous *cheesy* material surrounded by squamous epithelium. They enlarge slowly and should be removed. The most common inclusion dermoid is the external angular dermoid at the orbital margin (see in the succeeding text).

Less common varieties include the sublingual dermoid in the floor of the mouth between the mylohyoid and genioglossus muscles. It may interfere with speech and swallowing and is usually excised through a submental incision. It may be confused with a ranula or mucocele of the floor of the mouth, a lesion that contains mucus.

A rare developmental anomaly found in this region is the midline cervical cleft, a vertical open groove that results from failure of fusion of the branchial arches. Operative repair should be undertaken.

Periorbital cellulitis

Infection in the soft tissues and sinuses around the eye may cause periorbital cellulitis with rapid extension across the face [Fig. 16.3]. The danger with this infection is that it may spread to the cavernous sinus, which

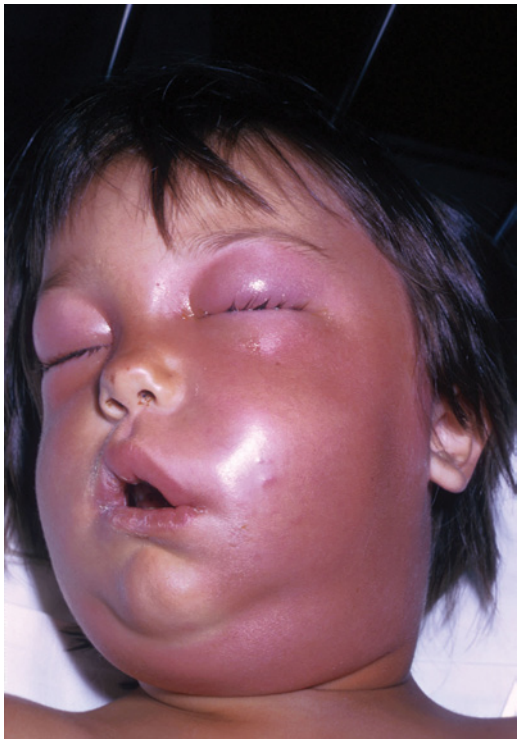

Figure 16.3 Periorbital cellulitis: sinusitis is a common source of infection.

is potentially lethal. Children with periorbital cellulitis should be admitted to the hospital for treatment with intravenous antibiotics and ophthalmological assessment (see Chapter 13) and warrant an opinion from an ear, nose and throat surgeon regarding drainage of pus from the sinuses abutting the orbital cavity (see Chapter 14).

Diseases of the lymph nodes

Infection is the most common cause of lymph node enlargement in childhood. It may be caused by bacteria, viruses or non-tuberculous mycobacteria. In many cases, the lymph nodes are reacting to an upper respiratory tract or ear infection leading to non-specific reactive hyperplasia. Lymph nodes also may become enlarged in primary or secondary malignancy. A surgical biopsy is indicated when the diagnosis is in doubt or if persistently enlarged lymph nodes (>3 cm) are present for longer than 4–6 weeks.

Reactive hyperplasia

Persistently enlarged lymph nodes are seen in many children with frequent upper respiratory tract infections. These nodes are not painful and are a normal response to infection. Occasionally, a markedly enlarged hyperplastic node (>3 cm) may require an excision biopsy to exclude tumour or other diagnoses.

Acute lymphadenitis

Acutely tender enlarged lymph glands are commonly seen during upper respiratory tract infections. Lymphadenitis usually settles with rest, analgesia and – if bacterial infection is suspected – intravenous antibiotics.

Acute lymph node abscess

Lymphadenitis may progress to an abscess, particularly in children aged 6 months to 3 years. The swelling enlarges over 3 or 4 days and may become fluctuant. An abscess in deeper nodes may not exhibit fluctuance, mainly because it is beneath the investing deep cervical fascia. The overlying skin eventually becomes red and, if untreated, the abscess will finally point and discharge. The management of an abscess is incision and drainage under general anaesthesia, taking care not to damage the mandibular branch of the facial nerve when submandibular abscesses are incised.

Non-tuberculous (*atypical*) mycobacterial adenitis

Mycobacterium avium-intracellulare, *Mycobacterium scrofulaceum*, *Mycobacterium fortuitum and Mycobacterium chelonae* cause chronic cervical lymphadenitis and *collar-stud* abscesses in children. Although human TB and bovine TB strains have been nearly eradicated in most Western countries, non-tuberculous mycobacterial lymphadenitis is still a problem in preschool children. Often termed *atypical*, such mycobacteria are found in the soil, and infection is from the child's dirty hand to the mouth and then to a tonsillar or parotid lymph node. Initially, the node is enlarged and firm but non-tender. Over 4–6 weeks, the node erupts to produce a collar-stud abscess in the subcutaneous tissue, thus causing the overlying skin to become a characteristic blue–purple colour [Fig. 16.4]. Untreated, the collar-stud *cold* abscess will ulcerate through the skin with multiple chronic discharging sinuses. Nontuberculous mycobacteria respond poorly to antibiotics and require operative excision to remove the infected lymph nodes.

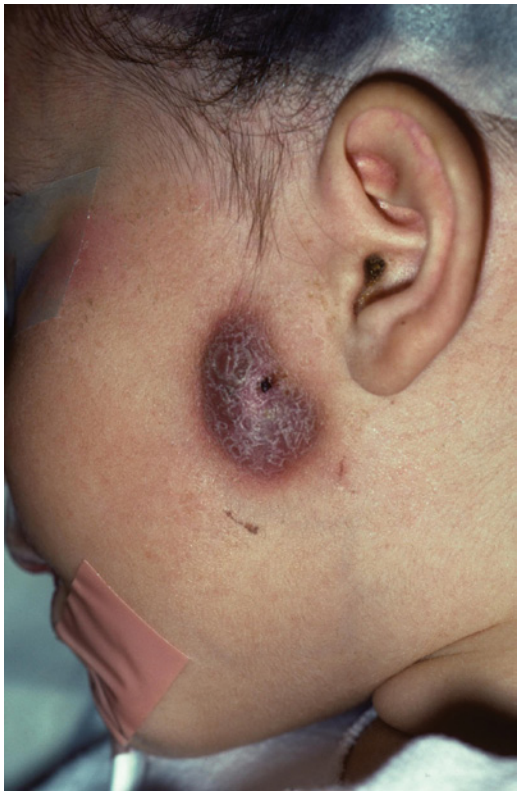

Figure 16.4 Atypical mycobacterial *cold* abscess.

The mandibular branch of the facial nerve may be at risk during excision of an affected jugulodigastric lymph node. The diagnosis of atypical lymphadenitis is confirmed by histological examination of the lymph node and culture of the pus and lymph node tissue.

Lymph node tumours

Primary neoplasia

Hodgkin's and non-Hodgkin's lymphomas may occur in cervical lymph nodes in older children. They tend to grow rapidly, and several contiguous nodes may feel spherical and rubbery. There may be associated malaise, night sweats and weight loss.

Secondary neoplasia

Nasopharyngeal and thyroid tumours and neuroblastoma may present with cervical node enlargement. In most cases, the marked enlargement and rocky hardness of the lymph nodes make the diagnosis of neoplasia obvious, but sometimes, the differential diagnosis between a large hyperplastic lymph node and a neoplastic node is difficult and necessitates an excisional biopsy.

The submandibular gland

The most common cause of enlargement is a small calculus in the submandibular duct, which produces rapid and painful swelling during eating. The gland becomes hard and tender and fluctuates in size. The submucous part of the duct in the floor of the mouth should be inspected for a tiny calculus impacted near the orifice under the tongue. An x-ray of the floor of the mouth may show an opaque calculus, which may be removed by simple incision of the duct.

The parotid gland

Recurrent enlargement of the parotid gland is due to recurrent parotitis associated with sialectasis, a condition analogous to bronchiectasis, which affects the lesser ducts and their tributaries. Parotid calculi are extremely rare.

Symptoms of sialectasis usually commence at 2–4 years of age, and the first attack may be misdiagnosed as mumps, although both sides are seldom swollen at the same time. The gland becomes enlarged and mildly tender, and the attacks may alternate from side to side. Fever and malaise are mild or absent.

Purulent saliva may issue from the orifice when the duct is compressed, and *Streptococcus viridans* or other weakly pathogenic organisms may be found on culture.

The diagnosis is clinical. However, if a sialogram is performed, it will show a snowstorm of sacculations 2–4 mm in diameter along the radicles of the gland [Fig. 16.5] but no duct obstruction. The changes are often present in both glands, even when the symptoms are confined to one side.

The condition is self-limiting and treated by massage of the parotid, tart drinks to promote the flow of saliva and chewing gum. The attacks typically last for 3–4 days, but the symptoms may persist intermittently for several years. Most children improve by about 10 years of age, and sialograms during adolescence often show that the sialectasis has disappeared. Parotidectomy is not necessary.

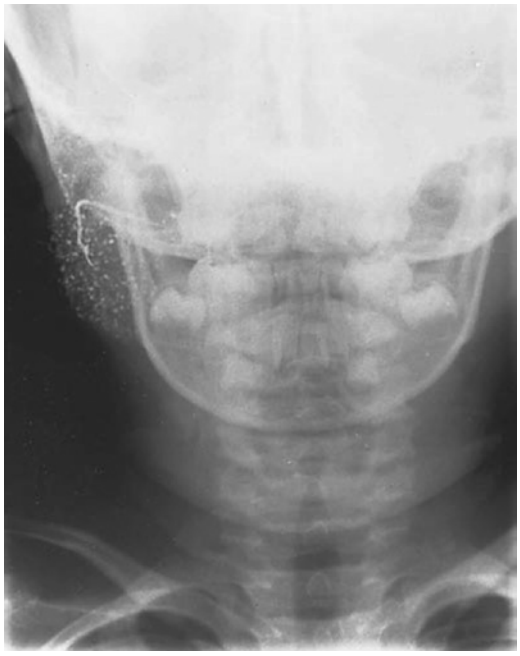

Figure 16.5 Sialogram showing sialectasis. A contrast x-ray of the parotid duct showing a *snowstorm* of saccular dilatations of the lesser ducts in the enlarged parotid.

Torticollis

The common causes of torticollis in infants and children are, in order of frequency:

1. Fibrosis in the sternomastoid muscle (often following an injury at birth)
2. Postural torticollis (a legacy of the neck position in utero)
3. Cervical hemivertebrae
4. Imbalance of the ocular muscles (producing a squint)

A posterior fossa tumour presenting with torticollis is a rare cause, although usually there would be obvious CNS signs and symptoms.

Postural torticollis is present from birth and disappears in a few months. Likewise, the associated plagiocephaly (Chapters 12 and 15) and scoliosis do not require treatment, for they are caused by intrauterine moulding.

Cervical hemivertebrae produce a mild angulation of the head and neck. The cause is readily seen in x-rays, which should be taken in all cases of torticollis where the sternomastoid muscle is not tight. No treatment is necessary, for the degree of torticollis is mild and the course is not progressive.

Ocular torticollis is not detectable until the age of 6 months and is usually not noticed until the child is at least 1 or 2 years old. Strabismus is the cause, but it is not always obvious and may be latent or intermittent. An ocular imbalance is the most likely cause of torticollis in a child without hemivertebrae, with normal sternomastoid muscles and a full normal range of passive rotation (i.e. the chin may be made to touch each acromion). Treatment is the correction of the imbalance by adjusting the attachment of the eye muscles to the globe.

Sternomastoid fibrosis

Sternomastoid muscle pathology may present in two groups of patients:

1. Neonates 2–3 weeks old present with a localised swelling in one sternomastoid muscle, that is, a sternomastoid *tumour* [Fig. 16.6]. It is often associated with plagiocephaly because the relatively malleable skull bones become distorted as a consequence of the head being consistently turned to the one side.
2. Older children present with torticollis and a tight, short fibrous sternomastoid muscle. Rotation of the head towards the affected side is limited, growth of the face on the side of the affected muscle is reduced (hemihypoplasia of the face [Fig. 16.7]) and the ipsilateral trapezius muscle may be wasted.

The aetiology is unknown, but birth injury is suspected. On histology, there is endomysial fibrosis around individual muscle fibres, which undergo atrophy.

Clinical features

In the infant, the *tumour* is so characteristic that it is diagnostic, a hard, painless spindle-shaped swelling 2–3 cm long within the sternomastoid muscle.

Due to shortening of the affected muscle, the infant's head adopts a characteristic position: rotated to the *opposite* side and often also angulated to the *same* side of the *tumour*. Plagiocephaly (Chapters 12 and 15) becomes evident during the first 3 months as a result of this preferred position and may be limited by putting the infant down to sleep on each side in turn.

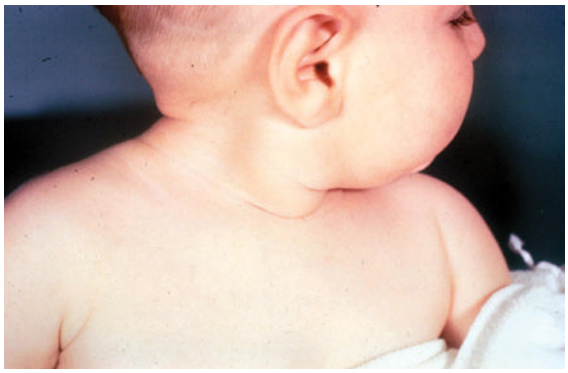

Figure 16.6 Sternomastoid *tumour* in an infant.

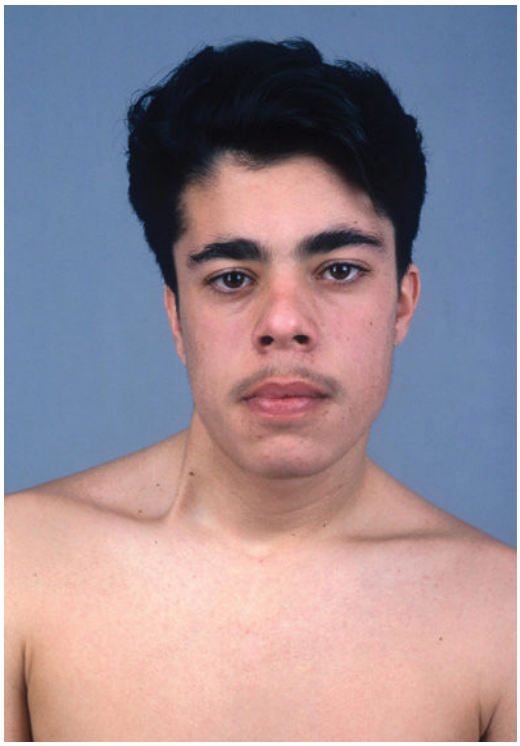

Figure 16.7 Sternomastoid torticollis. A tight (scarred) right sternomastoid muscle is apparent along with secondary hypoplasia of the right side of the face.

Hemihypoplasia of the face [Fig. 16.7] describes the decreased growth of one side that may occur as a non-specific result of any type of long-standing immobilisation and is not directly attributable to fibrosis in the sternomastoid.

Treatment

Infants with a sternomastoid *tumour* initially should be managed non-operatively, because in 90% it will heal completely and without complications in 3–6 months.

In the remaining cases, the resultant fibrosis causes permanent muscle shortening and persistent torticollis. Surgical division of the sternomastoid muscle is indicated if torticollis persists beyond 12 months of age or if there is progressive hemihypoplasia of the face. Following surgery, symmetry of the face improves over several years, but may never recover completely.

Developmental anomalies of the face

External angular dermoid

As introduced earlier, this is a common anomaly of fusion between the frontonasal and maxillary processes during formation of the head and face (Chapter 12). The cyst is noticed in infancy as it enlarges gradually [Fig. 16.8]. Often, it is beneath the pericranium, giving it a firmer consistency than may be expected. Occasionally, it is misdiagnosed as a bony lump. Excision through a lateral supra-eyebrow incision is curative.

Many varieties of facial clefts have been described and classified, but most of them are rare. Cleft lip and

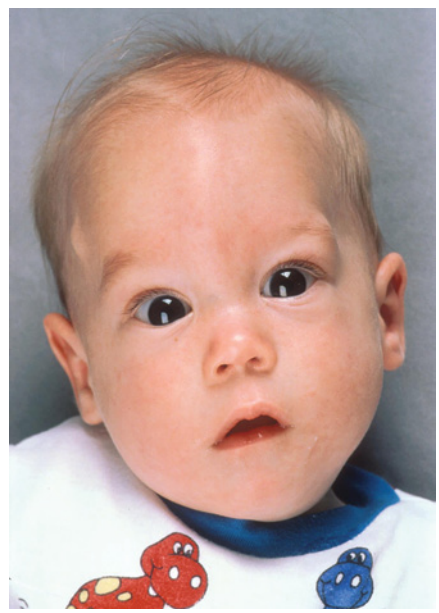

Figure 16.8 External angular dermoids, just above and lateral to the right eyebrow, and the left under the lateral edge of the left eyebrow..

palate, by contrast, are very common. Clefts are described in Chapter 15.

Microstomia

Malformations of the structures derived from the first pharyngeal arch may cause microstomia, a misshapen ear, absence of the external auditory canal, a rudimentary middle ear, hypoplasia of the mandible and its teeth, hypoplasia of the malar–maxillary complex and, sometimes, facial paralysis. Complex defects of the facial skeleton, such as this, are managed by craniofacial surgery (see Chapter 15).

Deformities of the ear

Accessory auricles

Small tags of skin and cartilage may be present, usually close to the tragus, but sometimes along a line extending to the angle of the mouth. They are excised for cosmetic reasons.

Pre-auricular sinus

This is a common condition (1/50 of Asian and 1/200 of Caucasian children) and is often bilateral and asymptomatic. There is a tiny hole just in front of the upper crus of the helix, from which an epithelial track extends deeply forwards and downwards. The track is often short, but sometimes extends deeply towards the pharynx.

Where there are no symptoms, or only an occasional bead of watery discharge, it is best left alone. If it becomes infected with purulent discharge and the opening becomes sealed, an abscess may develop. In such cases, the abscess should be incised and drained and the sinus then excised once the infection has settled.

Microtia

A rudimentary ear of irregular skin and cartilage is associated with absence of the external auditory canal, a rudimentary middle ear and a small mandible on the same side [Fig. 16.9]. When the site of the ear is acceptable, it may be used as the basis for reconstruction, which is preferable to a prosthetic ear. Only when the condition is bilateral is it necessary to create an external auditory canal and provide a hearing aid within the first few months of life to enable the infant to hear and develop speech. Further operations are required in later years.

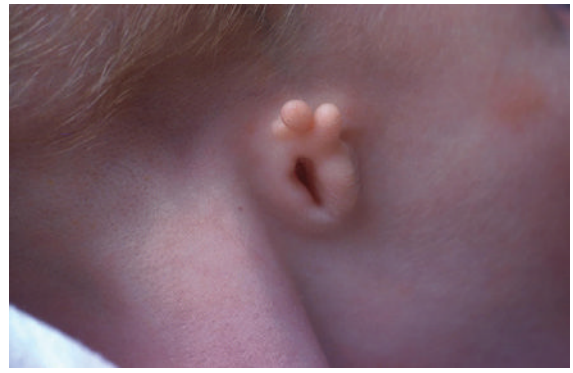

Figure 16.9 Microtia, associated with a maldevelopment of the dorsal ends of the first and second branchial arches. The external auditory canal is a shallow pit.

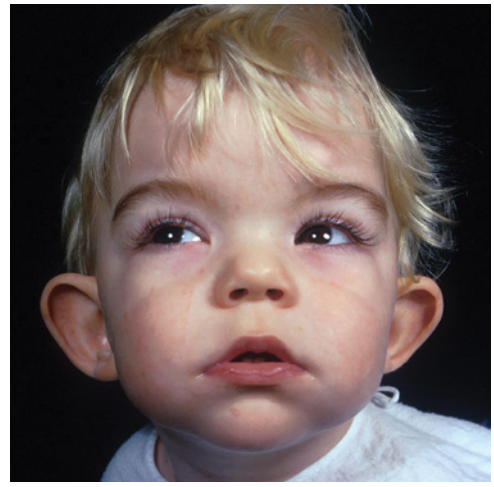

Figure 16.10 Bat ears.

Bat ears

Bat ears, both unilateral and bilateral, are common and often familial [Fig. 16.10]. The concavity of the concha extends to the rim that stands out farther than normal. The ear is often bigger than normal, as well as more protuberant.

Corrective surgery is advisable when there is gross protrusion, particularly when there are adverse comments from other children. Strapping in the neonatal period achieves nothing, and removal of skin from the postauricular groove is inadequate. The fold of the antihelix must be fashioned, shaping and fixing the cartilages in a new relationship and holding them in position for the approximately 3 weeks required for union of the cartilages. Operative correction may be done any time after infancy.

Shell ears

Shell ears are similar to bat ears in protruding from the scalp, but they are small, and the rim of the helix is so short that the ear cannot be readily folded back into the normal position. Operative correction is much more difficult than in bat ears and typically performed as a staged procedure.

KEY POINTS

- Thyroglossal cyst typically presents as a midline neck lump fluctuating in size and *that may become inflamed*.
- Lymphatic malformations (previously known as cystic hygromas) are a congenital anomaly of the jugular lymph sacs that frequently become infected. Early referral to the regional, multidisciplinary Vascular Anomalies Clinic is recommended.
- Enlarged lymph nodes need excisional biopsy if greater than 3 cm in diameter and present for more than 4–6 weeks.
- Sternomastoid fibrosis with torticollis may need operative division to prevent facial asymmetry.
- External angular dermoids require excision.

Further reading

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PART IV
Abdomen

CHAPTER 17
The Umbilicus

CASE 1

A few weeks after birth, the mother noticed an intermittent swelling at her baby's umbilicus, covered with skin. It became quite large on crying, making her concerned about rupture. The lump often gurgled if compressed, but did not particularly upset the neonate.
Q1.1 Is this lesion dangerous, and what is its natural history?
Q1.2 Is an operation needed?
Q1.3 Why is there a hole in the abdominal wall?

CASE 2

A week or two after separation of the cord stump, the umbilicus remained slightly red and damp. Despite careful drying, the dampness persisted, and by 6 weeks, there was a cherry-red mass protruding from the umbilical scar.
Q2.1 Why has this occurred and how is it treated?

CASE 3

A 6-year-old boy presented with a small, mildly tender lump 5 cm above the umbilicus. He has a history of recurrent epigastric pains, particularly after meals. His mother first noticed the lump in infancy but did not seek attention as it appeared to be harmless.
Q3.1 What are the contents of the lump?
Q3.2 Is it dangerous?
Q3.3 Does it need treatment?

Embryology

The umbilicus is first formed by folding of the embryonic disc to form a three-dimensional embryo, with fusion or *zippering* in the ventral midline towards the umbilical ring. The ring remains necessarily widely open during early fetal life, allowing physiological midgut herniation. Abnormalities in either folding or midgut herniation may produce the more serious but rare conditions of exomphalos and gastroschisis (see Chapter 8). The open umbilical ring is also a conduit for connections between the midgut and yolk sac (termed vitello-intestinal or omphalo-mesenteric duct) and between the bladder and allantois (urachus). These vestigial connections may persist after birth producing clinical features in accordance with the nature of persistence.

The umbilicus at birth

At birth, two umbilical arteries (branches of the internal iliac arteries) and one umbilical vein (via the falciform ligament and ductus venosus) form the umbilical cord. Routine assessment of umbilical cord vascular anatomy is an important element of the postnatal examination as abnormalities can be associated with other important congenital conditions. In the hours and days after birth, the cord vessels are obliterated, becoming the medial arcuate ligaments (umbilical arteries) and ligamentum teres (umbilical vein). Also after birth, the cord desiccates and separates. The umbilical ring closes. However, delayed contraction of this fibromuscular ring may allow the peritoneum and abdominal contents to bulge through the defect. Colonisation of the residual necrotic cord stump tissue bacteria may produce a low-grade infection resulting in granulation tissue formation.

The range of umbilical abnormalities is summarised in Table 17.1.

Umbilical hernia

Some degree of umbilical herniation is present in almost 20% of newborn babies: the incidence is higher in premature neonates or in conditions in which intra-abdominal pressure is increased, for example, ascites,

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Table 17.1 Abnormalities of the umbilicus

Skin-covered swelling	Umbilical hernia
Large membrane-covered swelling	Exomphalos (or omphalocele)
Uncovered bowel protruding at birth	Gastroschisis
Infection of cord stump	Omphalitis
Mucous discharge	Umbilical granuloma heterotopic bowel mucosa
Air and faecal discharge	Patent vitello-intestinal (omphalo-mesenteric) tract
Urine discharge	Patent urachus

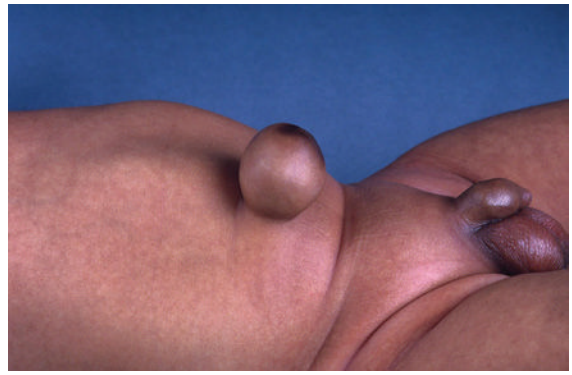

Figure 17.1 Umbilical hernia in a baby.

Down syndrome or congenital hypothyroidism. Because the anomaly occurs after involution of the umbilical cord, associated anomalies are rare, and the hernia is covered by skin.

While the infant lies quietly, the umbilical skin merely looks redundant, but on crying or straining, bowel fills the hernia and the lesion enlarges to become tense and bluish beneath the thin shiny skin [Fig. 17.1]. The bowel may be reduced easily, often with an audible gurgle.

While most umbilical herniae close spontaneously, there are several practical points that must be explained to the parents:

1. The time of natural closure: in the first 3–4 months of life, the bulge may actually increase a little before getting smaller. Resolution usually occurs in the first 12 months, but may take up to 3 years.
2. Importance of defect size: even defects that appear very large at birth may close spontaneously. A defect diameter less than 1 cm at 12 months of age is considered an indicator of likely closure, although spontaneous closure of larger defects may still occur.
3. The skin never ruptures, and the thin skin evident in the first 4 weeks gradually becomes thicker.
4. Strangulation is remarkably uncommon and it is safe to wait. The size and tenseness of the hernia when the infant cries is often interpreted incorrectly as causing pain: umbilical herniae probably are symptomless.

Treatment

For the majority, no treatment is required. Strapping is contraindicated because it is ineffective and may cause complications. Umbilical hernia repair is reserved for defects that fail to close spontaneously and is normally deferred until after 3 years of age.

Para-umbilical (or supra-umbilical) hernia

This is a defect in the linea alba separate from, but adjacent to, the umbilical cicatrix. Most are just above the umbilicus. The defect is a transverse elliptical slit with sharp edges, in contrast to the rounded shape and blunt edges of a central umbilical hernia. This morphological distinction imparts two key differences compared with umbilical herniae. First, there is an increased risk of strangulation. Second, spontaneous closure is less likely. For these reasons, operative repair is more often required, usually as an elective procedure after the third year of life.

Epigastric hernia

Extraperitoneal fat from within the falciform ligament may protrude through a tiny defect in the decussating fibres of the linea alba. It produces a lump in the epigastrium, which may be noticed incidentally. Typically, an epigastric hernia is otherwise asymptomatic, but some cause recurrent, vague epigastric tenderness or abdominal pain, particularly after eating and during exercise. A firm fatty swelling is palpable in the midline of the epigastrium, usually midway between the xiphisternum and umbilicus. Treatment is by excision or reduction of the protruding fat and closure of the defect in the linea alba.

Umbilical sepsis

Umbilical sepsis (omphalitis) is a potentially dangerous infection of the exposed cord stump occurring in the neonatal period. The most common causative organisms are *Staphylococci*, *Escherichia coli* and *Streptococci*. Consequently, an important aspect of preventive medicine is to keep the umbilical stump clean and dry. In minor infections, the umbilical region is red and swollen with a seropurulent discharge, but usually responds well to local and/or systemic antibiotics.

Proactive treatment of omphalitis is important as there may be little superficial evidence that infection has spread further via the lymphatics or the umbilical vessels. Infecting organisms may exploit superficial vessels or the recently patent umbilical arteries and vein to gain access to the bloodstream, with resultant septicaemia. Dissemination of infection via the umbilical arteries may cause abscesses in the distribution of the internal iliac arteries. Ascent of infection along the umbilical vein may involve the portal vein and via the ductus venosus to the vena cava. Clinically, overt infections in these venous structures are rare but serious, and latent infection may lead to thrombophlebitis and portal vein thrombosis. Portal hypertension may ensue, ensues with recanalisation and the opening of collaterals vessels, leading to a cavernomatous malformation of the portal vein (see Chapter 26).

The infant with a discharge from the umbilicus

A discharge from the umbilicus may be pus, urine or faeces.

Umbilical granuloma

An umbilical granuloma is a common lesion that presents as a small mass of heaped granulation, accompanied by a seropurulent discharge. Granulation tissue is most likely produced in response to subacute bacterial infection of the cord stump. Lesions with a definite stalk may be ligated without anaesthesia. Larger or broad-based granulomas are better treated with topical application of silver nitrate. This facilitates epithelialisation of the granuloma and is usually curative.

Heterotopic mucosa

Sometimes, a small focus of ectopic bowel mucosa at the base of the umbilicus masquerades as a granuloma with mucous or seropurulent discharge. This typically spherical lesion can be recognised by its deeper, cherry-red colour and location in the depths of the umbilical cicatrix. Presence of a small opening on its surface suggests a persistent vitello-intestinal duct and a sinugram is indicated (see below). In the absence of any such opening, distinction from a granuloma is a moot point as both are effectively treated by silver nitrate. One or two applications are usually all that are needed to remove the gut epithelium, which is replaced rapidly by normal skin. Very occasionally, operative excision is required.

Persistent vitello-intestinal remnants

The vitello-intestinal duct is a communication between the midgut and yolk sac, which normally obliterates and then disappears around the sixth week of gestation. Persistence of all or part of the vitello-intestinal duct results in various pathologies. These may present in early infancy, but on occasions are not recognised until some years later, for example, haemorrhage from a Meckel's diverticulum.

A true persistent vitello-intestinal duct indicates patency of the whole tract, from ileum to umbilical skin. Discharge of ileal contents through the small mucosal opening at the umbilicus causes inflammation, which may in turn confound clear visualisation of the duct orifice [Fig. 17.2]. Very rarely, when the duct is short and broad, the ileum may intussuscept through it

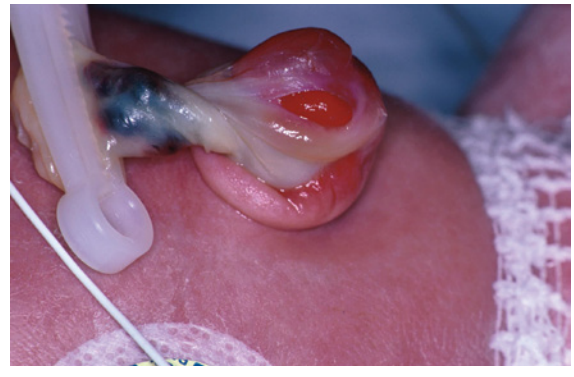

Figure 17.2 A patent vitello-intestinal duct and patent urachus may look similar: the diagnosis depends on whether the discharge is urinary or faecal.

onto the surface of the umbilicus. The Y-shaped segment of prolapsing bowel, inside out, with two orifices is diagnostic.

A vitello-intestinal band (or Meckel's band) is the result of a duct that has obliterated but not disappeared. Accordingly, the band runs from the ileum to the deep surface of the umbilicus. It may cause no symptoms throughout life or it may, at any age, cause intestinal obstruction when a loop of bowel becomes entangled beneath it.

Partial vitello-intestinal duct obliteration causes a sinus, cyst or Meckel's diverticulum, according to which part of the tract persists. Sinuses indicate the persistent duct segment communicates with the skin. Cysts and sinuses may become infected, form an abscess and discharge pus at the umbilicus. Meckel's diverticulum is a patent vitello-intestinal duct remnant with communication with the lumen of the ileum. There may be an associated Meckel's band tethering the diverticulum and umbilicus. The presenting features and complications of Meckel's diverticula are described in Chapter 23.

All remnants of the vitello-intestinal duct are best excised, which may necessitate a laparoscopy or laparotomy to search for discontinuous segments of the tract.

Urachal remnants

The urachus, the vestigial communication between the allantois and bladder, normally obliterates and remains as the median arcuate ligament. Urinary discharge from the umbilicus indicates a rare, persistent of the urachus [Fig. 17.2]. In such infants, concomitant lower urinary tract obstruction is a recognised but rare association. Treatment is excision of the patent urachus after investigation and relief of any underlying anomalies.

More commonly, the urachus is partly obliterated and presents as a tender mass or abscess in the midline, at or below the umbilicus. The diagnosis is confirmed on ultrasonography prior to treatment by drainage and excision as a single or staged procedure.

KEY POINTS

- Umbilical hernia typically only needs treatment beyond 3 years of age.
- Infection of exposed necrotic umbilical cord stump leads to umbilical granuloma.
- Epigastric hernia contains extraperitoneal fat.
- A discharging umbilicus may indicate a patent vitello-mesenteric duct or urachus.

Further reading

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CHAPTER 18
Vomiting in the First Months of Life

CASE 1

A 4-week-old breastfed boy was completely well until a few days earlier, when he began vomiting all feeds. He was otherwise well and keen to feed despite the non-bile-stained vomiting. He had lost weight and had few wet nappies.

Q 1.1 What physical sign would confirm the diagnosis you suspect?
Q 1.2 If you were unable to demonstrate this sign, what would you do if you still suspected the diagnosis?
Q 1.3 What initial investigation would you perform to assist you in resuscitation?

CASE 2

A 9-day-old girl who had been completely well suddenly began vomiting bile-stained fluid. There were no groin swellings. She would not feed.

Q 2.1 What diagnosis would you wish to exclude urgently?
Q 2.2 How would you do this?
Q 2.3 If this diagnosis was confirmed, how urgent is the treatment?

CASE 3

A 3-month-old boy was always vomiting, irrespective of how frequently he fed. Initially, he had been breastfed, but he was now on the bottle, and his grandmother assisted with night feeds. He was vomiting small volumes of milk. He was not distressed by the vomiting, weighed 6 kg and was growing well.

Q 3.1 What is the most likely diagnosis?
Q 3.2 What measures could be suggested to reduce the vomiting?

Vomiting is common in the first months of life, when the evaluation of its significance is particularly important. The temptation to disregard it must be resisted: it is a symptom, not a diagnosis, and its cause must be established [Table 18.1].

Vomiting is significant when it is:

1. Bile stained
2. Persistent
3. Projectile
4. Blood stained, that is, *coffee grounds*, flecked with altered blood
5. Accompanied by weight loss or failure to gain weight
6. In a child who is unwell, lethargic or listless and/or disinterested in feeding

Most vomiting is due to non-surgical conditions or feeding difficulties. Neonatal infections (e.g. septicaemia, meningitis or urinary tract infection) may present with a variety of clinical features, including vomiting, convulsions, diarrhoea, pallor, lethargy, listlessness, cyanosis, and pyrexia or hypothermia. Gastroenteritis may be seen in bottle-fed babies but is uncommon in fully breastfed infants. In approximately 25% of those with the rare syndrome of congenital adrenal hyperplasia, there is a salt-losing metabolic disturbance that produces severe vomiting and genital abnormalities in females (Chapter 10).

Malrotation with volvulus usually presents in the first week or so of life with bile-stained vomiting, but may occur at any age. The possibility of malrotation with volvulus must be entertained in any child with sudden onset of green vomiting for which there is no other obvious cause. An urgent upper gastrointestinal contrast study will diagnose malrotation if it shows that the duodenojejunal flexure is to the right of normal and below the level of the pylorus. It may also be diagnosed on ultrasonography, if there is sufficient radiological experience. If volvulus goes unrecognised, the entire midgut may be lost from ischaemia when blood flow

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Table 18.1 Causes of vomiting at 1 month of age

Septic	Urinary tract infection
	Meningitis
	Septicaemia
Mechanical	Gastro-oesophageal reflux
	Pyloric stenosis
	Strangulated inguinal hernia
	Malrotation with volvulus (bile-stained vomitus)
Other	Congenital adrenal hyperplasia
	Overfeeding

through the superior mesenteric artery is compromised. Untreated, the child will die: consequently, a child with volvulus needs an urgent operation.

Strangulated inguinal herniae may occur in infants and are easily diagnosed on examination. A hard, tender irreducible swelling at the external inguinal ring will confirm the diagnosis.

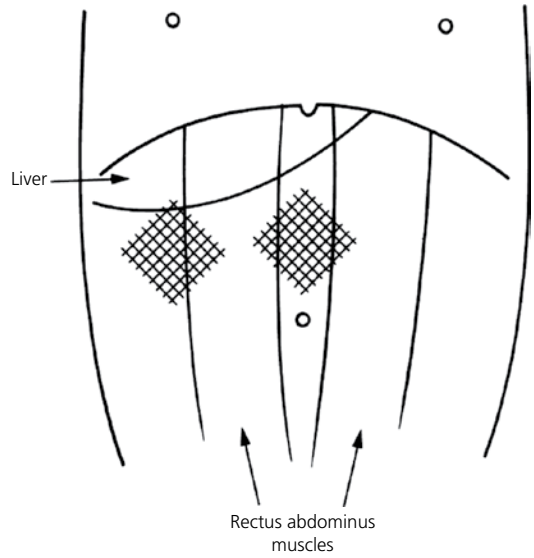

Figure 18.1 Schema showing the two places to palpate a pyloric tumour.

Pyloric stenosis

Congenital hypertrophic pyloric stenosis is the most common cause of vomiting that requires an operation in infants and affects 1:300 children, of whom 80% are boys.

Pyloric stenosis is important because it is common, there is a risk to life and permanent relief is obtained by a relatively simple operation. The aetiology remains obscure and is partly genetic, as almost 20% of those affected have a family history.

Symptoms

The usual presentation is with severe vomiting that commences between 3 and 6 weeks of age in an otherwise well baby. Pyloric stenosis is exceptionally rare in neonates younger than 10 days, or infants older than 11 weeks.

Copious vomiting occurs after all feeds; the vomitus contains curdled milk and is practically never bile stained. It may contain some brown coffee-ground flecks of altered blood, reflecting the gastritis secondary to the gastric outlet obstruction. Often, the vomiting is forceful or projectile and may occur well after the last feed. Initially, the child is active and hungry, and a key feature is his readiness and ability to feed again immediately after vomiting. Later, with increasing dehydration and electrolyte imbalance, he becomes weak, listless and lethargic, not unlike the clinical picture seen in a child with sepsis. He loses weight and looks scrawny. If untreated, he may ultimately die from dehydration and metabolic alkalosis.

Signs

Peristaltic waves of gastric contraction indicate hypertrophy of the gastric muscle secondary to progressive outlet obstruction: their observation makes pyloric stenosis likely. Palpation of the thickened pylorus in the epigastrium, however, is pathognomonic and confirms the diagnosis. The hypertrophic and thickened pylorus is traditionally called a *pyloric tumour*: it feels like an olive, a small pebble or the terminal segment of the little finger. It is relatively mobile. It is palpable most easily in the angle between the liver and the lateral margin of the right *rectus abdominis* muscle or in the gap between the two recti midway between the umbilicus and the xiphisternum [Fig. 18.1]. It is felt most easily when the baby is relaxed and not crying and when the stomach is empty. When difficulty is experienced feeling the *tumour*, a nasogastric tube may be passed to empty the stomach.

Failure to palpate the pyloric tumour

If the initial palpation is not conclusive, further observation is necessary and a second examination is made a few hours later. Other manoeuvres that may assist in pal-

pating an elusive pyloric tumour are summarised in Table 18.2. When symptoms suggest pyloric stenosis but no tumour may be palpated, and septic causes of vomiting have been excluded, a paediatric surgeon should be consulted, and imaging of the pylorus may be required. Real-time ultrasonography may identify the hypertrophied pylorus [Fig. 18.2]. A contrast meal performed under fluoroscopic control will reveal gastric outlet obstruction [Fig. 18.3] and may show other pathologies, including gastro-oesophageal reflux, but is not often needed. These investigations are required in a minority of cases only.

Table 18.2 Tips for palpating a pyloric tumour

1. Ensure the baby is relaxed	Be patient Palpate gently; avoid hurting the infant Flex the hips Wait until crying stops or infant is asleep Allow infant to feed or suck dummy Palpate at start of feed Repeat examination
2. Empty overfull stomach	Pass NG tube to empty stomach
3. Is the diagnosis wrong?	Check for sepsis/inguinal hernia if peristaltic waves absent and pylorus not palpable
4. Pyloric stenosis suspected but not proven	Ultrasonography Upper gastrointestinal contrast study

Initially, the diagnosis may be attributed to pre-existing gastro-oesophageal reflux (which is present in many infants) or feeding problems. Often, there is a history of several changes in feeding patterns before the diagnosis is made. However, palpating a tumour is the *sine qua non* of diagnosis and excludes all other causes. Other features, such as visible gastric peristalsis and projectile vomiting, are supporting evidences, but not in themselves diagnostic.

Investigation

The history (duration of vomiting, amount of vomiting, reduction in wet nappies) and clinical findings (anterior fontanelle and skin turgor) reveal the degree of dehydration. The extent of electrolyte and acid–base imbalance must be determined to guide resuscitation before operation. These infants often have an

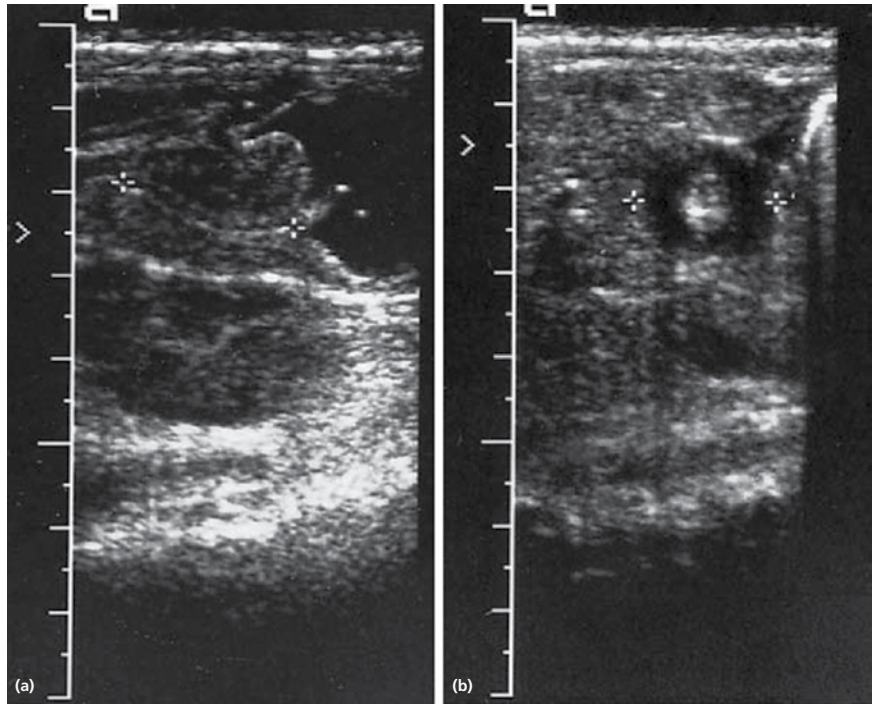

Figure 18.2 Ultrasonographic diagnosis of pyloric stenosis shows the thickened circular pyloric muscle in (**a**) longitudinal and (**b**) transverse plane.

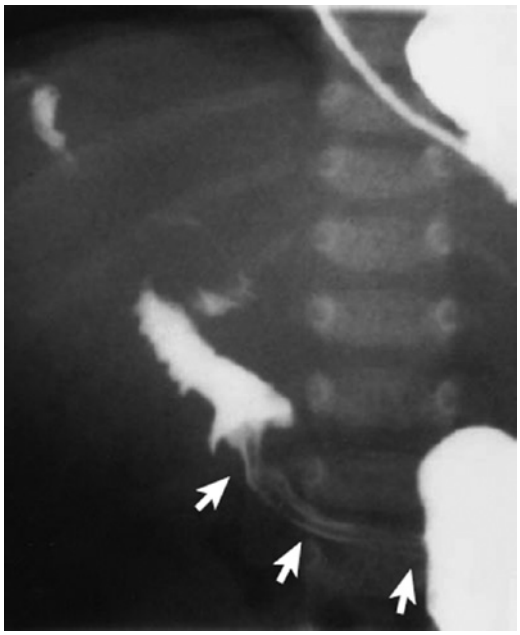

Figure 18.3 Pyloric stenosis demonstrated using an upper gastrointestinal contrast study, showing the *string sign*. The markedly narrowed pyloric canal (arrows) causes gastric outlet obstruction.

hypochloraemic, hyponatraemic, hypokalaemic metabolic alkalosis. Further estimations of the serum electrolytes and acid–base parameters after resuscitation should confirm complete correction of the disturbance prior to an operation.

Treatment

Treatment involves correcting the fluid and electrolyte abnormality followed by pyloromyotomy, the Ramstedt operation. There are numerous resuscitation protocols available, such as intravenous administration of 0.45% sodium chloride in 5% dextrose and supplementary potassium chloride. The infusion rate is determined after estimating the percentage dehydration, the infant's weight and maintenance requirements.

The Ramstedt operation may be done via a supra-umbilical incision, via a right transverse incision or laparoscopically. The hypertrophied pyloric muscle is split longitudinally allowing the pyloric mucosa to bulge through the gap, thus providing a wider channel into the duodenum. Oral feeds may be commenced within 24 h of surgery, and babies rapidly regain their lost weight.

Gastro-oesophageal reflux

An incompetent sphincter at the oesophago-gastric junction allows vomiting in the neonatal period and tends to improve as the infant gets older. Vomiting occurs at any time during or between feeds and usually is neither projectile nor bile stained. If oesophagitis is present, bleeding may produce bright blood or *coffee-ground* flecks of altered blood in the vomitus or be revealed as anaemia, but more commonly, the infant seems unsettled and irritable. In some infants, severe gastro-oesophageal reflux leads to repeated episodes of aspiration pneumonia, failure to thrive or an oesophageal stricture.

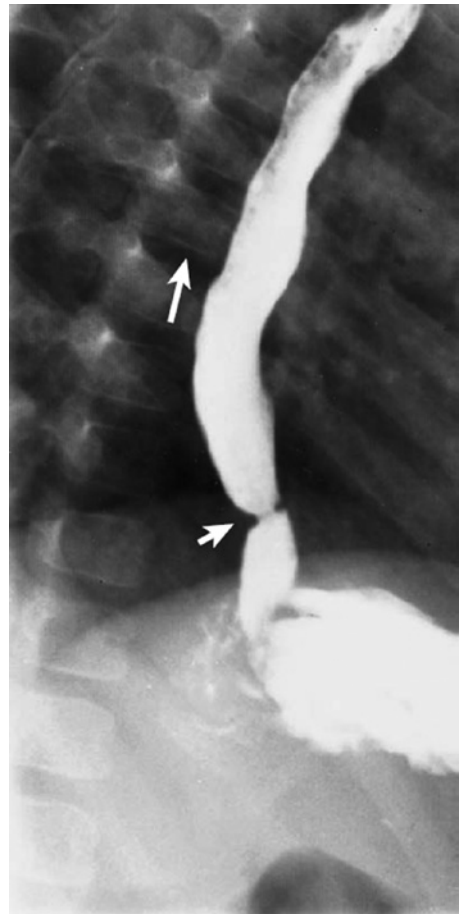

Figure 18.4 Severe gastro-oesophageal reflux with an oesophageal stricture secondary to reflux oesophagitis (arrow head). The contrast may be seen to flow freely up the oesophagus (arrow).

Management

Gastro-oesophageal reflux affects many infants and the diagnosis is made on clinical grounds. There is a natural tendency towards spontaneous improvement with age, so initial treatment should be conservative, with the head of the cot elevated on blocks. Thickening of feeds and the use of mild antacids, may also be helpful. Increasingly, proton pump inhibitors are employed to decrease the acidity of the reflux, reducing the discomfort of oesophagitis.

Where there is oesophagitis, anaemia, respiratory symptoms or failure to thrive, an upper gastrointestinal contrast study is advisable to document the severity of gastro-oesophageal reflux and to demonstrate any hiatus hernia or oesophageal stricture [Fig. 18.4].

Oesophagoscopy and oesophageal biopsy should be performed if haematemesis or anaemia is present, to assess the severity of the peptic oesophagitis, or when an upper gastrointestinal contrast study demonstrates oesophageal obstruction. Further information may be gained from 24 h pH monitoring and oesophageal manometry.

Surgical intervention to control the reflux is indicated if conservative treatment fails, if there is an oesophageal stricture or if a hiatus hernia is present. Operation involves plicating the gastric fundus around the lower oesophagus [Nissen fundoplication], usually laparoscopically. The oesophageal hiatus is reinforced at the same time. Oesophageal strictures secondary to reflux normally resolve spontaneously once the reflux has been eliminated.

KEY POINTS

- Vomiting in babies is significant when it is blood or bile stained, persistent, projectile or accompanied by weight loss.
- Bile-stained vomiting in babies without sepsis should be treated as possible malrotation with volvulus and referred as an emergency to a surgeon.
- If pyloric stenosis is suspected, the epigastrium should be observed for gastric peristalsis.

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CHAPTER 19
Intussusception

CASE 1

A 5-month-old boy has a 48 h history of being unwell and vomiting. At times, he appears to have been in severe pain. He looks pale and lethargic. There is a vague impression of a mass on the right side of his abdomen.

Q1.1 What is the likely diagnosis?
Q1.2 How may the diagnosis be confirmed?
Q1.3 Once treated, is it likely to recur?

CASE 2

A 7-month-old girl has been unwell for 5 days: she initially seemed irritable and vomited her feeds, refused further feeds and soon became listless and dry. Her mother measured her temperature at 37.8°C. She has had few dirty nappies and has developed a distended and tender abdomen. There are no herniae.

Q2.1 Which conditions would be in your differential diagnosis?
Q2.2 What would be the initial management of this child?
Q2.3 What is the likely definitive treatment that will be required?

In intussusception, one segment of the bowel (the *intussusceptum*) passes onwards inside the adjacent distal bowel (the *intussuscipiens*). The apex of the intussusceptum is termed the *lead point*. Once this telescoping phenomenon becomes established, intestinal obstruction follows. Ileo-colic intussusception represents one of the more common surgical emergencies in the first 2 years of life.

Aetiology

In 90% of all ileo-colic intussusceptions, there is no pathological lesion at the lead point (so-called idiopathic intussusception). In such cases, the apex of the intussusception is most likely an enlarged submucosal lymphoid tissue in the distal ileum (Peyer's patches), which has undergone reactive hyperplasia. This may be the result of a viral infection. With continued peristalsis, the lead point moves through the ileo-caecal valve into the colon and rarely may even reach the anus.

Examples of pathological lead points include Meckel's diverticulum, lymphoma, polyp and duplication cyst. The likelihood of such a lesion increases with age, such that in the small number of children presenting after 5 years of age, a pathological lead point is present more than half of the time.

In school-age children presenting to emergency departments with colicky abdominal pains, it has been recognised recently that many have ileo-ileal intussusception, presumably secondary to gastroenteritis causing a disturbance of normal peristalsis. Ileo-ileal intussusception is usually very transient, with spontaneous resolution corresponding with cessation of colic. It is extremely rare for it to progress to ileo-colic intussusception, which needs specific treatment and potential surgical intervention, as in infants between 3 months and 3 years.

Incidence

The peak incidence of ileo-colic intussusception is in infants 5–7 months old, and 70% of patients are between 3 and 12 months of age. Boys are affected more frequently than girls.

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Clinical features [Box 19.1]

Symptoms

Pain is the most important symptom (85%). It typically commences as a colicky pain lasting 2–3 min, during which time the infant screams and draws up his knees. Spasms typically occur at intervals of 15–20 min. The infant becomes intermittently pale and clammy (similar to a syncopal episode in older children), exhausted and lethargic between spasms. After 12 h or so, the pain becomes more continuous.

Vomiting almost always occurs as well: usually once or twice in the first hours and then again if intestinal obstruction is fully established.

Signs

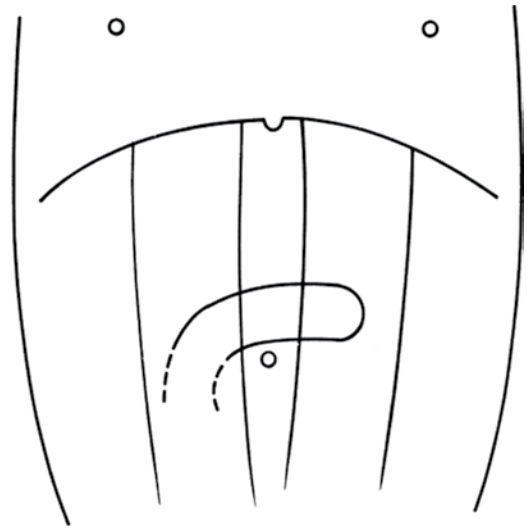
Children with intussusception look pale and lethargic, except when they are aroused by a spasm of severe pain. A mass (sometimes described as sausage shaped) is palpable in more than half the infants and is usually found in the right hypochondrium, although it may be anywhere between the line of the colon and the umbilicus [Fig. 19.1]. The intussusception mass is most likely to be felt early, before being concealed by abdominal distension and increasing abdominal tenderness.

Normal or loose stools are often passed shortly after the onset of symptoms, and any diarrhoea tends to be of small volume and transient. About half the patients pass a stool containing blood and mucus (*red currant jelly*), formed by the diapedesis of red cells through the congested mucosa of the intussusceptum. Blood may be identified on the glove following rectal examination, which may disclose the apex of the intussusceptum within the rectum.

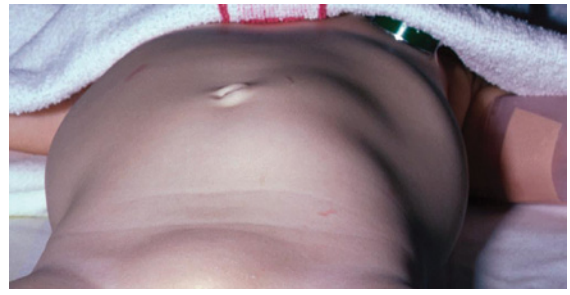
The infant is pale, limp and tired and has tachycardia. If there is delay in diagnosis, the infant becomes dehydrated, listless and febrile, has abdominal distension and looks ill. These are late signs and ideally the diagnosis should be made before they appear.

Box 19.1 Presenting features of intussusception

1 Vomiting
2 Abdominal colic
3 Pallor
4 Lethargy
5 Abdominal mass
6 Rectal bleeding



(a)



(b)

Figure 19.1 The site of the sausage-shaped mass of intussusception in the abdomen, shown schematically in (**a**) and in a patient (**b**).

Differential diagnosis

Wind colic is common in the first 3 months after birth but rarely lasts more than an hour and usually is not accompanied by vomiting. Persisting severe colic for more than 1–2 h should arouse suspicion of an intussusception, particularly if accompanied by vomiting.

Colic and the passage of blood and mucus in severe cases of gastroenteritis may mimic intussusception, except that the volume of diarrhoea is greater. In intussusception, the small loose stools passed early in the course of the disease simply represent evacuation of the stimulated colon beyond the obstruction. Persistent vomiting and pain without diarrhoea is unlikely to be gastroenteritis.

A strangulated inguinal hernia may present with abdominal pain, vomiting and distension but is recognised

easily when an irreducible lump is seen on examination of the inguinal region.

Investigations

A plain x-ray of the abdomen may be normal, show non-specific abnormalities or reveal a small bowel obstruction with air–fluid levels in the dilated small bowel. Occasionally, the apex of the intussusceptum may be seen.

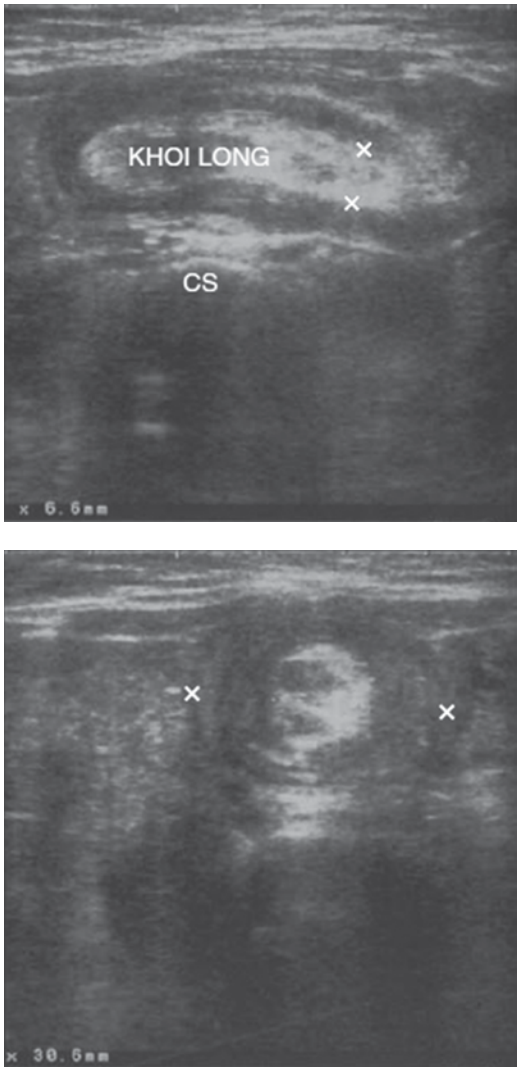


Figure 19.2 Ultrasonography is the first investigation in infants and children with suspected intussusception. Upper panel shows longitudinal view of intussusception, while lower panel is a transverse section.

The diagnosis is confirmed on ultrasonography, which is usually the first investigation when intussusception is suspected [Fig. 19.2]. An air or contrast enema study will also confirm the diagnosis and may be therapeutic (see the following).

Treatment

Enema reduction of intussusception should be attempted in most cases, unless there is clinical evidence of dead bowel, as demonstrated by peritonitis or septicaemia. Gas (air or oxygen) is the most commonly used enema medium and is preferred over the historical barium enema because of its superior efficacy and safety [Fig. 19.3]. Enema reduction is slightly less likely to be successful if there are prolonged symptoms (>24 h), if the patient is outside the usual age range (<3 months or >24 months) or if there is an established small bowel obstruction with air–fluid levels on x-ray. Providing there is no peritonitis, it is still worth attempting enema reduction in these circumstances. If the child remains stable clinically after attempted, incomplete reduction, a delayed repeat enema 30 min to 2 h later is performed. Overall, 80–90% of intussusceptions should be reducible using a gas enema.

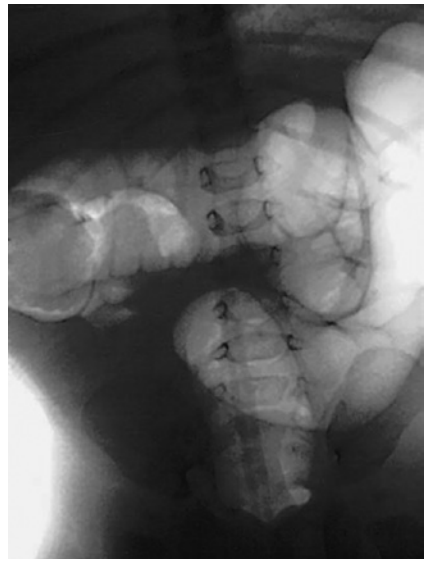


Figure 19.3 Gas enema showing the end of the intussusceptum, which confirms the diagnosis of intussusception (Reproduced with permission from Phelan *et al.* (1988)).

Technique of gas enema reduction

The patient is resuscitated with intravenous fluids and kept warm. A Foley catheter is inserted into the rectum. To achieve a seal, the catheter balloon is inflated and/or buttocks are strapped tightly together. Gas (usually oxygen from the wall supply) is introduced into the colon through the catheter, the pressure being controlled by a manometer [Fig. 19.3]. Under continuous fluoroscopic control, progress of the reduction is monitored. Sudden filling of the small bowel with gas suggests reduction is complete. The infant may be fed within hours of the procedure.

Surgery

Operative intervention is indicated when the delayed repeat enema fails to reduce the intussusception, where there is peritonitis clinically or where there is strong evidence of a pathological lesion at the lead point, for example, circumoral pigmentation of Peutz–Jegher syndrome. Laparotomy via a transverse right supra-umbilical incision, or laparoscopy, may be used. The intussusception is reduced by manipulation, although segmental resection may be required if there is gangrene or where there is a pathological lesion at the lead point, for example, Meckel's diverticulum or polyp.

Recurrent intussusception

Intussusception recurs in about 7% of patients and is more likely after enema reduction than surgery. It usually occurs within 2 or 3 days of the first reduction. Recurrence usually presents early and is treated the same way as a first episode, although the possibility of a pathological lead point should be considered.

KEY POINTS

- Intussusception should be suspected when children between 3 months and 3 years present with vomiting, colicky abdominal pain and intermittent pallor.
- Intussusception is diagnosed by palpating a central/right-sided mass and confirming with x-ray, ultrasound scan or gas enema.
- Hydrostatic enema reduction corrects intussusception in about 85%, with 7% risk of recurrence.

Further reading

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CHAPTER 20
Abdominal Pain: Appendicitis?

CASE 1

A 6-year-old boy developed central abdominal pain and high fever yesterday. Today, he has pain in the right iliac fossa. He looks flushed and has enlarged, tender cervical lymph nodes. There is tenderness in both iliac fossae (right more so than left), but no peritonism.

Q 1.1 What is the likely diagnosis?

Q 1.2 What tests are required?

CASE 2

Alex is a 5-year-old who has been vaguely unwell with abdominal pain for 2 days. Today, he has pain in the right iliac fossa. He limps into the consulting room but does not look flushed, and his temperature is normal (37.2°C). There is tenderness and guarding very lateral in the right flank.

Q 2.1 What is the likely diagnosis?

Q 2.2 What investigations, if any, will aid you making your diagnosis?

Improved diagnosis of appendicitis has been a major factor in reducing its morbidity in children. As a rule, abdominal pain in childhood lasting more than 4 h should be regarded as evidence of a potential abdominal emergency, until proven otherwise. Likewise, diarrhoea lasting more than 24 h should raise the possibility of pelvic, retrocaecal or retro-ileal appendicitis, particularly if associated with lower abdominal pain. An illness may start with gastroenteritis and diarrhoea, but later, appendicitis may supervene. Importantly, only a minority of children presenting with acute abdominal pain are found to have a genuine surgical cause. Where there is a high index of suspicion of appendicitis, the child must be reassessed frequently, with early referral to a paediatric surgeon.

Interpreting abdominal pain in children

The interpretation of abdominal pain can be difficult in children who are young or have developmental delay. Significant pain is often recognised late by parents and clinicians alike, because of children's inability to voice their symptoms adequately, but also because the pain of appendicitis may not be as dramatic in the younger age group.

Features suggesting that movement exacerbates the pain heighten the concern that peritonitis is present, for example, when an infant prefers to lie still, refuses to be cuddled, wants to be left untouched and is reluctant to be examined.

If abdominal pain persists, active observation must be continued and examination repeated until there are definite signs that indicate a need for an operation or until the pain has subsided. In most children whose pain is subsiding, an operation should be deferred if the physical signs are not completely diagnostic.

Assessment of physical findings

Examination of the older cooperative child is relatively straightforward, whereas in a young, sick and frightened child, physical examination requires great skill. The examination must be unhurried, gentle and performed with warm hands with the examiner seated beside the child. The child should be supine and straight, with arms resting alongside. Useful assessment may sometimes be made if the child is asleep. In the toddler, the abdomen may be palpated from behind with both hands while the parent cuddles the child's front against

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themselves. However, with a continually crying child, adequate assessment is difficult and re-examination must be undertaken later. Failure to acknowledge inadequate examination may result in serious diagnostic error.

Abdominal tenderness

Localised tenderness is found in conditions ranging from excess flatus, a faecally loaded colon or inflamed mesenteric lymph nodes to acute appendicitis and strangulated gut; consequently, tenderness *alone* is an insufficient reason for operation. Direct palpation over any distended or inflamed loop of the gut may elicit localised tenderness. This is a particular feature of the solitary distended loop in *closed-loop* intestinal obstruction but may also be observed in gastroenteritis. Inflamed visceral peritoneum will produce tenderness by direct, rather than by referred, pathways.

Localised tenderness in the right iliac fossa, without guarding or a strongly suggestive clinical history, may be due to causes other than appendicitis. Although localised tenderness is often the earliest sign of appendicitis, children requiring operation soon develop signs of peritonism to support the diagnosis. In contrast, tenderness due to non-surgical conditions typically subsides in the following days.

Peritonitis: Guarding and percussion tenderness

Children with localised or generalised peritonitis rarely display the board-like rigidity so often found in adults. More often, there is a variable degree of involuntary increased muscle resistance, referred to as *guarding*. Small differences in resistance in the lower and upper abdomen, or between right and left sides, may be significant when the findings are consistent.

Assessment of percussion tenderness is a sensitive test of peritoneal irritation in children and should be used in preference to *rebound tenderness*, which is unkind and unhelpful. Likewise, pain in the abdomen during micturition (as opposed to *dysuria*) is common in pelvic appendicitis, as the appendix lies close to and irritates the bladder.

There are no pathognomonic symptoms or signs of appendicitis itself, but when signs of peritonitis are localised to the right iliac fossa, appendicitis is likely.

Acute appendicitis may be missed when it presents with signs of local peritonitis outside the right iliac fossa. The interposition of other structures between the appendix and the anterior abdominal wall in pelvic, retrocaecal and retro-ileal appendicitis may delay the appearance of abdominal signs until relatively late, and even then, they may be atypical.

In retrocaecal appendicitis, tenderness is maximally high and lateral on the right side, whereas in pelvic appendicitis, or in retro-ileal appendicitis, the signs are more central and lower and may even be predominantly left sided.

A palpable mass

Apart from faecal masses and symptomless masses in the loin (Chapter 25), the most common mass in a child's abdomen is an appendiceal abscess (or phlegmon), particularly in children under 5 years of age, in the developmentally delayed child or when the appendix is in an unusual position.

Scrotal examination

Torsion of the testis or appendix testis occasionally presents with referred pain in the iliac fossa and may be mistaken for appendicitis. The scrotum must always be examined to exclude this important finding.

Rectal examination

Rectal examination is not indicated if appendicitis can be diagnosed on history and anterior abdominal signs alone. However, it may aid diagnosis of a pelvic mass with a perforated pelvic appendicitis should an ultrasound scan be unavailable or equivocal. When required, rectal examination should be performed by a member of the surgical team, having first gained informed consent from the carers and ensured a chaperone is present.

Signs arising in other systems

Infections in the ear, tonsils or respiratory passages may be accompanied by abdominal pain and vomiting and simulate an abdominal emergency. Measles or chickenpox may produce abdominal signs, as may many other viral infections, by causing mesenteric lymphadenitis. Acute appendicitis may coexist with other conditions, so that proven pneumonia, tonsillitis or generalised lymphadenopathy should not divert attention from any abdominal signs, which also may be present. Similarly, gastroenteritis may progress to appendicitis, so that even a well-established and undoubted diagnosis of gastroenteritis should be subject to review. Special

diagnostic difficulties may be presented by the abdominal crises of diabetic ketoacidosis, Henoch–Schönlein purpura and various haematological disorders, including sickle cell disease and haemophilia. In these conditions, an operative intervention is usually contraindicated. Active observation and re-examination for signs of peritonitis are essential.

Acute appendicitis

The mortality of acute appendicitis in children is less than 0.2%, due to early diagnosis and the management of fluids and electrolytes before and after operation.

The clinical diagnosis is determined by localised tenderness and objective signs of localised or generalised peritonitis. The value of history lies in arousing suspicion that an abdominal emergency is present and in determining its most likely cause. However, acute appendicitis is not only a common abdominal emergency, but also a great imitator, and it may appear in a variety of guises [Box 20.1].

Differential diagnosis

A perforated appendix is the only common cause of general peritonitis in childhood. In most children with an appendiceal *abscess*, there are signs of localised or generalised peritonitis. Occasionally, however, an appendiceal *mass* may be present with little or no constitutional upset or localised signs of peritoneal irritation.

Infections of the urinary tract may mimic appendicitis and vice versa. Lower urinary tract infection may give rise to tenderness in the right iliac fossa, but does not exhibit the guarding typical of peritoneal irritation. Conversely, a high retrocaecal appendicitis occasionally imitates acute pyelonephritis with prominent loin tenderness. In pelvic appendicitis, the child may complain of lower abdominal pain during micturition. Mild pyuria (20–50 white cells/mm^3) may occur in appendicitis due to an inflamed appendix adjacent to the ureter or bladder.

Referred pain to the right abdomen may occur with right lower lobe pneumonia or right testicular torsion.

Peritonitis in the young child

Peritonitis is a frequent complication of appendicitis that may be difficult to recognise in infants and young children. Tenderness may be diffuse rather than localised and marked guarding may be absent, even with advanced generalised peritonitis. Often, a lesser degree of involuntary muscular rigidity or guarding is encountered. Differences in muscle tone between the right and left sides, or between the lower and upper abdomen, are highly significant.

As localised peritonitis progresses, the signs become more definite in the right iliac fossa. Paradoxically, as peritonitis becomes more generalised and the abdomen more distended, the right iliac fossa signs may appear to diminish in some children. In this situation, abdominal distension and the child's reluctance to allow abdominal palpation are signs of great significance.

Box 20.1 The various presentations of acute appendicitis: Presentation and differential diagnosis

- Local tenderness in the right iliac fossa
  - Simple colic
  - *Bilious attack* (food poisoning)
  - Gastroenteritis
  - Acute constipation
  - Mild mesenteric adenitis
  - Urolithiasis
  - Deep iliac lymphadenitis
- Local peritonitis (mostly in the right iliac fossa)
  - Severe mesenteric adenitis
  - Primary peritonitis
  - Meckel's diverticulitis
  - Ruptured luteal cyst
  - Ovarian torsion
  - Omental torsion
  - Suppurating deep iliac lymph nodes
- Generalised peritonitis
  - Primary peritonitis
  - Perforated Meckel's diverticulum
- *Urinary tract infection*
  - Urinary tract infection
  - Acute pyelonephritis
- An inflammatory mass
  - Intussusception
  - Duplication of the gut
  - Ectopic kidney
  - Retroperitoneal masses (Chapter 25)
- Intestinal obstruction
  - Adhesive bowel obstruction
  - Internal hernia
  - Meckel's band
- Acutely painful scrotum (Chapter 29)
  - Torsion of testis
  - Torsion of appendix testis
- *Gastroenteritis* (from the retro-ileal or pelvic appendix)
  - Gastroenteritis

Even without distension, however, persisting pain with or without accompanying diarrhoea demands careful assessment, re-examination of the abdomen and sometimes a rectal examination.

Treatment

The management of acute appendicitis involves:

1. Adequate preoperative replacement of fluid and electrolyte deficits
2. Operative removal of the appendix
3. Irrigation of the peritoneal cavity to remove pus and contaminated free fluid
4. Effective antibiotic treatment to cover aerobic and anaerobic organisms commencing prior to the operation

A dehydrated, toxic child is an anaesthetic risk. Obstructed airways by inhaled vomitus, unexpected cardiac arrest and prolonged surgical shock with circulatory collapse are all minimised by adequate preoperative resuscitation. This includes intravenous replacement of fluid and electrolyte deficits and, in children with marked abdominal distension, nasogastric tube decompression of the bowel.

An operation by laparoscopy or laparotomy aims to remove the appendix and perform intraperitoneal lavage.

Antibiotics reduce the septic complications of appendicitis, which is a polymicrobial infection caused by bowel organisms. A combination of an anti-aerobic agent (e.g. cephalosporin derivative) and an anti-anaerobic agent (e.g. metronidazole) is effective, but complicated cases may require broader cover. Antibiotics should be commenced at induction, or earlier if there is significant preoperative delay or the child is unwell. Preoperative antibiotics can reduce wound infection resulting from intra-operative soiling by infected peritoneal fluid.

Abdominal pain of uncertain origin

There are some children [Box 20.1] where the final diagnosis remains in doubt. *Indigestion*, wind pains, acute constipation and other minor bowel disturbances are not objective diagnoses, but which tend to be attached to a number of children.

Mesenteric adenitis *non-specific viral infection* of the ileum causes difficulty in distinguishing some cases from acute appendicitis. The combination of high fever, mild abdominal tenderness that varies in location, the absence of guarding (the inflammation is confined to mucosa only) and failure of the signs to progress suggests mesenteric adenitis. A succussion splash in the right iliac fossa on palpation is typical and reflects an ileus with stagnant loops containing air and fluid without guarding from peritonitis.

Abdominal emergencies in developmentally delayed children

Acute appendicitis is still the most common abdominal emergency in developmentally delayed children, but perforation by an ingested foreign body is more frequent than in children with normal development.

The diagnostic difficulty depends on the severity of delay. In the most severely affected children, with hyperkinesia, hypertonia, inability to speak and a high threshold of pain, there may be few symptoms, and abdominal signs – for example, tenderness or rigidity – may be difficult to detect or evaluate. Abdominal distension and absent bowel sounds, although late developments, are usually present once attention is first drawn to the abdomen. Vomiting, fever and tachycardia may also be present.

Similar to preschool children, appendicitis in developmentally delayed children more often presents with a local abscess or spreading peritonitis. A pelvic mass or intestinal obstruction from the small bowel adhering to the wall of an appendiceal abscess is common. The delayed diagnosis leads to increased complications and, consequently, an increase in mortality.

Intestinal obstruction

A common cause of intestinal obstruction is a strangulated inguinal hernia [Box 20.2], which, if recognised, presents few problems in diagnosis or management.

In children with no previous abdominal operation, the cause of obstruction may be a volvulus (Chapter 7), Meckel's band or diverticulum (Chapter 23), an intestinal duplication (Chapter 7) or, very rarely, an internal hernia [see Box 20.2].

Most cases of obstruction in older children are due to adhesions following a previous operation. Recurrent pain, accompanied by bile-stained vomiting, generally causes these patients to present early. Visible or palpable

Box 20.2 Causes of bowel obstruction in children after the neonatal period

Common
- Adhesive bowel obstruction
- Intussusception
- Appendicitis
- Strangulated inguinal hernia

Uncommon
- Malrotation with volvulus
- Meckel's band (closed-loop obstruction or localised volvulus)
- Duplication cyst
- Internal hernia

distended intestinal loops, or air–fluid levels on an abdominal x-ray, confirm the diagnosis.

Bowel obstruction is less obvious and so more likely to be overlooked in (i) the early post-operative period after abdominal surgery, when pain is difficult to interpret and the expected delay in recovery from paralytic ileus may mask an early adhesive bowel obstruction, and (ii) proximal small bowel obstruction, in which pain and abdominal distension may be minimal and vomiting alone is the presenting feature.

Treatment

The mainstay of management is non-operative: oral intake is withheld, the stomach is aspirated by nasogastric tube, and resuscitation (and maintenance) intravenous fluids are commenced. Some children respond promptly to this regimen and symptoms may subside within a few hours.

Most adhesive bowel obstructions will self-resolve within 1–2 days of initiating the aforementioned non-operative management. However, continuing or increasing volumes of aspirate or persistent localised abdominal tenderness are sufficient grounds for surgery, preferably before a rising pulse rate, severe pain and increasing abdominal tenderness that suggest impending strangulation of the bowel.

Meckel's diverticulum

A Meckel's diverticulum may cause abdominal pain in a variety of ways. The tip of a Meckel's diverticulum may still be joined to the umbilicus by a fibrous band, which may entrap the bowel, causing a closed-loop obstruction or a localised volvulus. Therefore, A Meckel's diverticulum may also be suspected in intestinal obstruction in a child with no previous abdominal operation.

Alternatively, Meckel's diverticulum may become inflamed. The clinical manifestations are rarely distinguishable clinically from appendicitis, and the diagnosis often only becomes apparent at operation.

Finally, Meckel's diverticulum is the most common cause of major gastrointestinal bleeding in childhood and presents when the child passes dark-red stools associated with dull abdominal pain and tenderness. Often, the child looks pale and anaemic (Chapter 23).

Primary peritonitis

Primary peritonitis (also known as spontaneous bacterial peritonitis) presents with a sudden onset of high fever greater than 39°C with diffuse abdominal distension and tenderness with guarding. This is a primary infection of the peritoneum and the abdominal contents are *normal*.

The condition is more common in girls, but often, it is not related to tubal infection. It may occur in patients with ascites secondary to nephrotic syndrome. Laparoscopy is performed as the presumed clinical diagnosis is appendicitis with secondary peritonitis. Odourless peritoneal fluid with a *soapy* feel is found, along with a normal appendix. Sometimes, the infecting organism is *Streptococcus pyogenes*, but in most cases, the culture is negative. The prognosis is good and the peritonitis resolves rapidly.

Paediatric gynaecologic emergencies

In menarcheal or pubertal girls, a gynaecologic disorder may present with acute abdominal pain. This group of conditions includes intraperitoneal bleeding at the normal time of ovulation (Mittelschmerz bleeding) or rupture of a small luteal cyst. Tubal menstruation, torsion of the ovary and acute salpingitis are all uncommon. Physical examination of girls with any of these conditions will typically reveal lower abdominal tenderness and guarding. Pelvic inflammatory disease may also present with fever and vaginal discharge.

Rectal examination is occasionally performed after informed consent and with a nurse chaperone, but a pelvic/abdominal ultrasound scan will diagnose most conditions.

Ovarian torsion demands urgent surgery, and the ovary is usually preserved after untwisting the torsion.

KEY POINTS

- Abdominal pain requires careful physical assessment to determine the cause.
- Peritonitis is hard to diagnose in preschool children: beware of a toddler who refuses examination.
- Localised peritonitis in the right iliac fossa is likely to be appendicitis.
- Pelvic appendicitis (by ultrasonography or rectal exam) and testicular torsion (by scrotal exam) need exclusion in children with vague pains in the right iliac fossa.

Further reading

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CHAPTER 21
Recurrent Abdominal Pain

CASE 1

A 10-year-old girl presents with recurrent abdominal pain.

Q 1.1 What underlying fears may the parents have about the pain?

Q 1.2 How may the history and examination distinguish the causes of recurrent abdominal pain?

Q 1.3 What is the role of the surgeon in this situation?

Recurrent abdominal pain is a common problem in children. The child usually has frequent short-lived episodes of peri-umbilical colic. The attacks are unpredictable in onset and usually last only a few minutes, although they may last for longer periods of time. Often, they have gone on for many months, or even years. Sometimes, they may be exacerbated by stress at school or home or by intercurrent illness. Despite these psychological triggers, the pain itself is very real, and while its exact nature remains uncertain, it may be due to intestinal colic. It may be compared with the psychosomatic stress headaches or non-specific gastric problems seen in adults. Constipation and intestinal upset brought on by food allergies may also cause recurrent abdominal pain. A frequent reason some families present to a surgeon with these symptoms is because of an underlying parental fear of a serious cause for the pain, such as appendicitis, cancer or a *twisted bowel*. In fact, it is quite unusual to find a serious underlying cause for recurrent abdominal pain. Despite this, it is important to exclude these uncommon but more serious causes for abdominal pain: only then can the family recognise the true nature of the problem, which sometimes is stress induced. The diagnosis depends on a careful history and physical examination, augmented as required by targeted investigations. In children presenting to an emergency department with more severe colic, pains may be caused by transient ileo-ileal intussusception. This form in intussusception is probably triggered by disordered small bowel peristalsis in response to an enteric pathogen and usually resolves spontaneously. It can be distinguished easily from the more severe ileo-colic intussusception in infants by its transient nature.

History

The nature, severity and periodicity of pain is the key to the diagnosis. Recurrent abdominal pain is mild to moderate in severity. The pains come on suddenly and without warning in short-lived episodes lasting often only a few minutes and are often peri-umbilical. The episodes of pain are unpredictable and frequent and scarcely a day goes by without any. In contrast, pain due to surgical causes such as appendicitis, obstructive hydronephrosis or malrotation with volvulus is severe and persistent.

A child may find it difficult to quantify the severity of pain, which is best established by other factors. Severe pain will stop the child from normal activities such as play, or the child may be sent home from school. Severe pain will wake the child from sleep and may induce vomiting. Bile-stained vomitus is of particular significance in relation to the possibility of malrotation with volvulus. Surgical pain tends to be more continuous and prolonged, lasting for some hours, and may be localised in relation to the underlying cause: the pain of an obstructed kidney in the older child will be localised to one loin, while the pain of appendicitis will typically

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localise to the right iliac fossa. However, younger children find it difficult to localise pain. The periodicity of surgical pain is different from that of recurrent abdominal pain. A child with obstructive hydronephrosis may be well for many months and develop severe prolonged episodes of pain lasting for a week, followed by many pain-free months.

The family and social histories are critical. If a relative has recently developed cancer, the parents may have an underlying fear of malignancy in the child and be looking for reassurance. On the other hand, a strong family history of renal anomalies may suggest the possibility of hydronephrosis in the child. As stress may be a key factor in recurrent abdominal pain, the social history is of great importance: family breakdown, financial distress and moving to other schools are common problems. Stress at school may be due to many factors, such as poor student–teacher relations, bullying or unrealistic parental expectations. Sometimes, when one asks the question, 'How does your child get on at school?', the parents answer that there is no problem because the child is always a *straight-A* student. The stress of trying to meet these high parental expectations is often the trigger for recurrent abdominal pain, or at least exacerbate it. Many parents spend considerable amounts of time away from home due to work: the stress of separation may manifest itself in the child as recurrent abdominal pain. Occasionally, abdominal pain may be a presentation of child abuse.

Physical examination

In most patients with recurrent abdominal pain, the physical examination is entirely normal. The child appears to be perfectly well, and a careful and complete physical examination, including measurement of the child's height and weight on a growth chart, offers a powerful reassurance to the parents. The most common abnormal physical finding is of a faecal mass palpable in the left iliac fossa. Although this may not be the complete explanation of the pain, correction of the constipation may be helpful in reducing its severity or frequency.

A loin mass due to an enlarged hydronephrotic kidney is an uncommon finding. Weight loss associated with malaise and lethargy may indicate a more serious underlying cause for the pain.

Special investigation

In the majority of children, special investigation is often not clinically helpful, although it may be useful to allay specific parental anxieties. Normal blood investigations, including full blood count, electrolytes, liver function tests, amylase, screening for celiac disease and measuring C-reactive protein may help to exclude organic disease. Of all investigations, abdominal ultrasonography is the most useful and least invasive: the kidneys, bladder, ovaries, gall bladder, liver, spleen and pancreas may all be examined. The presence of appendicitis should be excluded. Further investigation should be reserved for patients with a possible surgical cause of the pain. The return on the investigation of recurrent abdominal pain is not particularly high, but occasionally, a child is treated for recurrent abdominal pain for many years before hydronephrosis is diagnosed on ultrasonography.

Gastroscopy or colonoscopy may be indicated in a select group of children who have upper or lower gastrointestinal symptoms. Children with Crohn disease may have weight loss, growth failure, delayed puberty and perianal disease. Where this is suspected, or where there is a family history of, inflammatory bowel disease, a faecal calprotectin should be obtained. Those children with oesophagitis, gastritis and/or duodenitis will usually have symptoms that distinguish them from other children with recurrent abdominal pain.

Treatment

Following exclusion of significant pathology, recurrent abdominal pain is treated by reassurance, by identification of the stress factors (if any) and by helping the family to understand the possible link between stress and the symptoms. It is hard to *cure* the pain, but if the family understands the pathology, they may help the child live with the symptoms. It may last many months or years but eventually subsides.

It is important to uncover any possible hidden fears the family may have, such as the risk of cancer. These underlying fears must be dispelled by demonstrating to the parents' satisfaction that the child is free of these problems.

Constipation may be a factor aggravating recurrent abdominal pain. It is a simple matter to use laxatives to

clear the faecal load, and in some children, this may be helpful in reducing the colic.

Some parents worry that their child has a *grumbling appendix* or *chronic appendicitis*. While diagnostic laparoscopy and appendicectomy are performed sometimes in a small, highly selected group of the more severely affected children, the results are variable, and if the selection is poor, the symptoms may be made worse. The best results are obtained in those children who were previously well and who undergo frequent hospitalisation for pain, which is typical of acute appendicitis (with few abdominal signs), that settles quickly. This diagnosis of *chronic appendicitis* (repeated short-lived episodes of acute appendicitis) is uncommon, and operative exploration should be a rare event.

The management of children with recurrent abdominal pain is a test of clinical acumen and counselling skills. The cases presenting for surgical opinion are usually more severe, and it is important to exclude any underlying *surgical* cause. A thorough clinical history and physical examination is the basis of diagnosis; extensive, traumatic or invasive investigations are rarely indicated, and if imaging is required, ultrasonography is most useful.

KEY POINTS

- Recurrent abdominal pain may be psychogenic, but occult organic causes must be excluded.
- Detailed history and examination is usually sufficient to exclude significant underlying pathology.
- Abdominal ultrasonography may reassure patients and their families.

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CHAPTER 22
Constipation

CASE 1

A 6-month-old baby presents with pain and rectal bleeding with defaecation.

Q 1.1 What is the likely diagnosis and management of this common problem?

CASE 2

A 6-year-old boy presents with a long history of faecal impaction and soiling.

Q 2.1 Discuss the diagnosis and treatment of this condition.

Constipation is a common problem in infancy and childhood. Severe acute constipation presents to the surgeon with abdominal pain or rectal prolapse. Chronic constipation may present with soiling or an abdominal mass.

Acute constipation

This is mainly seen in babies between 6 and 12 months. Dietary problems lead to the passage of a hard stool, which tears the sensitive anal lining to cause an acute anal fissure with pain and bleeding. The pain on defaecation makes the baby hold onto stool, and a cycle of constipation is established. Importantly, an anal fissure is now recognised to be a common symptom of milk allergy. The difficulty with stooling is easily treated with dietary advice, including reduction or elimination of cows' milk. Dietary change is the long-term solution, but relief of the acute problem is obtained by laxatives, such as lactulose or paraffin, to soften the stool. Disposable enemas or suppositories are useful to clear the initial hard stool from the rectum. Parents also may have an underlying fear that the rectal bleeding is caused by cancer, so this subject should be explored and the parents reassured. Acute constipation is sometimes seen in older children following an intercurrent viral illness or with bed rest after surgery. The results of treatment for acute constipation are excellent. In a child with previously normal bowel habits, the rectum maintains its muscle tone and recovers rapidly with treatment, though laxatives should be continued until the precipitating factors are corrected.

Chronic constipation

This is an increasingly common and debilitating problem in children, and the treatment may be difficult and prolonged. In most cases, the anorectal mechanism and bowel are normal, but in rare cases there may be an underlying cause, such as Hirschsprung disease [Box 22.1].

Chronic constipation presents with a history of many months or years of soiling, abdominal pain and abdominal distension. Generally, the diagnosis of constipation is made by the presence of hard faecal masses in the abdomen. These are felt along the line of the colon and especially in the sigmoid colon. These masses may be indented with digital pressure, which is a characteristic feature that helps differentiate faeces from other abdominal masses. Inspection of the anus may reveal faecal soiling with a lax, open anal canal.

Rarely, other features on physical examination may indicate a serious underlying disease [Box 22.2]. Hirschsprung disease usually presents with neonatal bowel obstruction (Chapter 7), but occasional cases present at a later age with chronic constipation. These children are usually sick, with

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Box 22.1 Predisposing factors in chronic constipation

1. Holding back – behavioural problems with toilet training
2. Dietary factors – low fibre and fluid intake and food allergy/intolerance
3. Post-operative – bed rest, inactivity and narcotics
4. Intercurrent illness, for example, chickenpox
5. Emotional upset at home or school
6. Uncommon organic causes – Hirschsprung disease, slow-transit constipation, spina bifida and anorectal malformations

Box 22.2 Organic causes of constipation

Neurological anomaly
Slow-transit constipation
Hirschsprung disease
Spina bifida
Spinal cord anomaly
Sacral agenesis
Anatomical anomaly
Anal stenosis
Pelvic tumour
Anorectal anomaly (post-operative)

poor nutrition and marked abdominal distension. The anal canal in Hirschsprung disease is tight, as against the lax anus seen in other causes of chronic constipation. Slow-transit constipation describes a group of conditions with (presumed) congenital defects in bowel motility due to functional anomalies of the neural plexuses of the bowel wall. These children present with chronic, unremitting constipation, which fails to resolve with normal treatment.

An early clue in both Hirschsprung disease and slow-transit constipation is delayed passage of the first meconium stool beyond 24 h after birth. Another useful clinical feature is that, despite infrequent bowel actions, the retained stool in patients with slow-transit constipation is usually soft. Many patients with slow-transit constipation have a deficiency of substance P in the myenteric nerves supplying the colonic muscle, or abnormalities of the interstitial cells of Cajal, although a small number have hyperplastic or hypoplastic ganglia.

Congenital anorectal anomalies usually present at birth as an anorectal malformation. However, some minor anomalies may present later with anal stenosis. The anus in this situation will be tight and anteriorly placed. Spina bifida anomalies are usually apparent at birth, but some cases of spinal dysraphism are not so obvious and may present later with constipation. Diastematomyelia, sacral agenesis and spinal cord lipoma may all be diagnosed by careful clinical examination of the spine. Digital examination of the rectum is useful in assessing constipation, but should only be undertaken after discussion with the patient (if appropriate) and parents and in the parents' presence.

Special investigations

In most cases of constipation, special investigation is unnecessary. A plain abdominal x-ray is sometimes performed to assess the extent of faecal loading. Contrast enema studies usually are rarely indicated.

If, on clinical history and examination, a specific underlying cause is suspected, further tests may be indicated, particularly if standard diet and laxative therapy has failed. Hirschsprung disease may be diagnosed with an open rectal biopsy showing aganglionosis. Diagnosis of slow-transit constipation entails a 3-day nuclear transit study to confirm abnormally delayed proximal colonic transit. The scintigraphic study is also helpful in diagnosing unrecognised food allergy, as this is associated with rapid proximal transit in the colon and holdup in the rectum.

Spinal dysraphism is diagnosed on a plain x-ray of the lower spine, and anorectal anomalies are often best assessed with an examination under anaesthetic.

Treatment

While chronic constipation is easy to diagnose, it is difficult to treat, and the course of treatment is often more prolonged than the parents expect. Treatment aims to empty the rectum and to keep it empty as often as possible for weeks or months until colonic and anorectal tone returns [Box. 22.1]. The normal rectum is empty most of the time. However, when a mass reflex of the colon conveys faeces into the rectum once or twice per day, the rectal receptors are stimulated, resulting in the urge to defaecate. This urge is usually suppressed until socially convenient. If the faeces remain too long in the rectum, water resorption makes the faeces hard and more difficult to pass. As more faeces accumulate in the rectum, distension of the rectal smooth muscle reduces its contractility and sensation. Eventually, this process causes faeces to bank up in the colon, and stools are only passed by overflow incontinence past a lax anal sphincter, which dilates in response to a chronically distended rectum.

Treatment follows four lines:

1. Dietary advice, with reduction or elimination of cows' milk and increase of fibre with a normal mix of the main food groups.
2. Behavioural training is required to establish normal toileting. This is initially difficult as the child has diminished rectal sensation and motility due to chronic distension.
3. Laxatives both soften the stool and stimulate the bowel. The child may be quite dependent on continuous laxatives for many weeks. New oral formulations containing polyethylene glycol have proved useful in severe, chronic constipation.
4. Enemas are the most invasive form of treatment but are only necessary in severe cases. Initially, full bowel washouts in the hospital may be required to clear gross faecal impaction. Less severe degrees of constipation respond well to small disposable enemas. In very severe cases, enemas may be given antegradely through an appendix stoma.

Treatment must be carefully supervised until a normal bowel habit is established. Constipation is often underestimated and undertreated. Soiling and abdominal pain in primary school children are all too common, as both parents and medical practitioners have an inadequate understanding of the problem. If the symptoms persist after 6 months of adequate therapy, the child should be referred to a tertiary centre for assessment and further investigation. Recent trials in our own department suggest that transcutaneous electrical stimulation, similar to TENS, may significantly improve colonic motility in slow-transit constipation and may become a standard therapy in the near future.

Rectal prolapse

Rectal prolapse is a particularly distressing consequence of constipation. Straining to pass a hard stool leads to prolapse of the poorly supported rectum in the young child. The rectum may reduce spontaneously, leaving blood and mucus around the anus, or the parents may need to reduce the prolapse. In children, this problem resolves quickly by treating the underlying constipation with laxatives and enemas. Rarely, rectal prolapse may indicate an underlying anomaly such as malabsorption due to coeliac disease or cystic fibrosis. Rectal prolapse is also seen in spina bifida because of paralysis of the *levator ani*.

KEY POINTS

- Acute constipation is common and easily treated.
- Chronic constipation (with or without soiling) is very distressing for families and often difficult to treat.
- Most children respond to good laxative and behaviour treatments.
- Persisting symptoms despite 6 months of adequate therapy need referral for investigation for possible organic disorder.
- Delayed meconium stool beyond 24 h of birth is suggestive of an organic bowel disorder.

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CHAPTER 23
Bleeding from the Alimentary Canal

CASE 1

A 2-year-old girl is being toilet-trained by her mother when a small amount of bright blood is seen at the anus.

Q 1.1 What are the common causes of minor rectal bleeding in childhood?

Q 1.2 What is the likely cause in this case?

Q 1.3 Describe the management of a fissure.

CASE 2

A previously well 8-month-old infant presents with hypovolaemia after passage of two very large bowel motions containing dark-red blood.

Q 2.1 What are the causes of major rectal bleeding in children?

Haemorrhage, large or small, may occur from any part of the alimentary canal and at any age. Sometimes, the haemorrhage threatens the child's life; on other occasions, it is an important sign of other medical or surgical pathologies.

Alimentary tract bleeding may present with a variety of symptoms, according to the level within the tract and the rate of haemorrhage. It may be *occult* and present as iron-deficiency anaemia, or it may be seen as blood passed per rectum; in this instance, there may be melaena (dark, changed blood) or bright red bleeding [Box 23.3].

If bleeding occurs into the oesophagus, stomach or duodenum, it may present as either *coffee grounds* or frank blood in the vomitus.

Coffee-ground vomiting

A small amount of blood mixes with the gastric contents, is denatured and changes to a brown colour. When vomited, these flecks of blood may have the appearance of coffee grounds. It is seen in a variety of conditions, including pyloric stenosis, reflux oesophagitis, non-specific oesophagitis and Mallory–Weiss syndrome.

Pyloric stenosis

Obstruction of the pylorus results in gastritis, where small amounts of blood mix with the gastric contents and then may be vomited. Hypertrophic pyloric stenosis, which occurs in approximately 1:300 infants, causes vomiting at about 1 month of age. Cardinal clinical features are projectile vomiting, visible gastric peristalsis and a pyloric *tumour* palpable in the epigastrium (Chapter 18).

Reflux oesophagitis

Gastro-oesophageal reflux into the lower oesophagus sometimes causes ulceration of its mucosal surface. Small vomits occur after meals and when lying flat, often with epigastric discomfort. Initial treatment of this common condition includes general measures such as thickening of the feeds and elevating the infant's cot end. More specific medical measures to reduce gastric acid include antacids, H_2-receptor antagonists (ranitidine) and proton pump inhibitors (omeprazole, lansoprazole). Surgical treatment (fundoplication) is sometimes necessary for complications (especially oesophageal stricture) (Chapter 18).

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Box 23.1 The causes of blood in vomitus

Coffee grounds
- Pyloric stenosis
- Reflux oesophagitis

Frank blood
- Oesophageal varices
- Peptic ulcer

Others
- Nose bleeds with swallowed blood
- Nasogastric tube ulceration

Rare causes
- Aneurysm of the bed of the tonsil
- Foreign body perforation of the aorta

Non-specific gastritis

This condition may be due to a viral infection and usually responds to measures which reduce gastric acidity.

Haematemesis

The vomiting of large amounts of frank blood means that there has been significant loss of blood into the stomach.

Oesophageal varices

Oesophageal varices are the result of portal hypertension, and in children, this occurs in two main groups:

1. Extrahepatic portal hypertension: thrombosis of the portal vein in the neonate (e.g. after umbilical sepsis) results in *cavernous malformation* of the portal system.
2. Intrahepatic portal hypertension, due to (i) cirrhosis of the liver caused by biliary atresia; (ii) inborn errors of metabolism, such as alpha$_1$-antitrypsin deficiency; (iii) chronic viral hepatitis; or (iv) cystic fibrosis.

Oesophageal varices are treated by prophylactic endoscopic sclerosant injections to prevent rupture and haemorrhage. However, they may bleed torrentially, requiring tamponade with a Sengstaken–Blakemore tube and followed, if necessary, by emergency surgical treatment. Emergency endoscopic treatments are effective but require an experienced endoscopist. Operative management consists of oversewing the varices and/or creating a shunt to join the portal system to the systemic venous system, aiming to lower the pressure in the portal system.

Peptic ulcer

Peptic ulcer disease is rare in children, but a *stress ulcer* may occur in a child of any age with severe burns, cerebral tumour, head injury or other forms of severe stress. In all these conditions, there tends to be an increased production of gastric acid, with resultant diffuse ulceration of the gastric lining or more localised ulceration in the duodenum. Peptic ulceration also may be a complication of drug treatment, for example, after administration of steroids. In adolescents who develop peptic ulceration, the aetiology is the same as in adults (i.e. *Helicobacter pylori* infection). There may be a strong family history of ulcer disease.

Management

For a bleeding peptic ulcer, the management is as follows:

1. Adequate resuscitation of the patient with blood replacement.
2. Commencement of proton pump inhibitor infusion.
3. Endoscopic upper gastrointestinal tract examination and injections as required.
4. Operative intervention to oversew the bleeding point is rarely required.

Mallory–Weiss syndrome

This may cause alarming haematemesis with bright blood and may occur in any child who vomits or retches continually. It is thought to be due to the formation of small longitudinal splits in the upper gastric mucosa. Fortunately, it responds to medical treatment if the vomiting can be stopped.

Iron-deficiency anaemia

Iron-deficiency anaemia in children is usually caused by poor dietary intake, but may be due to reflux oesophagitis or one of the other causes mentioned in Box 23.2.

Box 23.2 Occult bleeding causing iron-deficiency anaemia

- Reflux oesophagitis
- Haemangioma of the bowel
- Polyps of the bowel
- Inflammatory bowel disease

Box 23.3 Rectal bleeding in children

- **Neonatal**
  - Necrotising enterocolitis
  - Volvulus with ischaemia
  - Haemorrhagic disease of the newborn
  - Gastroenteritis
  - Anal fissure
  - Maternal blood
- **Sick child with an acute abdominal condition**
  - Intussusception
  - Gastroenteritis
  - Henoch–Schönlein purpura
- **Major haemorrhage from the gastrointestinal tract**
  - Oesophageal varices
  - Acute peptic ulcer
    - – gastric erosions
    - – duodenal ulcer
  - Meckel's diverticulum
  - Tubular duplications
- **Small amount of bright blood in well child**
  - Fissure
  - Polyps
  - Unrecognised prolapse
  - Haemorrhoids
  - Idiopathic
- **Chronic illness with diarrhoea**
  - Crohn disease
  - Ulcerative colitis
  - Non-specific colitis

Rectal bleeding

Rectal bleeding in children may be considered under various distinct clinical groups [Box 23.3].

Neonatal bleeding

There are two important surgical conditions and several important medical conditions that lead to rectal bleeding in children.

Necrotising enterocolitis (Chapter 7)

This is an important condition that has become more common as modern neonatal nurseries care for increasing numbers of extremely premature neonates. Most neonates with necrotising enterocolitis respond to supportive treatment, consisting of adequate ventilatory care, circulatory support, resting of the gastrointestinal tract and intravenous antibiotics. Some patients require operative intervention for full-thickness necrosis of the intestine, as revealed by free intraperitoneal gas on x-ray or by continued clinical deterioration despite intensive supportive care.

Volvulus neonatorum with ischaemia (Chapter 7)

Volvulus of the midgut occurs at any age, but is more likely in the neonatal period. Malrotation is associated with abnormal attachment of the midgut to the posterior abdominal wall via a narrow-based mesentery, which allows easy twisting of the entire midgut. The initial sign of green bile-stained vomiting results from luminal obstruction of the bowel; however, the most serious event, which may occur, is ischaemia of the midgut, due to obstruction of the vessels in the twisted mesentery. Bleeding from the bowel is a late sign, and very urgent surgical treatment is necessary at this stage if there is to be any hope for the baby. Such blood may be evident in the vomitus, gastric aspirate and/or per rectum.

Non-surgical causes

Haemorrhagic disease of the newborn is due to vitamin K deficiency and is prevented by routine administration of vitamin K_1. Gastroenteritis may occur in the neonatal period, resulting in blood mixed with diarrhoea. An anal fissure may occur at any age and is common in the neonate who has required a rectal examination. Swallowing of maternal blood, either during delivery or from a cracked nipple, may give rise to blood in vomit or stool, masquerading as alimentary tract haemorrhage.

A small amount of blood in a well child

A small amount of fresh blood may be passed rectally in a well child. This is by far the most common clinical group, and the cause of the bleeding often may be distinguished on the history alone.

Anal fissure

Anal fissure occurs at any age and usually is due to constipation (Chapters 22 and 27). The child passes a large, hard stool, which tears the anal mucosa, usually in the midline, either posteriorly or anteriorly. If old enough, the child complains of pain on defaecation, and there is bright blood on the surface of the stool [Fig. 23.1]. The fissure may be seen by gently parting the anus. Rectal examination causes severe pain and is ill advised. The fissure heals quickly, and even when a fissure is not seen, the history may be quite diagnostic. Sometimes, a *sentinel tag*, a mound of oedematous skin just external to the

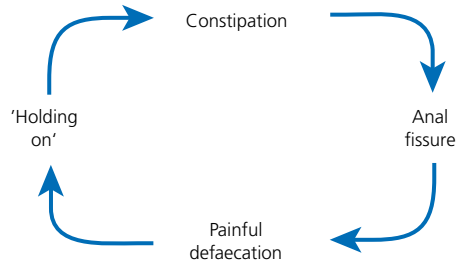

Figure 23.1 The cycle of anal fissure. The treatment aims to break the cycle.

fissure, is visible. Anal fissures in children almost always respond to adequate treatment of constipation, often with elimination of dairy protein. Local anaesthetic gels achieve little, and an operation on the anal sphincter is rarely indicated.

Polyps

Juvenile polyps are relatively common in children and should be suspected when there is no constipation or no pain on passage of a stool (Chapter 27).

Rectal prolapse

Prolapse of the rectum is easily diagnosed on the history or by direct observation (Chapter 27). Sometimes, the rectal prolapse may become congested or traumatised, bleed and then reduce spontaneously; the parents observe the bleeding without knowing its cause. Rectal prolapse may occur with malabsorption or chronic diarrhoea, straining with constipation and rarely as the presenting symptom of cystic fibrosis.

Haemorrhoids

Symptoms from haemorrhoids are rare in children but do occur. The presence of a venous malformation of the rectum and rectal varices should both be considered. In older children, haemorrhoids may cause bleeding, but may be treated conservatively. In some children, no cause for rectal bleeding may be found.

An ill child with an acute abdominal condition

In these children, the symptom of bleeding is not important in its own right, but points to another significant condition.

Intussusception

Intussusception presents with vomiting, colic, pallor and lethargy. In advanced cases, the stools become blood stained (the typical *red currant jelly stool*) due to a mixture of blood and mucus (see Chapter 19).

Gastroenteritis

Patients with severe gastroenteritis also often have vomiting, colic and specks of blood mixed with the stool. The separation of these patients from those with intussusception may be difficult in the child under 2 years of age (Chapter 19).

Henoch–Schönlein purpura

This condition causes arthralgia and a typical non-blanching rash over the extremities and buttocks. Submucosal haemorrhages in the bowel cause abdominal pain as well as passage of blood rectally. Henoch–Schönlein purpura needs to be distinguished from intussusception as it rarely requires operative intervention.

Chronic illness with diarrhoea

Crohn disease may occur anywhere in the alimentary tract and should be suspected in a patient with a chronic illness, unexplained fever, weight loss, bowel symptoms and chronic blood loss in the stools (Chapter 24). In patients with ulcerative colitis, the diarrhoea is more prominent, and again, it may contain blood. In non-specific colitis, there is usually involvement of only the lower part of the large bowel with less general symptomatology.

Major haemorrhage per rectum

In these patients, the haemorrhage is enough to cause anaemia or to require acute transfusion. The causes range from oesophageal varices and peptic ulcer (as discussed under the heading of vomiting) to Meckel's diverticulum and tubular duplications (both latter anomalies may contain heterotopic gastric mucosa).

Meckel's diverticulum

Meckel's diverticulum occurs in 2% of the population. In a small proportion of these heterotopic gastric mucosa forms part of the lining of the diverticulum [Fig. 23.2]. Acid produced by the gastric mucosa causes ulceration of the adjacent ileal mucosa. Bleeding usually presents as painless *brick-red* stools with associated

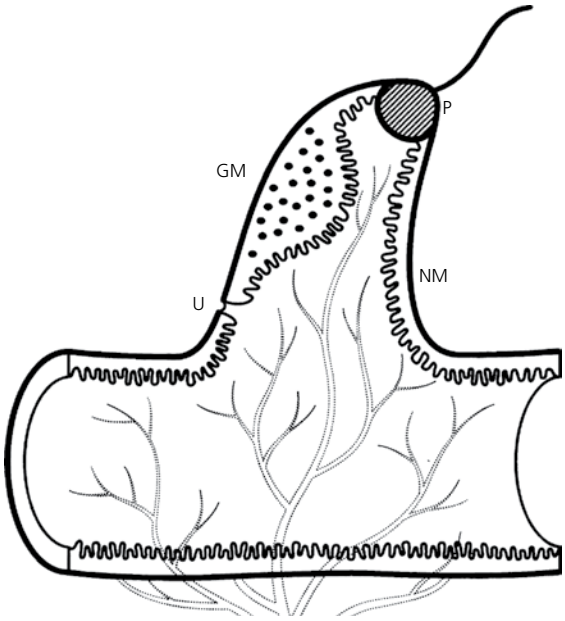

Figure 23.2 Meckel's diverticulum. A composite diagram showing heterotopic pancreas (P) and gastric mucosa (GM). An ulcer (U) lies in the adjacent normal ileal mucosa (NM). The site of attachment of a vitello-intestinal or Meckel's band is shown at the tip.

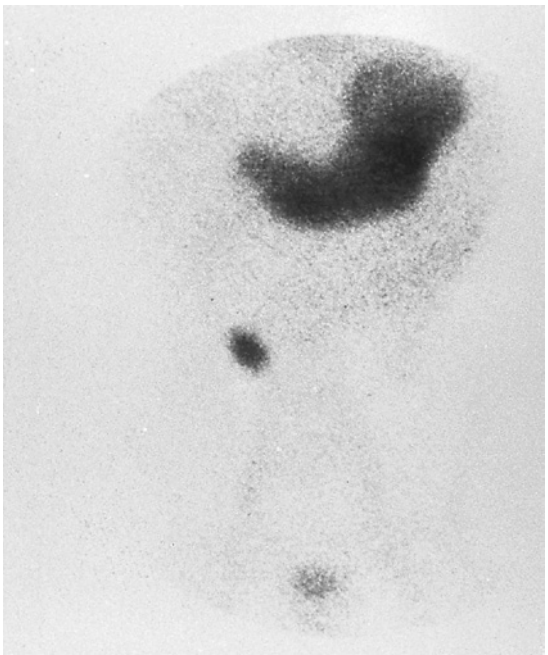

Figure 23.3 A technetium scan showing heterotopic gastric mucosa in a Meckel's diverticulum.

Box 23.4 Complications of Meckel's diverticulum

1. Bleeding
2. Intussusception with an inverted diverticulum
3. An associated fibrous band causing a small bowel obstruction
4. Diverticulitis (rare in children)
5. Peptic ulceration with ileal perforation
6. Strangulation of diverticulum by its own band
7. Strangulation of diverticulum in an inguinal hernia

marked anaemia. The patient may require transfusion, but the bleeding usually stops spontaneously without an emergency operation. The definitive investigation is laparoscopy, but a technetium scan may show the heterotopic gastric mucosa [Fig. 23.3]. A Meckel's diverticulum may result in a variety of other complications [Box 23.4].

Tubular duplications

These are much less common than a Meckel's diverticulum. Tubular duplications of the small bowel occur in the mesenteric side of the bowel and communicate proximally or distally with the bowel. They may be lined by heterotopic gastric mucosa and cause bleeding when adjacent small bowel mucosa becomes ulcerated. Like a Meckel's diverticulum, they may be demonstrated by a technetium nuclear scan.

KEY POINTS

- The cause of bleeding may often be determined from the site, colour and volume of blood passed.
- *Coffee grounds* in vomitus suggest gastritis.
- A small volume of bright blood passed rectally is characteristic with anal fissure.

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CHAPTER 24
Inflammatory Bowel Disease

CASE 1

A 6-year-old girl presents with a 1-month history of weight loss and mild diarrhoea, containing blood and mucus.

Q 1.1 What is the likely diagnosis and how is it confirmed?

CASE 2

A 12-year-old boy presents with vague pains in the abdomen, some weight loss and a perianal abscess.

Q 2.1 How is the diagnosis made?

Q 2.2 How are the different forms of inflammatory bowel disease distinguished?

Three categories of inflammatory bowel disease are encountered in childhood:

1 Crohn disease
2 Ulcerative colitis
3 Inflammatory bowel disease of indeterminate pathology

Incidence

Crohn disease is by far the most common category of inflammatory bowel disease. During the past 25 years, there has been a dramatic increase in the incidence of Crohn disease worldwide. This disease was almost unknown in childhood prior to 1980. Ulcerative colitis has also increased in incidence in the last 2–3 decades.

Crohn disease

Crohn disease is a chronic inflammatory disorder of unknown aetiology that can affect any part of the gastrointestinal tract from mouth to anus. It is a transmural inflammatory process, which most commonly occurs in the terminal small intestine and colon. There is a high incidence of involvement of the large bowel and rectum in children and adolescents.

Clinical features

Age of onset of symptoms

The mean age of onset of Crohn disease in Victoria is currently 11 years.

Symptoms and signs

Crohn disease presents with a broad spectrum of symptoms and signs. The most common symptoms include recurrent abdominal pain and bowel disturbance, usually diarrhoea together with rectal bleeding. However, these symptoms may be relatively mild, and patients may present with long-term effects of the disease such as weight loss, growth failure and delayed onset of puberty.

Delay in diagnosis may occur because not all doctors are aware of the relatively high incidence of Crohn disease in childhood and the variety of non-specific presenting symptoms.

Perineal inflammation

One of the most common modes of presentation is perineal inflammation, occurring in one-third of paediatric patients with Crohn disease, and this is usually associated with rectal disease. Accordingly, children or adolescents who present with a perianal abscess and associated anal fistula should have the abscess wall biopsied at the time of drainage, to exclude underlying

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Crohn disease. In addition, they need to be referred for full upper and lower gastrointestinal endoscopy, with measurements of faecal inflammatory indices and possible small bowel imaging studies.

Extra-intestinal manifestations of Crohn disease

Examples of extra-intestinal manifestations include arthritis and erythema nodosum, which may be presenting symptoms.

Unusual modes of presentation of Crohn disease

Occasionally, Crohn disease may present with acute right-sided abdominal pain and gastrointestinal disturbance, mimicking acute appendicitis. The diagnosis is then made at laparoscopy/laparotomy.

Cheilosis

Uncommonly, patients may present with chronic inflammation of the mouth and lips, so-called cheilosis, manifested by oedema, erythema and fissuring of the lips, as part of the syndrome of orofacial granulomatosis. Biopsy reveals evidence of chronic inflammation including granulomas consistent with Crohn disease.

Investigations

Role of endoscopy

Endoscopy has a crucial role in diagnosis, initial evaluation and continuing assessment of Crohn disease.

Upper and lower (*top and tail*) gastrointestinal endoscopy with biopsies is the key investigation for the diagnosis and initial assessment of the extent and severity of the disease. Colonoscopy through to and including the ileum, together with biopsy, provides a good chance of diagnosing Crohn disease because there is a high incidence of macroscopic colonic involvement in children under 6 years of age. Gastroscopy is performed as well as colonoscopy because involvement of the upper intestinal tract is common. Periodic endoscopy is important to assess the response to treatment and disease distribution.

In Crohn disease, the inflammation is typically segmental, and the characteristic appearance is single or multiple ulcers with normal intervening mucosa. In some instances, the diagnosis may be made by serial biopsies even when the macroscopic appearances are normal. Histological diagnosis depends on finding granulomas, in association with other chronic inflammatory changes in the bowel wall.

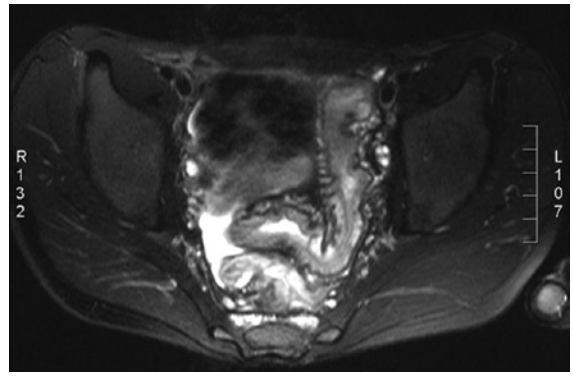

Figure 24.1 MRI is used to assess the small bowel affected by Crohn disease that is not accessible to endoscopy.

MRI

This imaging is used to assess Crohn disease of the small bowel after full bowel preparation as for colonoscopy and also as a filling agent, which will distend bowel loops [Fig. 24.1]. Examination of the pelvis and the anorectum is carried out without the need for bowel preparation. It also demonstrates sphincter anatomy and distortion or damage related to the inflammation.

CT scan with oral contrast

Abnormal findings include an irregular bowel contour, longitudinal ulcers and fissures, luminal narrowing by oedema and separation of loops by mural thickening. There may be evidence of stricture formation associated with dilated proximal bowel. Bowel loops may be displaced by an inflammatory mass.

Other investigations

FBE detects evidence of anaemia, and liver function tests exclude associated liver disease. Stool cultures are performed to rule out chronic infection due to such enteric pathogens such as *Clostridium difficile*, *Salmonella*, *Shigella*, *Campylobacter* and *Yersinia*. Faecal calprotectin is a very useful measure to determine the likelihood of significant gastrointestinal inflammation.

Treatment

The aetiology of Crohn disease is unknown, although it is recognised to represent an abnormal immune response directed against the gut. Treatment is directed at suppressing the immune response.

Medical treatment

High doses of oral steroids are used to induce remission, for example, prednisolone 2 mg/kg (maximum 60–75 mg/day) for 4 weeks, with gradual reduction to nil after 8 weeks. Sulphasalazine, to prevent relapses, is introduced and is built up to a dose of 50 mg/kg/day. In moderate to severe disease and if there is steroid dependency, agents such as azathioprine, are introduced early. Metronidazole may be helpful for perianal Crohn disease. Biological therapies such as infliximab appear to be very effective.

Nutrition

Children with inflammatory bowel disease fail to grow because of the disease and its effect on appetite and caloric intake. High caloric dietary supplements have their place in management. Enteral feeds can be used for either nutritional supplementation or direct treatment, as exclusive enteral feeding has a direct anti-inflammatory effect almost equivalent in potency to steroids. Rarely, parenteral nutrition may be required.

Surgical treatment

The indications for surgical treatment of Crohn disease are as follows:

1 Perianal disease
2 Intestinal complications
3 Acute abdomen – possible acute appendicitis

1. Perianal disease

Perianal disease is the most common indication for surgical intervention in Crohn disease [Fig. 24.2], which is invariably associated with rectal and colonic involvement. There may be extensive inflammation of the soft tissues of the perineum, scrotum, penis or vulva.

Skin tags and anal fissures are common and usually do not require surgery. Perianal abscess is also common and requires incision and drainage. There is usually an associated anal fistula Occasionally, insertion of a seton suture along a chronic fistula tract may be appropriate to drain an infection associated with it and promote healing. More extensive suppuration may produce an ischio-rectal abscess. Appropriate surgical management of this complication is drainage of the abscess and possible faecal diversion with a colostomy or ileostomy to help control the infection. Faecal diversion is usually effective, but it does not necessarily influence the underlying Crohn disease. The potential end result of ischio-rectal sepsis is damage to the sphincters. Once surgical drainage is achieved, medical therapy can be instituted with biological agents such as infliximab or adalimunab in the long term.

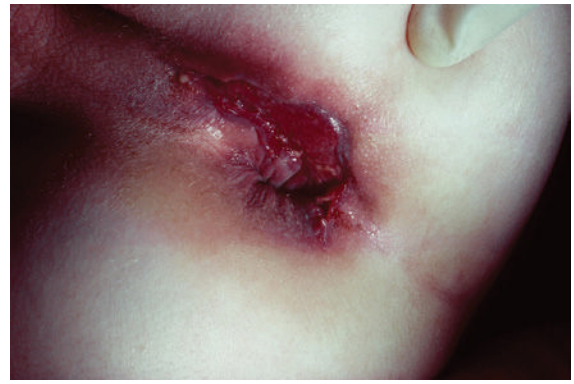

Figure 24.2 Perianal disease in a child with Crohn disease.

Perineal disease

Occasionally, there may be extensive involvement of the perineal soft tissues extending into the scrotum, penis or vulva. This may be manifest by unsightly, painful oedema and inflammation of the scrotal or vulval tissue.

Surgical treatment for complicated perineal and perianal Crohn disease aims to control infection and promote healing, which is often protracted despite appropriate therapy.

2. Intestinal complications

These complications are due to transmural inflammation and include the following:

Localised stricture formation
Localised disease unresponsive to medical treatment
Localised disease associated with growth delay and often delay in pubertal development
Inflammatory mass
Intestinal fistulae
Rectal stricture

Surgery for these complications aims to preserve as much intestine as possible.

Localised strictures may be treated by simple stricturoplasty (without loss of bowel) or resection. An important role for surgery is resection of localised disease associated with growth delay, delayed pubertal development or

both, despite maximal medical treatment. In most of these patients, a sustained remission can be expected, together with catch-up growth and resumption of normal schooling. The best results can be anticipated with resection of localised ileo-caecal disease before or at the onset of puberty. Resection may be necessary for inflammatory masses and fistulae.

Rectal strictures are common in paediatric Crohn disease and are often associated with perineal inflammation. Their management includes steroid therapy, both systemic (as previously mentioned) and local steroid medication as administered in enema form. The surgery includes regular dilatations of the stricture under GA, usually in association with endoscopic evaluation. The associated perineal inflammation tends to resolve with control of the rectal stricture.

Ulcerative colitis

Ulcerative colitis is a chronic inflammatory disease of the rectal and colonic mucosa, the aetiology of which is unknown.

Clinical features

The onset is usually insidious, but an acute onset similar to a *Salmonella* infection can occur. The onset of disease is usually after 5 years of age, but can be as early as the first year of life.

The typical features are as follows:

1 Unexplained bloody diarrhoea, with mucus, lasting more than 2 weeks
2 Anaemia
3 Fever
4 Weight loss

All degrees of severity are encountered, and the predominating symptom varies from one patient to another.

Perianal complications occur in a small percentage of patients and include ulcers, abscesses and fistulae and raise the suspicion that the diagnosis was actually Crohn disease. Rarely, perianal complications may be the presenting problem, but more usually, perianal disease is preceded by a period of diarrhoea.

Investigations

1. Colonoscopy

Colonoscopy accurately assesses the extent of macroscopic disease, and biopsies achieve the diagnosis. In ulcerative colitis, inflammatory changes are seen in the rectum and extend for varying distances proximally in the colon. The changes range in severity from loss of the normal mucosal sheen and vascularity with associated mucosal friability to diffuse ulceration with blood and pus in the lumen. Numerous biopsies at colonoscopy will confirm the histological diagnosis and indicate the severity of inflammation at various levels. The histology can be reported, at best, as *consistent with* ulcerative colitis, for there is no pathognomonic lesion. At diagnosis, the inflammation is confluent, but after therapy has started, it may become patchy and potentially confused with Crohn disease.

In some instances, even when macroscopic appearances at endoscopy are normal, multiple biopsies will provide diagnostic histological changes.

2. Contrast imaging

This investigation may show a *sawtooth* or marked irregularities in the mucosa, with deep ulceration. Later, the colon becomes narrow, rigid and devoid of visible peristalsis or haustration [Fig. 24.3]. Finally, there may be pseudopolyps or stenosis due a fibrous stricture.

3. Other tests

Other tests include an FBE, to demonstrate anaemia. Bacteriology tests should include a careful search for enteric pathogens, including *Clostridium difficile*, *Salmonella*, *Shigella*, *Campylobacter* and *Yersinia*. Blood for *Yersinia* antibodies should be collected.

Natural history

Improvements in medical treatment, with more aggressive use of immunosuppressants, such as azathioprine, have produced better control of the disease. Many children have remissions lasting several years, to the extent that the diagnosis is subsequently questioned. A small proportion continue to have recurrent lapses and may require colectomy.

Risk of malignancy

The incidence of carcinoma is directly proportional to the duration of disease. The risk in the first 10 years of disease is very low, but after 10 years, the rate increases by 10% for each decade and may be even higher if associated with primary sclerosing cholangitis.

Surveillance by regular colonoscopy to detect premalignant dysplasia is an important component of management of paediatric patients with ulcerative colitis.

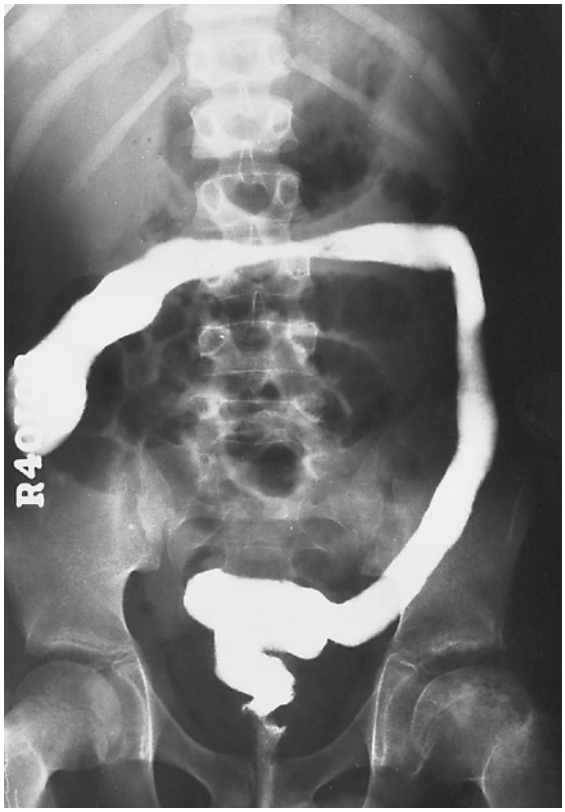

Figure 24.3 Ulcerative colitis. Featureless colon with *sawtooth* outline on barium enema.

Medical treatment of ulcerative colitis

The principles of medical treatment are similar to those for Crohn disease, which have been enumerated previously. However, many of the drugs for ulcerative colitis can be given as enemas, if there is significant inflammation in the rectosigmoid.

Surgical treatment of ulcerative colitis

Procto-colectomy is curative, but other surgical procedures have a place in the treatment of this disease.

The indications for surgical treatment are as follows:

1. Severe inflammation unresponsive to medical treatment
2. Severe disease associated with growth delay and delayed pubertal development
3. Long-term risk of malignancy (see previous text)
4. Acute haemorrhage
5. Perforation
6. Toxic megacolon

It is most important to realise that absent symptoms should not be taken as evidence of quiescent or inactive disease or of healing. Surveillance by regular colonoscopy should continue in proven cases of ulcerative colitis.

Surgical options

Subtotal colectomy with ileo-rectal anastomosis. This procedure may be considered where there is minimal rectal inflammation, particularly in an older adolescent. This procedure may avoid the need for an ileostomy. However, this option requires continuing endoscopy surveillance, together with biopsies every 6 months. Removal of the rectum will be necessary after 10 years of disease.

Procto-colectomy. This procedure may be achieved with a number of techniques as follows: ileo-anal anastomosis with ileal reservoir and ileo-anal anastomosis without reservoir – Soave pull-through procedure.

Chronic inflammatory bowel disease of indeterminate pathology

In a small number of patients, the pathology is uncertain, and a diagnostic dilemma arises as to whether the patient has ulcerative colitis or Crohn disease. The principles of medical treatment are similar to those outlined previously, but surgical treatment depends on what is considered to be the most likely condition as judged on clinical, endoscopic and histological evidence.

KEY POINTS

- Crohn disease is now relatively common in children and adolescents.
- Recurrent abdominal pain with intermittent diarrhoea suggests IBD.
- Perianal sepsis in adolescents suggests Crohn disease.
- IBD diagnosis requires sophisticated imaging, endoscopy and biopsy.
- Surgical treatment is required if medical therapy fails and for prevention of colonic cancer.

Further reading

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CHAPTER 25
The Child with an Abdominal Mass

CASE 1

A 9-year-old boy presents with a 2-week history of intermittent pain in the left loin and flank. Physical examination reveals a large, smooth, left-sided mass in the abdomen. The mass is firm, but not solid, and is *ballottable*.

Q 1.1 What is the likely diagnosis?

Q 1.2 What investigations might be needed?

CASE 2

A previously well 4-year-old girl presents with a large, smooth and solid mass in the right side of the abdomen, noted incidentally during examination. Her blood pressure is 110/80.

Q 2.1 What is the differential diagnosis?

Q 2.2 What treatment might be needed?

CASE 3

A 5-year-old girl has been unwell and pale in recent weeks. The school nurse finds a large, hard and craggy mass in her upper abdomen.

Q 3.1 What investigations are needed, and what is the likely diagnosis?

Q 3.2 What would you tell the parents?

The following points need to be considered when assessing a child with an abdominal mass:

1. Age of the patient and the most likely pathological process arising in that organ at that age
2. Length of history and type of symptoms, which may also implicate a particular pathological process (e.g. tenderness of the mass suggests infection or bleeding)
3. *Site* of the mass and its precise characteristics, which will suggest the probable organ of origin

Normal and abnormal masses

The most common abdominal masses in infancy and childhood are non-pathological. They are usually accounted for by the liver, which normally extends below the right costal margin until 3–4 years of age; faeces in the colon; or full bladder [Table 25.1].

In addition, three common pathological conditions often present with an abdominal mass in childhood: Wilms tumour (nephroblastoma), abdominal neuroblastoma and hydronephrosis. These pathological conditions have certain features in common:

1. They are most common in infants and toddlers between 1 and 3 years of age.
2. The mass is typically large when first detected as the normally protuberant infantile abdomen may conceal masses of smaller sizes.
3. The mass itself is usually the presenting feature, while general or local symptoms are typically minimal or absent.

Neuroblastoma is an exception to the latter point: a significant number of children with neuroblastoma present systemically unwell, including failure to thrive.

As stated earlier, the site of the mass assists in formulating a differential diagnosis. A mass situated in the midline in the upper abdomen is most likely a primary abdominal neuroblastoma, particularly if the child is less than 4 years of age. Other possible causes include massive hepatic metastases (e.g. from a primary neuroblastoma) and primary hepatoblastoma. Masses arising in the loin can be palpated

Jones' Clinical Paediatric Surgery, Seventh Edition. Edited by John M. Hutson, Michael O'Brien, Spencer W. Beasley, Warwick J. Teague and Sebastian K. King.

Table 25.1 Common normal and abnormal abdominal masses in children

Normal	Abnormal
Liver	Hydronephrosis
Faeces	Wilms tumour
Bladder	Neuroblastoma
Lower pole of kidneys	

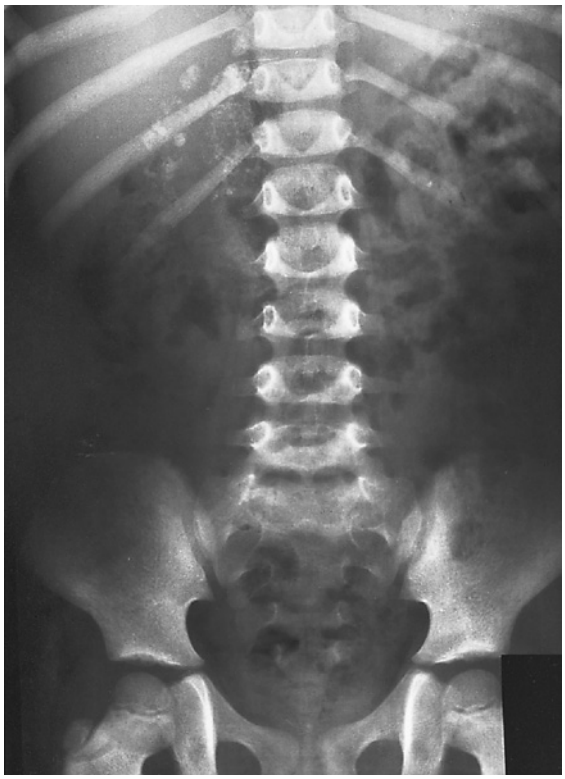


Figure 25.1 Neuroblastoma, showing calcification in the right paravertebral region.

extending below the rib margin towards the iliac fossa and are most likely due to hydronephrosis or Wilms tumour.

Investigations

Simple imaging can assist in diagnosis, for example, plain abdominal x-ray and abdominal ultrasonography. Plain x-ray may show calcification within the mass, which is more common in neuroblastoma [Fig. 25.1] than Wilms tumour, and does not occur in hydronephrosis, unless there is a renal stone. Also, blood tests can be indicated, for example, full blood count, electrolyte analysis and tumour markers as appropriate.

Abdominal ultrasonography will determine whether a mass is cystic or solid. Cystic masses may be seen with hydronephrosis, multilocular or simple renal cysts, multicystic dysplastic kidneys or a dilated renal pelvis from high-grade vesicoureteric reflux. Ultrasonography documents the size, position and extent of a solid tumour and may demonstrate blood vessel involvement (e.g. extension of a Wilms tumour into the inferior vena cava). Lymph node involvement and metastases may be demonstrated, but these and other oncologically relevant imaging findings are ordinarily investigated using abdominal computerised tomography (CT) scan. Some renal masses warrant a nuclear scan to quantify relative function of each kidney and, with some isotopes, obstruction of the renal tract, for example, MAG3 renogram to investigate hydronephrosis. Magnetic resonance imaging (MRI) and angiography to determine vascular supply have select roles. The management of hydronephrosis is described further in Chapter 33.

Neuroblastoma

Neuroblastoma is the most common extra-cranial solid tumour of childhood. It is an embryonal tumour that arises from fetal neural crest cells and may occur at any site in the sympathetic nervous system. The most common sites are the adrenal gland [Fig. 25.1], elsewhere in the abdomen, and the sympathetic chain or the sympathetic plexus in the mediastinum or pelvis.

Metastases are present in 70% of patients at diagnosis and may be in the bone marrow, the cortex of long bones, the regional or distant lymph nodes, skull, eyes, liver and skin. The numerous sites of primary tumour and propensity to early metastasis account for the wide variety of possible presentations. Examples include:

1. Proptosis and periorbital ecchymoses due to ocular metastases
2. Long bone pain and tenderness due to bony metastases
3. Rubbery lymph nodes in the neck or axilla due to lymph node metastases
4. Bone marrow involvement manifesting as any or all of pain, limping, paralysis or weakness and failure to thrive with or without anaemia

5 Palpable skull nodule due to skull metastases
6 Paraplegia of rapid onset due to intraspinal extension of a paravertebral primary
7 Horner syndrome due to involvement of the stellate ganglion (sympathetic chain)
8 *Blueberry muffin spots* on the skin of infants due to skin metastases
9 Diarrhoea caused by tumour metabolites, for example, vasoactive intestinal peptide (VIP)

In view of the invasiveness and malignant potential of neuroblastoma it is a paradox that, in a minority of cases, the tumour regresses completely with a spontaneous cure. This unusual tumour behaviour is restricted to children younger than 18 months who present with metastases strictly confined to the skin, liver and/or bone marrow: clinical stage *MS* (previously called stage *4S*). The striking skin metastases in such infants are pathognomonic for this stage and are described as *blueberry muffin spots*. Resection of the primary tumour in these children has no impact upon local relapse of the tumour nor the patient's overall survival.

Diagnostic criteria

The diagnosis of neuroblastoma requires one or both of the following criteria:

1 Unequivocal pathologic diagnosis from tumour tissue *or* raised serum catecholamines (i.e. dopamine, adrenaline, noradrenaline) *or* raised urinary catecholamine metabolites (VMA, HVA)
2 Unequivocal pathologic evidence for bone marrow involvement (evident in 65–75% cases at presentation) *together with either* raised serum catecholamines *or* raised urinary catecholamine metabolites

Biopsy may be of the suspected abdominal mass or metastases, for example, lymph nodes. This is now often performed percutaneously under radiological guidance, but open tumour biopsy has select indications. Bone marrow assessment requires bone marrow biopsy and trephine. Previously, catecholamine metabolites were measured from a 24 h urine collection sample, but this is cumbersome, and spot testing is now considered preferable.

While the diagnosis of neuroblastoma may be made without tumour biopsy, tissue biopsy for tumour histology and biological features is essential for risk stratification. Further assessment of patients with confirmed neuroblastoma includes an abdominal CT to look for image-defined risk factors as a part of clinical staging and a metaiodobenzylguanidine (MIBG) nuclear scan. The MIBG isotope is avidly taken up by 90–95% of neuroblastomas and so identifies both primary and metastatic lesions. Other imaging routinely performed to inspect for metastases include a CT chest and bone scan.

Treatment

Treatment of patients with neuroblastoma is coordinated in a comprehensive oncology service and protocolised in accordance with the child's risk stratification. There is a wide range of possible treatment options, including:

1 *Observation only*: this is limited to a select group of patients, typically in the under 18-month age group with stage *MS* disease.
2 *Operative excision alone*: also limited to a select group of patients.
3 *Operative excision with neo-adjuvant and adjuvant chemotherapy*: this is the most common treatment pathway and may be complemented by autologous stem cell transplant.
4 *Radiotherapy*: usually reserved for high-risk disease in addition to operative excision and chemotherapy.
5 Biological and immunological therapies are an increasingly important component of the treatment regimen in neuroblastoma – usually limited to high-risk patients and patients with recurrent disease.

When undertaken, the goal of operative excision may not be complete excision. Rather, the surgeon must weigh the benefits of complete resection against the risks of what can be prolonged and highly morbid surgery to achieve complete resection. Preoperative CT image-defined risk factors for non-resectability include tumour encasement of the aorta, vena cava or iliac vessels. The presence of such risk factors may be an indication for neo-adjuvant (preoperative) chemotherapy prior to restaging and consideration for tumour debulking surgery.

The prognosis is highly dependent upon the stage of disease at presentation. High-risk disease is currently associated with a 5-year survival rate of 50%.

Wilms tumour

Wilms tumour, or nephroblastoma of the kidney, is the most common renal malignancy in childhood. It arises from primitive embryonic cells and produces a mixed

histological picture of epithelial structures resembling tubules and a variety of mesenchymal tissues, including striated muscle fibres.

Wilms tumour is typically sporadic in origin. However, it may be associated with a number of syndromes, including Beckwith–Wiedemann; Wilms tumour, aniridia, genitourinary malformations and mental retardation (WAGR); and Denys–Drash. A genetic basis is not uncommon, with tumour suppressor gene WT-1 deleted in WAGR and Denys–Drash and WT-2 associated with Beckwith–Wiedemann. Wilms tumour is also seen in association with hemihypertrophy, aniridia and familial Wilms tumour. Bilateral disease is present in 6% of affected children.

Clinical features

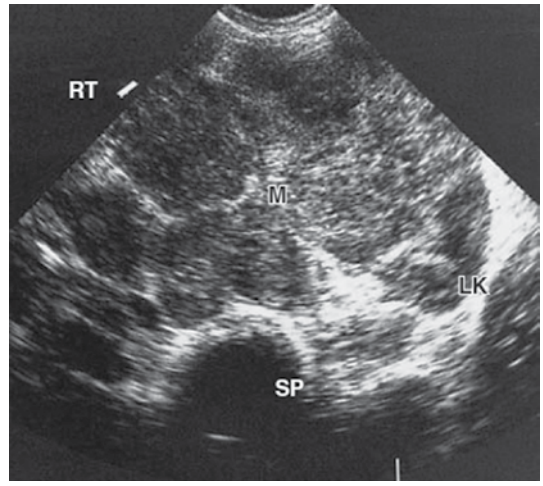
A Wilms tumour usually presents as a smooth loin mass that seldom crosses the midline, but extends down into the iliac fossa and up under the costal margin. A right-sided Wilms tumour may extend behind the liver, which, if pushed down, may present as hepatomegaly. On the left side, a Wilms tumour may be mistaken for an enlarged spleen. Children with Wilms tumours are typically well at presentation (80% of cases), unlike those patients with neuroblastoma.

Haematuria, often following minor trauma, is the presentation in some children, but does not indicate a poorer prognosis. Clinical examination of the child should always include a blood pressure measurement, as this may be elevated due to unusual metabolites from the tumour tissue or compression of the renal vessels

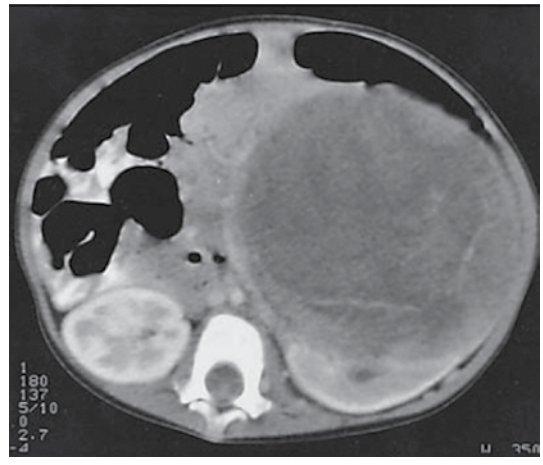
Investigation

Ultrasonography provides detailed information of the site, size and extent of the tumour [Fig. 25.2a and Table 25.2]. Doppler ultrasonography also identifies renal vein and inferior vena cava involvement. The liver is examined for the presence of metastases, and a chest x-ray excludes the presence of pulmonary metastases. A contrast CT scan of the abdomen and chest is utilised to assess the extent of disease, the involvement of the contralateral kidney, the effect on surrounding tissues and the extent of metastatic disease [Fig. 25.2b].

There remains controversy regarding the role of a preoperative tissue diagnosis. In some countries, particularly the United States, primary resection without a prior tissue diagnosis is the routine. The operative and histological findings then direct the need for subsequent chemotherapy and radiotherapy. In other countries, particularly in Europe, preoperative chemotherapy is initiated *without* a tissue diagnosis. The rationale is to downstage the disease and reduce the risk of intra-operative



(a)



(b)

Figure 25.2 **(a)** Ultrasound scan showing a large Wilms tumour (M) arising from the left kidney (LK) and **(b)** CT scan showing the same tumour in cross section.

Table 25.2 Staging of Wilms tumour

Stage	Tumour spread
I	Confined to kidney and removed completely
II	Microscopic local disease after resection
III	Macroscopic residual disease after resection
IV	Distant metastases
V	Bilateral renal tumours

tumour spillage. However, this approach carries the potential risk of treating a child with an inappropriate chemotherapy regimen for a non-Wilms renal tumour.

Treatment

As introduced earlier, the timing of Wilms tumour surgery relative to biopsy and/or chemotherapy remains controversial. This notwithstanding, tumour nephrectomy is an integral component of successful Wilms tumour management. This is typically undertaken by an open and transabdominal approach, but select indications for laparoscopic Wilms tumour surgery are being recognised. Bilateral Wilms tumour presents particular challenges, and in this uncommon setting, renal-sparing surgery may need consideration.

Chemotherapy regimens vary from 18 to 34 weeks, depending upon the tumour stage and the international protocols, and may utilise two or three chemotherapeutic agents. Radiotherapy is used in patients with spilt or residual tumour, as well as in metastatic disease.

Prognosis

Wilms tumours, in contrast to neuroblastoma, are associated with excellent rates of survival. Cure rates for stage I are as high as 95%, and even in those with stage V disease, the cure rates approach 75%. Late recurrence is rare, but patients must be followed for the late effects associated with both chemotherapy and radiotherapy, including:

1. Cardiotoxicity – occurs in 5% of patients treated with doxorubicin.
2. Musculoskeletal disorders – increased risk of scoliosis.
3. Reduced fertility – females may experience early primary ovarian failure, while males may have hypogonadism and azoospermia.
4. Development of secondary tumours – typically associated with the field of radiation. Most common forms of malignancy are leukaemia and lymphoma.
5. Renal failure – patients must be followed for up to 20 years to monitor the risk of renal failure in the remaining kidney.

Liver tumours

Hepatoblastoma is the most common malignant tumour presenting as a right upper quadrant mass in children less than 1 year of age. Alternative diagnoses include an haemangioendothelioma (the most common benign vascular tumour of the liver in infancy, typically presenting before 6 months of age) and mesenchymal hamartoma (a benign tumour that almost exclusively occurs in children under 2 years of age). The mass in an infant with hepatoblastoma is typically large, but the infant may also present with systemic features such as weight loss, vomiting and anaemia. Elevation of the tumour marker serum alpha-fetoprotein in a patient with a liver mass strongly suggests hepatoblastoma.

Accurate preoperative imaging is necessary, including ultrasonography, CT scan, MRI and angiography. The staging of liver tumours is according to the Pretreatment Extent of Disease (PRETEXT) system, which comprises four levels based upon the degree of liver involvement. Operative resection of the lesion, either locally or by lobectomy, remains the main treatment for all primary liver tumours, but preoperative chemotherapy significantly improves survival. There is almost no chance of cure in hepatoblastoma patients without complete resection. Liver transplantation is an option for those patients whose tumour is limited to the liver but remains unresectable after neo-adjuvant chemotherapy.

KEY POINTS

- A child with an abdominal mass needs immediate clinical assessment and investigation to exclude malignancy.
- Abdominal masses in toddlers may be huge before diagnosis.
- Ultrasonography is an effective screening test for malignancy.
- A patient with a presumed abdominal tumour needs immediate referral to the regional surgical and oncology centre.

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CHAPTER 26
Spleen, Pancreas and Biliary Tract

CASE 1

A 4-year-old girl presents with a distended epigastrium and paralytic ileus. On physical examination, there are several old fractures with callus formation.

Q1.1 What are the principal causes of pancreatitis?

Q1.2 How may pancreatitis be diagnosed?

Q1.3 What is a pancreatic pseudocyst?

CASE 2

A 3-week-old neonate develops gastroenteritis from her older siblings. After resolution of diarrhoea, she is noted to be jaundiced.

Q2.1 How would you determine whether obstructive jaundice was present?

Q2.2 What are the differences in management between a patient with a bile duct stone and a patient with biliary atresia?

The spleen

Elective splenectomy, usually done laparoscopically, is indicated for a variety of conditions in childhood [Box 26.1].

Overwhelming post-splenectomy infection

Overwhelming post-splenectomy Post-splenectomy infection (OPSI) occurs in 1–5% of children following splenectomy and is typically caused by encapsulated organisms such as *pneumococcus*, *meningococcus* and *Haemophilus*. The risk of OPSI is lifelong (though it more commonly occurs in the first year post-splenectomy) and has been associated with mortality rates in affected patients ranging from 10 to 70%. OPSI is more common when the spleen is removed for haematological conditions and when splenectomy is done in younger children. Preoperative immunisation against encapsulated organisms is essential, while the role for post-operative penicillin remains controversial.

Hereditary spherocytosis

Hereditary spherocytosis is an autosomal dominant condition resulting in variable degrees of haemolytic anaemia. In spherocytosis, the red cells are abnormally spherical and are destroyed within the spleen. This may result in:

1 Chronic anaemia
2 Episodic haemolytic jaundice
3 Increased tendency to form pigment gallstones

These complications may be controlled by splenectomy. However, unless complications are severe, splenectomy is delayed until at least the age of 7 years of age to minimise the risk of OPSI. Cholecystectomy may be required at the same time.

Idiopathic thrombocytopenic purpura

Idiopathic thrombocytopenic purpura is an autoimmune condition of unknown aetiology that causes destruction of platelets. It presents most commonly in the acute form, which usually resolves spontaneously. Administration of gamma globulin is sometimes required in severe episodes to control the platelet count, and splenectomy is rarely indicated. A small percentage of children will develop a chronic form of the condition in whom splenectomy may be required if medical therapy is unsuccessful.

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Box 26.1 Indications for splenectomy in childhood

Hereditary spherocytosis
Idiopathic thrombocytopenic purpura
Thalassaemia
Sickle cell anaemia
Metabolic storage diseases
Hypersplenism
Neoplasms
Congenital and acquired cysts
Trauma

Thalassaemia

In thalassaemia major, a homozygous, autosomal recessive condition, abnormal haemoglobin chains result in chronic haemolytic anaemia. In the past, chronic anaemia, blood transfusions and subsequent increasing iron stores have resulted in the patient developing a secondary hypersplenism with a very large spleen. The enlarged spleen tended to destroy all cellular elements in the blood.

Transfusions keep children healthy, while regular parenteral desferrioxamine chelates excess iron liberated from haemolysed red cells and maintains normal serum iron levels. In thalassaemia, splenectomy is indicated if the splenomegaly causes abdominal symptoms or difficulties with respiration or if the secondary hypersplenism is difficult to control.

Sickle cell anaemia

In sickle cell disease, the abnormal haemoglobin S causes abnormally shaped red cells during hypoxia. These *sickle cells* tend to impede blood flow through small vessels, causing ischaemia in the organ involved. Splenic infarcts may occur. Splenectomy is usually contraindicated as the resulting higher haemoglobin causes more *sickling* of the red cells in other organs.

Trauma

Splenectomy for trauma is now exceptionally rare (see Chapter 38).

The pancreas

Acute pancreatitis

Acute pancreatitis in childhood is an uncommon clinical entity. The known aetiologies in the paediatric age group are extensive. Some of the more common causes of pancreatitis in childhood are listed below [Box 26.2].

Box 26.2 Common causes of pancreatitis in childhood

1. Trauma (handlebar injury, motor accident, child abuse)
2. Drugs (steroids, azathioprine)
3. Viral
4. Biliary tract disorders (choledochal cyst, gallstones)
5. Hereditary
6. Metabolic (hyperlipidaemia)

Traumatic pancreatitis

The pancreas is the fourth most common abdominal organ injured in childhood trauma. Nearly all cases result from blunt abdominal trauma, as penetrating trauma in Australasian children is rare. The most common cause in most Western countries is a handlebar injury, but child abuse may present this way, secondary to a kick or punch to the abdomen. The morbidity associated with blunt pancreatic trauma is determined by whether there is disruption of the pancreatic duct. The best initial investigation is a CT scan: if this suggests disruption of the pancreatic duct, then further investigations to confirm this may be indicated. In this situation, a magnetic resonance cholangiopancreatogram (MRCP) or endoscopic retrograde cholangiopancreatogram (ERCP) may be useful. Most cases do not involve a duct injury and are managed conservatively, while management of duct disruption remains controversial. Some advocate conservative management, accepting the risk of a pseudocyst (see following text), while others advocate an early distal pancreatectomy.

Pseudocyst

A pancreatic pseudocyst is a collection of pancreatic fluid within a non-epithelial-lined cavity that forms at least 6 weeks after the initial pancreatic insult. Although it may complicate pancreatitis from any cause, it is most often the result of trauma. Most pancreatic pseudocysts lie in the lesser sac. Treatment is initially non-operative, as many resolve spontaneously. The progress of the collection is followed by serial ultrasound scans. Intervention is indicated if the pseudocyst is enlarging or causing symptoms. Internal drainage of the pseudocyst into the stomach via an endoscopic cystogastrostomy or open cystogastrostomy is the preferred technique.

Hyperinsulinism (causing hypoglycaemia)

Excessive production of insulin may occur in several situations:

1. In babies of diabetic mothers as a temporary response to high maternal sugar levels.
2. Beta-cell hyperplasia – an idiopathic condition in which there is excessive production of insulin, which usually resolves with drug treatment (diazoxide). Near-total pancreatectomy is only necessary occasionally.
3. Beckwith–Wiedemann syndrome – a condition of newborn babies that is characterised by exomphalos, organomegaly (large tongue and abdominal organs), hemihypertrophy and transient low blood sugar from excessive insulin and insulin-like growth factor production.
4. Islet cell tumours – rare cause of hypoglycaemia. They may be cured if the tumour (usually benign) is localised and excised.

The biliary tract

Neonatal jaundice

Jaundice in the neonatal period most commonly results from a prehepatic or hepatic cause. Posthepatic causes of jaundice result in conjugated hyperbilirubinaemia and may require surgical treatment. These causes include:

1. Biliary atresia (represents 80% of cases)
2. Choledochal cyst
3. Inspissated bile syndrome
4. Bile duct stricture
5. Spontaneous biliary perforation

All cases of conjugated hyperbilirubinaemia in the neonatal period should be promptly investigated.

Biliary atresia

Biliary atresia is a condition of progressive obliteration of the extrahepatic ducts as a result of an, as yet, undiagnosed inflammatory condition. This process may involve part of or the entire extrahepatic biliary tree. The ducts may shrivel and disappear or, more commonly, persist as a fibrous cord.

The incidence of biliary atresia is about 1 per 15,000 live births. The aetiology remains unknown, although there is a wide range of hypotheses based on infective, embryological, metabolic and vascular studies. In up to 20% of cases, biliary atresia is associated with a distinct syndrome known as the biliary atresia splenic malformation syndrome (BASM). Associated anomalies in this syndrome include splenic abnormalities (asplenia or polysplenia), situs inversus, intestinal malrotation, cardiac anomalies and a preduodenal portal vein.

Biliary atresia presents as prolonged jaundice after transient neonatal physiological jaundice should have resolved. Progressive obstructive jaundice occurs in the first 6 weeks of life with pale stools and dark urine. Typically, these infants are thriving at the time of presentation.

Diagnosis

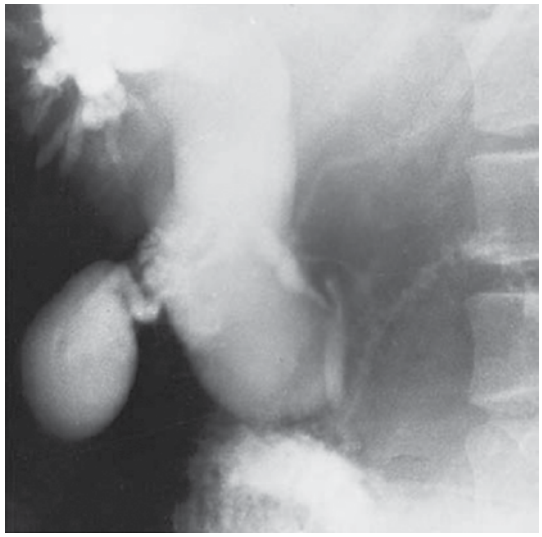
Prompt diagnosis is essential in biliary atresia as the long-term results of surgery are correlated with the timing of operation. Ultrasonography is the most important initial investigation and will exclude other surgical causes of jaundice (see preceding text). In biliary atresia, a fasting ultrasound scan will usually show a small and contracted gall bladder. A presumptive diagnosis is best made by liver biopsy (typically percutaneous), which in experienced hands will yield a positive diagnosis in up to 95% of cases. The definitive diagnosis is made at laparotomy by confirming non-patency of the extrahepatic biliary tree. HIDA scans are of limited value as their interpretation in neonates is difficult.

Treatment

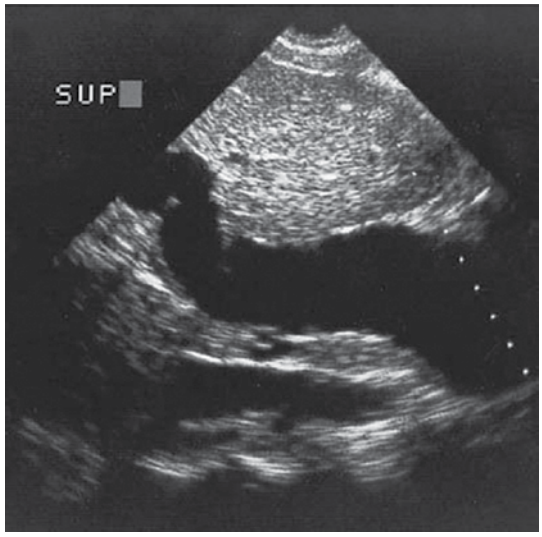
Biliary atresia is treated by portoenterostomy (Kasai procedure). This operation involves dissecting out the obliterated extrahepatic ducts up to the portal plate and shaving off the inflammatory tissue at the portal plate flush with the liver surface. A Roux-en-Y loop of the jejunum is then anastomosed to the edges of the portal plate. The operation relies on bile draining into the Roux loop from microscopic bile ductules in the portal plate. Drainage rates of up to 60% may be achieved in experienced centres. There is a dramatic decrease in drainage rates if the operation is performed after 100 days of life. It is not possible to determine at the time of operation whether the operation will be successful in the long term. Liver transplantation is required if the operation fails.

Choledochal cysts

Choledochal cyst is a congenital dilatation of the extrahepatic biliary tree. There are various forms, but in the most common variant, there is dilatation of the gall bladder, cystic duct, common hepatic duct and common



(a)



(b)

Figure 26.1 Contrast x-ray **(a)** and ultrasonography **(b)** of the biliary tract showing the massive tubular dilatation of a choledochal cyst.

bile duct. The dilatation is usually cystic, but fusiform variants are well described [Fig. 26.1].

The incidence in Western countries is between 1 in 10,000 and 1 in 15,000 live births, but a higher frequency is seen in Asia. The cause remains unclear, but in China and Japan, most patients with a choledochal cyst have an anomalous junction between the terminal common bile duct and the pancreatic duct. The ducts unite outside the duodenal wall and the ampulla of Vater in the third part of the duodenum and the resultant abnormal common channel is not surrounded by the normal sphincter mechanism. This situation encourages reflux of pancreatic juice into the common bile duct, which is thought to result in progressive damage of the biliary tree.

The most common presenting features of a choledochal cyst are obstructive jaundice and recurrent abdominal pain. Pancreatitis is not uncommonly the presenting complaint. Rarely, a choledochal cyst may present as an abdominal mass. The majority of choledochal cysts present before the age of 10 years and increasingly are being detected antenatally. Antenatal diagnosis may be made as early as 15 weeks' gestation. In some situations, differentiation from other congenital cysts in the upper abdomen such as duplication cysts, ovarian cyst and the rare cystic form of biliary atresia may be difficult.

Diagnosis

The diagnosis is easily made on ultrasonography. More detailed information of the nature of the dilatation is now obtained using an MRCP.

Treatment

Treatment is excision of the cyst and drainage of the proximal common hepatic duct by a Roux-en-Y loop of the jejunum. Choledochal cysts that have been diagnosed antenatally and remain asymptomatic are electively excised from 6 months of age.

Inspissated bile syndrome

Inspissated bile syndrome is a condition causing obstructive jaundice in neonates resulting from inspissation of bile in the lower third of the common bile duct. This condition most commonly occurs in premature neonates requiring prolonged total parenteral nutrition (TPN). It is also associated with extravascular haemolysis and may occur in otherwise normal neonates after gastroenteritis and dehydration.

The diagnosis is made on ultrasonography, which reveals a dilated proximal biliary tree in association with biliary sludge or stones.

Most cases resolve spontaneously. Resistant cases may be cleared by percutaneous transhepatic irrigation of the bile ducts or retrograde irrigation by ERCP. Operative intervention is only rarely required.

Bile duct strictures

Bile duct strictures are a rare but well-known cause of obstructive jaundice in neonates. Most strictures are idiopathic, and the most common site of obstruction is the distal common bile duct. A small proportion occur in association with a long common channel (see preceding text), where the presumed aetiology is reflux of pancreatic juice into the common bile duct. Diagnosis is made using percutaneous transhepatic cholangiography or ERCP. The obstruction may be relieved by balloon dilatation using the same modalities. If this is unsuccessful, then operative biliary diversion is required.

Spontaneous biliary perforation

Spontaneous biliary perforation is a rare condition resulting in progressive obstructive jaundice and ascites in neonates. Most cases present between 1 week and 2 months of age. The site of the perforation is almost always at the junction of the cystic and common hepatic duct. The aetiology is unknown and the presentation is usually insidious. Ascites results from a localised biliary peritonitis, and the jaundice occurs as a result of both proximal biliary obstruction secondary to oedema and reabsorption of bile through the peritoneum. The diagnosis is made on ultrasonography, which demonstrates a loculated collection in the portal region. Operative intervention with drainage is required.

Cholelithiasis in children

In children, the cause of gallstone formation may be divided into the following groups:

1. *Haemolytic disorders*. Conditions such as sickle cell anaemia, thalassaemia and hereditary spherocytosis cause pigment stones due to increased red cell breakdown.
2. *TPN*. The association of TPN and biliary sludge and cholelithiasis is well recognised. The exact cause is unknown, but biliary stasis due to impairment of the enterohepatic circulation of bile is probably important.
3. *Ileal resection*. This is a well-known risk factor for cholelithiasis. In children, the most common reason for ileal resection is necrotising enterocolitis in premature neonates. The traditional explanation for gallstones in this setting is that normal reabsorption of bile salts in the terminal ileum is impaired, leading to depletion of bile salts in the enterohepatic circulation. This promotes lithogenic bile. The neonates affected by necrotising enterocolitis will have also required a period of TPN during their recovery.
4. *Mechanical causes*. Any condition that leads to biliary stasis is associated with the formation of gallstones, such as bile duct strictures, choledochal cysts and congenital gall bladder abnormalities.
5. *Specific conditions*. Certain conditions such as cystic fibrosis, Crohn disease and diabetes are associated with an increased incidence of cholelithiasis.
6. *Adult causes*. Adolescents with typical adult-type risk factors, including obesity, oral contraceptive pill and family history, have the same tendency to develop gallstones.

The ways in which a child with gallstones may present are summarised in Box 26.3.

Box 26.3 Presentation of the child with gallstones

1. Biliary colic: pain from a stone in the neck of the gall bladder or common bile duct
2. Cholecystitis: chemical or bacterial inflammation of the gall bladder, usually associated with cystic duct obstruction
3. Obstructive jaundice: dark urine and pale stools due to a stone obstructing the common bile duct
4. Pancreatitis

Treatment

Treatment involves laparoscopic cholecystectomy with removal of stones in the common bile duct, if present.

KEY POINTS

- Splenectomy is avoided in small children because of the risk of overwhelming post-splenectomy infection (1–5%).
- Ruptured spleen rarely requires a splenectomy, as bleeding stops.
- Pancreatitis may occur with trauma.
- Prolonged jaundice in neonates needs investigation to exclude biliary atresia.
- Gallstones are increasingly common in children.

Further reading

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CHAPTER 27
Anus, Perineum and Female Genitalia

CASE 1

A 3-month-old boy presents with a tender, red, indurated area (2×2 cm) adjacent to the anal verge. Twice in recent weeks, antibiotics were prescribed for a similar problem that resolved. On palpation and compression of the mass, a drop of pus appears at the anus.

Q1.1 What is the diagnosis, and how would it best be managed?

CASE 2

A worried mother rushes her 8-month-old daughter to the emergency department after noticing that no vaginal opening is visible. She is frightened something serious is wrong with the child's genitalia.

Q2.1 What is the diagnosis?

Q2.2 How is it treated and recurrence prevented?

Anorectal problems occur commonly in children, with abscesses, fistulae and fissures affecting infants, while pilonidal disease, haemorrhoids and polyps tend to affect older children.

Anal fissures

These are confined mostly to infants and toddlers in whom the passage of a hard stool splits the anal mucosa. There is a sharp pain on defecation and a few drops of bright blood on the surface of the stool (see Chapter 23).

Anal fissures are superficial and may heal so rapidly that the fissure is already healed by the time it is brought to medical attention and examined. If not healed, it is usually visible anteriorly or posteriorly. A chronic fissure may be associated with a sentinel skin tag. Multiple fissures and those that are not in the midline may be due to other pathological processes such as inflammatory bowel disease, infection or trauma.

Treatment

An acute anal fissure is of no consequence in itself, and treatment is directed to the underlying constipation (see Chapter 22), which is commonly associated with food allergy. Cow milk protein is usually the trigger food.

Chronic and/or prominently symptomatic anal fissures may warrant specific treatment, with both operative and non-operative options available. Many of these therapies target relief of the internal sphincter spasm commonly associated with anal fissures. This spasm is thought to result in a degree of local ischaemia and contribute to the accompanying anal pain.

Historically (and more so in adults), various surgical strategies have been proposed to treat chronic anal fissures. These range from simple anal dilatation to open or closed internal sphincterotomy and even skin flap coverage of the fissure. Operative management achieves cure in 90% but at the expense of incontinence (usually to flatus) in 10%. Interestingly, anal stretch has higher reported rates of incontinence than limited lateral sphincterotomy and so should be used cautiously in children.

The risk of postoperative incontinence drove development of non-operative therapies, particularly topical agents to provide anal analgesia and/or internal sphincter relaxation. A wide variety of agents have been investigated including lignocaine, hydrocortisone, calcium channel blockers such as nifedipine, nitroglycerin ointment (GTN), and more recently botulinum toxin (Botox). Trials show GTN, nifedipine and Botox to be only marginally better than placebo, with chronic fissure healing in 55% versus 35% for placebo. Unfortunately, topical GTN causes headaches in up to 40% of patients, which may be severe enough to lead to treatment cessation. Late recurrence following medical therapy occurs in 50% of patients.

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In summary, most anal fissures can be managed by treating the underlying constipation if the fissure does not heal spontaneously, and other modalities can be reserved for the problematic or persistent symptomatic fissure. Persistence of a fissure may cause the child to experience pain and distress during defecation and even inappropriately *hold on* to stools because of their association of pain with defecation. The emotional tension built up around the act is more difficult to treat than the fissure itself and may be the forerunner of the whole vicious circle of constipation (Chapter 22).

Perianal abscess

This is fairly common in infants and arises from infection in the anal glands, which open into the crypts of the anal valves. Although the abscess almost always presents superficially, the fistulous tract passes through the most superficial of the internal sphincter fibres to open inside an anal valve.

Treatment involves identifying and laying open the fistula and draining the abscess [Fig. 27.1]. Failure to deal with the fistula may result in recurrent infection.

Sometimes, young children may develop a superficial subcutaneous abscess in the buttock or near the anus, which is often secondary to a nappy rash and infection with skin organisms. In these cases unrelated to anal gland infection, simple drainage and antibiotics are curative.

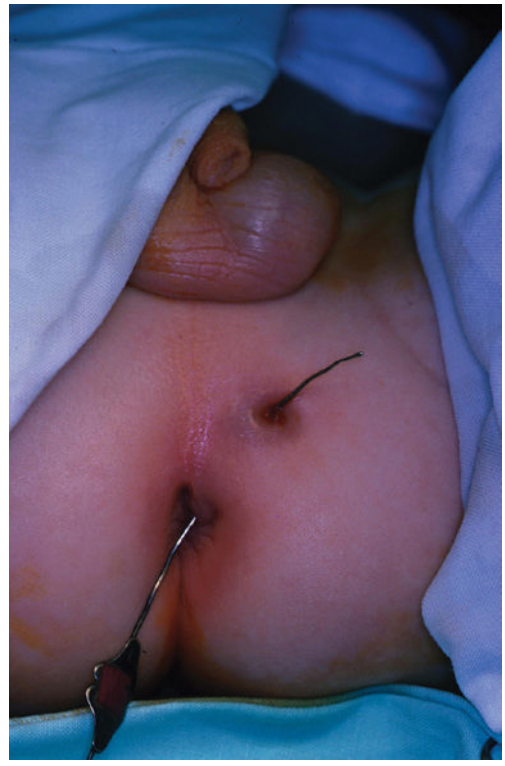


Figure 27.1 In perianal abscess, there is a fistula running from the abscess to the anus inside an anal valve. The tract is displayed by a lacrimal probe.

Rectal prolapse

Rectal prolapse is not uncommon in toddlers and is an alarming experience for the parents [Fig. 27.2]. However, in most cases, it disappears spontaneously after a few weeks or months without residual damage.

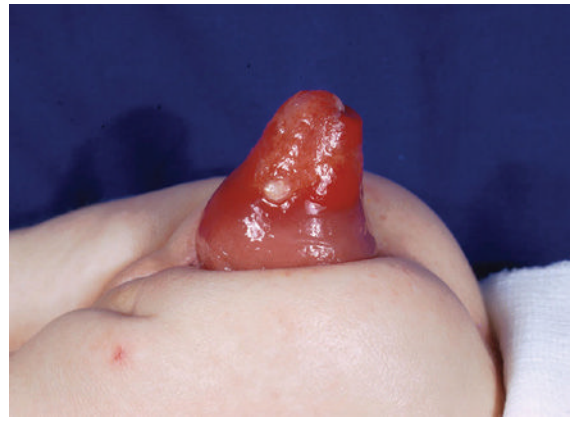


Figure 27.2 Rectal prolapse. The mucosa is congested and oedematous and may bleed.

Aetiology

The two common predisposing factors are:

1. Straining at stool by a child with constipation. Less frequently, straining may occur paradoxically with diarrhoea, for example, as part of a malabsorption syndrome such as cystic fibrosis or coeliac disease.
2. Explosive or reluctant defaecation. A healthy child occasionally develops a rectal prolapse following an explosive defaecation in which there is little time for moulding of the stool by the muscles of the pelvic floor.

Reluctant defecation describes prolonged attempts to defaecate with excessive straining in the *absence of constipation*, often as a result of parents' ill advice during toilet training.

Rare organic causes include:

1 Paralysis of anal sphincters (spina bifida, sacrococcygeal teratoma)
2 Hypotonia or starvation
3 Ectopia vesicae (bladder exstrophy with abnormal pelvic girdle)
4 Complication of surgery for an anorectal malformation or Hirschsprung disease

Clinical features

Most children with rectal prolapse have normal pelvic anatomy. The prolapse rolls out painlessly only during defaecation and usually returns spontaneously; manual replacement is required infrequently. The mucosa may become abraded while it is prolapsed and cause minor bleeding. A digital photograph taken by the parents may help the surgeon sort out the differential diagnosis.

Differential diagnosis

1 Rectal polyp: Polyps may prolapse per rectum (see following text) and may be positively identified by observation, digital palpation or proctoscopy.
2 Intussusception: Rarely, the apex of an intussusception appears at the anus, albeit usually accompanied by its own and distinctive clinical features (Chapter 19).
3 External haemorrhoids: These are rare in childhood, but congestion of the submucosal venous plexus during straining at stool sometimes produces a bluish sessile bulge, the parents' description of which may be mistaken for rectal prolapse.

Treatment

Constipation is the most common cause, and treatment to ensure no straining at stool is required for at least several weeks (Chapter 22). In the absence of constipation, the possibility of a malabsorption syndrome should be investigated.

Any errors in the child's sitting posture at defecation will need to be addressed, for example, squatting over a potty on the floor to defaecate, which stretches the pelvic floor and anal sphincters to maximum disadvantage. A potty chair or an insert for an adult toilet seat enables the child to sit with support for the pelvic floor. Also, a reasonable time limit for defecation should be set to discourage straining.

Rarely, rectal prolapse proves refractory to conservative medical management, and operative treatment is required. Various unpalatable and invasive surgical techniques are described in adult patients, but have highly selective, if any, application in children. More commonly, sclerotherapy is performed by injection of a sclerosant into the submucosal plane of the rectum to cause fibrosis and contraction of the rectal wall, for example, 0.5 ml of 5% phenol in almond oil injected into the submucosa at three equally spaced points, 2 cm above the anal valves.

Rectal polyps

Juvenile rectal polyps are isolated benign hamartomas and are a relatively common cause of rectal bleeding. Bright bleeding is produced painlessly at the end of defecation and is typically intermittent over long periods. The polyp is almost always within reach of an examining finger, and occasionally prolapses through the anus [Fig. 27.3].

Treatment

On those uncommon occasions, the polyp protrudes through the anus; the base may be ligated without anaesthesia. Otherwise, under general anaesthesia, the polyp may be located through the proctoscope and withdrawn to demonstrate its stalk, which is transfixed with a suture ligature or transected with diathermy. Higher lesions may be similarly removed at colonoscopy. Recurrence is rare and malignancy unknown.

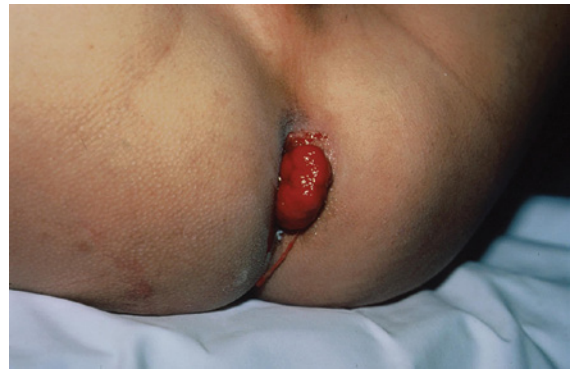


Figure 27.3 Prolapse of a benign rectal polyp through the anus.

Multiple polyposis

This familial condition with malignant potential is rarely seen in adults and even more rarely in children. It should be considered when more than three polyps are identified and/or when there is a family history of multiple polyposis. Colonoscopy is the principal form of investigation. In children, major fluid and electrolyte losses may ensue, and the colon may need to be removed.

Peutz–Jeghers syndrome

This is an even rarer condition that has gained prominence because of the external evidence of its existence – the presence of pigmented freckles on the mucocutaneous margins of the lips and the anus. Polyps are found anywhere in the gastrointestinal tract, especially in the jejunum, and may give rise to massive bleeding, intussusception and/or intestinal obstruction. The polyps may become malignant, but this is less common than in familial polyposis of the colon.

Postanal dimple (coccygeal dimple)

Many babies have a postanal dimple in the skin over the coccyx. These coccygeal dimples are typically shallow, small and of no significance. Occasionally, a coccygeal dimple is narrow, deep and so prone to infection, in which case it may be excised.

A coccygeal dimple should not be confused with a sacral sinus, which is rarer but also more dangerous due to its association with an underlying spina bifida occulta (Chapter 9). A sacral sinus can be distinguished by its situation over the sacrum, not coccyx, and its inability to demonstrate any base. Indeed, in the depths of the sinus is likely a small tract that communicates directly with the spinal theca or with an intraspinal dermoid cyst. As this tract is a potential source of recurrent meningitis, the child should be referred to a neurosurgeon for treatment.

Sacrococcygeal teratoma

Sacrococcygeal teratomas are the most common neonatal malignancy, but are still rare, occurring in 1:40,000 births and slightly more frequently in females [Fig. 27.4].

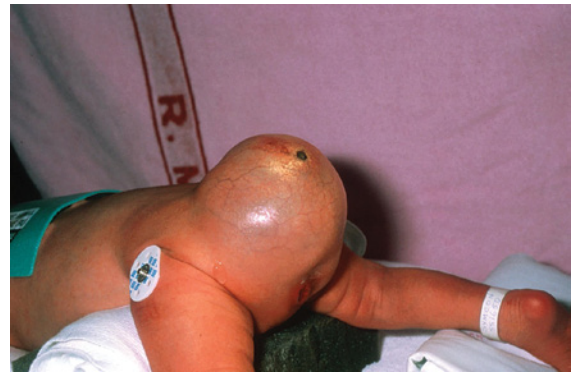


Figure 27.4 Sacrococcygeal teratoma of medium size; note distortion of the perineum and anal canal and a small ulcer on the surface of the tumour. The prognosis for faecal continence after operation, however, is excellent.

The tumour is thought to arise from primitive streak cells located in the region of the coccyx. A teratoma comprises solid and cystic areas arising from all three embryonic layers.

The majority of sacrococcygeal teratomas are diagnosed antenatally and may cause antenatal complications for both the fetus (e.g. hydrops) and mother (i.e. maternal mirror syndrome). The tumour is usually obvious at birth and may be so large as to cause obstetric difficulties. Occasionally, the tumour is in the pelvis and does not protrude from the perineum, such that diagnosis may be delayed or even missed. Malignant transformation occurs in between 5 and 35% of cases and is greatest for those lesions in which excision is delayed until after 3 months of age.

Management

Even very small sacrococcygeal teratomas should be referred to a tertiary paediatric centre for excision. Differential diagnosis includes other rare tumours (chordoma or ganglioneuroma) or an anterior sacral meningocele. Magnetic resonance imaging is now almost routinely employed to determine the extent of the intrapelvic tumour. Excision is typically undertaken soon after birth, assuming the neonate has no other anomalies that might take priority. In spite of gross stretching of the pelvic floor along with its nerve supply and the anal canal, these structures usually recover completely after careful operative excision without any long-term neural deficit or lack of function.

Prognosis

Local recurrence and/or metastases are uncommon, but more likely if:

1 The tumour is uniformly solid and devoid of cysts.
2 The coccyx is not removed *en bloc* with the tumour.
3 The diagnosis or operative excision is delayed.

The large benign teratoma presents at birth and could hardly be overlooked; it is removed in the neonatal period. However, a small malignant teratoma may escape diagnosis until a rapid increase in size brings it to notice later in the first few years of life.

Pilonidal sinus

The word *pilonidal* is derived from the Greek for *nest of hairs*. Pilonidal sinuses are uncommon in prepubescent children but are common in adolescents and adults. Contemporary understanding indicates the condition is acquired, with loose hair and the anaerobic environment of a deep natal cleft likely contributors to its pathogenesis. Most cases present with an uninfected sinus or an abscess, which may be chronic.

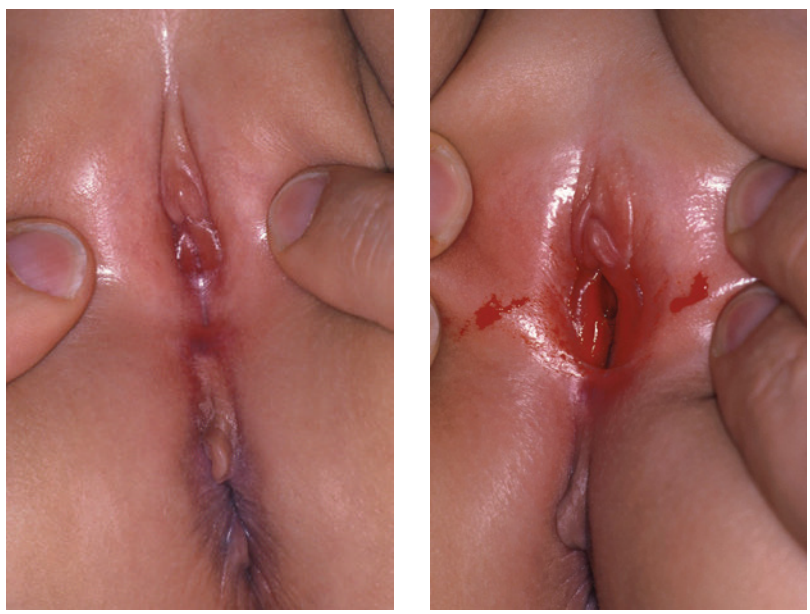
Pilonidal abscess treatment requires incision, drainage and curettage of the hair mass. Treatment of the sinus is more controversial with various proposed strategies, including excision and healing by secondary intention, excision with primary wound closure and excision with either subcutaneous tissue flaps (Karydakis flap) or rotational skin flaps (Limberg flap). Most attempt to provide both wound closure and minimise the risk of recurrence, which is unfortunately high. Recently, laser depilation has been added to the treatment algorithm in an attempt to reduce recurrence.

The female genitalia

Developmental anomalies of the external genitalia are rare in girls. The most common abnormality is an acquired defect, adhesion of the labia minora, which is caused by ulceration of the labia from nappy rash with secondary adhesion during re-epithelialisation.

Labial adhesions

This is a common condition, which may cause discomfort during micturition, but is more often discovered incidentally on routine examination [Fig. 27.5a and b].



(a) (b)

Figure 27.5 Adherent labia minora (labial adhesions). **(a)** The normal labia majora have been flattened by lateral traction to display the line of fusion. **(b)** Following the separation, the introitus is fully visible.

There is a delicate midline adhesion of the labia minora that partially closes the posterior introitus, overlying the opening of the urethra, and may extend as far anteriorly as the clitoris.

Unfortunately, many mistake the appearance of this common and benign condition for congenital absence of the vagina (i.e. vaginal atresia), causing the child's parents much unnecessary anxiety. Acquired labial adhesions are never present at birth.

Treatment

In infants, the fused labia may be separated by exerting gentle lateral traction on the labia minora without anaesthesia or by sweeping them apart with a suitably blunt instrument. Because of the tendency for the adhesions to recur, the parents should repeat separation of the labia daily for 2 weeks and apply petroleum jelly to the introitus to help prevent re-adhesion. Avoiding nappy rash also reduces recurrence. Separation under general anaesthesia is seldom required, but informed consent for the procedure without anaesthesia should acknowledge the possibility for pain due to separation. In older children, the labia may be left alone to resolve spontaneously at puberty.

Imperforate hymen

This is a rare condition, which presents either at birth (the vagina secretes mucus that accumulates beneath the bulging imperforate hymen to form a mucocolpos) [Fig. 27.6] or at puberty when apparent primary amenorrhoea, haematocolpos or even haematometrocolpos may be the presenting features, with cyclic attacks of abdominal pain. During childhood, the condition is usually symptomless, except for possible urinary symptoms such as *wetting* or dysuria when the cystic swelling distorts the urethra.

Treatment at birth requires removal of a circular disc or cruciate broad incision of the membrane to provide drainage.

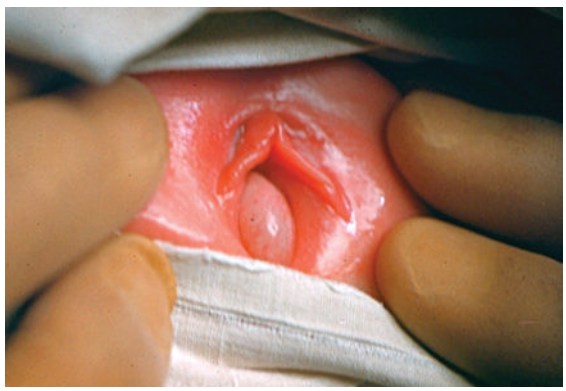


Figure 27.6 Imperforate hymen causing mucocolpos in a neonate.

Vaginal discharge

The chief symptom is vulval irritation, but in some cases, the discharge itself may be the only complaint. A profuse offensive or blood-stained discharge suggests the presence of a foreign body. Small objects may be successfully removed by irrigation using a soft rubber catheter, though sometimes instrumental removal under anaesthesia, through a miniature vaginoscope, is required.

The possibility of sexual abuse must be considered, and where suspected, the discharge should be sent for microbiological examination (Chapter 36).

KEY POINTS

- Painful defaecation associated with a few drops of bright blood suggests an anal fissure.
- Perianal abscess is common in the first year and often leads to a fistula-in-ano.
- Both rectal prolapse and anal fissure resolve spontaneously with correction of constipation and good toileting behaviour.
- Any baby with an abnormal mass near the coccyx should be referred to a paediatric surgical centre at birth, as the diagnosis may be sacrococcygeal teratoma.
- Labial adhesion is an acquired condition that is easily distinguished from imperforate hymen: it responds to simple separation and application of petroleum jelly to prevent re-adherence.

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CHAPTER 28
Undescended Testes and Varicocele

CASE 1

At birth, a baby boy was noted to have only one testis in the scrotum. Re-examination at 3 months showed that both testes were now in the scrotum.

Q 1.1 What is the likely natural history of testicular descent?

Q 1.2 Is treatment required later in childhood?

CASE 2

No testes were palpable in the scrotum or groin in a baby at the 6-week postnatal check.

Q 2.1 What is the differential diagnosis?

Q 2.2 What is the management?

CASE 3

Unilateral undescended testis (UDT) is diagnosed at birth and confirmed at the 6-week postnatal check, in a baby with no other anomalies.

Q 3.1 What is the recommended age for an operation?

Q 3.2 What is the prognosis for fertility and cancer risk?

Definitions

Congenital UDT

A UDT is one that has failed to reach the bottom of the scrotum by 3 months of age. It represents the second most common problem in paediatric surgery after indirect inguinal hernia. At birth 4–5% of boys have undescended testes, but postnatal descent may continue for the first 3 months, when the incidence of cryptorchidism falls to 1–2%, which is about double the incidence 50 years ago. Further descent after 3 months is rare. Most undescended testes have no recognisable primary abnormality, but degenerative changes increase with the age of the patient. These degenerative changes are likely to be secondary to the high temperature of the maldescended testis.

In the majority of patients, the cause of maldescent is unknown. Mechanical factors may be important as cryptorchidism is more common in boys with gastroschisis, exomphalos and prune belly syndrome, all of which have reduced intra-abdominal pressure in utero. A potential hormonal aetiology is suggested by association with intrauterine growth retardation, multiple pregnancies and previous stillbirth, all features suggestive of a degree of placental failure or insufficiency. It has been suggested that the increasing incidence of UDT, hypospadias, testicular cancer and low sperm counts in Northern Europe is caused by in utero exposure to environmental pollution with *endocrine disruptors* (synthetic molecules that mimic oestrogen).

Acquired UDT

The concept of acquired undescended testes is controversial, but it explains the high frequency of children presenting later in childhood. The cause may be failure of the processus vaginalis to disappear completely after descent. The processus vaginalis remnant in the spermatic cord may tether the testis by preventing normal cord elongation with age (the length of the spermatic cord increases from about 5 cm in infants to 8–10 cm in adolescents).

Retractile testis

A normal retractile testis may be manipulated to the bottom of the scrotum, regardless of its initial position, and remains in the scrotum after manipulation. It is a normal

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size and is present in the scrotum on some occasions, such as during a warm bath. Testes may retract into an extension of the tunica vaginalis outside the external oblique aponeurosis and under the superficial abdominal fascia, known as the *superficial inguinal pouch*. The position of the testis is controlled by the cremaster muscle, which helps to regulate testicular temperature by retracting the testis out of the scrotum when cold; it also protects the testis from trauma. Cremasteric contraction is absent in the first few months after birth and is maximal between 2 and 8 years. Retractile testes are typically normal but in severe cases may represent the development of acquired maldescent.

Ascending testis

An *ascending* testis is one that is in the scrotum in infancy, but the testicular position becomes progressively higher during childhood as the spermatic cord fails to elongate at the same rate as body growth. There is often a history of the testis descending into the scrotum some weeks after birth. This anomaly is thought to be a form of acquired undescended testes.

Impalpable testis

Approximately one in five (20%) undescended testes will not be palpable. Of these, 40% will in fact be absent, having undergone prenatal or perinatal atrophy, 30% will be found within the inguinal region, 20% will be intra-abdominal, and 10% will be in an ectopic location. Repeat examination, ultrasound and laparoscopy each have a select role in determining the exact nature and location of a UDT deemed to be impalpable.

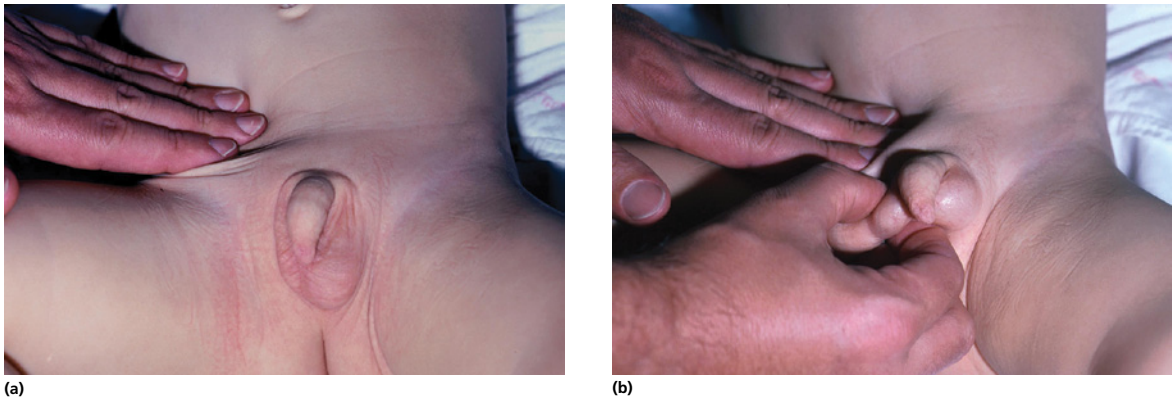
Examination

The examination of the testis should take place in warm and relaxed surroundings and is begun by placing one finger on each side of the neck of the scrotum to pull the scrotum up to the pubis and to prevent the testes from being retracted out of the scrotum by the other examining hand. Each side of the scrotum is then palpated for a testis; if it is not there, the fingertips are placed just medial to the anterior superior iliac spine and moved firmly towards the pubic tubercle [Fig. 28.1a], where the other hand waits to capture the testis if it appears [Fig. 28.1b]. Its range of movement is determined carefully, for the diagnosis depends on this. The precise classification of a palpable UDT is made by determining how far it may be manipulated into the scrotum.

More than two-thirds of undescended testes are located in the *superficial inguinal pouch* (i.e. they are palpable in the groin) [Fig. 28.2]. The testes are normal in size and are within the tunica vaginalis, which makes them deceptively mobile. Rarely, the testis may migrate to a truly ectopic position, such as the perineum, the base of the penis (prepubic), the thigh (femoral) or the opposite hemi-scrotum (crossed testicular ectopia).

Sequelae of maldescent

The higher temperature in which the extra-scrotal testis exists inhibits postnatal germ cell maturation and development of the seminiferous tubules. After puberty,



(a) **(b)**

Figure 28.1 Examination to locate the position of the testis: **(a)** the fingers of one hand push the testis towards the neck of the scrotum, while **(b)** the other hand *snares* the testis at the top of the scrotum to see whether it may be pulled right down to the bottom of the scrotum.

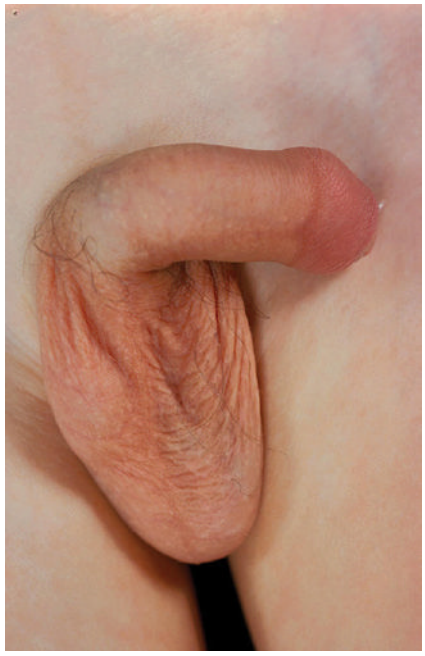


Figure 28.2 Undescended right testis. Once out of the scrotum, the testis is invisible, even though it is palpable in the groin.

there may be oligospermia or azoospermia. The incidence of paternity is not significantly decreased in males with a history of unilateral UDT compared with that of the normal population. However, the fertility rate for males with a past history of bilateral UDT is approximately half that of the normal population, and in some, there is complete infertility.

Maldescent is an accepted but difficult to quantify risk factor for testicular malignancy. Contemporary studies estimate the risk of malignancy due to maldescent to be two- to eight-fold higher than in a descended testis, with highest risk in men with bilateral UDTs.

The relative risk of malignancy in a unilateral UDT is reduced to twofold if orchidopexy is performed before 13 years of age. It is hoped, but not yet proven, that orchidopexy at earlier ages – as is accepted current best practice – further reduces the malignancy risk. Importantly, orchidopexy also places the testis in a location that is more amenable to self-examination, making earlier detection of testicular malignancy potentially easier.

A testis in the inguinal region is more liable to direct trauma and torsion.

Treatment of undescended testes

Treatment aims to preserve normal spermatogenesis, increase comfort and achieve normal appearance. Hormonal function at puberty (i.e. testosterone output) is usually normal regardless of treatment. The maturation of gonocytes to spermatozoa is adversely affected in undescended testes from the age of 4–12 months onwards and is, to a degree, proportional to the length of time testes remain undescended beyond this age. Testes with acquired maldescent may descend fully at puberty, but they are poorly developed and spermatogenesis is deficient, although less severely than congenital undescended testes.

Orchidopexy for congenital cryptorchidism is recommended at 6–12 months of age. Orchidopexy is usually performed as a day surgery procedure. Acquired undescended testes should have orchidopexy once they can no longer reside spontaneously in the scrotum. There is no current role for hormone treatment.

Determination of hormonal levels has no place in the investigation of unilateral UDT but should always be performed in boys with bilateral impalpable testes prior to surgical exploration. The best diagnostic test for an impalpable UDT is laparoscopic exploration. Laparoscopy will detect the 40% of absent testes, enable removal of a useless and potentially neoplastic testicular nubbin and permit the initial surgical management of the 20% of testes that are truly intra-abdominal. A coexistent indirect inguinal hernia is almost universal but usually latent; when it becomes apparent clinically, herniotomy is necessary regardless of the boy's age, at which time orchidopexy is performed.

The absent testis

Rarely, the testis is absent or excised because of torsion (and necrosis), tumours or dysgenesis. Affected boys may have psychological problems and suffer significant embarrassment in the locker room. The use of a prosthetic testis may be considered, but ideally, insertion should be delayed until adolescence when adult-sized implants may be accommodated in the scrotum.

Varicocele

A varicocele is an enlargement of the veins of the pampiniform plexus in the spermatic cord and usually appears in boys over 12 years of age or around the onset of puberty. The mass of veins is best seen and felt when the

patient is standing and feels like *a bag of worms*. Varicoceles are usually left sided (80–90%) but may be right sided in 1–7% or bilateral in 2–20%. There is sometimes a small secondary hydrocele and the hemi-scrotum is redundant. The varicosities typically empty when the boy lies down (but the left hemi-scrotum remains more pendulous than the right); thus, clinical examination should always include getting the boy to stand up. The varicocele is usually symptomless, though a dragging ache may develop when the varicocele is large.

The normal pampiniform plexus helps control the temperature of the testis, by cooling the arterial blood flowing to the testis through a countercurrent heat exchange. The loss of this cooling mechanism affects both testes and the consequent rise of scrotal temperature towards normal body temperature may cause oligospermia. While varicoceles are present in 10–20% of the normal male population, a subclinical varicocele is found in 40–75% of males investigated for infertility.

Treatment

The normal scrotal temperature is 33°C (4°C below body temperature). This is the optimal temperature for spermatogenesis. Relative infertility cannot be assessed until late adolescence, but secondary atrophy of the ipsilateral testis is well recognised, and if the affected testis is significantly smaller or softer in texture than the contralateral testis, then early operative intervention is indicated.

Laparoscopic ligation (Palomo operation) of the spermatic vessels should prevent recurrence in most patients, although there are many other techniques.

Very rarely, a varicocele is caused by obstruction of the renal veins by a retroperitoneal tumour (e.g. Wilms tumour, neuroblastoma). It is exceptionally rare for retroperitoneal tumours to present in this way: the tumour will more often present as a palpable mass with or without haematuria and hypertension.

KEY POINTS

- Undescended testes (UDT) are common.
- Congenital UDT should be confirmed at 3 months of age after postnatal descent is complete.
- Congenital UDT should be referred for an operation at 6–12 months.
- Acquired UDT should be screened for in 4–10-year-olds and referred for a possible operation if the testis does not reside in scrotum.
- Varicocele should be suspected in adolescents who have a pendulous left hemi-scrotum.
- Varicocele diagnosis is confirmed on standing and then referred for surgical assessment.

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CHAPTER 29
Inguinal Region and Acute Scrotum

CASE 1

A 6-month-old boy presents with an intermittent swelling in the left groin. Both testes are in the scrotum.

Q 1.1 What is the likely diagnosis?

Q 1.2 What is the treatment?

CASE 2

A 7-year-old boy complains of pain and swelling in the right scrotum for 6 h. He had mumps recently.

Q 2.1 What is the differential diagnosis?

Q 2.2 Could he have mumps orchitis?

Q 2.3 What is the treatment?

The inguinoscrotal region is the most common site for surgical conditions in childhood. As the area is readily accessible to inspection and palpation, accurate diagnosis is easy, but depends upon a knowledge of normal anatomy and the many conditions occurring in the area. The inguinoscrotal region is not isolated from the rest of the body. Symptoms and signs may arise here in systemic diseases, and vice versa, for example, blood or meconium in the tunica vaginalis from intraperitoneal haemorrhage or meconium peritonitis, or torsion of the testis presenting with pain referred to the abdomen. A careful examination of the inguinoscrotal region, and of the whole patient, is necessary to avoid diagnostic errors.

The acute scrotum

Several conditions cause a red, swollen and painful scrotum [Table 29.1], with wide variations in speed of onset, rate of progression and local signs and the severity of pain [Fig. 29.1].

Torsion of the testis

Testicular torsion is not the most common cause of an acute scrotum, but it is the most important. The spermatic cord undergoes torsion, obstructing the spermatic vessels, and is a surgical emergency because of the high incidence of testicular infarction if the cord is not untwisted promptly. The risk of torsion is greatest just after the testis enlarges at puberty in 12–16-year-olds. Also, the risk is increased in unoperated undescended testes.

Two kinds of torsion occur:

1. Intratunical (or *intravaginal*), the more common, is made possible by an abnormally narrow base of the mesenteric attachment of the testis and epididymis within the tunica vaginalis. The predisposing abnormality is almost always present on the contralateral side as well, and this testis should be fixed at the time of operation to prevent metachronous torsion. Rarely, torsion occurs between the testis and the epididymis, which are connected by a thin sheet of tissue. Unoperated undescended testes are at an increased risk, as their fixation within the tunica is commonly tenuous.
2. Extratunical (or *extravaginal*) torsion is rare. During testicular descent, a plane of mobility between the tunica vaginalis and surrounding areolar tissue permits testicular migration to the scrotum. The tunica becomes fixed to the scrotum after descent, but prior to fixation, an interval of torsion-permitting mobility exists. In accordance with this timing interval, extravaginal torsion typically occurs either just before birth or in the early neonatal period. The testis is almost always necrotic by the time the diagnosis is made.

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Table 29.1 Causes of acute scrotum in children

1.	Torsion of testicular appendage (Hydatid of Morgagni)	60%
2.	Torsion of the testis itself	30%
3.	Epididymo-orchitis	<5%
4.	Idiopathic scrotal oedema	<5%

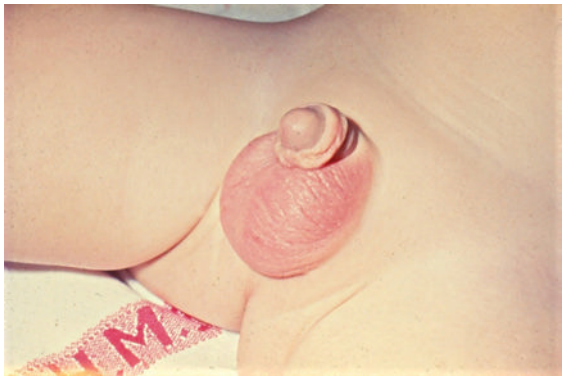

Figure 29.1 Acutely inflamed right scrotum in a prepubertal boy. The inflammation is confined by the right tunica vaginalis. Torsion of a testicular appendage and torsion of the testis itself are likely causes.

Clinical signs

The onset is usually sudden, with pain in the testis and/or ipsilateral iliac fossa, nausea and vomiting. Sometimes, the onset is more gradual, without severe pain, and the diagnosis will be delayed if this less typical mode of presentation is not recognised.

A previous history of similar but short-lived, even momentary, pain is suggestive of episodes of prior incomplete and spontaneously resolving torsion. A horizontal *lie* of the testis when the child stands, often referred to as a *bell-clapper testis*, indicates the possibility of torsion and should be taken as an indication for exploration and testicular fixation.

The swollen testis and epididymis are exquisitely tender (unless already necrotic) and may be partially obscured by overlying scrotal oedema and an effusion into the tunica (reactive hydrocele). The amount of swelling depends on the time that has elapsed and the rate of progression. The hydrocele and the exquisite tenderness may make precise palpation of the testis difficult. As the pathology is contained within the peritoneal membrane of the tunica vaginalis, the inflammatory signs are confined to the ipsilateral hemi-scrotum.

Treatment

Urgent exploration of the scrotum is arranged to untwist the testis and epididymis and to anchor (*pex*) both and the contralateral testis to prevent subsequent torsion. If the testis is completely necrotic, it should be removed.

Torsion of an appendage

Torsion of a testicular appendage (e.g. *hydatid of Morgagni*) is the most common cause of the acute scrotum in prepubertal boys. Testicular (or epididymal) appendages are vestigial remnants of the embryonic Műllerian ducts (that form the uterus and fallopian tubes in females) and are present in about 90% of boys [Fig. 29.2]. Recurrent attacks of pain also occur, sometimes very frequently, and the boy may present with a suggestive history, but few acute signs. A small tender lump at the upper pole of the testis is diagnostic.

Clinical signs

The boy complains of severe pain in his scrotum. A blue-black spot (the infarcted hydatid with secondary haemorrhage) may be seen through the skin of the scrotum near the upper pole of the testis: palpation of it causes extreme pain, whereas palpation of the testis itself causes minimal discomfort. It may be impossible to distinguish torsion of a testicular appendage from testicular torsion once a secondary hydrocele has developed.

Treatment

If testicular torsion cannot be excluded on clinical examination, urgent exploration is mandatory. At operation, the torted appendix testis is removed, which provides relief of symptoms and prevents recurrence. However, if a torted appendage can be diagnosed on the basis of clinical findings, non-operative treatment with effective analgesia is a valid alternative.

Epididymitis or epididymo-orchitis

Epididymo-orchitis is rare in childhood and virtually never occurs between 6 months of age and puberty. Although it is common practice to refer to inflammatory conditions in the scrotum as *epididymo-orchitis*, the inflammation is usually confined to the epididymis.

The most common causative bacterium in children is *Escherichia coli*, carried by retrograde flow along a patent vas deferens from the urinary tract. Predisposing factors for bacterial infection include abnormalities of the urinary tract and urethral instrumentation.

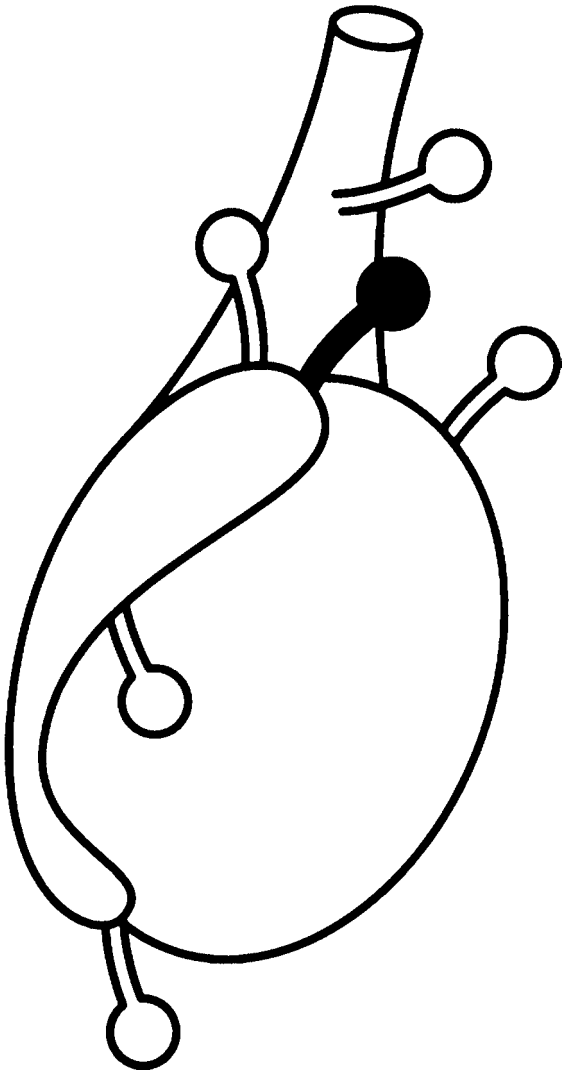

Figure 29.2 Torsion of a testicular appendage. The hydatid of Morgagni is the most common (remnant of the cranial Műllerian duct) and is at the upper pole. Rarely, there may be appendages on the spermatic cord and epididymis (upper or lower poles).

Clinical signs

The usual findings are those of an acute scrotum in a baby or adolescent. A lax secondary hydrocele is common, and bilateral signs are particularly suggestive of epididymitis. Examination of the urine may show pyobacteriuria.

Young children with epididymitis due to urinary organisms should have a renal ultrasound scan after the epididymitis has subsided, and some may require also a micturating cystourethrogram. These investigations aim to identify anomalies of the lower urinary tract before irreversible damage to the kidneys has occurred, as would be the case for a urinary tract infection in a male infant of the same age.

Differential diagnosis

The clinical picture may mimic torsion of the testes so closely that the diagnosis is appropriately made in the vast majority only after exploration of the scrotum. True acute orchitis is very uncommon, but may occur in mumps, Henoch–Schönlein purpura (HSP) or septicaemia. Mumps orchitis is extremely rare prior to puberty, and where the tell-tale parotid swelling is not obvious may be suspected due to a testis, which is larger and harder than expected in epididymo-orchitis. Malignant infiltration of the testis is rare also, but does occur occasionally in leukaemia or with a primary neoplasm (embryonic adenocarcinoma, seminoma or a benign tumour of the Leydig cells).

Treatment

Treatment of epididymitis consists of rest, antibiotics (e.g. co-trimoxazole, nitrofurantoin), a high fluid intake and alkalinisation of the urine. Severe or repeated infections may lead to an abscess or progressive destruction of the testis, but sterility is rare when only one side is affected.

Idiopathic scrotal oedema

In this condition, there is rapidly developing scrotal oedema, which may then spread to the inguinal region, penis and foreskin and/or the perineum.

The scrotum is symmetrically swollen, pale pink or red, and there is slight discomfort rather than acute pain. The pathology involves the skin (and therefore spreads beyond the tunica vaginalis) and may represent allergic inflammation.

Careful palpation reveals non-tender testes that are normal in size and position. The oedema subsides in 1–2 days, but may occasionally recur some weeks later. There may be a history of allergy or of playing outside at the onset; a bite from an insect or a spider is a probable cause in some, but as a rule, the history is inconclusive.

Differential diagnosis

It may be distinguished from other causes of the *acute scrotum* by the spread of oedema beyond the confines of the hemi-scrotum and by the complete absence of

tenderness in the epididymis or testis. Discomfort due to oedema of the scrotum per se may masquerade as testicular tenderness, and so, delivery of the testis into the non-oedematous inguinal region may allow separate assessment of the testis and epididymis versus the scrotum.

The spread of infection from a pustule in the perineum may produce an area of slightly reddened skin and subcutaneous oedema that extends beside or across one-half of the scrotum. A tender enlarged inguinal node at or near the external inguinal ring assists in the diagnosis of perineal lymphangitis.

A toddler who sustains a straddle injury or sits on a toy with a sharp projection may injure the urethra, causing extravasation of urine. Pain on voiding, blood at the urethral meatus and progressive oedema of the perineum, scrotum and suprapubic region are suggestive of urethral injury, which may be confirmed on urethrography (Chapter 38).

Fat necrosis of the scrotum

This extremely rare condition presents with tender, usually bilateral, comma-shaped lumps in the scrotal skin of overweight boys. Trauma may be responsible, but often, there is a history of swimming in very cold water, suggesting that cold injury is the cause. Treatment is supportive, as the necrotic fat gradually absorbs. If doubt exists, exploration is required.

Management of the acute scrotum

As a general rule, an urgent exploration is required in all cases of acute scrotum in which the possibility of testicular torsion cannot be positively excluded. The diagnosis of epididymitis or orchitis is unlikely, unless there is a history of urinary tract infection, a known developmental anomaly of the renal tract or significant pyobacteriuria.

A midline scrotal incision has advantages: when torsion of the testis is found, the testis may be untwisted and fixed, and exploration and fixation of the opposite testis are done through the same incision.

Inguinal lymphadenitis

The superficial inguinal lymph nodes drain the lower limbs, the perineum, the buttocks and the perianal region – all common sites of minor skin infections in the *nappy area* in infants. Infections often reach the inguinal nodes, which become enlarged and may form an abscess after the initial focus has disappeared. Occasionally, atypical mycobacterial infection in preschool children involves these nodes. The axilla, neck and spleen should be examined for evidence of a generalised lymphadenopathy. In small children, an inguinal abscess may be mistaken for a strangulated inguinal hernia. Treatment is incision and drainage when necessary.

Deep external iliac adenitis

The proximal drainage of the nodes of the femoral canal is a group of deep iliac nodes on the pelvic brim around the external iliac artery. For no apparent reason, an infection may pass inconspicuously through the more superficial inguinal nodes to form an abscess in these iliac nodes on the front of the external iliac artery and the iliopsoas muscle.

Clinical features

These are vague; general signs of toxaemia and fever are variable, and the hip may be held in slight flexion. The abscess is at first too deep to palpate clearly, and the diagnosis may be delayed until the abscess is large enough to appear above the inguinal ligament.

On the right side, it may resemble an appendiceal abscess, but a distinguishing point is that a deep iliac abscess is contiguous with the inguinal ligament, whereas in an appendiceal abscess, there is a gap between the two. The absence of vomiting and bowel disturbance is also helpful.

Treatment

Extraperitoneal drainage is required and pus is often present, even when fluctuation cannot be detected clinically because of the thickness of the intervening tissues.

Inguinal herniae

During the seventh month *in utero*, the testis descends into the scrotum inside a diverticulum of peritoneum, the processus vaginalis. This begins to obliterate shortly before birth and closure is normally completed during the first 6 months of life, leaving only the tunica vaginalis surrounding the testis [Fig. 29.3a].

Failure of obliteration of the processus vaginalis is associated with several clinical conditions in infancy and childhood: inguinal hernia, hydrocele and encysted

hydrocele of the cord (and also possibly acquired undescended testes).

A hernial sac may extend from the internal inguinal ring to the tunica vaginalis – the so-called inguinoscrotal hernia [Fig. 29.3b]. More commonly, there is a so-called incomplete sac proximal to an obliterated segment, which intervenes between the sac and the tunica vaginalis [Fig. 29.3c]. This accounts for the vast majority of inguinal herniae in children.

A hydrocele in childhood is a collection of the fluid that lubricates the intestines, formed by omentum within the peritoneal cavity; the fluid trickles down a narrow patent processus and collects inside the tunica vaginalis around the testis [Fig. 29.3d].

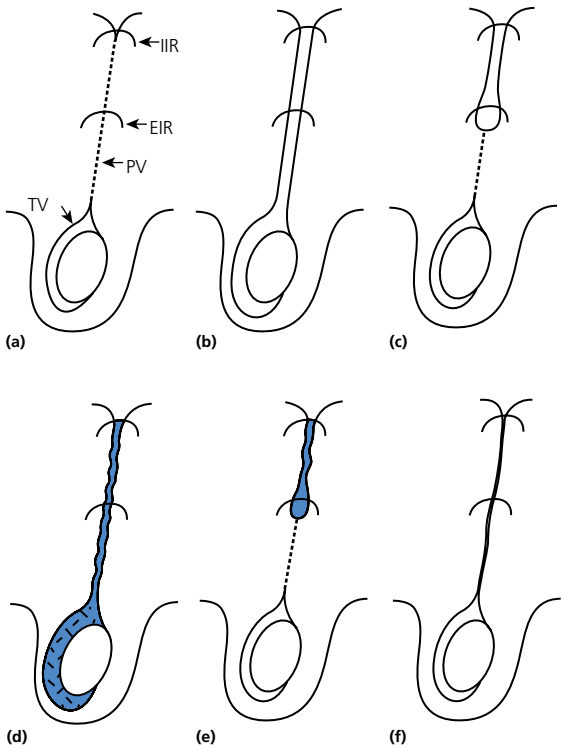

Figure 29.3 Herniae and hydroceles. **(a)** The normally obliterated processus vaginalis (PV) between the internal inguinal ring (IIR), external inguinal ring (EIR) and the tunica vaginalis (TV). **(b)** A completely patent hernia. **(c)** Incomplete hernia. **(d)** Hydrocele with narrow but patent PV. **(e)** Encysted hydrocele with fluid collecting as a *cyst* in the spermatic cord. **(f)** Residual fibrous remnant of PV, which may cause *ascending* testis (Reproduced with permission from Clarnette and Hutson (1997)).

An encysted hydrocele of the cord develops in the same way; the peritoneal fluid collects in a loculus of the processus at some point along the spermatic cord. This loculus usually retains its narrow communication with the peritoneal cavity [Fig. 29.3e]. In some children, multiple loculi or cysts develop along the processus, and it is not uncommon to find a proximal hernial sac communicating through a narrow tract with a distal hydrocele.

Finally, a residual fibrous remnant of the processus may prevent elongation of the spermatic cord with age, leading to an acquired undescended testis or *ascending testis*, later in childhood [Fig. 29.3f].

Failure of obliteration of the processus is more often right sided, perhaps because the right testis descends later than the left and the processus on the right side is therefore more likely to remain patent. Inguinal herniae are also more common in premature neonates, because the normally higher intra-abdominal pressure post-partum compared with the fetus makes it more difficult for the processus to close spontaneously.

In girls, the canal of Nuck undergoes the same obliteration as the processus vaginalis in boys. The obliteration is more likely to be complete, with a lower total incidence of inguinal herniae but a higher incidence of bilateral herniae.

Indirect inguinal hernia

Nearly all inguinal herniae in children are indirect, with an incidence of 1 in every 50 live male births. This is the most common condition requiring an operation during childhood and there is a high familial incidence.

About 10%–15% of indirect inguinal herniae occur in girls, in whom they appear more evenly throughout childhood than in boys. In boys, the greatest incidence is in the first year of life, especially the first 3 months [Fig. 29.4]: 60% are on the right side, 25% on the left and 15% bilateral. The sac usually contains loops of small bowel, and sometimes omentum. In girls, the ovary is often palpable in the sac and may be both difficult and unwise to reduce.

Diagnosis

The child's parent may report that there is an intermittent swelling overlying the external inguinal ring. It is usually painless but on occasions may cause discomfort. An infant's inguinal hernia is often seen at nappy changes as the hernia is made more obvious with crying

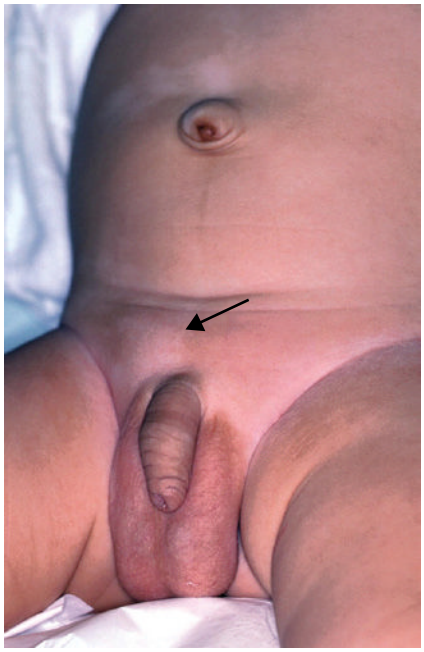

Figure 29.4 Right indirect inguinal hernia in an infant.

or straining. It may reach the bottom of the scrotum, as in the case of a *complete* sac, and there is an impulse on crying or straining.

When the history is suggestive but the hernia is not seen during examination, the index finger may be rolled transversely across the spermatic cord at the point where it lies on the pubic crest; when there is a hernial sac, the spermatic cord is thickened in comparison with the side on which no swelling has been seen, and the *rustle* of contiguous layers of peritoneum represents the empty hernial sac – sometimes referred to as a *silken sleeve*.

One source of confusion between the history and the clinical signs arises when the parents (or referring clinician) mistake a testis in the superficial inguinal pouch for a hernia, and the site of the reported swelling should be precisely indicated and the concurrent presence of a testis in the scrotum documented. If doubt still exists, a further examination is made a few days later, often after the parents have been asked to photograph the swelling.

The opposite side should always be examined, and both testes confirmed to be descended in the scrotum.

Differential diagnosis

The primary distinction to be made is between an inguinal hernia and a hydrocele; the latter is cystic, *brilliantly* transilluminable and irreducible (even though there is a connection, it is too narrow to squeeze the water out quickly), with no impulse on crying or straining, and the examining hand can *get above* the swelling – that is, a hydrocele's proximal pole is distal to the external ring.

Femoral and direct inguinal herniae are rare but should be kept in mind; a retractile or undescended testis may mislead the unwary, and in young children, an inguinal lymph node may be situated close to the external inguinal ring.

Treatment

An operation is necessary in all cases of an inguinal hernia because of the danger of strangulation, which occurs most commonly in the first 6 months of life. Operation should be performed as soon as practicable, unless there is an intercurrent condition, which requires more immediate attention, for example, a skin infection or bronchitis.

An open herniotomy is performed through an incision in the transverse inguinal skin crease and is a relatively simple operation in experienced hands, even in the neonate. Some centres now correct inguinal herniae laparoscopically. Exploration on the opposite side remains controversial and operator dependent.

Strangulated inguinal hernia

Strangulation is the most important complication of indirect inguinal herniae; it is common in infancy but somewhat less common in older children. If small bowel becomes trapped in the hernial sac, the hernia is irreducible and is termed *incarcerated*. In children, incarceration occurs at the external inguinal ring, unlike adults in whom the obstruction is typically at the internal ring. Due to incarceration, blood supply of the trapped contents (and testis) can become impaired, that is, *strangulation*. Untreated strangulation can be life-threatening. Therefore, any irreducible hernia should be considered as being potentially strangulated and effort made to ensure reduction as outlined below.

Strangulated herniae are seen more often in infants under 6 months of age, such that up to 30% of infants with an inguinal hernia initially present with a strangulated hernia.

Clinical features

The infant cries and cannot be pacified; when the parent changes the nappy, a swelling in the groin is noted – often for the very first time. There is a tense, tender

swelling at the external inguinal ring, and no impulse on crying. There may be generalised colicky abdominal pain, vomiting, abdominal distension and constipation when complete intestinal obstruction supervenes – but this may occur 12 h after the onset. With delayed diagnosis, there may be redness and induration overlying the lump, or signs of peritonitis, suggesting bowel ischaemia.

Differential diagnosis

The differential diagnosis includes an encysted hydrocele of the cord, which may appear suddenly – but the swelling is not tender, the cyst moves readily with traction on the cord and abdominal signs and symptoms are lacking.

Absence of a testis in the scrotum on the affected side may point to torsion of an undescended testis or torsion of a descended testis, which has been elevated out of the scrotum.

Lymphadenitis or a local inguinal abscess may be so confusing in young children as to warrant exploration to clarify the diagnosis.

Secondary effects

The testicular vessels may be severely compressed by a tense, strangulated hernia. Testicular atrophy has been reported in 15% of boys after an episode of irreducibility and strangulation. For this reason, early reduction is important for both the testis and the incarcerated bowel. Occasionally, in infant girls, the ovary may be strangulated inside the hernial sac.

Treatment

An incarcerated hernia may reduce spontaneously *en route* to the hospital, but more often than not persists. The strangulated hernia should be reduced by *taxis*. The tips of the fingers of one hand are applied to the distal extent of the hernia while the fingertips of the other hand are cupped at the external ring. Gentle pressure is exerted initially by the upper hand to disimpact the hernia from the external ring, and then, the contents of the hernia are reduced by the lower hand along the line of the inguinal canal. Nothing seems to be accomplished for a minute or two, and then, the bowel suddenly gurgles and returns to the abdomen. *Taxis* is a manipulative trick, not a matter of force, and if necessary may be attempted several times. As long as the necessary monitoring is available, a distressed child may be sedated with midazolam or opiate analgesia. Taxis is successful in over 90% of cases, with virtually no chance of complication by *en masse* reduction.

When it is successful, the patient should not return home until herniotomy has been performed, usually after 24 h, to give time for oedema of the sac and its investing tissue to subside.

When taxis fails, the child should be transferred immediately to a tertiary paediatric surgical centre for operation. The friable sac is difficult to handle and the surgery should always be performed by a paediatric surgeon. In exceptional cases, the bowel is gangrenous and a segmental excision with anastomosis may be necessary. The need for preoperative resuscitation and intravenous antibiotics will be obvious from the clinical findings.

Direct inguinal hernia

Direct inguinal herniae are rare in paediatrics, forming less than 1% of inguinal herniae. They are occasionally seen in premature infants who develop bronchopulmonary dysplasia after prolonged ventilation, and in teenagers with cystic fibrosis. Repair of the posterior wall of the inguinal canal medial to the epigastric vessels is required.

Femoral hernia

Femoral herniae are equally rare. The diagnosis is made clinically when the swelling is below the inguinal ligament and lateral to the pubic tubercle.

As in adults, femoral herniae are more common in females, usually between 5 and 10 years of age. The hernia is usually small and irreducible, for most of it is composed of a fibro-fatty investment of the fundus. The hernia may be repaired easily from below the inguinal ligament.

Hydroceles

Almost all hydroceles in infancy and childhood communicate with the peritoneal cavity via a patent processus [Fig. 29.3]. Much less common is the development of an *acute* hydrocele secondary to some affliction of the testis or epididymis, for example, torsion, infection, trauma or tumour.

Clinical signs

A hydrocele is a painless cyst containing peritoneal fluid, which has tracked down a narrow but patent processus vaginalis [Fig. 29.3d]. It is situated around the testis, is brilliantly transilluminable and cannot be emptied by pressure because of a *flap valve* at its junction with the processus. When the hydrocele is lax, the testis

within it may usually be palpated with ease, or, when the hydrocele is tense, its shadow may be demonstrated by transillumination.

The upper limit of the hydrocele is clearly demonstrable; that is, the palpating finger *can get above it* except in unusual varieties that extend up to the inguinal canal. There is no impulse on crying or straining.

Hydroceles in infants

Unilateral or bilateral hydroceles are common in the first few months of life. They are often large, lax and nearly always symptomless and have a strong tendency to close and absorb spontaneously. Most will have disappeared by the age of 1 year and an operation is only recommended if the hydrocele persists beyond 2 years of age.

An encysted hydrocele of the cord is a loculus of fluid located above and separate from the tunica vaginalis [Fig. 29.3e]. It does not require an operation in infancy and may be considered a variety of the natural process of obliteration.

Hydroceles in older children

In boys more than 2 years of age, there is often a diurnal variation in the hydrocele's size. It is small or absent in the mornings and at its biggest in the late afternoon, when it may cause a dragging ache. These changes reflect the narrow communication with the peritoneal cavity along which the fluid returns during recumbency and reaccumulates by the effect of gravity during the day. Despite the patency of the processus, almost never can the fluid be expelled by pressure.

A hydrocele in this age group rarely disappears spontaneously and an operation is recommended. The processus is transfixed and divided at the internal inguinal ring (i.e. herniotomy). The whole sac need not be removed but the fluid in it may be released.

KEY POINTS

- In acute scrotum, urgent exploration is mandatory unless testicular torsion may be excluded on clinical examination.
- The scrotum contains two peritoneal sacs (tunica vaginalis) that limit spread of inflammation arising from within one of them.
- The processus vaginalis normally closes shortly after testicular descent (i.e. perinatally).
- An inguinal hernia in a baby is potentially dangerous as incarceration is common.
- Hydrocele (with a narrow peritoneal connection) only needs an operation after 2 years of age if spontaneous closure does not occur.

Further reading

Clarnette TD, Hutson JM (1997) Is the ascending testis actually "stationary"? Normal elongation of the spermatic cord is prevented by a fibrous remnant of the processus vaginalis. *Pediatr Surg Int* 12: 155–157.

Glick PL, Boulanger SC (2012) Inguinal hernias and hydroceles. In: Coran AG, Adzick NS, Krummel TM, Laberge J-M, Shamberger RC, Caldamone AA (eds) *Pediatric Surgery*, 7th Edn. Elsevier Saunders, Philadelphia, pp. 985–1002.

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CHAPTER 30
The Penis

CASE 1

A mother brings her 6-week-old son to her general practitioner for advice on the care of the foreskin.

Q1.1 Should she retract and clean the foreskin?

Q1.2 At what age does the foreskin become easily retractable?

CASE 2

A 5-year-old boy presents with a non-retractable foreskin.

Q2.1 How do you differentiate between normal preputial adhesions and pathological phimosis?

Q2.2 What types of treatment are available for pathological phimosis?

CASE 3

A mother of a newborn baby asks for advice on the pros and cons of circumcision.

Q3.1 What advice would you give on neonatal circumcision?

Q3.2 What standards of surgery are required for circumcision?

Q3.3 What are the complications of circumcision?

CASE 4

A newborn baby presents with hypospadias.

Q4.1 At what age is corrective surgery best performed?

Q4.2 What are the aims of surgery for hypospadias?

Q4.3 What are the principles of hypospadias surgery?

The prepuce and glans penis

The prepuce (foreskin) is lightly adherent to the glans in the first few days of life, but becomes more densely adherent during the first year.

Forcible retraction of the prepuce should be avoided until spontaneous separation from the glans occurs. This is usually in infancy, but it may take 5 years or even longer. The normal process of separation of preputial adhesions is by a combination of pressure of urine, traction and stretch applied by erections and self-examination and the build-up of shed skin cells from the inner aspect of the foreskin (smegma). This sebaceous deposit of white cheesy material builds up under the foreskin to lift it off the glans; the discharge of this material is often mistaken for infection.

Accumulations of smegma may produce yellowish bulges under the preputial skin [Fig. 30.1], often referred to as *smegma pearls*, and may be mistaken for sebaceous cysts or even a tumour!

Care of the normal foreskin

The normal foreskin needs no special care in young boys. If the foreskin is healthy, it need not be retracted and does not have to be more specially cleaned than any other part of the body. After puberty, the foreskin should be retracted for cleaning. Young children do get problems with infection in the foreskin, but this is due to phimosis and the foreskin cannot be retracted in this pathological condition. Three abnormal conditions arise in the prepuce.

Phimosis

Phimosis is tightness of the preputial orifice that prevents retraction of the foreskin over the glans [Fig. 30.2]. It may be either physiological (the normal prepuce with residual adhesions) or pathological. In the case of normal preputial adhesions, gentle retraction opens out the foreskin to reveal the tip of the glans and urethral meatus, but the adhesions prevent further retraction. Pathological phimosis may be caused by ill-advised

Jones' Clinical Paediatric Surgery, Seventh Edition. Edited by John M. Hutson, Michael O'Brien, Spencer W. Beasley, Warwick J. Teague and Sebastian K. King.

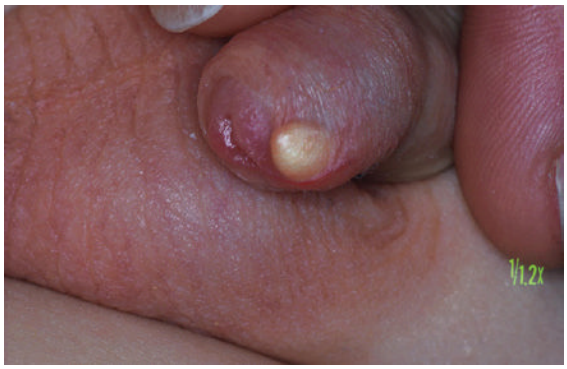


Figure 30.1 Accumulation of smegma beneath the foreskin appears as a yellowish bulge in the coronal groove.

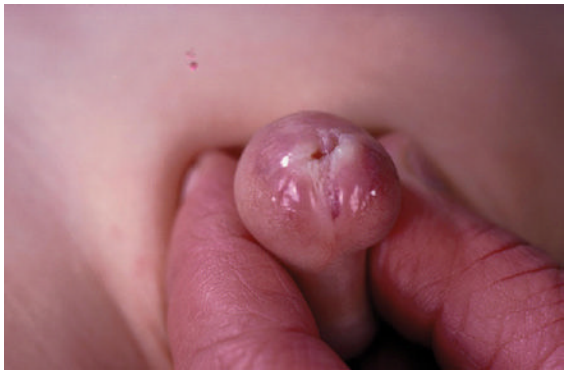


Figure 30.2 Phimosis. Scarring of the distal foreskin causes stenosis of the preputial opening.

forceful retraction of physiological phimosis, recurrent balanitis, an incomplete circumcision or balanitis xerotica obliterans (BXO). Phimosis may impair drainage of the space between the foreskin and the glans, leading to accumulation of stagnant urine and smegma. The urea in the stagnant urine becomes converted to ammonia and may cause an ammoniacal dermatitis of the prepuce, which may then become secondarily infected to cause balanitis.

Resolution of a persisting physiological phimosis may be hastened by the local application of steroid cream applied daily to the phimotic skin for 2–4 weeks. This steroid treatment is successful in many cases, but circumcision can be considered in older boys in whom steroid treatment is not successful. Persisting phimosis should be treated actively due to the likelihood of pain during sexual activity due to trauma to the non-retractile foreskin. Also, if chronic low-grade infection occurs under a non-retractile foreskin for many decades, it can lead to squamous cell carcinoma of the glans in later adult life. Originally described in chimney sweeps during the industrial revolution, this is uncommon now due to marked improvements in personal hygiene.

Paraphimosis

This occurs when a tight foreskin has been forcibly retracted. The retracted foreskin forms a constriction ring around the coronal groove of the glans, causing venous engorgement and painful swelling of the glans [Fig. 30.3]. Increasing pain and retention of urine occur, and the problems demand urgent resolution. Reduction of the paraphimosis may often be achieved by patient gentle compression in an awake patient but occasionally requires a general anaesthetic.

Balanitis

Balanitis is an infection of the glans penis, infection of the foreskin is called posthitis and the combination is referred to as balanoposthitis. It usually starts as an inflammatory process due to ammonia in urine that is retained under the prepuce, which becomes secondarily infected. This infection responds quickly to topical or systemic antibiotics. Recurrent infection may occur until the preputial adhesions have separated completely. Further episodes of infection may be prevented by simple hygiene measures, gentle retraction of the foreskin or the topical application of a steroid ointment to expedite preputial adhesiolysis. A topical barrier

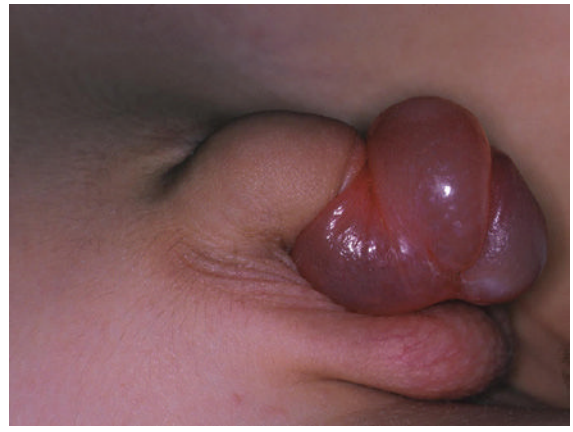


Figure 30.3 Paraphimosis. The penis is red, swollen and painful. The prepuce has been retracted behind the glans and is compressing the shaft, causing oedema and venous congestion.

ointment at the first sign of inflammation may prevent bacterial infection.

BXO

BXO is a scarring condition of the foreskin causing a pathological phimosis that, if left untreated, may extend onto the glans penis and into the urethra causing urethral stricturing. It is a chronic dermatitis of unknown aetiology, similar to lichen sclerosis et atrophicus. If detected early enough, it may respond to topical steroids but usually warrants circumcision.

Circumcision

Phimosis due to BXO and the complications of phimosis, paraphimosis and balanitis are some of the medical indications for circumcision. Persisting physiological phimosis is a relative indication, but resolution of phimosis augmented by topical application of steroid cream has reduced this need for circumcision [Box 30.1].

Circumcision may offer some protection against urinary tract infection (UTI), with a 10-fold reduction in the incidence of UTI in circumcised males from 1/100 to 1/1000. This is not of great significance for the normal child. However, infants with severe urinary tract abnormalities are at increased risk of UTIs and so are more likely to benefit from circumcision, especially in the first year of life.

Apart from these medical indications, circumcision is also performed for religious and social reasons. The pendulum of opinion has swung back and forth, but circumcision for social reasons is now performed less frequently.

Irrespective of the debate regarding the indications for circumcision, the most common reason given by parents is concern regarding cleanliness and convenience, in that the foreskin does not have to be retracted. However, general standards of hygiene are now so high that prophylactic circumcision is not recommended. Carcinoma of the foreskin does occur as a rare tumour in elderly men with long-standing phimosis, but the lesson here is that phimosis should be treated rather than the normal foreskin removed.

Box 30.1 Indications for circumcision

Medical	Phimosis
	Paraphimosis
	Balanitis
	Serious urinary tract anomaly
Non-medical	Religious
	Social

Box 30.2 Complications of circumcision

- Bleeding
- Infection (local or septicaemia)
- Ulceration of glans or meatus
- Penile deformity
- Penile obliteration

There may be complications after circumcision [Box 30.2]. The penis has a very good blood supply and post-operative bleeding may occur. In the neonatal period, any bleeding is of major concern as the blood volume of the average newborn is 80 mL/kg, which is 240 mL for a 3 kg baby. Any blood loss over 25 mL is potentially life-threatening. By the age of 6 months, a baby has doubled its birth weight and the tolerance for blood loss is greater. Infection of the open wound by coliforms in the nappy area may lead to septicaemia in the neonatal period as the baby's own immunity is poorly developed. By the age of 6 months, the baby is better able to cope with infection. The other common complication of circumcision is ulceration of the thin delicate epithelium of the glans. This may occur at any age and sometimes leads to meatal stenosis from scar contraction around the urethral meatus. The glans needs to be protected after circumcision with copious amounts of moisturising cream applied via nappy-liner cloths for 2–3 weeks after the surgery.

Circumcision for religious reasons will be performed at the age and situation decreed by the religion. Circumcision for medical or social reasons needs to be performed at the optimal time for the best standards of medical practice. The standards of safety and skill expected of surgery and anaesthesia are very high, and the previous methods of circumcision performed in the neonatal period do not meet these standards. Circumcision should be performed after the age of 6 months in an operating theatre under general anaesthesia, with careful surgical technique. The parents should receive consultation and education, so they may give informed consent. In years gone by, the large numbers of babies presenting for social circumcision gave rise to the practice of circumcision in the neonatal period without anaesthesia, with a surgical technique in which speed, rather than meticulous tissue handling, was the main

consideration. The situation today is quite different, and circumcision should be judged by the same standards that apply to any operation.

Meatal stenosis

This occurs as an acquired lesion caused by ulceration of the glans following circumcision. It leads to a thin urinary stream with dysuria and bleeding due to meatal ulceration. The problem may be prevented by protection of the glans with moisturising cream for 2–3 weeks after circumcision. Scar contracture after meatal ulceration leads to meatal stenosis, and this requires operative meatoplasty to correct the problem. It is more common if circumcision is carried out while the child is still in nappies.

Hypospadias

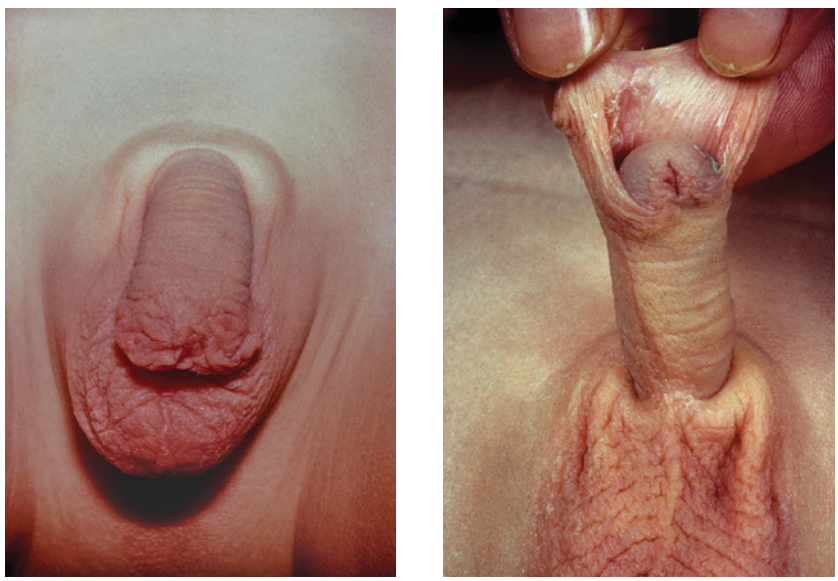
Hypospadias is caused by failure of development of the tissues forming the urethra, on the shaft of the penis. The urethral orifice opens on the ventral surface of the penis and does not reach the end of the glans. In severe cases, the urinary meatus may open in the scrotum or perineum, where a disorder of sexual development should be considered. There is deficiency of the ventral foreskin and the skin on the ventral penile shaft [Fig. 30.4]. The lack of tissue on the ventral surface of the penis leads to a tight *bow-string* effect, causing a ventral bending of the penis known as *chordee*. This chordee deformity is more marked during erection and will cause difficulty with intercourse in later life if not corrected.

The malposition of the urinary orifice and the chordee deformity are usually present together, but in some cases, severe chordee may be present with an orifice at the end of the penis. Hypospadias affects 1 in 150–200 boys.

Severe *hypospadias* with a bifid scrotum and undescended testes is actually a presentation of a disorder of sexual development with ambiguous genitalia (Chapter 10).

The clinical findings in hypospadias are:

1. Downward deflection of the urinary stream from the ventrally placed meatus.
2. The penis is bent ventrally with chordee, which causes difficulty with intercourse.
3. The foreskin forms a dorsal hood and is deficient ventrally, which gives an abnormal appearance to the penis.



(a) (b)

Figure 30.4 Hypospadias. Incomplete fusion of the inner genital folds leads to a proximal urethral meatus, a dorsal hooded prepuce and chordee. The penis may look fairly normal **(a)** until the foreskin (dorsal hood) is pulled upwards, revealing the proximal urethral meatus **(b)**.

These disabilities are primarily functional, as the boy may find it difficult to direct his urinary stream, and in later life, intercourse may be difficult if there is significant chordee. The disabilities are also psychological, as severe anomalies of the penis or a poor cosmetic result following operative repair may interfere with the development of his normal male body image. Therefore, the age for correction of hypospadias has been made younger, and now, the recommended age for repair is 6–12 months.

Investigation

Hypospadias is associated with some increase in other anomalies of the genito-urinary system (9% incidence of cryptorchidism, 9% inguinal hernia, 3% renal anomalies) and investigation with renal ultrasonography is recommended. More severe penoscrotal hypospadias is associated with a utriculus masculinus, a remnant of the vaginal anlagen that may predispose to recurrent UTI, epididymo-orchitis or stone formation. Severe hypospadias with bifid scrotum and/or undescended testes (ambiguous genitalia) requires full investigation for disorders of sexual development.

Treatment

The four aims of treatment are:

1 To correct the chordee
2 To bring the urinary meatus to the tip of the penis
3 To provide a good cosmetic appearance
4 To achieve the above aims with the minimum complications

Hypospadias surgery is one of the most difficult areas of surgery in children. As the primary defect is failure of tissue development, there is tissue missing from the ventral surface of the penis and any simple attempt at closing the defect has a high failure rate. There are over 250 different operations described for hypospadias, most of which are no longer performed having been superseded by procedures with better cosmetic and functional outcomes. However, with modern surgical techniques, the operative results are quite good and the success rate should be 95%. The principles of surgery are as follows:

1 Correct the chordee by releasing the ventral skin that tethers the penis.
2 Relocate the meatus to the tip of the penis using locally based skin flaps.
3 Achieve a cosmetic outcome with either a circumcised or uncircumcised appearance by reconstructing the prepuce.
4 Post-operative urinary drainage is usually aided by a urinary catheter or a urethral stent.
5 In most cases of hypospadias, surgery is performed in a single stage; however, in severe cases, the chordee may be corrected first and then the urethra is repositioned at the tip of the penis at a later operation to reduce the complication rate.

Complications

Failure of healing with complete breakdown, or a partial breakdown with urinary fistula formation, is a distressing problem. Strictures may occur in the neourethra, and poorly corrected chordee will lead to troubles in adult life. These complications used to be common, but the standards of surgery for hypospadias are now quite high, and one should expect good results.

Epispadias

In this condition, the urethra opens at the base of the penis on its dorsal aspect. It is part of the spectrum of lower abdominal wall defects in which ectopia vesicae (bladder exstrophy) is a more severe form (Chapter 8). Most boys with epispadias are incontinent of urine because the bladder neck is deficient; epispadias as an isolated abnormality in a continent child is exceptionally rare, even rarer than ectopia vesicae itself, which occurs in 1 in 30,000 live births.

Apart from the problem of the repair of the urethra, using the same type of urethral reconstruction as in hypospadias, there are many of the same major difficulties that arise in ectopia vesicae.

KEY POINTS

- The normal foreskin needs no special care in young boys.
- Indications for circumcision include phimosis, recurrent balanitis and complex urinary tract anomalies.
- Neonatal circumcision should be discouraged.
- Hypospadias requires operative intervention at 6–12 months.
- Severe *hypospadias* needs investigation for disorders of sexual development.

Further reading

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PART V
Urinary Tract

CHAPTER 31
Urinary Tract Infection

CASE 1:

Stacey is a 5-year-old girl who presents with dysuria, pyrexia and haematuria. There is no relevant past history.

Q 1.1 What investigations should be done?

Q 1.2 What is the likelihood of an underlying urinary tract anomaly?

Q 1.3 If there is no urinary tract anomaly, why has the infection occurred?

CASE 2:

Thomas is 6 months old and presents with fever, lethargy and smelly, turbid urine. He is not gaining weight.

Q 2.1 How would a urinary tract infection (UTI) be confirmed?

Q 2.2 What tests are needed to document a possible urinary tract anomaly?

A UTI is best defined as the symptomatic occurrence of pathogenic microorganisms, usually bacteria, in the urinary tract. It is a common cause of illness in infants and children, may herald an underlying urinary tract anomaly and may be associated with the occurrence of renal scarring and subsequently the development of hypertension. UTIs are commonly misdiagnosed in children. Dysuria and the passage of cloudy urine are common symptoms in children with a febrile illness and do not necessarily reflect UTI. On the other hand, many children with a UTI have non-specific symptoms or have unexplained fever, vomiting or even failure to thrive: in these patients, the diagnosis may be overlooked.

The diagnosis of UTI is based on the presence of a single species of bacteria growing in large numbers in an appropriately collected specimen of urine. The standard required for a significant culture is greater than 10^5 colony-forming units (cfu)/mL, based on samples of urine obtained from clean-catch voided specimens. Lesser counts are regarded as significant in specimens obtained in a more sterile manner, for example, 10^3 cfu/mL for specimens obtained by urethral catheterisation and 10^2 cfu/mL for specimens obtained by suprapubic aspiration. Asymptomatic bacteriuria has been reported in the urine of 8% of infants and 6.6% of children.

The diagnosis of a UTI is further supported by the detection of white blood cells (WBCs) in the urine ($>5\times10^6$/L in boys and $>40\times10^6$/L in girls). But this is not a prerequisite for the diagnosis. Children on immunosuppressant therapy may not be able to produce an immune response, and some infants with overwhelming sepsis may have bone marrow suppression. WBCs can also be found in the urine of patients without a UTI such as those with intra-abdominal infection (e.g. appendicitis) and other pyrexial illnesses; however, there will not be a significant bacteriuria.

Incidence/prevalence

There is considerable variation in the reported incidence of UTI. By the age of 7 years, approximately 8% of girls and 3% of boys will have been treated for a UTI. UTI is more common in neonates and decreases steadily after the first month of life. A large Swedish population-based study of infants under the age of 2 years reported an incidence of UTI in 2.2% of boys and 2.1% of girls. After this age, UTI becomes more common in girls such that by the age of 16 years, 3.6% of boys and 11.3% of girls will have been diagnosed with a UTI.

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UTIs are responsible for 1–5% of febrile illnesses in children under 2 years of age. A UTI is more common in children with higher temperatures, with UTI as the cause of pyrexia greater than 38° in 9% infants less than 2 months old. It was diagnosed in 7% of infants with a maximum temperature of less than 39° and in 16% of those whose temperature was 39° or higher.

Clinical presentation

The symptoms and signs of UTI vary in children of different age groups [Table 31.1]. In older children, a UTI presents with typical symptoms of cystitis (such as frequency, dysuria, hesitancy, secondary enuresis and suprapubic pain, or upper UTI) and pyelonephritis (such as fever, vomiting, malaise and loin pain). All children with unexplained pyrexia should have a UTI excluded.

History

A detailed history is important and should include antenatal and perinatal history, fluid intake and voiding patterns as well as bowel habits. A history of previous UTI or any previous episodes of unexplained fever is important. Bed-wetting or voiding disorders do not necessarily indicate a urinary tract abnormality, except in a child who has been previously continent, although bladder instability may often present with recurrent UTIs. On the other hand, a history of constant dribbling of urine is abnormal and requires investigation to exclude an ectopic insertion of a ureter. The family history is pertinent, as vesicoureteric reflux (VUR) and duplex kidneys are known to be common among siblings.

Table 31.1 Presentation of urinary tract infection

Infants	Older children
Pyuria of unknown origin	Abdominal pain
Septicaemia	Dysuria
Listlessness and lethargy	Pyrexia
Haematuria	Haematuria
Vomiting	Pyelonephritis
Failure to thrive	Dysfunctional voiding
Persistent neonatal jaundice	

Clinical examination

A general physical examination should include blood pressure measurement, because hypertension in a child with a UTI indicates significant renal pathology. The abdomen should be examined carefully for a renal mass or an overdistended or expressible bladder, which in a neonate is suggestive of a neurogenic bladder. The perineum should be inspected carefully to check perianal sensation and anal tone. Labial adhesions, phimosis, meatal stenosis (and even rarities such as prolapsing ureterocele in a female) can be diagnosed on inspection. *A urological examination includes a neurological examination*, as a neurogenic bladder is an important cause of UTI. The lower limbs are examined for signs of muscle wasting, sensory loss and orthopaedic deformities (e.g. talipes), which suggest neurological abnormality. The bony spine is inspected and palpated for occult forms of spina bifida or sacral agenesis. An overlying patch of abnormal skin (e.g. pigmented naevus, hair, vascular anomaly, lipoma or sinus) may indicate the presence of a serious spinal lesion.

Many abnormalities can be diagnosed from the history and physical examination, prior to organ imaging. Radiological investigations often confirm clinical suspicions.

Diagnosis

In the presence of pyuria, a definite diagnosis of UTI can be made when there is a pure culture of a urinary pathogen in an appropriately collected specimen before antibiotics were started or changed. The choice of method for sample collection will depend on the age and condition of the patient.

Children

There are considerable difficulties in collecting a midstream specimen of urine (MSSU) in infants and toddlers, but it should be possible to collect a clean midstream specimen in the older child. In circumcised boys, the glans should be cleaned with soap and water using a soft flannel rather than antiseptic solutions. The urine is collected midstream in a universal container during continuous voiding. Uncircumcised boys probably do not need to retract the prepuce to clean the glans. Similarly, in the older female child, the labia should be parted, cleansed with a flannel, soap and water from the

front to the back three times, and the child asked to void while holding the labia parted. A disposable funnel may facilitate sample collection in girls. The urine is collected midstream during continuous voiding. Alcoholic preparations should not be used, as these cause intense pain on delicate mucosa.

Younger children

Toddlers who have recently been toilet-trained are often reluctant to void on request into a container, but a reliable sample can be obtained by having the child void into a potty that has been cleaned with hot water and detergent, rather than an antiseptic, or that has a disposable insert.

Infants

Getting a usable sample from infants can be difficult, although a number of reliable methods can be used. A clean-catch specimen of urine obtained by stripping the child from the waist down and waiting for him/her to void provides a sample that is as reliable as that obtained by suprapubic aspiration and better than those obtained by pad or bag collection. Micturition in infants may be encouraged by tapping the suprapubic region or caught when the baby is first exposed to cold as he/she is undressed. Parents generally consider this to be a time-consuming and messy method.

A sterile adhesive urine collection bag is one of the most commonly used collection systems. The bag is applied to the skin around the genitalia after cleaning. Some bags are designed with a secondary inner bag into which the urine drains to minimise skin contact and potential contamination. The bag should be removed as soon as the child has voided and the specimen decanted into a sterile container by cutting a hole in a corner of the bag. Bag specimens are particularly prone to skin contamination but clearly in an appropriately processed specimen should not yield a false negative, and a false positive is unlikely in the presence of significant pyuria.

An absorbent pad can be placed inside the nappy, for those parents who do not like the erythema that adhesive bags produce, and has been shown to produce samples as reliable as bag specimens if properly monitored.

The most reliable technique of collecting urine is by suprapubic aspiration (or by *in/out* catheterisation). In infants up to about 18 months of age, the bladder is an intra-abdominal organ, making suprapubic needle aspiration of urine simple, quick and reliable. A *bladder tap* should be performed in any sick infant to exclude UTI, particularly if a urine specimen obtained by other means is inadequate. In a *septic workup*, it is important to do the suprapubic aspiration first, as infants will void during painful procedures, such as venepuncture or lumbar puncture.

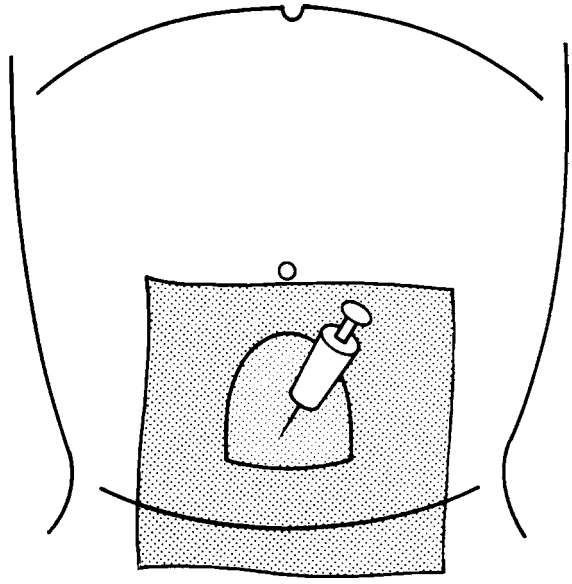

Figure 31.1 The method of suprapubic aspiration for urine culture. The shaded area is the area of aseptic skin preparation.

A 10 mL syringe with a 23 gauge 4 cm needle is used for the procedure [Fig. 31.1]. The child is nursed supine and restrained by an assistant. The suprapubic area is swabbed with skin disinfectant, and the needle introduced in the midline, 1 cm above the upper margin of the symphysis pubis. The needle should be introduced by aiming perpendicular to the floor: in the neonate, insert the needle about 2 cm and further in older infants. The needle is then withdrawn while aspirating on the syringe, until urine is drawn into the syringe. If the child starts passing urine, the urethra should be gently occluded or a clean-catch specimen obtained, so be prepared. It is sent for culture in a sterile container.

Suprapubic aspirates are the *gold standard*, as any concentration of bacteria is considered significant, although false-positive rates in the range of 10–30% have been reported. Furthermore, suprapubic aspiration does not always yield a sample with success rates from 25% to 90%, but this can be improved through the use of ultrasonography.

Once obtained, the specimen has to be processed as promptly as possible, to minimise overgrowth of contaminating bacteria. Samples should be refrigerated at 4 °C if there is to be any delay in processing. At 4°, the sample will remain suitable for culture for up to 2 days.

Sample analysis

Dipstick analysis

Urine dipstick test is now the most commonly used test for UTIs and is used to screen samples for further processing. The most useful components are the nitrite and leucocyte esterase tests. Most pathogenic bacteria produce nitrite by reduction of nitrate. There may be insufficient quantities to be detectable, hence the sensitivity is only 50%, but the specificity approaches 100%. False-positive tests may result from prolonged storage of urine. The urinary frequency in children with a UTI may lead to a false negative. Leucocyte esterase is a marker for WBCs and has similar false positives and negatives. Dipstick tests cannot be relied upon to confirm or exclude a UTI. They are most useful in children with vague symptoms in whom the clinical suspicion of a UTI is low. A negative dipstick suggests that the probability of a UTI is low and that patients can await the result of microscopy or culture before starting therapy. Regardless of the dipstick result, all children with a suspected UTI should have urine cultured to yield a definitive diagnosis.

Urine microscopy

The absence of bacteria or WBCs on microscopy makes a UTI unlikely. Bacteria are rendered more readily visible by either Gram staining or using phase-contrast microscopy, as now recommended in some renal units.

Urine culture is the definitive test for UTI and takes up to 24 h. A further 24 h subculture in the presence of antibiotic-impregnated discs is required to define antibiotic sensitivities.

Pitfalls in diagnosis

The urine specimen may be clear in a child with early pyelonephritis and upper tract obstruction. In this instance, the child should be treated empirically, and further specimens of urine should be taken during treatment, as it is common for bacteriuria to be detected on the second or third day.

The child with an infected urinary calculus may have more than one urinary pathogen cultured from the urine specimen.

Cloudy urine does not always signify UTI. In many instances, the cause of the cloudiness is simply precipitation of phosphate crystals when urine cools rapidly.

Organisms

Most UTIs are caused by a single organism originating from the bowel. *Escherichia coli* is the causative organism in approximately 75% of cases. More than 90% of upper UTIs are caused by *E. coli* possessing P fimbriae, which allow the bacteria to adhere to the urothelial lining and avoid elimination by micturition. Other causative agents include *Klebsiella*, *Streptococcus faecalis* and *Proteus mirabilis*. Proteus, a preputial commensal found in 30% of uncircumcised boys but only 2% of circumcised boys, produces urease and therefore promotes stone formation. Urease splits urea to form ammonia and increases urinary pH, which precipitates calcium and magnesium phosphate salts. Less common species such as *Pseudomonas*, *Staphylococcus aureus*, *Enterobacter*, *Citrobacter*, *Serratia marcescens* and *Acinetobacter* are more likely in children with urinary tract anomalies. *Candida albicans* rarely presents in the community at large but is now the second most common pathogen in hospital-acquired infections, especially those with indwelling catheters or on immunosuppressants.

There are a number of risk factors for UTI such as incomplete bladder emptying from dysfunctional voiding or VUR. UTIs are more common in uncircumcised boys (see Chapter 30) and those with constipation (Chapter 22).

Recurrence

Approximately a third of patients will have a further UTI within 3–6 months, especially younger infants and girls. Among girls who develop a second UTI, roughly half will go on to develop a further UTI. Recurrence is more common in children with high grades of VUR.

Management

Treating a UTI aims to eliminate the acute infection, providing symptomatic relief and reducing or preventing renal scarring. The American Academy of Pediatrics has made a number of recommendations in relation to the treatment of children with suspected or proven UTIs [Box 31.1].

Box 31.1 American Academy of Pediatrics recommendations for UTI management

- Suspect UTI in infants with unexplained fever.
- Await culture results before treatment if non-toxic.
- In unwell child, start treatment before culture result in hospital with IV, especially if less than 1 year old.
- Reassess with repeat culture if not better in 48 h.
- Antibiotics should be given for 7–14 days.

Treatment

Choice of antibiotics

The choice of antibiotics is governed by the sensitivities of the urinary pathogen, usually *E. coli*. Trimethoprim, nitrofurantoin and cefalexin are first-line options for empirical treatment while awaiting the results of urine culture. If the patient has been taking antibiotics recently, then a change of antibiotic may be appropriate unless they are clinically responding. *E. coli* resistance to trimethoprim is increasing, and 15–40% of studies report resistance. Co-trimoxazole (trimethoprim and sulfamethoxazole) is now seldom used in children because of the association of sulfamethoxazole and Stevens–Johnson syndrome.

Nitrofurantoin is effective but more likely to cause nausea and vomiting so is best taken with meals. Resistance to nitrofurantoin is also on the increase and it is ineffective against *P. mirabilis*. For patients with a history of previous antibiotic resistance or with breakthrough infections while on antibiotic prophylaxis, second-line choices include co-amoxiclav, an oral cephalosporin or pivmecillinam. Amoxicillin alone is not suitable because 50% of urinary pathogens are resistant to it. Nitrofurantoin and nalidixic acid are poor antibiotics in the ill child, as they do not achieve adequate tissue levels. Similarly, the new quinolones, although highly effective for treating adult UTI, are not suitable for children, as they may cause erosion of articular cartilage. Aminoglycosides are useful in serious upper UTI, but need careful monitoring in the child with poor renal function, because of nephrotoxicity.

Investigations

Investigation of patients with UTI aims to prevent progressive renal scarring and its consequences – hypertension and renal insufficiency [Box 31.2]. Scarring is a recognised complication of upper UTI; therefore, imaging

Box 31.2 Urinary tract investigations

Renal ultrasonography
Good screening test for obstruction and anatomical variants
Radio isotope imaging
MAG3/DTPA
Excretory scans measuring function and degree of obstruction
DMSA
Static renogram showing state of parenchyma (scar/inflammation/dysplasia)
MCUG
Gold standard test for VUR
Plain radiograph
Useful for spinal anomalies + calculi

is aimed at detecting scarring and identifying children at risk of further scarring. Therefore, the first investigation should be to determine the location of the infection, that is, upper or lower urinary tract. Lower UTIs are not associated with the development of renal scars, and further investigations are less useful. Clinical suspicion based on symptoms and clinical findings may be suggestive of an upper UTI but not conclusive. The gold standard test for the detection of pyelonephritis is a nuclear medicine scan – DMSA. Power Doppler ultrasonography may be as effective as DMSA in detecting acute pyelonephritis and renal scars, but this is not proven. Routine ultrasound scanning is not as effective as DMSA in the detection of upper UTIs.

The incidence of urinary tract abnormality in children with one proven UTI is at least 30%, and higher in the first year of life. The most common abnormality found is VUR. The incidence of VUR in children less than 1 year old with a UTI is less than 50%. A causal association between VUR and renal scarring was first proposed in the 1960s, secondary to reflux of infected urine. In recent years, there has been a paradigm shift in our understanding of the significance of VUR, following the detection of renal *scars* in neonates without a documented UTI. These defects probably represent congenital renal dysplasia that has developed in association with an abnormal ureteric insertion into the bladder. While VUR is a significant risk factor for recurrent UTIs, it is a weak predictor of renal damage in children hospitalised with a UTI. Added to the significance of detecting or excluding VUR is the uncertain clinical benefit of treating children with VUR. While there is no doubt about the benefits of treating an acute UTI, there is no evidence of prevention

of renal scarring by long-term prophylactic antibiotics. A large systematic review has failed to find evidence to support the clinical effectiveness of routine investigation of children with a confirmed UTI. This is not because the investigations do not yield positive results but rather because of a paucity of evidence of the significance of those findings or evidence of a change in disease progression in response to therapy.

This suggests investigation of children with UTI should be targeted on those children at higher risk of renal scarring such as the very young (<2 years old), those with recurrent UTIs and those with known anatomical abnormalities. It cannot be overstated that adequate documentation of UTI is important, and a clinical diagnosis of *UTI* without urine culture is inadequate. Given the low-cost, low-risk nature of renal ultrasonography, it seems reasonable to perform a renal ultrasound scan with pre- and post-micturition images on all patients with a proven UTI. In infants, it is a useful screening tool for obstruction, duplication and other congenital anomalies and in older children may suggest a degree of voiding dysfunction with incomplete emptying of the bladder on micturition.

Renal ultrasonography

An ultrasound scan is a good study for children as there is no ionising radiation involved and there is no need for painful injections. This is an accepted preliminary investigation to exclude urinary obstruction. If the scan shows severe hydronephrosis with obstruction and pus, an emergency percutaneous nephrostomy should be considered to drain the infected urine. This is minimally invasive and, similar to draining an abscess, provides immediate relief of symptoms, enables antegrade studies to detect the level of obstruction and may save the kidney. Ultrasonography is also valuable in the diagnosis of double systems and ureteroceles.

Nuclear isotope imaging

Nuclear imaging of the renal tracts is useful for assessment of renal function, but does not give good anatomical information. The main renal isotope scans available are the *MAG3*, the *DTPA* and the *DMSA*.

The MAG3 and DTPA are excretory scans providing dynamic renography that measure differential renal function and an estimate of glomerular filtration rate. They suggest obstruction when the clearance after the administration of Lasix is delayed; however, they must be interpreted with caution as there is a high rate of false-positive detection of obstruction. The DTPA scan is unreliable in the neonates up to about 6 weeks post-term, due to the immaturity of the kidneys, and for this reason, the MAG3 is used in these patients. Dehydration interferes with assessment of obstruction, as low urine flow causes delayed excretion. Increasingly, the dynamic renogram is being extended to look for VUR but cannot accurately grade the degree of reflux.

The DMSA scan is a static renogram and a more useful test in the neonatal period. DMSA is taken up by functioning renal cortical tissue, but does not give any indication of the excreting or concentrating ability of the kidneys. It is useful in determining renal damage in reflux-associated nephropathy and whether there is any functioning renal tissue in the neonate with gross hydronephrosis.

Micturating cystourethrogram

A micturating cystourethrogram (MCUG) is performed by the insertion of a small catheter into the bladder, filling the bladder with conventional radiological contrast and screening the patient during voiding to detect abnormalities. An MCUG remains the *gold standard* for the detection and grading of reflux (see Chapter 32). In the male child, it is mandatory to examine the urethra during voiding to exclude outlet urethral obstruction.

Plain abdominal radiographs

These may be useful for showing spinal abnormalities, renal or ureteric calculi or faecal loading.

KEY POINTS

- UTI is common in infancy and needs confirmation by culture and screening for underlying anomalies.
- Recurrent pyelonephritis predisposes to hypertension and renal damage.

Further reading

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CHAPTER 32
Vesico-ureteric Reflux (VUR)

CASE 1

Melanie is a 5-year-old girl who presents with a history of recurrent urinary tract infection.

Q 1.1 Which further investigations should be performed?

Q 1.2 What are the pros and cons of the micturating cystourethrogram?

Q 1.3 Are there any alternatives to the micturating cystourethrogram?

CASE 2

A 1-year-old child with severe right-sided VUR and recurrent urinary tract infection (UTI) is found to have reflux nephropathy with defects in the upper and lower poles of the right kidney.

Q 2.1 Is reflux nephropathy congenital or acquired?

Q 2.2 If the recurrent urinary tract infections are kept under control, will further renal damage occur?

Q 2.3 What are the indications for corrective surgery?

Vesico-ureteric reflux (VUR) – the retrograde passage of urine from the bladder up the ureter – is the most common abnormality detected in children with a UTI. It is found in up to a third of all children presenting with a UTI and in greater than 50% of those less than 1 year old. Frequent and complete micturition protects against UTI by *flushing* the urinary tract and removing any bacteria. Children with reflux do not empty completely and are therefore at risk of UTI. Furthermore, reflux allows transfer of bacteria from the bladder to the kidney, with the risk of developing pyelonephritis and renal scarring.

Incidence

Micturating cystourethrogram (MCUG) demonstrates VUR in 1–2% of healthy children, although it is an active and intermittent phenomenon and may be missed in 15% of studies [Table 32.1]. VUR is five times more common in girls than boys and is up to 50 times more common in siblings of children with reflux.

Pathogenesis

VUR may be a primary, congenital anomaly or secondary to abnormal bladder function, which may itself be congenital or acquired.

Primary VUR is due to a failure of the one-way valve at the vesico-ureteric junction. The normal ureter runs inside the bladder muscle and under the epithelium for some distance before opening into the bladder cavity. This part of the ureter, known as the submucosal tunnel or intramural ureter, is compressed against the muscular bladder wall by the increased intravesical pressure associated with bladder filling or micturition. If the submucosal tunnel length is too short, then the ureter may not be adequately compressed to prevent reflux. It is the increasing length of this submucosal ureter with growth that is responsible for spontaneous resolution of low grades of VUR with age.

Secondary VUR describes reflux due to impaired bladder outflow. This impairment to outflow with a subsequent increase in intravesical pressure may result from physical or functional impediments to bladder emptying. Congenital anatomical causes of secondary

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Table 32.1 International Reflux Study Committee definitions of grades of VUR, percentage incidence of each grade together with likelihood of spontaneous resolution

Grade	Definition	Percentage incidence	Spontaneous resolution
I	Reflux into ureter only	7	83
II	Non-dilating reflux to the level of renal calyces	53	60
III	Mild to moderate calyceal dilatation with minimal blunting of calyces	32	46
IV	Moderate dilatation with loss of forniceal angles but preservation of papillary impressions	6	9
V	Gross dilatation and tortuosity	2	0

VUR include posterior urethral valve and neuropathic bladder in patients with spina bifida. VUR may develop secondary to voiding dysfunction seen in older girls or in patients with *dysfunctional elimination syndrome*, hence the association of VUR and constipation.

Consequences

The detection of reflux per se is of little significance; rather, it is the consequences of its presence that matter. It used to be thought that there was a clear association between VUR, UTI and renal *scarring*, but in recent years, the margins have become blurred (see Chapter 31 – UTI). We now know that renal dysplasia can exist prior to any infection, that sterile reflux does not produce scars and that pyelonephritis can cause scarring in the absence of reflux. In children found to have VUR after a UTI, static isotope renography (e.g. DMSA scan) reveals photopenic areas suggestive of inflammation or scarring in 25–40%. Some of these *scars* will not be due to infection but rather represent congenital renal dysplasia. Fifteen to thirty percent of infants born with antenatally suspected VUR (based on ultrasonographic findings) will have isotope evidence of renal dysplasia antenatally, usually in the form of a global reduction in renal size. By contrast, infective renal scarring tends to result in focal areas of renal damage, usually at the poles of the kidney where the renal papillae are most susceptible to reflux.

Some patients with renal scarring, regardless of the aetiology, will develop hypertension. Raised blood pressure has been found in about 15% of patients with VUR, UTI and dysmorphic kidneys. Reflux nephropathy is responsible for paediatric end-stage renal failure in about 22% of patients.

Presentation

Urinary tract infection

VUR is found in 30–50% of children presenting with a symptomatic UTI (see Chapter 31 – UTI).

Antenatal diagnosis

There is no accepted ultrasonographic definition of antenatal hydronephrosis (ANH), but we would investigate all infants in whom the anterior–posterior (AP) diameter of the renal pelvis is 5 mm or more. VUR is detected postnatally in 10% of all neonates with ANH and is more likely when the AP diameter is less than 15 mm; more severe ANH tends to be associated with anatomical obstruction. Postnatal confirmation of ANH is undertaken with an ultrasound scan within the first week of life (and again at 6 weeks of age). If hydronephrosis is confirmed, then an MCUGs is done to look for VUR (as well as to exclude urethral obstruction caused by posterior urethral valve). Interestingly, 25% of babies with normal postnatal ultrasound scans have reflux on MCUG, but mostly, this is of no consequence.

The diagnosis of reflux on an MCUGs at this early stage, before the development of UTI, enables administration of prophylactic antibiotics, which, it is hoped, by preventing reflux of infected urine will limit renal scarring. There is some evidence that long-term prophylactic antibiotics prevent recurrent UTIs but no evidence that renal scarring is reduced. So, while it is uncertain whether prophylactic antibiotics will reduce the long-term risks of scarring, hypertension and renal failure, the benefits of UTI reduction in infants are worthwhile, especially as these children are often hospitalised.

Family history

VUR has been found in a quarter to a half of siblings of children with VUR. Given the current debate regarding the significance of VUR, investigation of asymptomatic siblings is even more controversial. There is some evidence that a normal renal ultrasound scan obviates

further testing. VUR, if present, is likely to be low grade, and in these patients, the benefit of prophylactic antibiotics has not been proven.

Diagnosis

There are no clinical symptoms or signs specific to VUR; it can be diagnosed only by special investigations.

Lower tract studies

The MCUGs or MCUs is the gold-standard test for the diagnosis of VUR [Fig. 32.1]. The bladder is catheterised and filled with x-ray contrast, and the child is then screened while voiding. Although invasive and uncomfortable, as well as documenting the presence of reflux, MCUGs allows the severity of VUR to be graded [Table 32.1] – which has implications for prognosis and potential spontaneous resolution – and provides detailed anatomical information about the bladder and urethra. Because of the discomfort associated with urethral catheterisation and the risk of causing a UTI, MCUGs should not be requested in every patient. Some factors to consider when deciding on whom to order an MCUGs include:

1 Age: Urethral catheterisation is easier and the diagnosis more important in infants less than 12 months of age.

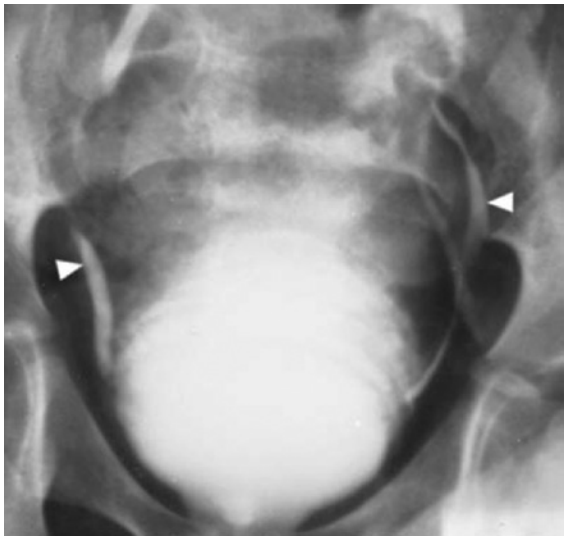


Figure 32.1 Bilateral Grade 1 VUR shown on MCUG. The contrast in the lower ureters is arrowed. There is a high chance that reflux of this grade will resolve spontaneously.

2 Recurrent UTI: A child with recurrent UTIs proven on urine culture should have an MCUGs to check for VUR or other associated anomalies. The zeal with which an MCUGs is sought will depend on the age of the child as VUR is probably less significant in older children in terms of further management.
3 First UTI: A child who has one documented UTI should have an MCUGs if the child (a) is under 12 months of age; (b) has clinical or sonographic evidence of pyelonephritis; (c) has abnormalities, for example, hydronephrosis, scarring, duplex on ultrasonography; and (d) there is a strong family history of urinary tract abnormalities (controversial).

If the patient is due for an examination under anaesthetic (e.g. cystoscopy) anyway, then a catheter can be inserted under GA and the MCUGs carried out later the same day.

If clinician or parental concerns relate to the use of radiation to the gonadal region, then a direct isotope cystogram can be performed. This test also involves urethral catheterisation and bladder instillation with a radioisotope. This test will allow for a longer period of assessment, making the detection of VUR more likely, but does not enable accurate classification.

The indirect isotope cystogram avoids the need for urethral catheterisation by extending the dynamic renogram using either DTPA or MAG-3 isotope, which having passed through the kidneys accumulates in the bladder and may indicate the presence of VUR by showing a second increase in radioactivity with the renal region of interest.

Upper tract studies

The performance of investigations to examine the upper tracts is less controversial. Routine renal ultrasonography is a well-tolerated, non-toxic, inexpensive investigation that can be repeated periodically to assess renal growth and scar progression.

Isotope renography, though more invasive, provides a more accurate assessment of the presence of renal scars, differential renal function and indirectly VUR.

Timing of investigations

Ultrasonography can be performed at any stage, potentially detecting pyelonephritis early or scars late in the clinical course of infection. The MCUG, if undertaken, is

usually delayed until the UTI has resolved, as VUR may be more likely to cause a UTI. The MCUGs is usually carried out prior to discharge. If the isotope study is carried out during the acute episodes, it may detect photopenic areas suggestive of either pyelonephritis or scars. Approximately 50% of these photopenic areas will disappear within 2 months. For long-term prognosis, it is the presence of permanent scars that is significant, and hence, the isotope is best delayed for at least 2–6 months after UTI.

Natural history

There is a strong tendency for primary VUR to resolve spontaneously in the preschool years, with the normal growth of the bladder muscle offering better support to the intravesical ureter. Nearly all cases of mild VUR without ureteric dilatation (Grades I and II) [Table 32.1] resolve spontaneously. More severe cases of VUR with dilatation of the ureter (Grades III, IV and V) [Fig. 32.2] have a lower rate of spontaneous resolution and may require surgical correction. As well as grade of reflux, the probability of spontaneous resolution is influenced by laterality and age of the patient at diagnosis. As the spontaneous resolution of reflux is associated with bladder growth, reflux presenting in older patients is less likely to resolve. Similarly, reflux is less likely to resolve in patients with bilateral, as opposed to unilateral, reflux.

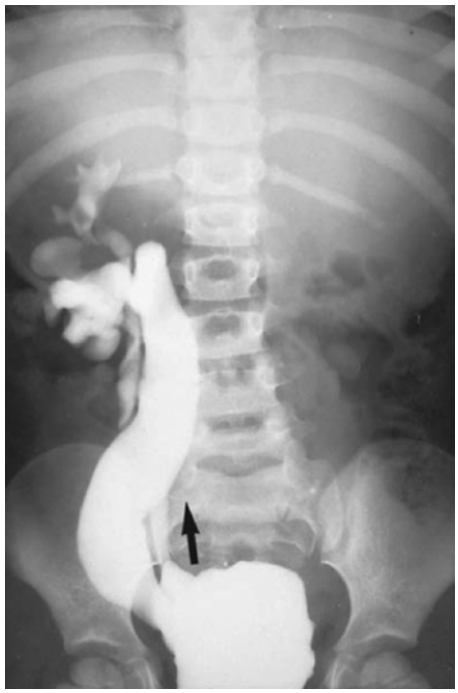


Figure 32.2 MCUGs showing gross right-sided VUR (arrow) up both ureters in a duplex system. There is no reflux on the left.

Management

Medical management

The initial management of VUR is always medical, which aims to prevent symptomatic pyelonephritis and renal scarring, while awaiting spontaneous resolution. Medical management is based on preventing or minimising UTIs on the premise that reflux of infected urine is harmful. This is achieved by ensuring a normal fluid intake and regular toileting, proper perineal hygiene – more important in girls, elimination of constipation if present and administration of low-dose prophylactic antibiotics. The optimum dose schedule and duration of treatment have not been established. Most clinicians will start newly diagnosed infants with VUR on low-dose continuous antibiotic (trimethoprim or nitrofurantoin) administered at night (as it is usually at this time that urine dwells in the bladder for long), stopping either when the child is toilet-trained or has been without a proven UTI for 12 months. Some clinicians would question the need for prophylactic antibiotics at all.

The critical factor in medical management is vigilance and prompt appropriate treatment of UTIs as they occur. This requires close medical supervision and well-informed, motivated parents with ready access to medical attention to prevent pyelonephritis leading to renal scarring and potential long-term damage.

Surgical management

Where medical management has been a failure, as evidenced by recurrent breakthrough UTIs, surgical intervention may be appropriate. Structural anomalies such as para-ureteric diverticulae, ureteric duplication and ureterocele may make spontaneous resolution of VUR less likely but do not negate the potential benefit of a trial of medical therapy. Secondary VUR such as that seen in association with a neuropathic bladder or

posterior urethral valve is best managed by treating the underlying condition rather than surgical reimplantation of the ureters.

There are a number of surgical strategies that may be employed in patients with VUR. Circumcision may be appropriate in boys with VUR, especially if the UTI is due to *Proteus mirabilis*, a known preputial commensal. A nephro-ureterectomy may be appropriate if the reflux is into a non-functioning dysplastic kidney. In the very young/small infant, a temporary vesicostomy – permitting the bladder to drain at low pressure onto the abdominal wall, decompressing the upper tracts and minimising reflux – may be appropriate. However, the primary aim of surgical therapy for VUR is to prevent reflux, and this can be achieved either endoscopically or surgically with ureteric reimplantation.

Endoscopic treatment (*STING* or *HIT*)

Endoscopic injection with synthetic polysaccharide is gaining increasing acceptance worldwide, with published success rates of 75% following a single injection, 85% following two injections and 95% following three injections. Endoscopic therapy offers a number of advantages over open surgery in that it is a day-case procedure, it can be easily repeated and it does not make surgery – for those patients in whom it fails – more difficult. Disadvantages are lingering doubts about its long-term safety and efficacy and some concerns about overtreatment in patients who may have resolved spontaneously anyway (i.e. Grades I and II VUR).

Ureteric reimplantation

For many years, this was the mainstay of surgical management of VUR. This is because the reported success rates for reflux resolution were in excess of 95%. There are a number of differing surgical approaches that traditionally have involved detaching the ureter from the bladder and creating a new submucosal tunnel and neo-ureterovesicostomy largely from within the bladder. More recently, it has been shown that minimally invasive ureteric reimplantation can be done with pneumovesicum (bladder filled with CO_2), although the merits of this new approach have yet to be demonstrated.

KEY POINTS

- VUR is associated with abnormal development of the kidney (dysplasia) and secondary scars of pyelonephritis.
- VUR is common in fetuses and babies, as the bladder (and ureteric valve) is small: resolution is common with growth.
- VUR may be diagnosed antenatally, but postnatal MCUGs is needed for confirmation.

Further reading

McQuiston LT, Caldamone AA (2012). Renal Infection, Abscess, Vesicoureteral Reflux, Urinary Lithiasis and Renal Vein Thrombosis. In: Coran AF, Adzick NS, Krummel TM, Laberge T-M, Shamberger RC, Caldamone AA (eds) *Pediatric Surgery*, 7th Edn, Elsevier Saunders, Philadelphia, pp. 1427–1440.

CHAPTER 33
Urinary Tract Dilatation

CASE 1

Antenatal ultrasonography at 18 weeks shows bilateral hydronephrosis in the fetus, which is still present in the third trimester, when oligohydramnios develops.

Q 1.1 What is the natural history of antenatal hydronephrosis?
Q 1.2 What conditions cause antenatal hydronephrosis?
Q 1.3 What treatment is required at birth?

CASE 2

An 18-month-old male infant presents with fever and dysuria. Urine culture shows an infection and an ultrasound scan shows hydronephrosis and hydroureter (bilateral).

Q 2.1 What causes *hydroureter*?
Q 2.2 What investigations are needed for UTI?

Hydronephrosis is defined as an abnormal dilatation of the kidney, specifically the renal pelvis, and sometimes referred to as pelviectasis. More severe cases have an associated dilatation of the calyces (caliectasis) and, possibly, also the ureter (hydroureter). The presence of hydronephrosis implies a degree of partial outflow obstruction (which may still be present or have resolved), but can also be found associated with retrograde flow of urine or vesico-ureteric reflux (VUR). Differentiating those patients with hydronephrosis secondary to a persisting and potentially harmful partial obstruction from those in whom the dilatation probably represents the sequelae of an obstruction that is now resolving or has resolved presents an interesting clinical challenge. Having determined the level of the likely obstruction, we must then ascertain the potential for renal injury or loss of function.

Hydronephrosis is diagnosed by ultrasonography. A normal kidney will not have any dilatation of its collecting system, and therefore, any dilatation is defined as hydronephrosis. The Society of Fetal Urology has proposed a grading system for hydronephrosis, but most units adopt descriptive documentation of the maximum anteroposterior renal pelvis diameter in a transverse plane at the level of the renal hilum, often referred to as the RPD or renal APD. By consistently measuring the renal pelvis at this point, it standardises repeated observations to look for trends towards progression or regression and also to compare with the published literature for prediction of outcome. A precise APD threshold above investigation should be pursued cannot be found, but most surgeons would investigate a patient with an APD greater than 5 mm.

Clinical presentation

Prior to the advent of routine antenatal screening, patients with urinary tract dilatation typically presented with pain or urinary tract infections (UTIs). Pain is the most common presenting feature in the older child and may be accompanied by infection or haematuria, especially after minor trauma [Table 33.1]. A distinguishing clinical feature is lateralisation of the pain to the loin and accompanying nausea or vomiting. Symptoms are exacerbated by a fluid load and sometimes by position. Intermittent loin pain precipitated by a fluid load (known as a Dietl's crisis) is caused by stretching the renal capsule with a sudden onset of hydronephrosis.

Jones' Clinical Paediatric Surgery, Seventh Edition. Edited by John M. Hutson, Michael O'Brien, Spencer W. Beasley, Warwick J. Teague and Sebastian K. King.

Table 33.1 Clinical presentation of urinary tract obstruction

Child	Infant/neonate
Pain	Antenatal hydronephrosis on ultrasound
Infection	Incidental finding
Haematuria	Infection
Loin Mass	Loin Mass
Incidental finding	Haematuria
	Pain

Nowadays, most neonates and infants with hydronephrosis are detected by antenatal ultrasonography. For that small proportion not detected antenatally, hydronephrosis in the neonate may manifest as a UTI or as a palpable abdominal mass. Presentation as a loin mass is unusual except in a neonate, in whom 50% of all abdominal masses are renal in origin. The most common renal abnormality detected on antenatal screening is hydronephrosis picked up at the 18–20 weeks of gestation scan. When defined as an APD greater than 5 mm, antenatal hydronephrosis was detected in 100 of 18,766 antenatal ultrasound scans or 0.59% of pregnancies. However, in approximately half of these patients, the postnatal ultrasound will be normal. The likelihood of significant pathology increases with increasing size of antenatal hydronephrosis, such that if the antenatal APD was greater than 20 mm, then the majority would require surgery or long-term follow-up; of those with an APD of 10–15 mm, half will have a significant abnormality, and of those with APD less than 10 mm, only 3% have an abnormality.

Another mode of presentation is where renal investigations are performed for suspected abnormalities in children with known multiple anomalies.

Investigations

The investigation for suspected or proven urinary tract dilatation aims to:

1. Demonstrate and document the nature and degree of dilatation
2. Assess renal function (on both sides)
3. Define the abnormal anatomy

Physical examination

Physical examination is aimed at detecting an abdominal mass (suggestive of obstruction or a large multicystic dysplastic kidney) or a palpable bladder.

Ultrasonography

Ultrasonography is the first investigation performed for suspected obstruction and will not only demonstrate any abnormal anatomy but also may determine the likely cause. However, an ultrasound scan will not prove that a dilated system is obstructed, nor will it demonstrate function in the dilated system. Given its non-toxic nature, efforts are continually being made to extend its role to hopefully replace other tests, hence the use of Doppler ultrasound and resistive indices for obstruction and scarring and contrast-enhanced ultrasound to demonstrate VUR (see Chapter 32).

Micturating cystourethrogram (MCUG)

An MCUG is essential in the investigation of children with dilated upper tracts, to exclude associated reflux, but also to exclude distal obstruction, for example, posterior urethral valve in boys. The fervour with which one pursues an MCUG will depend on the individual scenario; for instance, all newborn male infants with small thick-walled bladders and bilateral hydro-ureteronephrosis must have an MCUG. By contrast, a 7-year-old asymptomatic female sibling of a patient with VUR who is found to have mild unilateral hydronephrosis may not have her clinical management altered by the result of an MCUG and hence could be justifiably spared the trauma.

Renal isotope scan

Nuclear medicine or renal isotope scintigraphy may be useful in ascertaining differential renal function and even implied absolute renal function. Renal isotope scans are either static (DMSA), for demonstrating absolute renal parenchyma detection of scars, or dynamic (DTPA or MAG3). Dynamic isotope renography provides both differential renal function and evidence about obstruction or reflux. The interpretation of MAG3 or DTPA excretion curves is prone to significant error and should be left to experts.

A MAG3 scan can be used in the first few months of life when renal function is low (and DTPA scan is ineffective).

Intravenous pyelogram

Intravenous pyelography is used rarely today for the demonstration of function, but is still an excellent investigation where it is essential to demonstrate the anatomy, particularly in duplex systems where both moieties are functioning.

Retrograde and antegrade pyelography

Both techniques are employed to demonstrate anatomy or obstruction when this is essential to the management of the patient.

MR urography

MR urography is increasingly being employed as a non-toxic investigation for the determination of differential renal function as well as anatomical information.

PET

PET scanning, especially when combined with CT or MR, provides an excellent opportunity to locate the elusive upper pole of a duplex kidney in a young girl with urinary incontinence.

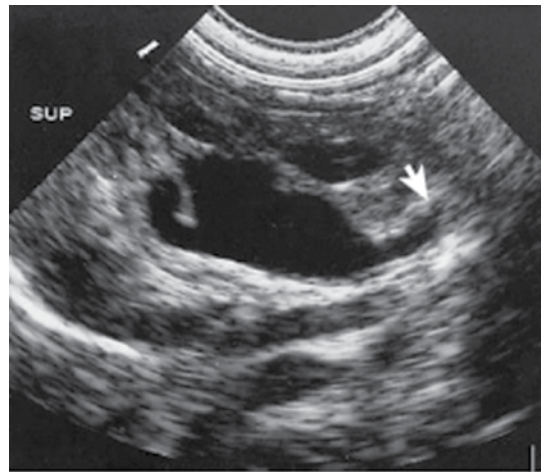
Pitfalls of investigations

The immaturity of the neonatal kidney presents difficulties in interpretation of functional tests in the first month of life. As the concentrating ability and total renal function is low in the neonate, it is likely that functional studies will give misleading results. For this reason, it is best to defer any functional study for at least 6 weeks post-term, although a MAG3 scan can be used at this time. Isotope renography is further prone to errors caused by the level of patient hydration and the regions of interest drawn by the radiographer.

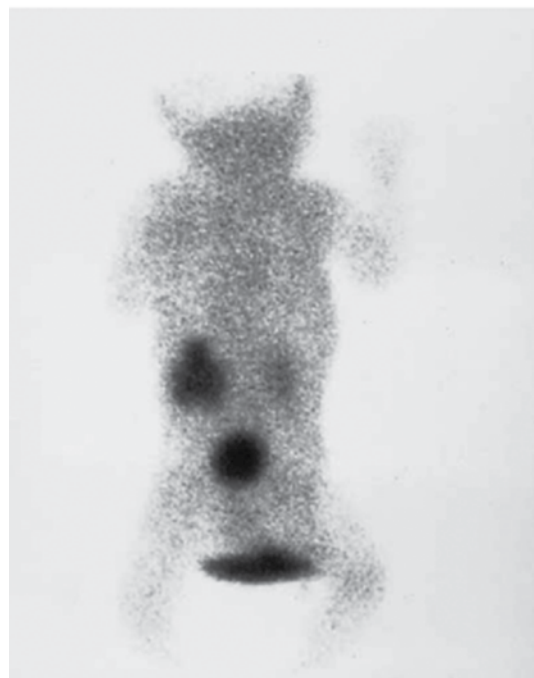
Aetiologic factors

Pelvi-ureteric junction (PUJ) obstruction

PUJ obstruction affects approximately 1 in 2000 children, is more common in boys and on the left side, but may be bilateral in 20–25%. Partial obstruction of the PUJ is caused by intrinsic stenosis (75%), congenital kinking or a lower pole vessel crossing the ureter as it joins the renal pelvis (20%). If the obstruction is intermittent, there is good preservation of renal function in the early stages [Fig. 33.1]. Infection and progressive obstruction lead to loss of renal function, unless severe blockage is relieved surgically. Occasionally, if progressive deterioration has been identified prenatally, early intervention is necessary after birth. However, less severe degrees of hydronephrosis in the newborn often resolve spontaneously. In a large series of babies with antenatal hydronephrosis, babies with postnatal APD less than 12mm rarely required surgery, those with APD greater than 50mm all required surgery and 25% of those with APD of 12–50 mm required surgery because of progressively increasing hydronephrosis or loss of function on repeated isotope renography.



(a)



(b)

Figure 33.1 Postnatal ultrasonography examination in an infant with antenatal hydronephrosis, showing (**a**) PUJ obstruction (arrow) with pelvi-calyceal dilatation, but good preservation of renal parenchyma; (**b**) nuclear renal scan (DTPA) showing holdup at the PUJ at 45 min.

Vesico-ureteric obstruction

Any degree of ureteric dilatation seen on ultrasonography is abnormal as the ureter is a conduit for urine and not a storage vessel. A dilated ureter or megaureter (>7 mm) may be due to obstruction, reflux or a combination of both. Obstruction is usually secondary to a stenosis, or valve in the lower ureter [Fig. 33.2]. Mild cases may resolve spontaneously, leaving a persistently dilated ureter that is no longer obstructed. A ureterocele is a cystic dilatation of the intravesical ureter, which may be associated with a duplex kidney and usually requires endoscopic surgery to relieve the obstruction and improve drainage. More severe cases of ureteric obstruction may require surgical correction in the form of a ureteric reimplantation.

VUR

VUR may present with a UTI and hydro-uretero-nephrosis on ultrasonography or may be found in 9% of neonates with antenatal hydronephrosis (see Chapter 32). Secondary PUJ obstruction due to increasing ureteric tortuosity and kinking may occur.

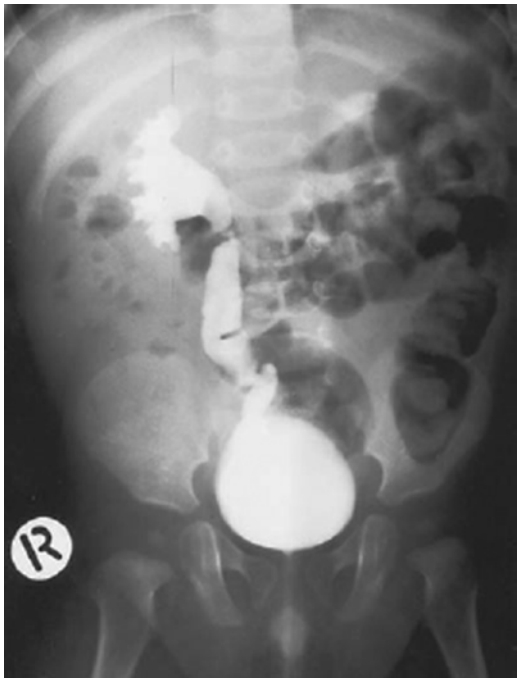


Figure 33.2 Right vesico-ureteric junction obstruction. Note the dilated ureter right down to the bladder.

Posterior urethral obstruction

Posterior urethral valve affects 1 in 8000 newborns and accounts for less than 1% of antenatally diagnosed hydronephrosis. In males, epithelial folds running down from the verumontanum in the posterior urethra form a membrane or *valve* that impedes the flow of urine with back pressure on the bladder, ureters and kidneys. When the obstruction is severe, intrauterine renal failure occurs with fetal death *in utero* or death soon after birth from Potter syndrome. Less severe obstruction allows the fetus to survive, but if the problem is not detected early, septic complications from UTI and metabolic abnormalities caused by renal failure soon occur. The majority of boys are detected or suspected on antenatal ultrasound. The postnatal features include a thick-walled, palpable bladder and a poor urinary stream in a newborn male infant. The diagnosis is confirmed on MCUG [Fig. 33.3]. Fetal intervention is often considered, but is seldom appropriate, and if it has any role, it is probably beneficial to lung development in severe oligohydramnios rather than to preserving or improving renal function. Up to a third of boys with a posterior urethral valve will develop renal insufficiency or end-stage renal failure.

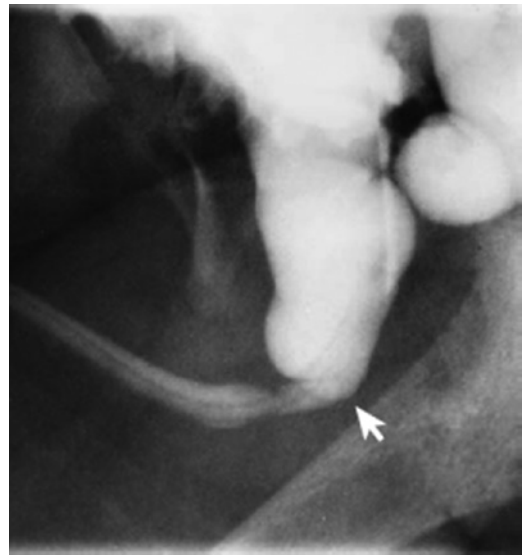


Figure 33.3 Posterior urethral valve (membrane) seen on a lateral view of the urethra on MCUG (arrow). Note reflux into a megaureter, massive dilatation of the posterior urethra and a urethral catheter.

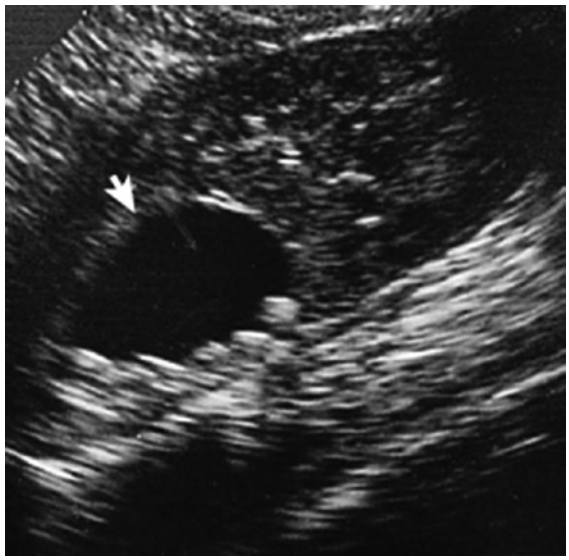


Figure 33.4 Duplex kidney with dilated upper moiety (arrow) on ultrasonography.

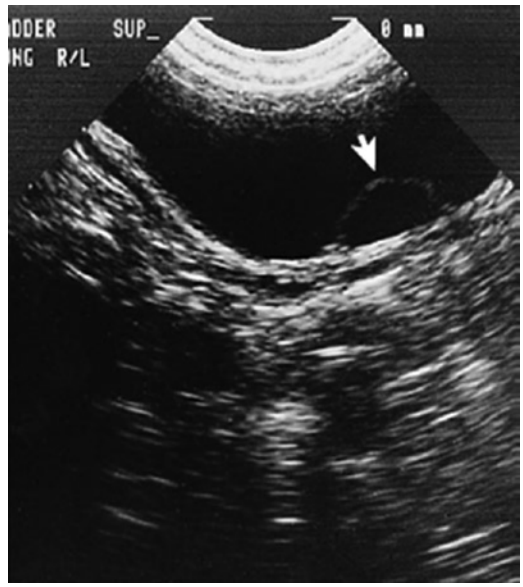


Figure 33.5 Ultrasonography of bladder showing ureterocele (arrow) in the same patient shown in Figure 33.4.

Neurogenic (neuropathic) bladder

Neurogenic bladder causes hydronephrosis in a number of ways. Patients may have a functional bladder neck obstruction from sphincter dysfunction, with upper tract dilatation secondary to high intravesical pressure. Many patients with neurogenic bladder have VUR secondary to the neuropathy, which further exacerbates the upper tract dilatation.

Double ureters and kidneys (duplex system)

Congenital duplex kidneys may develop hydronephrosis of either part of the duplex system. The upper moiety is usually the more abnormal [Fig. 33.4], and the dilatation is caused by dysplasia or distal obstruction (from ureterocele) [Fig. 33.5], or an ectopic position of the ureteric orifice (e.g. in the bladder neck). Ectopic ureteric insertion is often associated with dysplasia in a very poorly functioning upper renal moiety. Dilatation of the more normal lower moiety may be caused by PUJ obstruction or may be associated with high-grade VUR.

Stones (urolithiasis)

Rarely in children, a renal or ureteric calculus may cause an acute obstruction, resulting in hydronephrosis.

Management of obstructive lesions

It is best to divide the investigation and management of hydronephrosis into two age groups: those presenting in the neonatal period and those presenting later.

Antenatal hydronephrosis

Not all hydronephroses on antenatal examination turn out to be significant. In fact, approximately half do not have any abnormality detected on postnatal investigation and are labelled as having had transient hydronephrosis. However, when hydronephrosis is detected antenatally, it is important to follow it throughout pregnancy. If other urinary tract abnormalities are detected on scanning, this would suggest that the hydronephrosis is pathological. Increasing hydronephrosis with oligohydramnios is also pathological, suggestive of low urine output with a posterior urethral valve. The more severe the hydronephrosis, the more likely there will be a pathological cause: most cases with antenatal APD less than 10 mm will either be normal or have VUR, whereas PUJ obstruction is more likely if APD is greater than 15 mm.

Despite lack of good randomised evidence of benefit, most urologists/nephrologists commence all neonates with antenatally diagnosed hydronephrosis on prophylactic antibiotics from birth while awaiting full

evaluation, as there is significant risk of severe UTI developing in these children. They usually receive trimethoprim 2 mg/kg at night.

Preliminary investigations should include a careful clinical evaluation to exclude abdominal masses and inspection of the perineum to detect clinically obvious abnormalities, such as prolapsing ureteroceles.

All children with antenatally diagnosed hydronephrosis should undergo a postnatal ultrasound examination and an MCUG within the first week. It is important that the ultrasound scan is not carried out too early (<48 h), as the neonate is relatively oliguric at this stage and ultrasonography may underestimate the severity of the dilatation. The ultrasound scan will confirm the degree of hydronephrosis [Fig. 33.1] and an MCUG will exclude distal obstruction or VUR, which accounts for 10% of hydronephroses in the antenatal period.

Functional evaluation is of limited value at birth because of the relative immaturity of the kidney; it is best to defer a renal DTPA scan until the baby is 6 weeks old. A DMSA of MAG3 nuclear scan, however, can be very useful in this period, as this shows up any functioning renal tissue.

Except for a posterior urethral valve, definitive treatment can be deferred in most cases until full evaluation of the degree of obstruction is completed. A significant number of apparent neonatal PUJ obstructions improve spontaneously. However, severe obstruction in the neonatal period will require early surgery.

In posterior urethral valve, the bladder is drained by urethral or suprapubic catheter. The metabolic and septic complications are treated before endoscopic resection of the valve is performed. The patient's creatinine is allowed to reach its nadir prior to undertaking surgery to ablate the valve, relieving the obstruction.

Children with severe obstruction usually have gross hydronephrosis on postnatal ultrasound scan. The kidney is tense and usually palpable. A DTPA scan may show a non-functioning kidney, but if the DMSA scan shows an appreciable amount of renal cortical tissue, early repair will lead to significant recovery of renal function.

Management of older children with obstructive lesions

In the older child, the preliminary investigations should always include a renal ultrasound and dynamic renography to determine function, drainage and possibility of obstruction. An MCUG may be indicated, especially if surgery is planned. Unless renal function is severely impaired (<10%), surgical relief of the obstruction should be undertaken. Where there is minimal function, the kidney should be removed [Fig. 33.6], and this can be done laparoscopically.

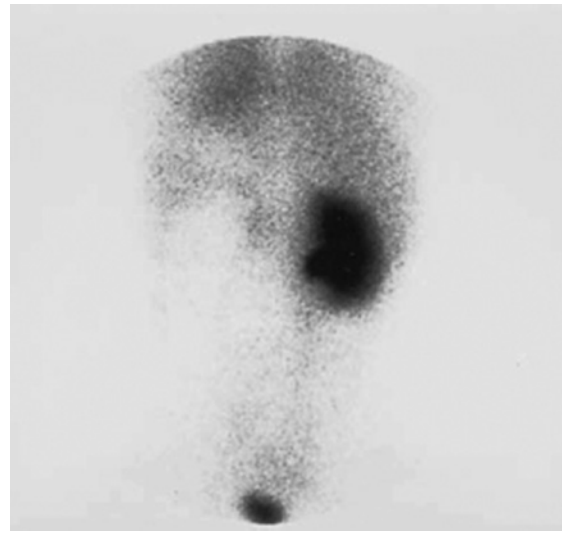


Figure 33.6 Nuclear renal scan showing no function on left side at 5 min (image taken from behind).

Percutaneous nephrostomy

This is a useful emergency measure to drain an obstructed kidney, particularly in the presence of infection. In a sick child with pyelonephritis, it leads to rapid clinical improvement, as well as significant improvement in renal function. Percutaneous nephrostomy also allows evaluation of overall function and delineation of the anatomy by antegrade pyelography.

Pyeloplasty

The standard operative procedure to relieve a pelvi-ureteric obstruction is an Hynes–Anderson pyeloplasty. This requires excision of the narrowed segment and anastomosis of the spatulated ureter to the renal pelvis. The functional results of this operation are good, but these kidneys may retain their dilated appearance permanently. Laparoscopic pyeloplasty is gaining popularity, but the long-term results are not yet known. Attempts at endoscopic management of PUJ obstruction in children have had limited success and have not been widely undertaken given the success of open pyeloplasty.

Total nephrectomy

Nephrectomy may be considered where the back pressure from obstruction has destroyed the kidney, which usually has a function of 10% or less. A poorly functioning kidney will not prevent the need for dialysis were the patient to lose the other kidney, and carries with it a significantly increased risk of sepsis and hypertension.

Partial nephrectomy

Duplex kidneys draining into an ectopic ureter or ureterocele (secondary to ureteric stenosis) are similarly likely to be very poorly functioning and a potential source of recurrent infections. Again, if these moieties provide less than 10–12% of overall renal function, they are treated by partial nephrectomy and excision of the ectopic duplicated ureter.

Obstructed megaureters

Where the obstruction is at the uretero-vesical junction, excision of the stenotic segment, and reimplantation of the ureter into the bladder, is accepted treatment, with good results. In small infants, a temporary stent may be placed endoscopically across the VUJ to relieve the obstruction and removed after 6 months. In our limited experience, this may obviate the need for early reimplantation; however, long-term follow-up is required.

KEY POINTS

- Hydronephrosis diagnosed antenatally is common and often resolves, but all babies need immediate investigation in the first week of life.
- Prophylactic antibiotics are widely recommended to prevent urosepsis while postnatal assessment occurs.
- Surgery is required for severe and/or progressive obstruction, especially if renal function is compromised.

Further reading

Groth TW, Mitchell ME (2012) Ureteropelvic Junction obstruction. In: Coran AG, Adzick NS, Krummel TM, Laberge J-M, Shamberger RC, Caldamone AA (eds) *Pediatric Surgery*, 7th Edn. Elsevier Saunders, Philadelphia, pp. 1411–1426.

McQuiston LT, Caldamone AA (2012) Renal infection, abscess, vesicoureteral reflux, urinary lithiasis and renal vein thrombosis. In: Coran AG, Adzick NS, Krummel TM, Laberge J-M, Shamberger RC, Caldamone AA (eds) *Pediatric Surgery*, 7th Edn. Elsevier Saunders, Philadelphia, pp. 1427–1440.

CHAPTER 34
The Child with Wetting

CASE 1

A 6-year-old girl presents with severe day and night wetting and urinary tract infections (UTIs). She can have dry days and her symptoms are worse with infection.

Q1.1 What is the relationship between UTIs and wetting?
Q1.2 How would you investigate this case?
Q1.3 Discuss further treatment.

CASE 2

A 7-year-old girl presents with continuous mild wetting (a few drops leak out every few minutes) every day without fail and no other symptoms.

Q2.1 Of what condition is this a classic history?
Q2.2 How is this diagnosis confirmed by investigation?
Q2.3 What treatment is required?

CASE 3

A 4-year-old boy presents with severe wetting day and night. When his doctor examines the lumbosacral spine, he finds a previously undiagnosed anomaly.

Q3.1 Discuss the *hidden* variants of spinal dysraphism that may be missed in the neonatal examination and present at a later age with wetting.
Q3.2 Why does the further investigation of these anomalies become much more difficult and costly if not performed in the first few months of life?
Q3.3 How does the management of major neuropathic incontinence differ from other types of incontinence?

Urinary incontinence is the most common disorder of the urinary tract in childhood. It causes immense distress to both the patient and parent. Childhood urinary incontinence forms a spectrum of disease ranging from benign self-limiting nocturnal incontinence to neuropathic incontinence with potential renal impairment. Fortunately, the majority of children do not have any underlying pathology and will achieve dryness even without treatment. Despite these reassuring facts, most families seek medical attention because of the stress and anxiety associated with urinary incontinence. Initially, help is sought from family doctors, continence advisors or paediatricians. Surgeons tend to see those patients who have overt neurological signs or who have failed previous therapeutic intervention.

Development of continence

The bladder (together with the urethra and pelvic floor) has two main functions, namely, the storage of urine at a low pressure and emptying of urine at a socially appropriate time. These functions are achieved because of the visco-elastic properties of the interlacing network of smooth muscle fibres in the bladder wall and the integration, within the brain, of both somatic and autonomic nervous systems that are relayed both to and from the bladder. In infants, who void approximately 20 times a day, micturition is a reflex act coordinated in the pons. Over the next 2 years, the frequency of micturition reduces to around 11 times a day – mainly due to increase in bladder capacity. It is around this time that children also begin to recognise symptoms

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of bladder fullness. By 3 years of age, children have some conscious control and most have daytime control with occasional accidents. Most children are dry by day and night by the age of 4 years old.

The attainment of voluntary control of micturition is dependent on a maturation of communication between the pontine micturition centre, the pontine storage centre and the cerebellum, which receives sensory input from the bladder and pelvic floor, the basal ganglia and the frontal lobes. This development allows the socially appropriate inhibition of reflex voiding and the initiation of micturition at any stage of bladder filling.

Definitions

Previous confusion around urinary incontinence can be attributed to inappropriate use of terminology such as *diurnal incontinence,* which has been used to mean either isolated daytime or both day- and night-time incontinence. The International Children's Continence Society (ICCS) has published standardised definitions and terminology that are descriptive, unambiguous, neutral and in line with adult terminology. The emphasis is on describing and quantifying the patients' symptoms, rather than attempting to pigeonhole patients in subgroups [Table 34.1]. The ICCS terminology is relevant in patients over 5 years of age and/or those who have attained bladder control.

Table 34.1 ICCS recommended terminology for patient symptoms

Storage symptoms	
Increased voiding frequency	Consistently voiding eight times or more a day
Decreased voiding frequency	Voiding three times or less in a day
Incontinence	Uncontrollable leakage of urine, which may be continuous or intermittent
Urgency	The sudden and unexpected sensation of an immediate need to void
Voiding symptoms	
Hesitancy	Difficulty initiating micturition
Straining	Application of abdominal pressure to aid micturition
Intermittency	Micturition that is not a continuous stream but rather as several discrete spurts
Weak stream	The observed ejection of urine with a weak force

Prevalence

Urinary incontinence is a major health-care problem said to affect 10 million Americans, of whom 85% are women. Occasional daytime urinary incontinence has been reported in about 10% of 11-year-old British schoolchildren (7% of boys and 16% of girls). The prevalence decreases with age, with incontinence reported by 3% of 15–16-year-olds (1% of boys and 5% of girls). However, less than half of these patients have wetting of sufficient severity or frequency to seek treatment.

Nocturnal enuresis has been reported in 15–20% of 5-year-olds, 5% of 10-year-olds and 1–2% of 15- to 16-year-olds. Boys are more commonly affected than girls. Less than 3% of children will have an organic cause for their bedwetting. However, 25% will have daytime symptoms in addition to their bedwetting.

Assessment

Surgical assessment of children with urinary incontinence is directed towards the diagnosis or exclusion of an organic aetiology such as abnormal anatomy or neuropathy. This is usually possible based on history and physical examination with little need for aggressive investigation.

History

A full medical history is required to document the nature of the urinary incontinence, such as timing, frequency and pattern, periodicity, severity, precipitating factors, associated urinary symptoms of urgency or dysuria. A detailed voiding history should also be taken, looking at the frequency/urgency of micturition, nature of urinary stream (i.e. continuous/intermittent, strong/weak) any withholding manoeuvres such as crossed legs, squatting with the heel pressed into the perineum (Vincent's curtsey) or holding the penis. An assessment of fluid intake should be made with emphasis on the volume and nature of fluids. Patients should be asked about the bowel habit as constipation predisposes to UTIs, which may precipitate or exacerbate incontinence. The success or failure of previous treatment strategies should be recorded.

A detailed past medical and social history is also important, such as previous UTIs and major events within the family (parental separation/death, birth of new sibling, moving home, changing school, etc.), as these will impact on bladder function. The history should also include pre- and perinatal events such as

birth trauma, neonatal anoxia, prematurity and seizures that are associated with voiding disorder. The impact of wetting on both the child and parents, their reactions to it and the family dynamics should be noted.

The most difficult part of taking a history is getting reliable and accurate details of the voiding pattern, fluid intake, frequency and severity of incontinence. This information is most reliably obtained by asking the parent/child to complete a detailed intake and output diary or bladder diary. Clearly longer periods are associated with reducing compliance and incomplete recording. The minimum for detailed fluid intake and output is 48 h.

Clinical Examination

Physical examination is aimed at detecting organic disease. In addition to a routine physical examination, which should include blood pressure measurement, specific attention should be paid to the examination of the abdomen, looking for a palpable/enlarged bladder from which urine may be expressed. An expressible bladder is strongly suggestive of underlying neurological disease, especially if associated with severe faecal loading. The spine should be inspected and palpated, looking for subtle evidence of occult spinal dysraphism such as hairy patch, cutaneous haemangioma, sinus or a lipoma. The sacrum should be palpated and buttocks examined to exclude sacral agenesis. A limited neurological examination looking at gait, lower limb symmetry, calf muscle wasting, foot deformity, tone, power and lower limb sensation together with lower limb reflexes and the presence or absence of clonus must be carried out. Perineal sensation and anocutaneous reflex must be assessed.

The genitalia must be examined to look for evidence of skin excoriation consistent with incontinence and to detect anatomical abnormalities such as meatal stenosis, epispadias, pathological phimosis in boys and labial adhesions, urogenital sinus and, rarely, ectopic ureter in girls.

Investigations

Urinalysis and urine culture, though rarely positive, are routinely undertaken to look for evidence of UTI and to screen for renal disease. For a child with monosymptomatic nocturnal enuresis, analysis of osmolality of the first urine voided in the morning together with overnight urine volumes may help to direct therapy.

A plain abdominal film is not often indicated and is unlikely to yield usable information on the urinary tract, but it may provide information about faecal loading or occult spinal abnormalities.

Ultrasonography provides a simple, non-invasive, inexpensive look at the urinary tract. It may detect evidence of neuropathic bladder with a thick-walled bladder (>3 mm in distended bladder, >5 mm in empty bladder) or upper tract hydro-ureteronephrosis, suggestive of high-pressure urine storage. It may also detect evidence of duplication, which in girls may herald an ectopic ureter as the cause of continuous urinary incontinence. Assessment of bladder volumes both pre- and post-micturition may reveal significant (>10% of estimated bladder capacity or 25 mls) post-void urine residuals that may indicate outlet obstruction and underlying neuropathy or may influence therapeutic options.

In patients with functional incontinence, uroflowmetry is the simplest and the most commonly performed urodynamic investigation. Patients void into the uroflow apparatus that measures the volume of urine voided over time and plots the result as a graph of volume versus time. From this study one can comment on the shape of the flow curve and hence the nature of the urinary stream. It may be a normal smooth bell-shaped curve, the flattened plateau curve seen in outflow obstruction, the staccato or irregular flow curve seen in patients with incoordination between the sphincter and bladder or the interrupted flow pattern seen with patients with detrusor failure who void by abdominal contraction. The computer will also produce a number of parameters that describe the curve, of which the most useful are the voided volume, voiding time and maximum flow rate, for which nomograms are available to tell whether the flow rate is within the normal range or not. In paediatric urological practice, the uroflow assessment typically consists of three voids with ultrasound assessment scan of post-void residual. The recent addition of pelvic floor surface electromyography to uroflow assessment facilitates the easier detection of bladder sphincter incoordination.

Formal urodynamics assessment or cystometry is undertaken in a very small proportion of patients in whom a clinical diagnosis has not been made, who have failed medical therapy, those with a proven or suspected neuropathy or those patients with high-risk bladders, for example, posterior urethral valve. Correctly performed cystometry requires the simultaneous measurement of intravesical and intra-abdominal pressure, together with pelvic floor electromyography while filling the bladder at a rate close to physiological filling with x-ray

contrast, under image-intensifier screening. It is a time-consuming, intimidating test fraught with potential misinterpretation and should only be carried out by experienced personnel in a dedicated setting. Often, a suprapubic catheter is inserted under anaesthesia, with urodynamic assessment done using the catheter the next day.

Conditions

The simplest and most valid classification, based on onset, is into *secondary*, which refers to children who have previously been dry for 6 months, or *primary* for those who have not. Subdivision into patients with nocturnal or daytime urinary incontinence is also valid, but remember that one in four children with nocturnal incontinence will also have some daytime symptoms.

Nocturnal urinary incontinence or nocturnal enuresis

It is best classified as *monosymptomatic* for those children without any other urinary symptoms and *non-monosymptomatic* for who have concomitant daytime symptoms.

Daytime urinary incontinence

Classification of daytime symptoms is more problematic as there is a great deal of overlap: the ICCS advocates symptom description with reference to incontinence, voiding frequency, voided volume and fluid intake. There are some recognised patient subtypes that are still clinically applicable.

Functional urinary incontinence

Overactive bladder or urge syndrome

This was previously called *bladder instability* and is probably responsible for greater than 80% of children with non-organic daytime urinary incontinence. The critical feature is that of urgency, but urinary incontinence, increased frequency of micturition and reduced voiding volumes may also be present. The symptoms usually worsen as the day goes on. Patients may have identified triggers such as cold, running water, sports or carbonated/caffeinated drinks that will induce detrusor contraction and imminent urinary incontinence that may be averted by one of several withholding manoeuvres – classically squatting with the heel of one foot pressed into the perineum – Vincent's curtsey. Most patients will resolve spontaneously with final resolution often precipitated by moving away from home to live independently as young adults. Only 2–3% of patients are affected in adult life.

Dysfunctional voiding

Dysfunctional voiding occurs when there is a failure to relax the pelvic floor/external sphincter during bladder contraction. This results in a staccato stream with variable urine flow and usually does not result in complete bladder emptying. Girls are almost exclusively affected. UTIs are almost universal, and approximately 30% have vesico-ureteric reflux. These patients also often suffer quite severe degrees of constipation and have therefore been labelled – dysfunctional elimination syndrome. These patients are thought to represent the severe end of those with urge syndrome, who having relied so heavily on voluntary pelvic floor contraction to prevent incontinence are now unable to relax during micturition.

Underactive bladder

Previously referred to as *lazy bladder*, these patients rely on increased abdominal pressure to void and do so with an interrupted urinary stream and are prone to large post-void residuals and recurrent UTIs. It is believed to result from bladder decompensation in patients with prolonged dysfunctional voiding.

Voiding postponement

Typically these patients are infrequent voiders who defer voiding due to either pleasurable distractions; for example, computer games/television (younger children) or due to some behavioural disturbance or psychological co-morbidity. The patients will often void to completion and may or may not suffer from urgency.

Giggle incontinence

A rare condition principally affecting girls who void to completion when giggling/laughing. These patients typically lack other symptoms. It does not tend to resolve, but patients adjust their lifestyle to enable to avoid or limit provocative situations.

Structural urinary incontinence

There are a number of anatomical abnormalities that may predispose to urinary incontinence.

Epispadias/exstrophy

This congenital malformation of the lower urinary tract that will result in incontinence if the epispadias extends sufficiently proximally through to the bladder neck. Will result in incontinence. The diagnosis of bladder exstrophy is obvious and often detected antenatally, as is epispadias in a boy. More subtle degrees of epispadias may be missed in a female patient unless the perineum is specifically examined.

Persistent urogenital sinus

This failure of embryological separation of the urethra and vagina may be associated with an incompetent sphincteric mechanism.

Ectopic ureter

In girls with duplex kidneys, the ureter that drains the upper moiety may enter the urinary tract in an ectopic position, which if below the bladder neck or into the vagina will result in constant low-flow urinary incontinence. This does not happen in boys as the ectopic ureter always enters the urinary system above the level of the external sphincter.

Bladder outlet obstruction

The most common cause of this is a posterior urethral valve in a boy. Nowadays, the majority of boys with valves are detected prenatally, but prior to the advent of antenatal ultrasonography, a third of patients would present late with urinary incontinence and a minority still do.

Neuropathic bladder

This may be present in a patient with known neuropathy such as myelomeningocele, or patients at high risk of neuropathy such as those post-surgery for anorectal malformations or pelvic tumours, or patients with a history of spinal trauma. It may also occur in patients with previously undetected neuropathy as in spina bifida occulta, tethered spinal cord, diastematomyelia or sacral agenesis. These patients may present in any number of ways, and their detection is based on a high index of suspicion and appropriate investigation.

Management

The management of children with urinary incontinence depends on the aetiology of their incontinence. For those with a structural cause, surgery may be appropriate, and, in the case of ectopic ureter, curative. For all other patients, the main thrust of treatment is supportive and educational, as the majority will resolve spontaneously even without intervention.

Urotherapy

Urotherapy is the general term for all forms of non-surgical, non-pharmacological treatment of lower urinary tract malfunction. It has a large number of components including:

1. Education – Providing parents and children with an explanation of how the normal urinary tract functions and the natural history and likely progression of their condition.
2. Voiding education (bladder retraining) – This involves teaching the patient correct voiding posture (mainly applicable to girls). Girls need to sit in a comfortable position with their feet resting on the floor or a step and their hips abducted to open up their perineum/pelvic floor. Voiding needs to occur in a relaxed, unhurried manner. For patients with large post-void residual urines, initiation of *double voiding* may be appropriate (voiding is attempted again a few minutes after completion). Girls need to wipe in a backward direction after micturition. Patients need to be taught to avoid postponing micturition or implementing withholding manoeuvres. A programme of regular, timed voids with an initially short interval that is progressively increased until a normal pattern of five to six voids a day is attained.
3. Lifestyle education – Patients are advised regarding the avoidance/management of constipation and appropriate fluid intake.
4. Support – Regular and intensive follow-up and support is critical to the success of any urotherapeutic strategy.

More aggressive forms of urotherapy are available and are becoming more prevalent [Box 34.1].

Box 34.1 Specialised urotherapies available

- Pelvic floor training by physiotherapist
- Biofeedback, for example, pad and bell bedwetting alarm
- Electrical stimulation (transcutaneous or with implanted electrodes)
- Intermittent catheterisation

Pharmacotherapy

The management of urinary incontinence is a billion dollar industry, with a huge array of medications and appliances available, although many are not licensed for use in children. Bladder emptying is under the control of excitatory parasympathetic fibres that originate in the sacral segments of the spinal cord and act via muscarinic receptors. Currently, five different subtypes of muscarinic receptors (M1 to M5) have been identified. In the bladder, as in most tissues, there is a heterogeneous population of receptors with a predominance of the M2 subtype and a smaller population of M3 receptors (ratio of 3:1). Despite this, it is the M3 receptors that are primarily and directly responsible for bladder contraction. M2 receptor stimulation indirectly facilitates bladder contraction by inhibiting sympathetic-mediated detrusor relaxation. The majority of drug therapy is directed at reducing detrusor overactivity with a number of potential pharmacological targets [Box 34.2].

Box 34.2 New potential drugs for urinary incontinence

- Muscarinic receptor antagonists (oral oxybutynin, tolterodine, trospium chloride, solifenacin, darifenacin)
- Vanilloid receptor antagonists (intravesical, e.g. capsaicin, resiniferatoxin)
- Botulinum toxin (injected into detrusor muscle endoscopically)

KEY POINTS

- Wetting is common and stressful but rarely needs surgery.
- Clinical assessment aims to identify the surgical causes needing referral.
- The mainstay treatment for functional wetting is education, bladder training, laxatives and anticholinergics.

Further reading

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CHAPTER 35
The Child with Haematuria

CASE 1

A recently circumcised baby is noted to have a spot of blood on the tip of the penis after micturition.

Q 1.1 What is the likely problem?

Q 1.2 How is it treated or prevented?

CASE 2

Red water is passed after a 3-year-old falls off a chair onto the side of a toy box. Physical examination reveals a fullness in the upper abdomen on the left.

Q 2.1 What is the differential diagnosis?

Q 2.2 What is your plan of management?

Macroscopic haematuria is very uncommon in children, with a reported incidence of less than 0.2%. It usually causes such alarm that the child is brought early for medical attention. Confirmation of the presence of red blood cells should be obtained, because haemoglobinuria, ingested dyes and plant pigments occasionally can be misleading. Because of the potential causes, frank haematuria should be investigated promptly. Unfortunately, haematuria has often ceased by the time the child is examined, and the decision to investigate the child may be based solely on the observations of the parents or colleagues.

Microscopic haematuria has been detected in 0.5–1.6% of asymptomatic schoolchildren, and its presence should be confirmed on repeat testing. Isolated microscopic haematuria persists beyond 6 months in less than 30% of patients.

History, physical examination and urine microscopy will yield a diagnosis in the majority of patients. The causes are many, and in some, the diagnosis is readily made [Table 35.1]. Urine infection accounts for 50% of cases, perineal irritation (10%), trauma (7%), acute nephritis (4%) with stones, coagulopathy and tumours among the other rare causative factors. Twenty-five percent of renal tumours, potentially the most worrisome cause, present with haematuria, but account for less than 0.7% of all cases of frank haematuria in children. There are usually other signs such as a palpable mass.

Hydronephrosis and other malformations of the upper urinary tract often present with haematuria. It is seldom the sole presenting feature, and the clinical findings, examination of the urine and renal ultrasound or nuclear scan, usually make a diagnosis possible.

History

Frequency, dysuria, abdominal pains and fever point to a urinary tract infection (UTI). Injuries severe enough to damage the kidneys, ureter or lower urinary tract will nearly always present with an obvious history of trauma.

A history of a recent sore throat or skin lesions suggestive of streptococcal infection will be present in most of those with glomerulonephritis.

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Table 35.1 Plan of investigation of a patient **with** haematuria

1. Cause obvious or readily determined	
Renal mass	Overt glomerulonephritis
Bleeding disorders	UTI
Hereditary haematuria	Meatal ulcer
2. Cause apparent on simple radiological investigation	
(a) Plain film	Urinary calculi
(b) Renal ultrasound	Hydronephrosis and hydroureter
	Cystic or malformed kidneys
(c) MCUG	Vesico-ureteric reflux
	Vesical diverticulum
	Urethral polyp
3. Cause obscure without resort to more extensive investigation	
(a) Endoscopy	Urethral valve
	Vesical diverticulum
	Vascular anomalies
(b) Retrograde pyelography	Small benign neoplasms of ureter or pelvis
(c) Renal biopsy	Atypical nephritis
(d) Selective renal arteriography	Vascular anomaly

Pain on micturition and a few drops of blood at the end suggest urethral abnormality or meatal ulcer.

Severe colicky loin pain radiating to the groin and preceding the haematuria is very suggestive of ureteric colic associated with the passage of a renal calculus.

A detailed family history will yield information suggestive of familial causes such as an inherited coagulopathy or an association with familial deafness (Alport disease).

A history of frank haematuria occurring at the end of micturition in adolescent boys is consistent with a diagnosis of posterior urethritis.

Clinical examination

Physical examination is rarely helpful in determining the cause of haematuria. In boys who have been circumcised recently, the first thing to look for is a meatal ulcer. In these boys, appropriate local measures will prevent unnecessary investigation. Occasionally, haematuria is seen in boys with phimosis after attempted forceful retraction of the foreskin. The abdomen should be palpated for the presence of a renal mass and the skin examined to look for a rash suggestive of either systemic lupus erythematosus or Henoch–Schönlein purpura.

Hypertension may point to chronic glomerulonephritis, and a palpable mass in the loin will focus attention on three conditions – hydronephrosis, Wilms tumour and neuroblastoma – which are considered in greater detail in Chapter 25.

Investigations

Microscopy and culture of a mid-stream or catheter specimen of urine is the basis of diagnosis. Granular and cellular casts or persistent proteinuria in addition to *glomerular* red cells will lead to the diagnosis of glomerulonephritis, while pyuria and bacteriuria indicate infection as the cause of bleeding.

Phase contrast microscopy may show crenated and dysmorphic red cells to distinguish atypical focal glomerular lesions from lesions elsewhere in the urinary tract, which tend to give rise to more uniform red cell patterns.

Sterile pyuria accompanied by haematuria raises the possibility of a tuberculous infection.

If MSU and physical examination do not reveal the cause of haematuria, more detailed investigation is warranted including blood tests for U+E, creatinine, pH, albumin, ASOT, C3, C4, immunoglobulins, ANF, anti-DNA antibodies, FBC and clotting factors and urine tests for protein/creatinine ratio and calcium/creatinine ratio.

Plain radiographs

In selected patients, a plain x-ray may reveal a calculus in the urinary tract or a renal soft tissue mass.

Renal ultrasonography

Except in children with meatal ulcer or readily demonstrable glomerulonephritis, a renal ultrasound scan is necessary in every case. Ultrasonography may reveal a urinary calculus, a hydronephrotic kidney or a renal mass that may be either a tumour or an inflammatory condition of the kidney – xanthogranulomatous

pyelonephritis. CT scan and radioisotope studies will help to differentiate the two.

Micturating cystourethrogram

A micturating cystourethrogram (MCUG) will exclude vesical or urethral diverticula or urethral polyps. A plain x-ray prior to the MCUG may show a calculus.

Endoscopy

In some patients with haematuria, all investigations so far are normal. Cystoscopy may be undertaken next, preferably while haematuria is present, although this may be difficult in children, for bleeding is often of short duration. Occasionally, cystoscopy reveals a vesical cause, for example, a small haemangioma or a diverticulum not shown in an MCUG or a urethral cause, for example, urethral valve or posterior urethritis.

Renal biopsy

Most children with *idiopathic* or *essential* haematuria have histological evidence of a focal type of glomerulonephritis in which haematuria is precipitated by physical effort or by an intercurrent infection. Biopsy is not required routinely, but does have a place when haematuria is persistent or severe and all other investigations have not yielded a diagnosis. Children with persistent microscopic haematuria, proteinuria, hypertension or a family history of renal disease may warrant renal biopsy.

Arteriography/MRI

When haematuria is too persistent and severe to be explained by atypical focal glomerulonephritis, and the renal ultrasound, MCUG, cystoscopy and renal biopsy are all normal, renal arteriography or MRI may be needed to exclude the exceptionally rare vascular anomalies of the renal or ureteric vessels. This may also facilitate therapeutic intervention for conditions such as A–V fistulae.

Treatment

Haematuria is a symptom that leads to a variety of diagnoses, and the treatment of these conditions depends on the diagnosis (see related chapters).

KEY POINTS

- Haematuria in children is rare but causes parental alarm.
- Urine microscopy is important to determine cause.
- Renal ultrasonography is essential to exclude serious renal lesions (stone, obstruction, tumour, inflammation).

Further reading

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Pan CG (2006) Evaluation of gross hematuria. *Pediatr Clin North Am* **53(3)**: 401–412.

PART VI
Trauma

CHAPTER 36
Trauma in Childhood

CASE 1

A 10-year-old boy is brought to the emergency department 35 min after being knocked off his bicycle at an intersection. Eye witnesses saw the child bounce off the car onto the road. The boy was unconscious on arrival, cyanosed and shocked.

Q 1.1 What are the priorities in initial assessment and management?

Q 1.2 What is a secondary survey?

Q 1.3 Could there be a simple explanation for the loss of consciousness?

CASE 2

Ian is a 4-year-old who fell over in the backyard (no adult witnesses); he has a jagged puncture wound in the palm of his right hand.

Q 2.1 How would a deep visceral (nerve, tendon, arterial) injury be excluded?

Q 2.2 What management is required to prevent anaerobic infection?

Trauma is the most common cause of death in the paediatric population over 1 year of age. (In children younger than 1 year, congenital abnormalities, prematurity and sudden infant syndrome (SIDS) are the most common causes.)

The *Trimodal distribution* of trauma death was first described in 1983:

1. Immediate (minutes): major vascular or neurological insult
2. Early hours *golden hour* (1–2 h post-injury): 35% of deaths, secondary to brain, abdominal or chest injury
3. Late (days to weeks): 15% of deaths, from brain death, multi-organ failure and/or overwhelming sepsis

The majority of the immediate deaths are not remediable with medical intervention, and prevention of these injuries affords the best opportunity to reduce this mortality. There has been a dramatic reduction in trauma mortality over the last decade, particularly due to improvements in car safety and restraints. Many of the early deaths are potentially preventable with airway, ventilation and circulation intervention. This produced the concept of the *golden hour* where there is the opportunity to prevent early deaths with immediate and appropriate resuscitation, that is, primary survey with A, B, C, D.

It is important to remember that while trauma is the most common cause for deaths in childhood, the majority of paediatric trauma results only in minor injury, but may be associated with long-term morbidity that stretches into adulthood. Here again, injury prevention is the best strategy.

Injury prevention

There are three ways to lessen or prevent childhood injury:

1. Educate parents and children about potential accident situations.
2. Minimise injury in the actual accident, for example, use of car restraints or cycling helmets.
3. Limit injuries sustained after the accident, for example, by first-aid techniques. This requires an effective transport system, which allows early, accurate assessment of injuries by trained personnel and rapid resuscitation prior to transport to an appropriate institution.

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Table 36.1 The causes of accidents in children

Accident	Percentage
Falls	62
Bicycle accidents	12
Traffic accidents as pedestrians	9
Traffic accidents as passenger of motor vehicle	7
Other	10

Mechanism of injury

In paediatrics, it is particularly important to obtain a detailed history of the accident in order to ascertain the likely injuries and particularly if non-accidental injury is to be considered. Often, it is the ambulance personnel who provide valuable information relating to the time and mechanism of the accident.

Adult studies have supported that particular mechanisms may be associated with major trauma:

- Prolonged extraction time (>20 min)
- Motor vehicle accidents at high speed (>60 km/h)
- Ejection from the vehicle

These factors predict that a child may have a major, but as yet undetected, injury, and this must be taken into account during assessment.

The top causes of paediatric minor trauma are illustrated in Table 36.1.

Initial assessment and management

In assessing the injured child, many steps are accomplished simultaneously; for example, while conducting a rapid assessment of a patient's respiratory, circulatory and neurological status, the history and events relating to the injury are obtained.

It is particularly important when considering the paediatric population to nurse the child, if possible, with a primary carer present. Ensure that you explain in simple language to the child what you are doing. Always provide adequate analgesia, and if possible, for an alert child, use distraction therapy and involve the carers, as this will be an extremely frightening and anxiety-provoking situation for the child and family.

Establishing priorities

The Early Management of Severe Trauma (EMST) course provided by the Royal Australasian College of Surgeons has developed a set of priorities that apply to adults and children.

In summary, patient management consists of a rapid primary survey and resuscitation of vital functions, followed by a more detailed secondary assessment, then stabilisation and transfer to definitive care centre where required.

The primary survey: *A, B, Cs*

During the primary survey, life-threatening conditions are identified and treatment instigated immediately:

1. A – Airway maintenance with cervical spine control.
2. B – Breathing and ventilation.
3. C – Circulation with haemorrhage control.
4. D – Disability: neurological status. Do not forget the glucose.

Extensive discussion of paediatric trauma resuscitation is beyond the scope of this book, but guidelines exist in the form of Advanced Paediatric Life Support (APLS), EMST and Advanced Trauma Life Support (ATLS); see also recommended reading.

During the A, B, C, D assessment, once problems are identified, immediate intervention is taken.

Airway

- Check that the child's airway is open while protecting the C-spine.
- Ensure that an infant's neck is in a neutral position and not flexed due to the large occiput.
- A jaw thrust should be used to open the airway.
- Clear the airway with gentle suction under direct vision.
- An oropharyngeal airway may be required to maintain airway if necessary.

Breathing

- Assess the effort, efficacy and effect of breathing.
- Apply oxygen 10 L/min.
- Breathing adjuncts as necessary – bag and mask – endotracheal intubation.
- Monitor saturations/respiratory rate.
- Consider gastric decompression tube to prevent gastric dilatation.

Circulation

- Assess heart rate/blood pressure/capillary refill/mental status.
- Establish two large bore intravenous cannulae rapidly.

- Intraosseous needle insertion if unable to gain access in 60 s.
- If circulation inadequate – 20 mL/kg normal saline bolus.
- Ongoing circulatory support – if third bolus required use O-negative blood.
- Ensure cross-matched sample sent early.
- Ensure platelets and fresh frozen plasma (FFP) and cryoprecipitate available if on-going circulatory support required.
- All fluids should be warmed.
- Arrange early surgical consult.
- Consider hidden sources of bleeding: head, chest, abdominal, pelvis and femur.
- Establish haemorrhage control.

Children will compensate for hypovolaemic shock with tachycardia and vasoconstriction. Hypotension will not occur until more than 30% of circulatory volume is lost. Hypotension is a pre-terminal sign in paediatrics.

Note: Tachycardia may be a response to fear and pain or a normal anxiety response.

Disability

1. Assess mental state using the AVPU or the paediatric Glasgow Coma Scale (GCS).
2. AVPU
   a. A Alert child
   b. V response to voice
   c. P response to pain (equivalent to GCS < 8)
   d. U Unconscious
3. GCS [Table 36.2]. Paediatric-specific charts
   a. Eye opening
   b. Verbal response
   c. Motor response
4. Documentation of pupillary response and size
5. Do not forget the glucose: all children undergoing resuscitation should have the glucose level checked and be provided with normal maintenance fluids of dextrose and saline.

Trauma radiology

- CXR
- C-spine lateral
- Pelvis – if the child is awake, orientated with no other distracting injuries and there is no clinical suspicion of a pelvic fracture, then this x-ray may be omitted.

Monitoring

- Ensure continuous monitoring of all parameters. If any changes occur, the primary survey should be repeated.

Secondary survey

The secondary survey begins after the primary survey (A, B, C, D) has been completed, and the resuscitation phase (management of other life-threatening conditions) has begun. It is a comprehensive examination top to toe and front to back (including log roll), examining all orifices, with full documentation of all injuries and with instigation of first-aid management. Ensure that you explain to the child and carers what you are about to do and keep the child warm during exposure.

In paediatrics, rectal and vaginal examination are not routine and should only be performed once if deemed necessary by the appropriate specialist.

Analgesia

For all children involved in trauma, it is likely to be an extremely anxiety-provoking experience, and they should be nursed:

1. With parents or primary carer in attendance to provide support
2. In a paediatric-focused environment – toys and distractions provided
3. Provided with adequate analgesia
   a. For example, morphine IV 0.1 mg/kg.

Table 36.2 Glasgow Coma Scale (paediatric)

Score	Eye opening	Verbal	Grimace response	Motor response
1	None	None	None	None
2	To pain	Occasional whimper/moan	Mild to pain	Extension to pain
3	To voice	Inappropriate cry	Vigorous to pain	Flexes to pain
4	Spontaneous	Decreased verbal/irritable cry	Less than usual face movement	Withdraws
5		Alert babbles/words as normal	Spontaneous face movement	Localises pain
6				Obeys commands

Superficial soft tissue injuries

The basic management principles involve:
- Providing analgesia
- Cleaning the wound
- Inspection and assessment
- Wound closure
- Dressing

The extent and severity of soft tissue injuries tend to be underestimated. It is important to take into consideration the mechanism of injury, that is, high velocity / penetrating/is there a high risk of contamination?

The initial first-aid management of any wound is irrigation and cleaning – sterile water will suffice.

For the injury itself, a full neurovascular assessment must be performed, assessing movement, perfusion and sensation of all structures distal to the wound. The wound itself must also be examined. In the paediatric population, it may be difficult to fully assess a wound without a general anaesthetic. However, it may be possible with the use of analgesic and sedative adjuncts to inspect a wound and avoid an unnecessary anaesthetic. Such adjuncts include topical adrenaline/lignocaine/amethocaine (ALA) ointment which when applied locally to a wound may be as effective as as lignocaine infiltration, and use of a sedative agent such as inhalation nitrous oxide or intravenous or intramuscular ketamine. These agents must be given in a controlled environment by senior medical staff only.

Always involve senior colleagues or plastic surgeons in the assessment and management of any wound where there is:
- Neurovascular compromise.
- Too large a wound to close under local anaesthetic.
- A large degree of contamination has occurred – gravel or dirt can leave a *tattoo* if not properly removed with scrubbing.
- Involvement of the face or lip across the vermillion border where a good cosmetic result is necessary.

These patients are best managed having their wounds surgically assessed, cleaned and closed under an anaesthetic.

Any wound where there is a risk of retained glass should have a soft tissue x-ray performed.

Wound closure

Superficial wounds can be cleaned and managed with a simple dressing. Simple small lacerations that are not under tension can be closed with either a tissue adhesive (*Dermabond glue*) or adhesive strips (*Steri-Strips*). Other wounds will require surgical closure with sutures.

Specific injuries

The tongue

Despite initially vigorous bleeding and major deformity of the tongue contour, suture of the tongue is rarely needed. Most lacerations should be left to heal and remodel naturally. Infection of intra-oral lacerations is rare. Consider referral to a specialist for review if there is a full-thickness injury or if there is a large flap.

The straddle injury

A slip on to the edge of the bath, bicycle bars or a fence may cause injury to the perineum. Where adequate assessment is not possible in the emergency room, children should be admitted for examination and definitive management under anaesthesia. In females, a straddle injury often causes a tear in the posterior fourchette, often with significant bleeding; minor splits do not require sutures. Injuries through the hymen need careful repair. Where a laceration has penetrated into the rectum, a colostomy for faecal diversion is required.

In boys, a straddle injury may tear the bulbar urethra and cause extravasation of urine into the scrotum and lower abdominal wall. A retrograde urethrogram demonstrates leakage of contrast and the need for catheter drainage of urine or primary urethral repair.

Antibiotics

The most important factor in the prevention of infection is the use of primary first aid and cleaning and, if necessary, surgical debridement of extensive wounds. Antibiotics should be used when wounds are contaminated. However, antibiotics are ineffective in the presence of dead tissue or foreign matter and should never be relied upon to prevent infection in contaminated wounds.

Tetanus

Successful prophylaxis against clostridial infections rests upon the triad of: (i) immunisation, (ii) antibiotics and (iii) adequate surgical cleansing of wounds: removal of dead tissue, foreign material and blood clot.

Active immunisation

Tetanus immunisation should be part of routine childhood immunisation. Primary immunisation of infants is achieved with three doses of triple antigen (diphtheria, tetanus, pertussis) and booster doses at 4 and 15 years.

The new vaccination schedule recommends that the 10-yearly tetanus booster is no longer required up until the age of 50 years, provided that the primary series of three vaccinations plus two boosters has been given.

It is important to be aware of the current schedule of childhood immunisations.

Passive immunisation

Tetanus immunoglobulin is available for the passive protection of individuals who have sustained a tetanus-prone wound and those who have received less than three doses of tetanus vaccination.

Tetanus-prone wounds

Types of wounds likely to favour the growth of tetanus organisms include:

- Compound fractures
- Deep penetrating wounds;
- Wounds containing foreign bodies (especially wood splinters)
- Wounds complicated by pyogenic infections
- Wounds with extensive tissue damage (e.g. contusions or burns)
- Any wound obviously contaminated with soil, dust or horse manure (especially if topical disinfection is delayed more than 4 h)
- Reimplantation of an avulsed tooth (which is also a tetanus-prone event) as minimal washing and cleaning of the tooth is conducted to increase the likelihood of successful reimplantation.

Wounds must be cleaned, disinfected and treated surgically if appropriate.

Child abuse and neglect

Whenever an injured child attends for treatment, clinicians have a duty of care to exclude child abuse [Box 36.1].

Incidence

The incidence of intentional injury is difficult to determine, but it is probably far higher than is generally realised, and has been estimated to be from 0.3 to 3.0% of all injuries in childhood. Infants and children less than 3 years of age are particularly vulnerable.

Box 36.1 Observations suggestive of child abuse

1. Bizarre scars, scabs, weals, circumferential limb abrasions and hemispherical bite marks.
2. Multiple retinal haemorrhages.
3. Periosteal thickening of long bones in unusual areas.
4. Symmetrical burns or scalds in unusual areas.
5. Multiple insect bites and/or infestations, for example, pediculosis.
6. Abnormal behaviour of the child in hospital, for example, withdrawal or stark terror alternating with effusive affection.
7. An abnormal attitude of the parents to the injury. This varies considerably, for example, lack of affect, indifference, panic, guilt or belligerence. Their reactions may conceal an appeal for help.
8. An apparently unrelated developmental abnormality: handicaps, both physical and neurological, that make the child *different* may lead them to becoming objects of abuse.

Clinical features

Certain clinical signs and other features may raise the index of suspicion of abuse and point to the need for a closer examination of the psychosocial climate of the patient and family. The social and psychiatric aspects are often more important than the trauma itself, but lie outside the scope of this book.

Some key features are as follows.

History

- The history is variable but may be quite reasonable and acceptable.
- The mechanism is not in keeping with the child's developmental age.
- The story is inconsistent between the care givers/historians.
- The version may change.
- The child may implicate an adult.

Examination

- The injuries present do not fit the history given of the *accident*.
- Pattern of bruising unlikely accidental on abdominal wall/scapula area.
- There may be linear bruises.
- Torn frenulum.
- Bite marks consistent with adult-sized teeth.
- Retinal haemorrhages.
- Fractures of different ages.
- Rib fractures, especially posterior fractures.

Investigation

Investigation of suspected non-accidental injury should be overseen by a specialist familiar with paediatric forensic medicine. Consider coagulation studies and a full blood count.

It is important to x-ray all bones where a clinical fracture is suspected. Investigations will involve a bone scan, which will identify *hot spots* that may suggest healing fractures. In children less than 1 year old, a skull x-ray should be taken because the bone scan may be unreliable in determining a skull fracture. Often, a full skeletal survey will also be performed.

Diagnosis

This depends to a large degree on an awareness of the possibility that serious injuries in young children may not be accidental. Thus, it is important that where there is some suspicion of non-accidental injury, the child is admitted to the hospital for assessment by the child protection unit.

Management

The full management and treatment of non-accidental injury is beyond the scope of this text. The basic plan of management is to ensure the diagnosis and appropriate treatment of the child's injuries, and this occurs in a place of safety and comfort. Always ensure early involvement of senior colleagues and the local government child protection unit.

Trauma scores and injury severity scores

There are various scoring systems, which exist to allow the quantification of the magnitude of single or multiple injuries, for example, the Champion Trauma Score and Paediatric Trauma Score (PTS). These scores can be a useful predictor of outcome and allow comparison of groups of trauma patients.

Injury Severity Score

The Injury Severity Score (ISS) is based on the extent of tissue injury, which changes little following the initial insult. The ISS indicates increasing severity of injury on a scale from 0 to 75. Severe injury is defined by an ISS greater than 15.

Champion Trauma Score

To quantify the severity of trauma of different types, a numerical value is assigned to five physiological parameters: systolic blood pressure, respiratory rate, respiratory effort, capillary refill and GCS. The sum of the assigned values is the trauma score. On a 0–16 range (where 16 is the least injured) [Table 36.3], a trauma score less than 13 is an indication for transfer to a major trauma centre.

PTS

This quantifies the severity of multiple injuries in children to enable speedy triage and dispatch to an appropriate institution. It measures six different parameters: patient weight, patency of the airways, systolic blood pressure, neurological state, cutaneous wounds and the extent of bony injury. Each parameter is scored −1, 1 or 2, with low scores indicating severe trauma [Table 36.4].

There are three categories of mortality risk: PTS greater than eight should have no mortality; PTS between eight and zero has increasing mortality and PTS less than zero has 100% mortality.

Table 36.3 Champion Trauma Score

Score	Respiratory rate (breaths/min)	Respiratory effort	Systolic BP (mm/Hg)	Capillary return(s)	GCS
5	—	—	—	—	14–15
4	10–24	—	>90	—	11–13
3	25–35	—	70–89	—	8–10
2	>35	—	50–69	<2	5–7
1	<10	Normal	<50	>2	3–4
0	None	Shallow/retraction	No pulse	Nil	—

Table 36.4 Paediatric Trauma Score

Score	Body weight	Airway	Systolic BP (mm/Hg)	CNS	Skeleton	Skin
+2	>20	Normal	>90	Awake	None	None
+1	10–20	Controlled	50–90	Obtunded/LOC	Closed fracture	Minor wound
−1	<10	Unmaintainable	<50	Coma/decerebrate	Open/multiple fracture	Major/penetrating

KEY POINTS

- Systematic response to trauma (primary survey A, B, C, D; resuscitation; secondary survey; definitive treatment) saves lives.
- Airway, breathing and circulation are paramount for survival.
- Secondary survey needs comprehensive top-to-toe and front-to-back examination.
- Penetrating wounds need examination under anaesthesia (LA/GA) to exclude important visceral injuries.
- Surgical debridement (removal of dead tissue, foreign bodies and blood clot) is essential to prevent anaerobic infection.

Further reading

Advanced Life Support Group (2004) *Advanced Paediatric Life Support: The Practical Approach*, 4th Edn. Blackwell publishing, Hoboken, NJ.

ATLS Advanced Trauma Support/EMST Emergency Management of Severe Trauma, 7th Edn. American College of Surgeons.

Lukish JR, Eichelberger MR (2012) Accident victims and their emerging management. In: Coran AG, Adzick NS, Krummel TM, Laberge J-M, Shamberger RC, Caldamone AA (eds) *Pediatric Surgery*, 7th Edn. Elsevier Saunders, Philadelphia, PA, pp. 265–274.

CHAPTER 37
Head Injuries

CASE 1

An anxious mother brings her 6-year-old son to the emergency department after he fell off the fence.

Q 1.1 Which children should be referred to the emergency department after a head injury?

Q 1.2 Which children should have a CT scan after a head injury?

CASE 2

The air ambulance brings a 4-year-old boy who was hit by a car at 70 km/h when he ran on to a busy street. He has been unconscious since the accident (20 min).

Q 2.1 What are the principles of management of a child with a severe head injury?

CASE 3

A 4-year-old child falls out of a tree 2 m to the ground, striking his head. There is a 5 min loss of consciousness. There is a boggy scalp haematoma on the left side and the child is pale, drowsy and confused with a thready, rapid pulse, complaining of headache and has a sluggish, dilated left pupil and right-sided limb weakness.

Q 3.1 As the medical officer who receives the child, what is your diagnosis?

Q 3.2 What urgency do you place on the child?

Q 3.3 What management do you propose?

CASE 4

A 6-month-old baby presents with a history of falling out of a pram. Skull x-rays show a large fracture.

Q 4.1 What would lead you to suspect that a head injury was non-accidental?

CASE 5

A 6-year-old boy falls over at school, hitting his head on concrete. He cannot remember the accident.

Q 5.1 What is concussion?

Traumatic brain injuries (TBI) are the leading cause of morbidity and mortality in children in the developed world. Head injuries account for one of the most frequent causes of emergency department admissions. Most children recover spontaneously but about 10% suffer from sequelae such as changes in behaviour, impaired intellectual performance and post-traumatic epilepsy. Prompt resuscitation to restore adequate circulating volume, oxygenation and blood pressure is necessary to prevent secondary brain injury. Careful and frequent neurological examination along with appropriate neuroimaging is vital if the many reversible aspects of head injury are to be treated and the outcome optimised.

Determinants of injury

The pattern of head injuries in childhood is similar to that in adults, but there are some important differences related to the nature of the injury and the physical characteristics of the child's skull and brain.

The most frequent causes of injury are accidents in the home including falls, playground injuries, sporting accidents, pedestrian- or bicycle-related injuries, motor vehicle accidents and non-accidental injuries (NAI). NAI resulting from child abuse occur most frequently in the 0–4-year age group and is the most common cause of severe head injury at this age.

Jones' Clinical Paediatric Surgery, Seventh Edition. Edited by John M. Hutson, Michael O'Brien, Spencer W. Beasley, Warwick J. Teague and Sebastian K. King.

The vault of the skull in children is thin and pliable and capable of much greater deformation than in adults. The increased elasticity permits more energy to be absorbed by the skull, which dampens the acceleration or deceleration of the head after impact and reduces the concussive effects. Children can tolerate blows of considerable severity without immediate loss of consciousness. The increased skull elasticity also causes local indentation resulting in Pond's depressed fracture. A child's skull can sustain considerable distortion without fracture, but when the limit is reached, the fracture that results is often extensive and frequently of the *bursting* type. The skull sutures do not close until the fourth year, so that marked diastasis of the sutures may follow bursting injuries.

At the point of maximal distortion, a fracture line may lacerate the underlying dura, allowing brain tissue to herniate through the fracture. In the subsequent days and weeks, the pulsations of the underlying brain may cause this brain herniation through the skull defect to progressively enlarge, resulting in further widening of the fracture. This will produce what is called a growing fracture of childhood, which requires operative repair.

Children more frequently present with diffuse injury and cerebral swelling with resultant intracranial hypertension compared to adults. Children as a group survive TBI more often than adults and also tend to have better functional outcomes. This is felt to be due to the plasticity of the paediatric brain.

Mechanisms of brain injury

Sudden acceleration/deceleration of the head causes shearing of axons, which constitute white matter tracts. This results in diffuse parenchymal and vascular injuries. The clinical manifestation of this insult can vary from a transient loss of consciousness, called concussion, to profound and persistent neurological deficits or death resulting from diffuse axonal injury (DAI).

Direct impact against the calvarium and brain causes *coup* injuries such as parenchymal petechiae, contusions, intraparenchymal haematomas and extra-axial haematomas such as extradural and subdural haematomas [Fig. 37.1]. Sometimes the viscoelastic brain rebounds within its rigid confines resulting in injuries on the side opposite to the impact, i.e. *contrecoup* injuries..

Penetrating injuries to the brain can produce cerebral lacerations and damage to the cerebral vessels.

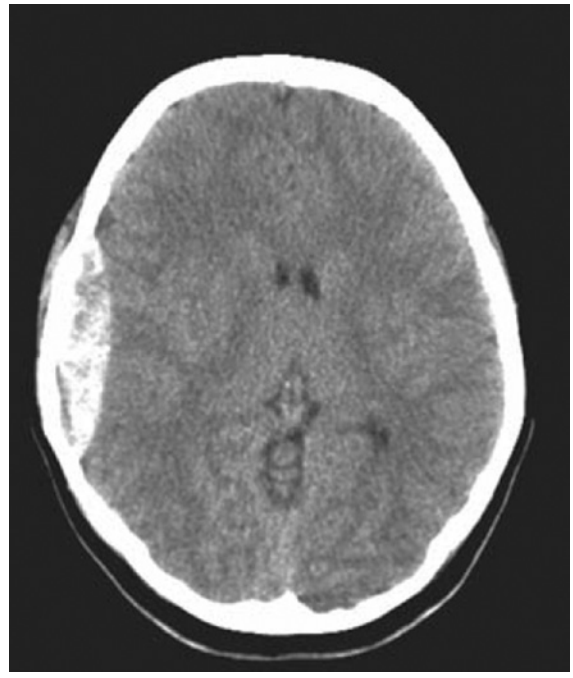

Figure 37.1 Extradural haematoma.

Pathophysiology of TBI

All of the aforementioned brain injuries constitute primary brain injury. Subsequent to the primary insult, a cascade of biochemical and vascular events occurs, collectively called secondary brain injury. This results in further compromise of the cerebral blood flow, producing cerebral ischaemia and cytotoxic damage. These occur due to cerebral oedema, disturbed cerebral autoregulation, release of excito-toxic neurotransmitters and calcium entry into cells causing cell death. To date, no effective pharmacological intervention has been successful in preventing or mitigating the effects of secondary injury. However, secondary insults such as raised intracranial pressure from hydrocephalus, haematomas, hyperthermia, dyselectrolytaemia, hyperglycaemia, hypotension, hypoxia and seizures should be anticipated and treated promptly and aggressively to improve the outcome of head injuries.

General management of head injury

The National Institute of Clinical Excellence (NICE) in the United Kingdom has set out guidelines to identify those patients who need referral to the

Box 37.1 Indications for referral to hospital after head injury in children

1. GCS < 15 at any time since the head injury
2. Any loss of consciousness as a result of injury
3. Any focal neurological deficit
4. Any seizure since the injury
5. Persistent headache since injury
6. Any suspicion of a skull fracture or penetrating head injury
7. CSF leak from the ear or nose
8. Multiple injuries in association with head injury
9. Suspicion of NAI or poor social circumstances
10. Amnesia for events before or after the injury

Box 37.2 Principles of management of a child with a severe head injury

1. Airway, breathing and circulation must be secured.
2. Adequate resuscitation.
3. Accurate neurological assessment.
4. Investigation with CT scan.
5. Prevention of secondary brain injury:
   - **a** Correction of hypoxia and hypercapnoea with intubation
   - **b** Initial hyperventilation to reduce intracranial pressure
   - **c** Replacement of blood volume to maintain adequate cerebral perfusion pressure
   - **d** Urgent surgery to remove mass lesions
   - **e** Anticonvulsants, anti-pyrexial treatment and antibiotics for compound injury
   - **f** Intracranial pressure monitoring and continued ventilation and sedation in an intensive care unit
   - **g** Vigorous control of intracranial pressure with sedation, paralysis, CSF drainage and decompressive craniectomy

emergency department in a hospital [Box 37.1.]. The principles of management of a child with a head injury include the fundamentals of trauma resuscitation with restoration of adequate airway, oxygenation and circulation being of the utmost importance [Box 37.2.]. Following this, the establishment of the level of consciousness according to the Glasgow Coma Scale [Table 37.1] and identification of focal neurological deficits are carried out. In certain cases, a CT scan of the head is required [Box.37.3.]. Some types of head injury raise suspicion of child abuse [Box 37.4.].

Table 37.1 Glasgow Coma Scale (paediatric)

Score	Eye opening	Verbal response or grimace response		Motor response
1	None	None	None	None
2	To pain	Occasional whimper/ moan	Mild to pain	Extends to pain
3	To voice	Inappropriate cry	Vigorous to pain	Flexes to pain
4	Spontaneous	Decreased verbal/ irritable cry	Less than usual face movement	Withdraws
5		Alert babbles/ words as normal	Spontaneous face movement	Localises pain
6				Obeys commands

Note: The paediatric version of the Glasgow Coma Scale is scored between 3 and 15 with 3 being the worst and 15 being the best. It is composed of three parameters: best eye response, best verbal or grimace response and best motor response, as described.

Box 37.3 Indications for CT scan after head injury

- GCS < 13 at any point since injury
- Suspected open or depressed skull fracture
- Any sign of basal skull fracture (haemotympanum, panda eyes, Battle's sign, CSF otorrhoea)
- Post-traumatic seizure
- Focal neurological deficit
- Persistent vomiting
- Amnesia greater than 5 min of events before impact (assessment of amnesia not possible in children <5 years old)

Box 37.4 Indications of suspected non-accidental head injury in a child

Suspicious and inconsistent story
History of previous injuries
Multiple cutaneous bruises of different ages
Bilateral retinal haemorrhages
Acute subdural haemorrhage and brain swelling on CT scan
Chronic subdural haemorrhage
Bilateral skull fractures
Old fractures of long bones and ribs
Evidence of malnourishment
Subdued behaviour
Fear

Characteristic injuries

Skull fractures

Linear skull fracture

Linear fracture results from low-energy blunt trauma over a wide surface area of the skull. It runs through the entire thickness of the bone and, by itself, is of little significance except when it runs through a vascular channel, venous sinus groove, or a suture. In these situations, it may cause epidural haematoma, venous sinus thrombosis and occlusion, and sutural diastasis, respectively.

Depressed fractures

Depressed skull fractures result from a high-energy direct blow to a small surface area of the skull with a blunt object such as a golf club [Fig. 37.2]. A depressed fracture may be open or closed. Open fractures have either an overlying skin laceration or the fracture runs through the paranasal sinuses and the middle ear structures, resulting in communication between the external environment and the cranial cavity. Open fractures may be clean or contaminated. The indications for surgery are cosmesis, underlying intracranial haematoma requiring evacuation, suspicion of a dural breach with an open fracture or a dirty contaminated wound.

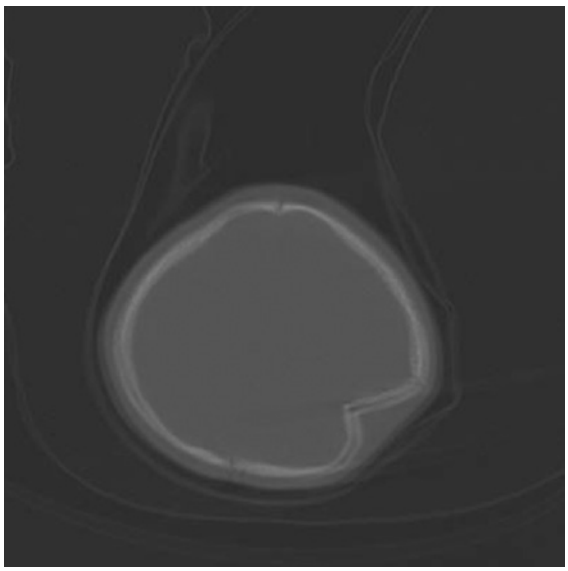

Figure 37.2 Depressed fracture of the skull.

Basilar skull fracture

A basilar fracture is a linear fracture at the base of the skull, commonly in the temporal bone.

Temporal bone fracture is encountered in 75% of all skull base fractures. The two main subtypes of temporal fractures are longitudinal and transverse classified according to the orientation of the fracture line in relation to the direction of the petrous temporal bone. Longitudinal fracture is the more common of the two subtypes (70–90%) and usually causes a haemotympanum with conductive deafness. Transverse fractures are more commonly implicated in seventh nerve palsies and sensorineural deafness.

Intracranial haemorrhage

Several types of intracranial haemorrhages can occur, including the following.

Extradural haematoma

Extradural (or Epidural) haematomas are more common in teenagers than in young children. These occur due to a direct blow to the skull with associated laceration of the dural arteries or veins (often by fractured bones) and sometimes by diploic veins in the skull's marrow. More often, a tear in the middle meningeal artery causes this type of haematoma. When haematoma occurs from laceration of an artery, blood collection can cause rapid neurologic deterioration.

Subdural haematoma

Acute subdural haematoma tends to occur in patients with injuries to the cortical veins or pial arteries in severe head injury usually following acceleration/deceleration. The mortality rate may be high due to an associated severe brain injury.

Intracerebral haemorrhages

Intracerebral haemorrhages occur within the cerebral parenchyma secondary to lacerations or contusions of the brain. Injury to the larger, deeper cerebral vessels results in extensive cortical contusion.

Intraventricular haemorrhage

Intraventricular haemorrhage tends to occur in the presence of very severe TBI and is, therefore, associated with an unfavourable prognosis.

Subarachnoid haemorrhage

Subarachnoid haemorrhage may occur in cases of head injury caused by lacerations of the superficial microvessels in the subarachnoid space. If not associated with another brain pathology, this type of haemorrhage could be benign. Traumatic subarachnoid haemorrhage also may lead to communicating hydrocephalus if blood products obstruct the arachnoid villi.

Coup and *contrecoup* contusions

A contusion is a bruise in the brain. A combination of vascular and tissue damage leads to cerebral contusion.

Coup contusions occur at the area of direct impact to the skull. *Contrecoup* contusions are similar to coup contusions but are located opposite the site of direct impact. This occurs when the force impacting the head is not only great enough to cause a contusion at the site of impact but also is able to move the brain and cause it to strike into the opposite side of the skull, which causes the additional contusion. The amount of energy dissipated at the site of direct impact determines whether the ensuing contusion is of the *coup* or *contrecoup* type. Most of the energy of impact from a small hard object tends to dissipate at the impact site, leading to a coup contusion. On the contrary, impact from a larger object causes less injury at the impact site since energy is dissipated at the beginning or end of the head motion, leading to a *contrecoup* contusion.

Concussion

A concussion is the most common type of TBI.

A concussion is caused when the brain receives trauma from an impact or a sudden momentum. Concussion is caused by deformity of the deep structures of the brain, leading to widespread neurologic dysfunction that can result in impaired consciousness or coma. A person may or may not experience a brief loss of consciousness. A person may remain conscious but feel *dazed* or *punch drunk*.

Concussion is considered a mild form of DAI and is thought to be due to a transient arrest of axoplasmic function.

DAI

DAI is a frequent result of traumatic high-speed acceleration–deceleration injuries and a frequent cause of persistent vegetative state in patients. DAI is widespread damage to the white matter of the brain and is produced by a shearing injury due to rotational forces. The injury to tissue is greatest in areas where the density difference is greatest. For this reason, approximately two-thirds of DAI lesions occur at the grey–white matter junction such as parasagittal frontal lobe and basal ganglia. The remainder occur where the brain impacts against a rigid structure such as the tentorium or falx cerebri, producing lesions in the corpus callosum or the cerebral peduncles. Characteristic CT findings in the acute setting are small petechial haemorrhages that are located at the grey–white matter junction, within the corpus callosum and in the brainstem. The result of shearing forces histologically is trauma to the axons. This focal alteration of the axoplasmic membrane results in impairment of axonal transport. Neuropathologic findings in patients with DAI include axoplasmic swelling, retraction balls, Wallerian degeneration and glial scars. This widespread disturbance in the white matter can produce a variety of functional impairments depending on where the shearing occurred in the brain. Clinically, these injuries may manifest as temporary or permanent brain damage, coma or death.

NAI

NAI is defined as any form of physical injury where the injury is not consistent with the account of its occurrence or where there is definite knowledge or a reasonable suspicion that the injury was inflicted by any person having custody, charge or care of a child.

Non-accidental head injury or shaken baby syndrome is a deliberate violent shaking of a young child. The forceful whiplash-like motion causes the brain to be injured due to acceleration–deceleration forces. Blood vessels between the brain and skull rupture and bleed. The accumulation of blood causes the brain tissue to compress while the shearing injury causes the brain to swell. This can cause seizures, lifelong disability, coma and death.

Suspected NAI in a child always requires urgent referral. Consider admission and/or referral to the local social services department according to local policy. A high index of suspicion is required to first diagnose and then manage these injuries appropriately.

Indicators of possible NAI are detailed in Box 37.4.

Cranial birth injuries

Injuries to the infant that result from mechanical forces (i.e. compression, traction) during the birth process are categorised as birth injuries. Most birth traumas are self-limiting and have a favourable outcome.

Most birth injuries of the skull and brain come from excessive or rapid deformation of the skull as it passes through the birth canal or are due to compression by obstetric forceps. Distortion may produce surface lacerations of the brain or tearing of superficial vessels, the large veins, or dural sinuses.

Depressed fractures of the skull during birth may be due to *natural causes*, but most result from incorrect placement of obstetric forceps.

Extra-axial injuries

Cephalhaematoma

Cephalhaematoma is the most common birth injury and seen most frequently after vacuum extraction. It is a subperiosteal collection of blood secondary to rupture of blood vessels between the skull and the periosteum; suture lines delineate its extent. Most commonly parietal, cephalhaematoma may occasionally be observed over the occipital bone. The extent of haemorrhage may be severe enough to cause anaemia and hypotension, although this is uncommon. Usually, management solely consists of observation. Transfusion for anaemia, hypovolaemia or both is necessary if blood accumulation is significant. Aspiration is not required for resolution and is likely to increase the risk of infection. The resolving haematoma predisposes to hyperbilirubinaemia. Resolution occurs over weeks, occasionally with residual calcification.

Subgaleal haematoma

Subgaleal haematoma is bleeding in the potential space between the skull periosteum and the scalp galea aponeurosis. This space extends from the orbital ridges to the nape of the neck and laterally to the ears. Most cases result from vacuum extraction. The diagnosis is generally a clinical one, with a fluctuant boggy mass developing over the scalp. The swelling may obscure the fontanelle and cross suture lines (distinguishing it from cephalhaematoma). The swelling develops and spreads across the whole calvarium; its growth is insidious, and subgaleal haematoma may not be recognised for hours. Patients with subgaleal haematoma may present with haemorrhagic shock or develop significant hyperbilirubinaemia. In the absence of shock or intracranial injury, the long-term prognosis is generally good.

Management consists of vigilant observation over days to detect progression and provide therapy for such problems as shock and anaemia. Transfusion and phototherapy may be necessary.

Caput succedaneum

Caput succedaneum is a serosanguineous, subcutaneous, extraperiosteal fluid collection with poorly defined margins; it is caused by the pressure of the presenting part against the dilating cervix. Caput succedaneum extends across the midline and over suture lines and is associated with head moulding. Caput succedaneum does not usually cause complications and usually resolves over the first few days. Management consists of observation only.

Skull fractures

Fractures are rare and more commonly depressed than linear. The parietal and frontal bones are most frequently involved. Management is expectant and one needs to be aware of the possibility of a growing skull fracture [Fig. 37.3].

Intracranial haematoma

Bleeding into the subdural space or brain is uncommon in the newborn and the clinical picture varies according to the rate of bleeding, site and volume. The infant may show signs of obtundation, with a tense fontanelle and anaemia. There will also be signs of local pressure, for example, lack of movement or weakness of the contralateral limbs.

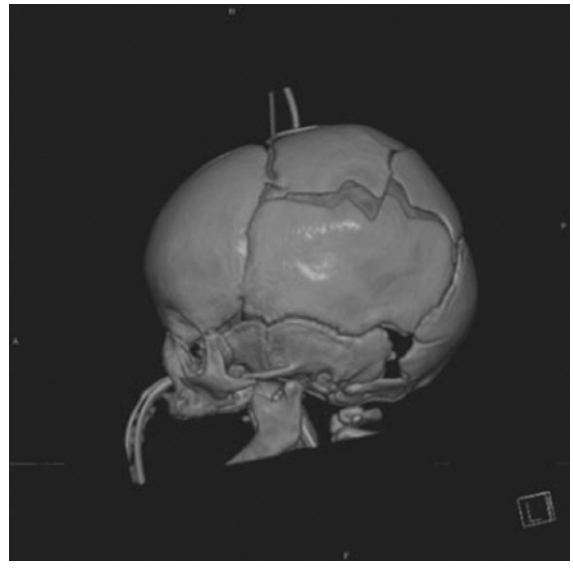

Figure 37.3 Growing skull fracture.

Subdural haematoma

Subdural haematoma is the most common intracranial abnormality following birth trauma. It arises from torn bridging cortical veins usually occurring during malpresentations or forceps delivery. The collection develops slowly, giving the brain and skull time to adjust. The clinical signs are an enlarging and sometimes asymmetrical head, delay in reaching the normal milestones, irritability, developmental delay, failure to thrive and occasionally convulsions.

The diagnosis is confirmed by ultrasound or CT scan or by needle aspiration of the fontanelle. Repeated aspiration may be sufficient, but persistent reaccumulation will require either open drainage via small burr holes, or internal drainage by means of a shunt (subdural to peritoneal).

Extradural haematoma

Extradural haematoma is rare following birth trauma. It is usually associated with a skull fracture. It may present with an enlarging head, obtundation and seizures or occasionally with focal signs. It is diagnosed with an ultrasound or MRI and management is conservative. If the mass effect worsens, then needle aspiration rather than a craniotomy is preferable in this age group.

Sequelae of head injuries in children

Neurological

Although children show a surprising capacity for recovery after head injuries, they may suffer permanent disabilities as a result of the more severe injuries. Brain injury in the younger child may disrupt the development of intellectual and physical milestones and result in reduced intellectual performance and psychopathological sequelae. Brain injuries are known to produce attention deficit disorders, problems with memory and learning and deficits of psychomotor, linguistic and executive skills. Cognitive deficits particularly affecting memory disrupt learning ability and produce enormous educational difficulties. There is good correlation between injury severity as assessed by Glasgow Coma Scale and post-traumatic amnesia, and neurobehavioural outcome.

Focal deficits appropriate to the injured part of the brain may become permanent. The cranial nerves may also be permanently damaged.

Post-traumatic epilepsy is common after birth injuries, especially those that affect the temporal lobe. In older children, it occurs after compound depressed fractures of the vault associated with a laceration of the cortex, or following intracerebral haematomas.

CSF leak

CSF rhinorrhoea may result from a fracture of the base of the skull involving the frontal, ethmoid or sphenoid sinuses, whereas a fracture of the temporal bone may cause CSF otorrhoea and/or rhinorrhoea. Both types are initially treated conservatively. In about 70% cases of CSF rhinorrhoea and almost all cases of CSF otorrhoea, the leak stops spontaneously. If the CSF leak persists, the communication with the exterior through a mucosal space is a potential source of meningitis, and this may occur months or years later. A skull defect should be suspected when meningitis occurs after a head injury. The treatment of a CSF leak persisting for more than 10 days after a head injury is operative repair.

KEY POINTS

- Principles of management of head injury include ABCs of trauma resuscitation, establishing level of consciousness (GCS) and identifying any focal deficits.
- CT scan is a regular part of initial assessment in any child with more than minor head injury.
- Non-accidental head injury is common in infants and toddlers and needs to be considered in any child with a suspicious history, where the injuries are unusual for the child's age or are out of proportion for the *accident*.

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CHAPTER 38
Abdominal and Thoracic Trauma

CASE 1

Jayden is a 7-year-old who fell off his bike, landing on his handlebar. He was winded initially, but recovered within a few minutes. Several hours later, he became pale and developed abdominal and shoulder pain.

Q 1.1 What is the likely diagnosis?

Q 1.2 Outline the management.

CASE 2

A 3-year-old girl stepped on to the road and was hit by a car travelling at moderate speed. On arrival in the emergency department, her femur appears bent and vital signs suggest hypovolaemic shock and central cyanosis.

Q 2.1 What is your approach to the management?

Intra-abdominal trauma

Abdominal trauma is second only to head injury as a cause of serious injury in children, and the third most common cause of trauma-related death. In Australia, most abdominal injuries in children result from blunt trauma, but knife and gun crime are important causes for penetrating abdominal trauma in other countries. The most common organs injured in blunt abdominal trauma are the spleen, liver and kidneys. A careful history of the trauma helps predict the likely injuries and so guides the clinical assessment. For example, a bicycle handlebar injury to the left upper quadrant of the abdomen or to the lower chest is often associated with splenic trauma [Fig. 38.1].

It is noteworthy that children are particularly prone to acute gastric distension following trauma, due to gastroparesis as well as air swallowing with anxious crying. Acute gastric distension may mimic the symptoms and signs of hypovolaemia and abdominal injury, thus confusing the clinical picture including the need for computerised tomography (CT) scanning. Also, unless the stomach is decompressed, these children are at risk of vomiting with aspiration (Mendelson syndrome). For these and other important reasons, stomach decompression with a gastric tube is an essential adjunct to abdominal assessment in trauma (the same may be said for urinary catheterisation to decompress the bladder). If a base of skull fracture is suspected, an orogastric, not nasogastric, tube is required.

When the spleen and liver are injured, intraperitoneal bleeding occurs (haemoperitoneum). The injured kidney bleeds into the retroperitoneal space, and if the urinary tract is disrupted, urine may also extravasate into the retroperitoneum.

Haemoperitoneum

Haemoperitoneum presents with signs of peritoneal irritation such as tenderness (often widespread) and a variable degree of reflex muscular rigidity (guarding). There may be accompanying signs of hypovolaemia, such as tachycardia, cool peripheries and poor capillary return. Abdominal distension results both from the volume of blood in the peritoneal cavity and the swallowed air and ileus, which develops rapidly.

Variations in posture and the time elapsed since injury result in wide variations in the signs of haemoperitoneum and the site of maximal tenderness. When the spleen is ruptured, the signs are usually maximal in the left upper quadrant with referred pain to the left shoulder tip (a sign of phrenic nerve irritation). If the bleeding has occurred for some time or has been rapid, it is not unusual for signs to be more widespread, even occurring on the right side.

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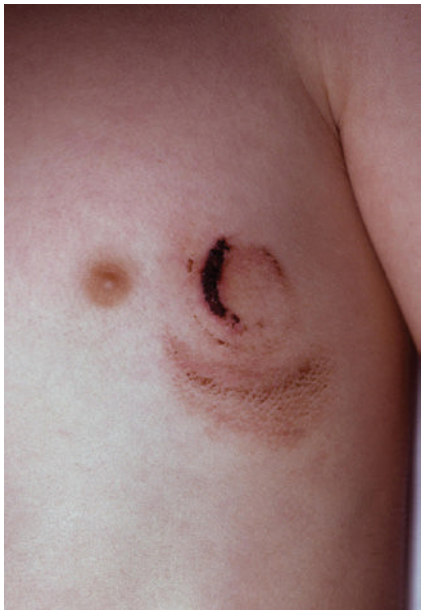

Figure 38.1 Handlebar injury with bruising over the lower ribs, which are so elastic that the underlying spleen has been torn without the ribs themselves being broken.

Focused Abdominal Sonography for Trauma (FAST) scanning in the emergency department is useful to demonstrate blood or free fluid in the peritoneum of adults. However, in children, sensitivity of FAST, that is, reliability of a negative result, is poor. Instead, a CT scan with intravenous contrast is preferred in those patients in whom imaging is indicated. As discussed later, the need for surgical exploration relies more on clinical signs and progression than on imaging.

Radiological investigation

Most children with minor blunt trauma to the abdomen require no specific imaging. CT scanning with intravenous contrast remains the gold standard where serious abdominal injury is suspected (e.g. ruptured spleen [Fig. 38.2], liver [Fig. 38.3] or kidney) or in an unconscious patient with multiple injuries where abdominal injury needs to be excluded. It should be noted, however, that no patient with major trauma should be taken to the imaging department for investigation unless he or she is haemodynamically stable. If stability cannot be achieved with resuscitation, it may be more appropriate for the patient to proceed directly to urgent surgery. As discussed earlier, FAST or other ultrasound scanning may have a select role in some trauma patients' management, but has not yet replaced need for either CT or surgical exploration.

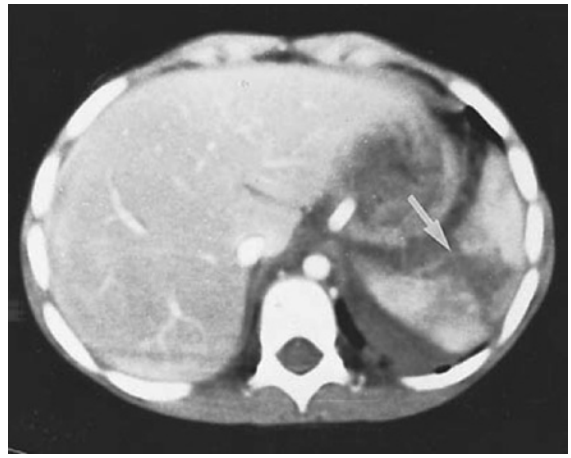

Figure 38.2 CT scan of the upper abdomen showing a ruptured spleen (arrow).

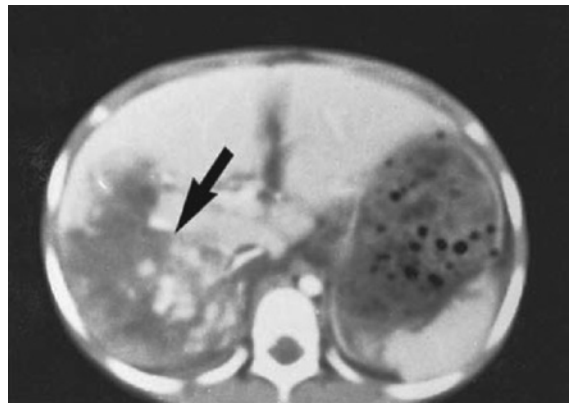

Figure 38.3 CT scan of the upper abdomen showing a ruptured liver (arrow).

Recognition of hypovolaemic shock

In children, hypovolaemic shock is characterised by tachycardia, poor peripheral perfusion and cool extremities. Importantly, unlike in adults, blood pressure is not a good guide to hypovolaemia in children. It will remain stable in the face of significant blood loss (up to 2/3 of total blood volume!), and then decompensation occurs quickly. Falling blood pressure therefore represents significant blood loss, and resuscitation should ideally be instituted before this occurs.

Two common errors confound paediatric blood pressure assessment. First, failure to recognise that normal blood pressure varies with age, for example, systolic pressure greater than 65 mmHg in infants and greater than 70 mmHg in preschool children. Second, accurate measurement of blood pressure in children also requires a correctly sized cuff, which, as a general rule, ought to be at least two-thirds of the length of the upper arm.

Non-operative management

Non-operative management is successful in more than 90% of solid visceral injuries in children, with injury-targeted bed rest, active observation and regular surgical reassessment. Such care is best delivered in a paediatric trauma centre with access to paediatric surgery and intensive care, should these be required.

Indications for operative intervention

Although failure of non-operative management is uncommon, it is important to recognise those patients in whom operative management is required. The three main indications for surgical intervention for blunt abdominal trauma are:

1. Persistently unstable circulation and/or hypotension despite appropriate resuscitation. Initial resuscitation is with two boluses of 20 mL/kg of normal saline. If the child remains unstable following this, a surgeon needs to be present. A further 60–80 mL/kg of blood may then be given as 20 mL/kg boluses. If the child fails to respond to this resuscitation, then operative intervention to control the bleeding should be strongly considered.
2. A proven or suspected intestinal perforation on clinical or radiological examination.
3. Severe, concomitant head injury in which an unstable circulation cannot be tolerated and where significant or ongoing intraperitoneal bleeding is suspected.

In the event of splenic trauma and ongoing haemorrhage, the options for surgical intervention include interventional radiology to embolise the involved vessel(s), and laparotomy. At laparotomy, some injuries will be amenable to oversewing the tear (*splenorrhaphy*) or performing partial splenectomy. This allows preservation of the spleen, thereby avoiding the significant risk of overwhelming post-splenectomy infection that may occur when the entire spleen is removed. If the spleen has to be removed completely, the child should be given the appropriate vaccinations and commenced on long-term prophylactic antibiotics (see also Chapter 26).

Diagnostic peritoneal lavage is not indicated in children because it is not specific, and free blood in the perineal cavity *per se* is not an indication for operative intervention.

Haematuria

In all suspected intra-abdominal injuries, the presence or absence of macroscopic blood in the urine must be established. The presence of macroscopic haematuria is strongly predictive of serious intra-abdominal injury (e.g. splenic and self-evidently renal trauma) and may form part of the basis for further investigation with a CT scan. The absence of haematuria virtually excludes a significant urinary tract injury except in the rare instance of a transected ureter. This rare false negative will be readily identified by CT scan showing extravasation in the flank or loin, which should prompt further investigation or operative exploration.

Urethral injury should be suspected in pelvic fractures and straddle injuries with:

1. Frank blood appearing at the external urethral meatus
2. Inability to void spontaneously (particularly if the bladder is palpable)
3. Urinary extravasation into the perineum

In these patients, a urethral catheter must not be inserted blindly into the urethra, as this may convert a partial tear into a complete urethral disruption. A retrograde urethrogram must be obtained first to delineate the injury. Depending on the type of urethral injury, either primary surgical repair of the urethra or temporary urinary diversion will be indicated, for example, suprapubic catheterisation.

Macroscopic haematuria following relatively minor trauma suggests a predisposing factor, such as hydronephrosis or Wilms tumour. All children with macroscopic haematuria, even after trivial injury, require a renal ultrasound scan to exclude these underlying lesions (Chapter 35).

Extraperitoneal extravasation

The signs of extravasation of blood or urine usually develop more slowly and less dramatically than those of intraperitoneal haemorrhage; they are also more consistently localised and less variable.

Renal injuries

Because the kidney is involved frequently in blunt abdominal trauma, tenderness and muscular rigidity should be sought in the loin. However, a peri-renal haematoma may

cause not only localised tenderness but also sometimes a mass that is palpable through the anterior abdominal wall.

When haematuria accompanies tenderness and rigidity in the loin, a CT scan with intravenous contrast should be performed. This will distinguish between a contusion, which may be managed conservatively, and a rupture of the kidney with urine extravasation, which may require operative exploration. Ultrasonography enables the renal injury to be monitored subsequently.

Early exploration (within 3 days of injury) rather than non-operative management may be required in some major renal injuries. In select cases of renal tract trauma without associated haemodynamic instability, cystoscopic retrograde placement of a ureteric stent is an appropriate alternative to open exploration. Delay in recognition and management of injuries to the renal pelvis or parenchyma, which results in urine extravasation, is followed by severe inflammatory changes that may prejudice attempts at conservation and later repair.

Bladder injuries

In extraperitoneal rupture of the bladder or membranous urethra, there may be signs of urinary extravasation into the perineum, scrotum and suprapubic region.

When haematuria or urethral bleeding accompany signs of intraperitoneal or extraperitoneal haemorrhage in the lower abdomen, a cystogram will establish whether the bladder is intact. However, if blood, as opposed to blood-stained urine, is seen at the urethral meatus, the catheter should be passed only after a carefully performed urethrogram has confirmed that the urethra is intact.

Ill-defined intra-abdominal injuries

Apart from those patients with intraperitoneal haemorrhage, extraperitoneal extravasation or haematuria, there is a difficult group with ill-defined symptoms and signs that may persist for several days after the injury.

Many probably have minor contusions of the abdominal wall, the intestine or its mesentery. Non-operative management is usually justified in these cases. Sometimes, lap-belt deceleration injuries may cause severe trauma to the bowel wall when it is crushed against the vertebral column. These injuries may only become apparent after several days when the bowel perforates and peritonitis suddenly develops. The same type of trauma may also cause a periduodenal haematoma and pancreatic injuries, some of which may require operative intervention. Severe lap-belt injuries with abdominal wall bruising±lumbar bruise (hyperextension tear of lumbar ligaments/vertebral fracture known as a Chance fracture) need immediate specialist referral.

Thoracic trauma

Major thoracic injuries account for less than 10% of trauma admissions in children. The vast majority result from blunt thoracic trauma, for example, motor vehicle accidents. Despite the relative rarity of major thoracic trauma, it is the second most frequent cause of paediatric trauma-related death. Major thoracic trauma often occurs in the setting of a multi-trauma, and the likelihood of death increases from 5% in isolated cases up to 40% for children with accompanying head and abdominal injuries.

The child's chest wall is very compliant, the chest wall structures being compressed rather than fractured due to the force applied. Therefore, most of the energy due to the trauma is transferred through to the intrathoracic structures, often without any external evidence of injury and without fractured ribs. For this reason, pulmonary contusions and direct intrapulmonary haemorrhage are more common in children, and rib fractures or flail segments seldom seen. Thankfully, tension pneumothorax and haemopneumothorax are also uncommon but potentially lethal unless recognised. Tension pneumothorax requires no further investigation, rather relief by emergency needle thoracocentesis prior to definitive intercostal chest drain insertion.

Very rarely diaphragmatic rupture may result from forceful crushing of the abdomen, contributing to respiratory embarrassment. In this setting, chest auscultation findings may be confusing, but the routinely performed chest x-ray should provide clarity, especially if a gastric tube has been inserted [Fig. 38.4]. Injury to the great vessels is also rare in children and reflects a lack of pre-existing vascular disease and fewer high-speed injuries.

Investigation

The mainstay in investigation of thoracic trauma is a chest x-ray, itself a standard component of best practice early management of the typically multiply injured child. Pulmonary contusion, pneumothorax, haemothorax, rib

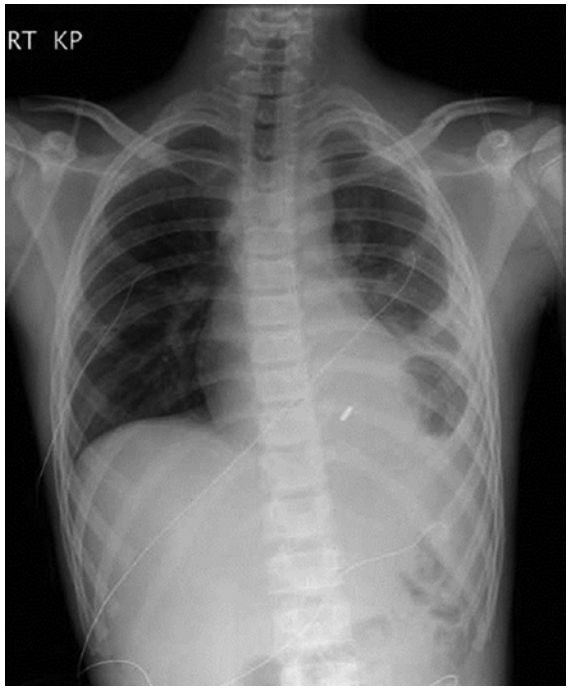

Figure 38.4 X-ray showing the abdominal viscera in the left hemithorax, following crush injury and rupture of diaphragm.

fractures when present, as well as the rare diaphragmatic rupture may each be demonstrated on chest x-ray. Don't forget that more than one injury may be evident. Also, trauma patients are typically supine when imaged, and care must be taken to recognise the more subtle appearance of intrapleural blood or fluid when the patient is lying flat.

Some patients will warrant further investigation with a CT scan, but not to the detriment of timely management of a known significant injury, for example, sizeable traumatic pneumothorax or haemothorax. ECG should also be considered in select patients.

Management

All paediatric major trauma patients should receive high-flow oxygen during primary survey, and this remains an integral component in the care of any thoracic trauma.

Pulmonary contusion in isolation requires supplemental oxygen, good analgesia, and close observation. Pneumothorax, with or without haemothorax, may accompany pulmonary contusion, and should be relieved by an intercostal chest drain with an underwater seal when of significant volume. Blood loss from haemothorax may be large, and so adequate prior intravenous access and resuscitation is mandatory. Occasionally, a traumatic pneumothorax is visible on CT scan but not chest x-ray. In this instance, chest drainage is reserved for those children requiring positive pressure ventilation and/or air transport.

Although multiple rib fractures are uncommon due to chest wall elasticity in children, a flail chest may occur, and when large enough to cause respiratory embarrassment, internal splinting by positive pressure ventilation is preferable. These and other major thoracic injuries are best managed initially in the high dependency or intensive care ward, where close observation and ventilatory support may be provided as required.

Penetrating thoracic trauma

On the rare occasion of penetrating thoracic trauma, injuries to the mediastinal structures and lungs should be assumed until proven otherwise. Particular attention should be given to the possibility of cardiac tamponade, with the need for emergency pericardiocentesis, as well as the potential for combined thoraco-abdominal injury.

KEY POINTS

- Bicycle handlebar injury may tear the spleen, and less commonly, the liver, kidney and pancreas.
- Ruptured spleen causes significant haemoperitoneum, but bleeding nearly always stops without surgery.
- Non-operative management is appropriate for most solid visceral injuries.
- Haematuria after trauma needs investigation to document the status of the kidneys and urinary tract.
- Blunt chest trauma leads to pulmonary contusions without rib fractures.

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CHAPTER 39
Foreign Bodies

CASE 1

A mother notices her 12-month-old child put a small safety pin in his mouth and apparently swallow it. An x-ray shows it to be in the stomach. He is asymptomatic.

Q 1.1 Are any further investigations required?

Q 1.2 Is an operation to remove the pin indicated?

CASE 2

A barefooted 8-year-old girl has trodden on a needle that has broken, part of it remaining in the sole of her foot. A small puncture wound is seen but the needle cannot be felt. It hurts her to walk on it.

Q 2.1 How is it best removed?

Young children's instinctive exploration and fascination with their environment may result in a wide range of foreign bodies lodging in a variety of places. Most are found in the alimentary tract; others may enter the aural or nasal cavities, the bronchial tree, or be accidentally driven into the soft tissues. The location of foreign bodies tends to vary with the child's age. Infants are more likely to ingest things into the alimentary canal, while older children are more likely to suffer puncture wounds.

Swallowed foreign bodies

In infants, oral exploration of the environment may lead to accidental swallowing of a variety of objects. Older siblings may feed the *new baby* inappropriate hardware. Accidental swallowing may be precipitated by a fall or a slap on the back. At any age, a bone hidden in food (e.g. fish bone) may be swallowed. Ingestion of a foreign body occurs most commonly between 6 months and 4 years.

The vast majority of swallowed foreign bodies pass through the gastrointestinal tract without causing any problem. With rare exceptions, objects that are first located below the diaphragm will pass naturally without hazard to the child.

The most common site (70%) of lodgement is in the oesophagus at the level of the cricopharyngeus muscle (the area between the clavicles on the x-ray). The other two common sites of lodgement are mid-oesophagus and at the gastro-oesophageal junction.

Objects

The size of coins (the objects that are swallowed most often) determines whether they are likely to become stuck in the oesophagus. Safety pins also may stick in the oesophagus, but if they enter the stomach, they will almost always be excreted without difficulty, even if they are open. Broken plastic toys are more dangerous, because they may be jagged or angular and their radiolucency may lead to a delay in diagnosis. Hair clips (*kirby grips*) pass easily as far as the duodenojejunal flexure but may be too long and rigid to negotiate this flexure in children less than 7 years of age and require endoscopy or laparotomy for their removal. In children more than 6 or 7 years of age, observation for up to 1 week is justified, although impaction at the duodenojejunal flexure should not be allowed to continue for more than 10–12 days.

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Button or *disc* batteries used in electronic toys, cameras and mobile phones may be particularly hazardous when swallowed. Their small size normally ensures their passage into the stomach, and the great majority will pass through the gastrointestinal tract uneventfully. However, if the battery is seen in the oesophagus on x-ray, referral should be made for urgent surgical endoscopic removal. Holdup in the oesophagus is hazardous because erosion and perforation may occur rapidly and may be life-threatening. The cause is thought to be related to discharge of current rather than breakdown of the battery. Even non-functioning batteries may discharge enough to erode through the oesophagus.

Beyond the stomach, the battery will invariably pass without issue. There has been concern in the past about leakage from the battery with the possibility of absorption of battery contents. However, with modern batteries, this is not a concern. An x-ray after a week to show it has passed may be useful to reassure parents but, generally, no intervention is required even if it takes longer for the button to pass, as long as the child remains asymptomatic.

The incidence of swallowed magnetic objects is rising due to increased prevalence of magnets within toys. Children who have swallowed *multiple* magnetic objects are at particular risk due to the opposition of multiple magnets through adjacent loops of bowel, which may injure, entrap or twist bowel loops. Untreated, this may cause a range of complications, including ulceration, perforation, fistulation, adhesions, intussusception, peritonitis, volvulus, sepsis and even death. Therefore, inpatient observation with a low threshold for removal of the magnets by endoscopic, laparoscopic or open approaches is indicated.

Prevention

It is important to ensure infants and young children have appropriate levels of supervision and that toys are age appropriate to prevent accidental ingestion. Access to *button* or *disc* batteries should be restricted in the same way as poisons in the household.

Clinical features

Often there are no symptoms; indeed, it is likely that innumerable small foreign bodies are swallowed and passed by children uneventfully and unnoticed. In some cases, an attack of gagging, coughing or retching is reported by parents, which may then be followed by the child being reluctant to eat or drink. This may progress to oesophageal obstruction with symptoms of excessive drooling and dysphagia. Sometimes children are witnessed with something in their mouth prior to the onset of the symptoms.

If the accident has not been reported or observed, there may be a delay in the diagnosis, and the first symptoms may be due to complications, for example, progressive dysphagia or dyspnoea caused by pressure of the swollen oesophagus on the trachea. In severe cases, the child may develop pain, swelling in the neck and fever, suggesting mediastinitis. Rarely, a pneumothorax or pleuritic pain may be the first indication of perforation of the oesophagus by a foreign body. Even less frequently, the object may erode directly into the trachea or aorta with resultant fistula formtation. An acquired tracheo-oesophageal fistula may manifest as a persistent cough or recurrent pneumonia. In the case of an aorto-oesophageal fistula, a child may have catastrophic gastrointestinal haemorrhage.

Investigations

If the object is radio-opaque or unknown, x-rays of the head, neck, thorax and abdomen are required, because a radio-opaque object may be located anywhere from the base of the skull to the pelvic floor. However, the site of the foreign body may also be indicated by the type of symptoms.

An x-ray will distinguish tracheal from oesophageal lodgement, for the maximum dimension of the trachea is in the sagittal plane, and that of the oesophagus is in the coronal plane [Fig. 39.1].

Some centres are now using metal detectors in emergency departments to detect metallic objects more accurately.

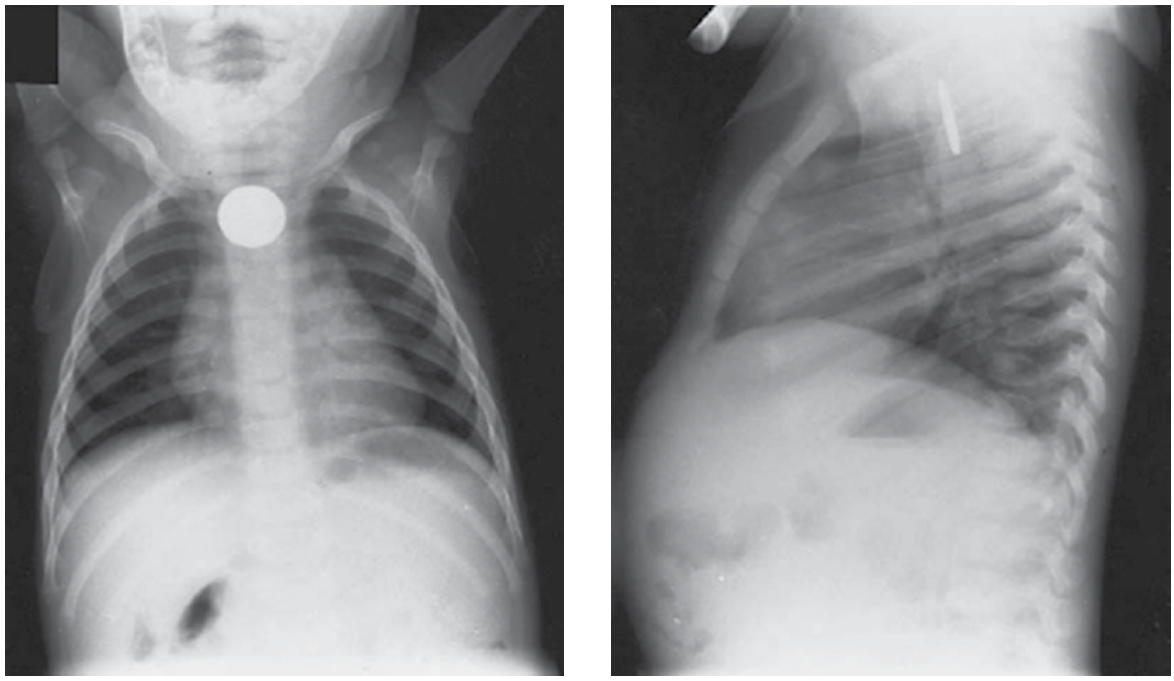
Radiolucent foreign bodies are more difficult to detect radiologically. In cases where ingestion of a radiolucent foreign body is suspected and the patient is symptomatic, consultation with an ENT or general surgeon will be required to decide the next appropriate investigation, for example, a contrast swallow.

Management

The vast majority of ingested foreign bodies do not need removal.

Located in oesophagus

Endoscopic removal is required for all objects impacted in the upper oesophagus. In the lower oesophagus, foreign bodies may pass with time, and some people



(a) (b)

Figure 39.1 The orientation of a foreign body (e.g. coin) will distinguish tracheal from oesophageal lodgement. Here, a chest x-ray shows a coin in the coronal plane characteristic of the oesophagus **(a)** AP view and **(b)** lateral view.

have advocated the use of medications to relax the lower oesophageal sphincter, such as glucagon. Allowing time for the foreign body to pass may be appropriate where the impacted foreign body is not hazardous, for example, a food bolus obstruction. A button battery, however, should never be allowed to remain in the oesophagus, even if it is in the lower oesophagus.

Beyond the oesophagus

Foreign bodies first located beyond the oesophagus have a good chance of being excreted without incident [Fig. 39.2]. Blunt objects small enough to enter the stomach will almost always be passed; the patient should return only if abdominal pain or vomiting occurs.

Further x-rays are seldom indicated, but typically show that the object has been passed, often unrecognised. On rare occasions, an ingested blunt foreign body will become stuck and so remains visible on repeated imaging weeks after ingestion. Such patients should be referred for paediatric surgical opinion as to whether or not removal of the suspected impacted foreign body is required.

Where the history and radiology indicates ingestion of *multiple* objects capable of magnetic attraction, proactive and prompt care is needed to prevent the complications outlined previously. Even complicated cases may lack acute abdominal signs, which may falsely embolden unsuspecting healthcare professionals to defer necessary intervention.

Even most sharp objects pass uneventfully and should be managed non-operatively initially, although arrest and failure to progress through the bowel may raise concerns of impending impaction, ulceration and perforation. If a sharp foreign body is being observed, parents should return to hospital if the child develops an unexplained fever or sudden abdominal pain.

A bezoar is a conglomeration of hair (trichobezoar) or vegetable material (phytobezoar), which may be swallowed by children as a habit. The mass forms in the stomach or proximal bowel and causes pain, vomiting or anorexia, malnutrition or unexplained anaemia. Less commonly, obstruction or perforation occurs. When x-rays indicate a mass of this kind, surgical removal is required, e.g. laparotomy and enterotomy.

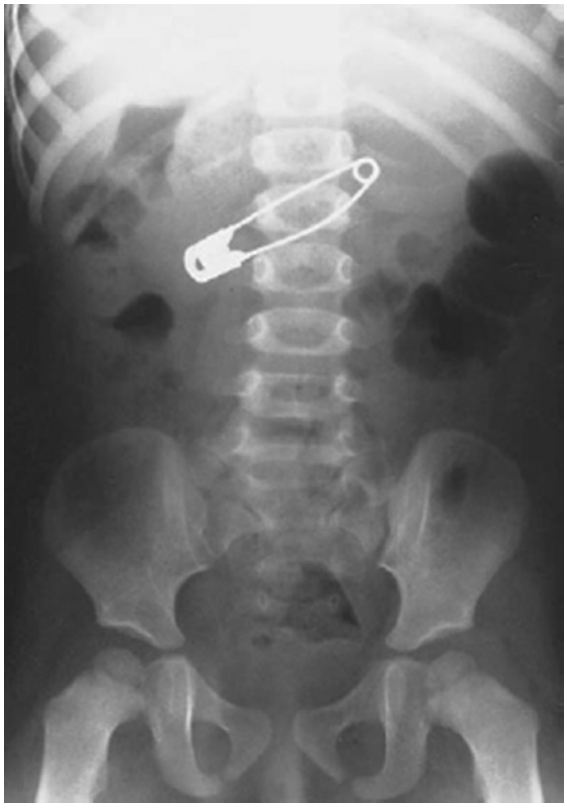


Figure 39.2 Abdominal x-ray showing a safety pin in the stomach. This should pass spontaneously.

Foreign bodies in the trachea and bronchi

Sudden onset of coughing, spluttering and gagging with a residual wheeze, are suggestive of an inhaled foreign body. The exact clinical picture varies with the size of the object, the site of lodgement and the time elapsed since the object was inhaled.

Large objects in the larynx or trachea produce obstruction, which may be complete if they impact in the narrow glottis. Inspiratory stridor and respiratory distress, with indrawing of the supraclavicular, substernal or intercostal areas, indicate that the object is in the larynx or subglottic area.

Foreign bodies in the trachea or bronchus cause a wheeze and may produce clinical and radiological evidence of an over-expanded (emphysematous) lung because of a ball-valve effect of the foreign body. Objects impacted more distally may present with symptoms of chronic chest infection from lobar or segmental consolidation.

The object may be demonstrated on x-rays if it is radio-opaque. X-rays taken in expiration and inspiration may show air trapping and assist in localising the presence and size of small radiolucent objects in the lungs, for example, a peanut. If the object is radiolucent, segmental pulmonary collapse and lobar emphysema may also be seen on the x-ray.

In infants, respiratory symptoms such as acute dyspnoea, stridor or cough may occur when the object is impacted in the oesophagus. This is due to the dilated oesophagus pushing on the adjacent trachea. The orientation of the object on x-ray may identify the site of lodgement because the maximum diameter of the trachea is in the anteroposterior plane, compared with the transverse plane in the oesophagus [Fig. 39.1].

Management

The foreign body is best removed at endoscopy under general anaesthesia. This may involve either a rigid or flexible bronchoscope. The foreign body may not always be easily seen because of mucosal swelling or infection. Organic material may also be very difficult to grasp. Mucosal abrasions and pulmonary changes caused by the foreign body, its complications or manipulations during removal are indications for a course of antibiotics. The earlier the diagnosis and treatment, the less the likelihood of residual pulmonary or mucosal damage.

Special consideration

Great care needs to be taken with peanuts, which is why they should not be given to young children. Not only may they obstruct the bronchus, but the oil content may produce a lipoid pneumonia, which is known to develop rapidly.

Foreign bodies in the ear, nose and pharynx

In the ear

Unless there is a definite history, children with a foreign body in the ear typically present with a blood-stained discharge, irritation or deafness. Cooperation of the child

is essential for the removal. Most may be removed with a good viewing instrument (auroscope) and illumination with a fine probe. However, if the first attempts fail, it is appropriate to refer to the ENT surgeon for removal under sedation or anaesthetic.

In the nose

The child, who is usually between 2 and 4 years of age, presents with nasal irritation and obstruction, or a purulent discharge. The object may often be seen on direct examination or with an auroscope; very occasionally, an x-ray may be useful. Prior to any procedure, the nose should be treated with lignocaine spray and phenylephrine for anaesthesia and to reduce local inflammation. In very small children, the foreign body sometimes may be blown out. This is achieved by asking the mother/carer to blow into the child's mouth and obstructing the other nostril. In older children, suction may be an effective method for removing objects, especially smooth round objects such as beads or buttons. Other methods involve the use of fine hooks or even a fine Foley catheter. If the child is not cooperative or the first attempts are unsuccessful, it is appropriate to involve the ENT surgeon or to use some form of sedation, such as ketamine, or occasionally an anaesthetic.

In the pharynx

The most common object to get lodged in the tonsil, the piriform fossa or the back of the tongue is a fish bone. Subjective localisation is poor, and pain may be constant or felt only during swallowing.

After the fauces and pharynx have been sprayed with a local anaesthetic, the usual sites are examined with the aid of a spatula or a laryngeal mirror and proper illumination, and the object is removed with forceps.

Foreign bodies in the urinary tract

These are rare but most frequently involve the bladder, when objects may be introduced into the urethra. Sometimes, a fragment of a ureteric catheter may break off and remain after surgery to the urinary tract. Any of these objects may cause infection, haematuria or pain, and may act as a nidus for the formation of a calculus.

Foreign bodies in the urinary or genital tract are removed cystoscopically, although occasionally an open procedure on the bladder may be required.

Other foreign bodies

X-ray may help identify foreign bodies deep in the soft tissue if radio-opaque.

In the hand

Apart from superficial splinters, small objects may be driven into the palm, such as wood, plant spikes or metal. If it is unrecognised, a painless swelling may develop on the palm, with the foreign body sitting inside a fibrous capsule. Sometimes, however, infection may occur around the foreign body leading to an abscess, which ultimately may drain, expressing the foreign body in the process. If the foreign body penetrates deep into the palm, it may go on to cause a deep web-space infection, with the potential for pus to track along the tendon sheaths.

Where a foreign body deep in the palm is suspected, exploration and removal under general anaesthetic is usually required. More superficial foreign bodies may sometimes be removed under sedation in the emergency department. However, prolonged attempts should be avoided, as this is distressing for the child and parents, and foreign bodies may sometimes be very difficult to identify in the absence of ideal surgical conditions.

In the lower limb

Nails, pins or needles driven into the foot or knee may be shown radiographically. When they are not visible through the skin, they are most easily and quickly removed under general anaesthesia and guided by an image intensifier.

KEY POINTS

- Most foreign bodies that reach the stomach pass spontaneously.
- Sudden coughing with a residual wheeze is suggestive of an inhaled foreign body.
- Foreign bodies in the palm and sole should be removed under anaesthesia.

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CHAPTER 40
The Ingestion of Corrosives

CASE 1

A 3-year-old girl drank from an unlabelled soft drink bottle that she found in her father's garage. She immediately developed severe mouth, throat and epigastric pain and had difficulty swallowing. The fluid ingested was identified as caustic soda.

Q 1.1 What should be your initial first aid?
Q 1.2 What investigation should be performed in hospital, and what major complication of this injury do you wish to prevent?

In children, swallowing corrosive fluids or solids nearly always is accidental, and the exploring toddler aged between 1 and 3 years is most often the victim. Symptoms of caustic ingestion include cervical and epigastric pain, irritability, excessive drooling, dysphagia and respiratory distress. However, about 20% present with no symptoms; in some, this is despite significant oesophageal injury.

Prevention

The most effective way to prevent such accidents is to keep all chemicals used in the home and garden out of reach of small children and in their proper containers. They should not be stored under the kitchen sink or in unlabelled containers. The introduction of safety caps for all bottles containing chemical materials has significantly reduced the incidence of corrosive ingestion in children.

Pathology

The oesophagus is the most common organ seriously injured by corrosive ingestion. Extensive or circumferential oesophageal burns may lead to severe strictures, which cause dysphagia within weeks of injury.

Burns of the buccal mucosa, soft palate or tongue suggest that the oesophagus has been damaged as well.

Mucosal injury and oedema of the larynx occurs in 15%, and may be life-threatening, requiring intubation or tracheostomy.

First aid

1. If ingestion has just occurred, wash off any excess corrosive material from the lips and skin, using plenty of water.
2. Immediately dilute any corrosive in the mouth, oesophagus or stomach by giving cold water or milk to drink. Do not attempt to use an *antidote* acid or alkali because the corrosive may have been identified incorrectly and the chemical antidotes themselves may cause damage. Do not promote vomiting, as the vomitus may include the corrosive material, leading to further insult to the oesophagus and oral cavity.
3. If ingestion of corrosive was not witnessed or confirmed by an adult, always assume it has occurred if the lips or mouth are blistered or if the toddler is drooling excessively and unable to swallow saliva.
4. Where the nature or composition of the corrosive is uncertain, consult the Poisons Information Service by

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telephone. Only induce vomiting with ipecac syrup if directed by a poisons service.

5 Send a sample of the corrosive agent with the child when transferring to hospital if identification has not been made with certainty.

6 All children should be sent to the nearest paediatric surgical centre as soon as possible after ingestion.

Definitive Management

Upper gastrointestinal endoscopy at about 24 h is performed to assess the severity and distribution of corrosive injury to the oesophagus and/or stomach. This will determine the need for prophylactic treatment to reduce the likelihood of subsequent oesophageal stricture formation. Where there is no damage to the oesophageal mucosa on endoscopy, no treatment is required.

Patchy oedema of the intact oesophageal mucosa is regarded as the minimal degree of burn and is not likely to cause a stricture. No treatment is required, and the patient may leave hospital as soon as normal feeding is re-established. A white mucosal slough or circumferential ulceration is more serious and may lead to subsequent stricturing (incidence varies from 5 to 50%). In these patients, antibiotics may be given to limit the effects of secondary infection. Steroids may diminish the extent of fibrosis, although their value in the early post-ingestion period is uncertain. Oesophagoscopy and dilatation of the oesophagus is performed under general anaesthesia in children with extensive and deep oesophageal burns. This procedure is usually commenced 2 weeks after the initial injury.

If the mucosa has healed and there is no evidence of narrowing, treatment is discontinued. If there are abnormal findings at 2 weeks, treatment is continued for a further 6 weeks. Where there is worsening dysphagia and the stricture cannot be dilated effectively by bouginage or repeated radial dilatation under fluoroscopic control over many months, placement of an oesophageal stent may be advantageous. If the stent fails to provide an adequate oesophageal calibre, then segmental resection and anastomosis may be required. Occasionally, an extensive resection and replacement of the oesophagus is inescapable; but in the long term, the best oesophagus is the patient's own, and repeated dilatations are justified to avoid an extensive oesophagectomy.

KEY POINTS

- Oesophageal injury may be minimised by rapid dilution of corrosive by drinking cold water or milk.
- Ulcers on lips or inside mouth suggest oesophageal burn – confirmed by drooling saliva.
- Upper gastrointestinal endoscopy within 24 h determines the severity of the burn and the treatment required.

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CHAPTER 41
Burns

CASE 1

At 5 P.M., an 18-month-old boy is rushed into the emergency department of a country hospital after tipping hot tea on to himself 10 min earlier. The tea was just boiled and no milk had been added. The area of scald estimated with a Lund–Browder chart is 15%.

Q 1.1 What first aid and primary management should be initiated in the country hospital?

Q 1.2 Does this child warrant referral to the regional specialist Burns Centre?

CASE 2

A 6-month-old infant brought to your clinic with red, weeping lower legs and feet after being scalded by a hot bath.

Q 2.1 What is the likely mechanism of injury?

CASE 3

Will and Ali are brought to the emergency department after they poured petrol on a campfire. Their faces are blackened and their hair and eyebrows are singed.

Q 3.1 What injury is likely to pose the most immediate threat to health and life?

A burn may be caused by extremes of temperature, friction, radiation (e.g. sunburn), electricity or chemical agents. In children, more than half of all burns are scald injuries, caused by exposure to hot liquids. Other common mechanisms of burn injury in children are contact with hot surfaces (~20%) and flame burns (~15%). Severity of a burn injury depends on the size and depth of burn, and its anatomical site. Depth of a burn is proportional to both the strength of the injurious agent (e.g. temperature, concentration of acid) and the duration of time the agent remains in contact with the tissues. Burn wound depth may be classified according to the skin layers involved:

1 **Superficial**: epidermal burns, superficial dermal burns, mid-dermal burns
2 **Deep**: deep dermal burns, full-thickness burns

Most burns are heterogenous with varying depths in the same burn. Epidermal burns are not included when estimating the percentage of total body surface area burned.

First aid and early primary management of even severe burns can be effectively delivered in the pre-hospital and non-specialist hospital setting. The ongoing care of a child with severe burns requires the services of a multidisciplinary specialist Burns Unit and may extend over many years. Alongside the sometimes more obvious need to manage a child's burn wound and associated systemic response to the burn insult, the psychological and social needs of a child (and family) with burns are significant. Therefore, health and allied health professionals (e.g. physiotherapy, occupational therapy, play therapy, dietician, psychologist, social

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worker) must work together as a team in the short- and long-term management of paediatric burns.

Prevention

Most burns in children

1. Occur at home (~90%), mostly in the kitchen or bathroom
2. Are 'self-inflicted' despite being in the care or supervision of an adult
3. Occur due to exposure to heat (temperatures >50 °C will produce necrosis)

The classic and commonest example of these observations is the toddler at home in the kitchen under parental supervision, who pulls a newly prepared hot beverage onto themselves, sustaining a scald. In older children, flame burns are more common than scalds and are most often the result of playing with matches and flammable fluids.

Prevention of burns rests on three main approaches:

1. *Education*: of both children and adults, concerning potential dangers, and the need for continual vigilance. Government-sponsored prevention programmes have stressed to the public that burns in children can be prevented by (i) supervising them, (ii) separating them from the hazard, (iii) reducing the hazard or access to it and (iv) removing the hazard. It is hoped that such messages will achieve a further reduction in the incidence of burn injuries in children: in the last 25 years, the frequency of paediatric burns has declined by more than 50% in the state of Victoria, Australia.
2. *Design*: for example, improvements in clothes, heating appliances, guards on stoves, temperature regulators in hot water systems.
3. *Legislation*: for example, government (legal) control of fireworks, nightwear materials and design regulations.

Treatment

Early, competent assessment and treatment of a burn is essential for good short- and long-term outcomes. The key phases of burns treatment are summarised in Table 41.1, with a multidisciplinary team approach being central to the success of each phase.

Table 41.1 Management of burns in children

1. First aid
2. Emergency management and assessment
3. Fluid resuscitation
4. Referral and transfer
5. Burn wound care
6. Early excision of devitalised tissue with grafting
7. Prevention and control of infection
8. Adequate nutrition
9. Psychological support and rehabilitation
10. Minimisation of scars and contractures (e.g. pressure garments)
11. Reconstructive surgery, if necessary

First aid

First aid aims to limit the extent and severity of the burn. The child must be removed quickly from the source of injury to stop the burning process, and the wound cooled. For example, any flames are extinguished, and clothing removed immediately as they may contain or concentrate latent heat. The burned area, or if necessary the whole body, should be cooled for 20 min, ideally with cool running water (15 °C). Ice or iced water is dangerous and should never be used to cool a burn wound. Ice may paradoxically increase the depth and extent of a burn by causing local ischaemia, as well increasing the likelihood of hypothermia.

Emergency management and assessment

Emergency management adheres to the priorities and principles of trauma patient management with primary (ABCDE) and secondary surveys, together with adjuncts such as intravenous fluid resuscitation, analgesia, as well as select insertion of gastric tubes and urethral catheters.

A central and unique aspect of the emergency management of burns is the accurate assessment of the extent and depth(s) of burned tissue. In addition, the distribution of the burn or specific anatomical sites that have particular implications for management should be noted, for example, any circumferential burns and/or burns to the face, hands, feet or perineum.

Extent

The 'Rule of Nines' (modified for use in children) is a widely available tool for rapid and sufficiently accurate estimation of the percentage of total body surface area

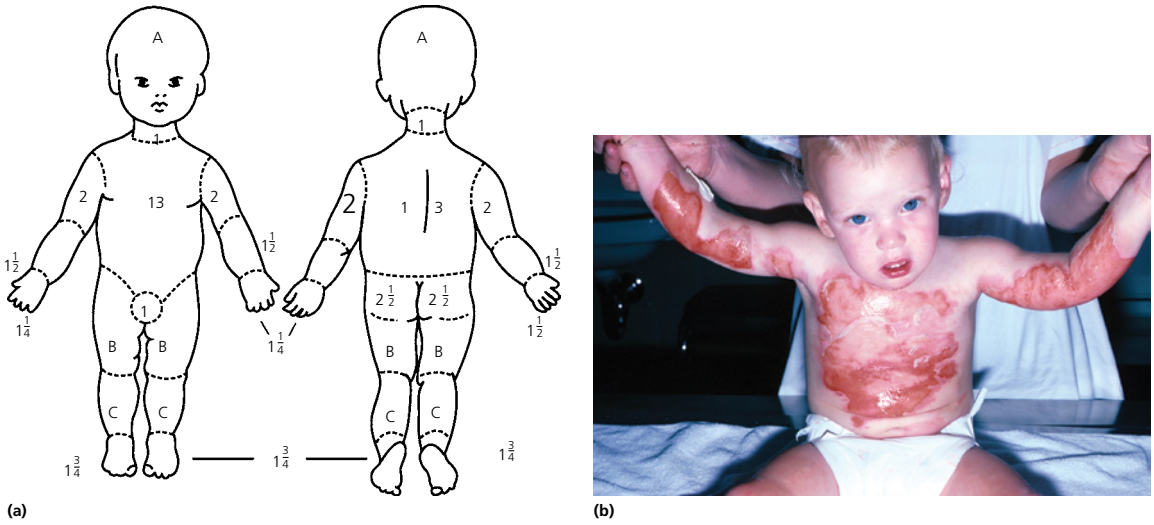

Figure 41.1 Figure 41.1 (a) Method of estimating the extent of burned surfaces, allowing for differences according to age (*Source*: Adapted from Lund and Browder (1944)). A is 10% at birth, decreasing to 4% at 15 years. B and C are 2.5% in babies, increasing to 5% and 3.5%, respectively, in adolescents. (b) A typical distribution of a scald in a toddler.

involved. Alternatively, a Lund–Browder figure chart [Fig. 41.1] allows a more accurate and age-specific estimation of area burned, as well as denotes burns of differing depth. Unless this estimate is charted carefully, the area of the burn may be overestimated leading to excessive and even unnecessary fluid resuscitation

Depth

A full-thickness skin burn (white, charred and painless) is usually obvious early. The exact depth of dermal burns may not be evident for 3 or more days, highlighting the need for reassessment of the burn over this period. A dermal burn which appears superficial at the time of injury (erythematous, blistering and painful white slough) may evolve to become deep (mottled, red and painless) – sometimes as a result of suboptimal management.

Fluid resuscitation

Major burns (>10% of total body surface area in children, >20% in adults) are associated with hypovolaemic shock. Causes of 'burn shock' include primary injury to the tissues and capillaries, as well as secondary to inflammatory mediators released due to the burn. The systemic effects of inflammatory mediators give rise to oedema, hypovolaemia and reduced cardiac output due to whole body changes in tissue and capillary permeability and depressed cardiac function. The pathophysiology of burns shock changes with time, particularly during the first 24 h after injury.

Resuscitation fluids. Fluid resuscitation is indicated for all children with burns greater than 10% of total body surface area, most commonly intravenous crystalloid Hartmann's solution. The volume and rate of fluid resuscitation is guided by the modified Parkland formula, which calculates the volume of fluid required in the first 24 h *from the time of injury*:

$$3-4\ \text{mL} \times \text{weight(kg)} \times \%\,\text{total body surface area burned}$$

Half this volume is to be given over the first 8 h *from the time of injury*, and the half over the remaining 16 h.

The modified Parkland formula guides but does not blindly dictate the ongoing rate of fluid administration. Rather, the rate of resuscitation fluids is titrated to achieve an optimal urine output of 0.5–1 mL/kg/h, obtained accurately with a urinary catheter and recorded. If the urine output is less than this, blood volume (and so perfusion) is likely reduced. Urine output exceeding 1 mL/kg/h may indicate excessive resuscitation, with resultant worsening of both oedema and, in turn, the burn wound.

Maintenance fluids. In addition to fluid resuscitation, children will also require maintenance fluids calculated in the usual fashion according to weight. The rate and type of these maintenance fluids is independent of resuscitation formulae. Initially, maintenance fluid therapy may be intravenous using 0.45% NaCl with 5% dextrose, but enteral maintenance fluids are favoured as soon as intake is tolerated. If oral intake is not possible or inadequate, early consideration should be given to insertion of a nasogastric (or other enteral feeding) tube.

Blood transfusion rarely is required in the first few days after the burn, but may be necessary to treat anaemia secondary to staged surgical excisions and skin grafting.

Referral and transfer

Once emergency primary care has been provided, children may require transfer to the regional specialist Burns Unit. The threshold for transfer is lower in children than adults. The *Australian and New Zealand Burn Association* criteria for referral for transfer to the Burns Unit include

1. Greater than 10% of the total body surface has been burned
2. Greater than 5% of the total body surface has sustained a full-thickness burn
3. Burns with an associated (or suspected) inhalation injury
4. Chemical or electrical burns
5. Burns of special areas: face, hands, buttocks and genitalia
6. Burns of the very young
7. Burns in which non-accidental injury is suspected

In major burns, transport is arranged to a hospital and intravenous fluids commenced in accordance with the principles of fluid resuscitation outlined earlier. Analgesia is provided by intravenous morphine given in titrated doses, and tetanus prophylaxis is ensured. The child is covered with a blanket or similar to prevent hypothermia.

Early burn wound care

Epidermal burns (e.g. sunburn without blisters) may need little more wound care than moisturising and simple analgesia. These burns will heal without scarring.

The aim of early wound care for dermal and full-thickness burns is to clean and cover the burn wound. Effective procedural analgesia and Play Specialist involvement is an essential adjunct to this care. After cleaning the wound with an antiseptic, loose or devitalised blistered skin is removed. The burn should then be covered, which significantly improves wound pain of dermal burns (full-thickness burns are insensate), and elevated. If transfer to the regional Burns Unit is imminent and the wound will be re-assessed on arrival, simple coverage by plastic film (e.g. Clingwrap®) is an ideal temporary dressing.

Various dressings can be used to definitively dress dermal and full-thickness burns. Nanocrystalline silver dressings such as Acticoat® are often favoured for their ability to prevent infection, and possibly thereby promote wound healing, in superficial and mid-dermal burns. These dressings can be left in place until review in 3–7 days.

Failure to recognise the need for escharotomies in circumferential burns may cost a burn patient their life or limb. Sometimes, escharotomies may need to be conducted prior to transfer to the regional Burns Unit by staff inexperienced with the procedure. Therefore, early consultation for advice from the specialist Burns Team is essential.

Surgical treatment

With competent burn wound dressing and care, most superficial dermal and mid-dermal burns will heal satisfactorily by re-epithelisation, and require no surgery. This healing is documented at dressing changes every 3–7 days, and if complete within 14–21 days is likely to be without significant scar formation. Deep burns, and those in whom healing is delayed or impaired, will benefit for operative burn wound care to reduce the risk of unsatisfactory scarring.

The aim of burns surgery is to excise dead skin and cover the burn wound with split-skin grafts as early as possible. Early skin coverage reduces the risk of many burn complications including infection, prolonged hospitalisation, scar formation and psychological disturbances. The trend in burns surgery is towards earlier primary debridement of even major burns; but care must be exercised when considering surgery in the patient with established ‘burns shock’.

When the area burned is so large that coverage with split skin grafts is limited by access to unburned donor skin sites, staged excision and grafting is required (e.g. twice a week until coverage is complete). Various options are available for interim coverage of a debrided burn

wound, including synthetic substitutes (e.g. Biobrane®) and biological dressings (e.g. cadoveric skin allograft from the Skin Bank).

Prevention of infection

Infection is the major cause of death following the initial burn injury. Empirical antibiotics are not effective in preventing infection – and may promote infection or colonisation by antibiotic resistant organisms. Burn wounds considered infected (or at risk of infection) are swabbed and targeted antimicrobial treatment commenced as required. Early enteral nutrition to prevent translocation of gastrointestinal tract flora into the bloodstream, early excision of devitalised burned tissue and the use of silver-based dressings have all reduced burn wound infection and systemic sepsis.

Nutrition

Good nutritional support of the severely burned patient is necessary to ensure an optimal outcome. Initiation of early enteral feeding (e.g. within 8 h of injury) improves nitrogen balance, reduces the hypermetabolic response and also reduces immunological complications. Hyperalimentation of calories, protein, trace elements and vitamins facilitates healing of the burn, as well as graft and donor wounds – despite the hypermetabolic response associated with major burns. Oral intake alone is seldom sufficient for this, particularly in young children, and a nasogastric (or other enteral) feeding tube is commonly required in these patients.

As with all severely injured children, children with major burns are at increased risk of stress peptic ulcers. Therefore, proton pump inhibitors (or other antacid therapies) are also indicated for children with burns exceeding 15% of total body surface area.

Psychological support and rehabilitation

Psychological support (ideally from parents, siblings and friends) is essential during hospitalisation and early rehabilitation, and may be required for years post injury. Discussion groups for parents and burn support groups should be available to assist the child and family return to a normal life. Children who have sustained a major burn are at increased risk of depression and suicide in later life. This is explained only in part by burns in children with pre-existing mental illness and/or significant social stressors.

Scars and contractures

Burn injuries, particularly deep burns, are notorious for healing with hypertrophic and functionally impairing scars. Hypertrophic scars require coordinated long-term care, with strategies including long-term pressure (e.g. silicon gels, taping, garments), splints and intralesional steroid injection. Active physiotherapy is an integral component of all scar management, to maintain and improve the range of movement at those joints and muscles implicated by scar formation. Such scar management is likely to extend for years, and in some cases may require specialist scar release surgery.

Reconstructive surgery

Contractures and scars can lead to functional disabilities and leave cosmetic blemishes. Surgical excision, inlay grafts and corticosteroid injections may be required until adolescence is reached and active growth has ceased.

KEY POINTS

- Burn severity depends on its size, depth and site.
- Early assessment, first aid and resuscitation is essential.
- Transfer to the regional burns unit all patients with full-thickness burn greater than 5%; total burn greater than 10%; inhalation burn; face, hand or buttock burns; non-accidental burn.

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PART VII
Orthopaedics

CHAPTER 42
Neonatal Orthopaedics

CASE 1

A 1-day-old baby, weighing 3.75 kg and born by a 'difficult' delivery, was not moving his right arm soon after birth. Examination revealed no deformities, but the baby cried when the arm was moved or when lifted. Two weeks later, a painless swelling was noted in the middle third of the right humerus.

Q 1.1 What is the swelling?

Q 1.2 What is the natural history?

CASE 2

A newborn baby was unable to leave the neonatal nursery because of being generally unwell with signs of sepsis. On the 10th day, lack of normal kicking movements were noted in the left lower limb. There was a low-grade pyrexia and the white cell count CRP and ESR were elevated.

Q 2.1 What might be seen on hip ultrasound?

CASE 3

A baby boy weighing 4.65 kg was born by a shoulder presentation to a diabetic mother. There were no left shoulder or elbow movements but normal grasp reflex and finger movements.

Q 3.1 What is the likely diagnosis?

Q 3.2 What is the likeliest outcome?

CASE 4

A newborn baby was noted to have both feet turned in to face each other immediately after birth. The father had had multiple operations for 'club feet' in childhood. On examination the feet could be corrected to the normal position with gentle pressure from one finger.

Q 4.1 What is the likely diagnosis?

Q 4.2 What is the prognosis?

CASE 5

In a country hospital, a newborn was noted to have both feet pointing upwards, lying along the lower tibia. The child was referred for an orthopaedic opinion, but the position improved before the consultation took place and then resolved without treatment.

Q 5.1 Is there a risk of other anomalies?

CASE 6

A male infant was born with the right foot turned down and inwards. The foot felt stiff and could not be placed in a normal alignment. There was a strong family history of club foot.

Q 6.1 What is the treatment and likely outcome?

Parents may bring their child to an orthopaedic surgeon in the first month of life because something looks wrong (club foot, bowed tibia) or because something is not moving or working properly (brachial plexus palsy, birth fracture). Alternatively, the paediatrician or orthopaedic surgeon may find something on examination of which the parent was not aware (developmental dislocation of the hip).

Newborn children have limited ways in which to respond to pain, be it from a birth fracture, osteomyelitis or a tumour. They often reduce or stop moving the limb, a condition known as 'pseudoparalysis'. The limb is not paralysed, but is held still because pain can be relieved by reducing or abolishing movements. The most common causes of 'pseudoparalysis' are fractures or infection.

Birth fractures

Birth fractures are quite common, with an incidence of 1–5 per 1000 live births and they are usually found in large, healthy babies after a difficult delivery, especially

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by the breech. The most common sites are the clavicle, the humerus and femur. Fractures of the clavicle may not be diagnosed until a painless swelling is noted because of callus formation. Not all birth injuries are fractures; separations of the humeral and femoral epiphyses also occur and can be difficult to diagnose without a high index of suspicion and special imaging techniques such as ultrasound. 'Dislocation' of the elbow or the knee probably never occurs in the newborn. However, the diagnosis of dislocation is often made incorrectly because the separation of the physis of the distal humerus or distal femur is an uncommon injury and on plain x-ray looks quite like a dislocation. Most birth injuries heal quickly with simple splinting and recover fully. Multiple fractures in a newborn suggest a bone fragility syndrome, such as osteogenesis imperfecta, or a generalised problem such as arthrogryposis multiplex congenita.

Neonatal musculoskeletal infection

Osteomyelitis and septic arthritis are difficult to distinguish in the neonatal period. The infection usually affects the end of the bone and the joint, and a better term is 'osteoarticular sepsis'. Infection in the neonate is the result of bacteraemia or septicaemia and presents with non-specific signs of generalised infection rather than signs of a localised bone and joint infection. Diagnosis may be delayed, during which time the growth plate or joint may be destroyed, with resulting life-long disability. A high index of suspicion is required in the neonate, with reduced limb movements and signs of sepsis. Fever may be absent or low grade.

The most common organism is *Staphylococcus aureus*, but a wide range of gram-positive and gram-negative organisms are recovered. Some are acquired from the birth canal. Identification of the organism from blood culture or joint aspirate is very important to direct antibiotic therapy appropriately.

Increasingly, intensive care units may be subject to colonisation with antibiotic-resistant organisms. Increasing numbers of neonates and children now present with methycillin-resistant staphylococcus aureus (MRSA) the majority of which are acquired in hospital but some may be acquired in the community (Community-Acquired Methycillin-Resistant *Staphylococcus Aureus* (CAMRSA)).

The most important factor in prognosis is the interval between onset and intervention. Growth plates and joints can be destroyed quickly and quietly in the neonate. Joint aspiration is a diagnostic procedure, but once pus is identified, the affected joint must be drained by arthrotomy and splinted for comfort. For the hip, abduction splintage is used to prevent or treat septic dislocation.

Birth brachial plexus palsy ('obstetric palsy')

A true paralysis of the upper limb may occur – usually in a large baby, and as the result of a difficult delivery. Injuries to the spinal cord are rare and present as a partial or complete quadriplegia or paraplegia. A partial palsy of the upper limb is usually caused by a traction injury to the upper trunk of the brachial plexus. The majority of injuries are neuropraxias, the nerve trunks are in continuity and 80% recover fully. As a general rule, a child who recovers elbow flexion by the age of 3 months will make a full recovery. These babies require careful evaluation at intervals to document recovery and to prevent secondary contractures and deformities by a simple programme of stretching exercises. In particular, internal rotation contracture of the shoulder can be prevented more effectively by physiotherapy than by splinting.

Microsurgical repair may be helpful for those infants with complete tears to the roots of the brachial plexus, where there is little or no recovery.

Neonatal foot deformities

Most parents have a good idea of what a baby's foot should look like, and are anxious and distressed when they see a deformity. The majority of deformities are postural variations or 'packaging defects'. In the womb, the foot has been compressed in an abnormal position, either because the baby is large, the womb is crowded (e.g. twin pregnancy) or lacking in amniotic fluid (oligohydramnios). Soon after birth, when the baby has room to move, the posture improves and the appearance becomes normal. Occasionally this process is sped up by a short period of stretching in a splint or cast, but surgery is not required. Postural club foot, metatarsus adductus and talipes calcaneovalgus are good examples of 'packaging disorders'.

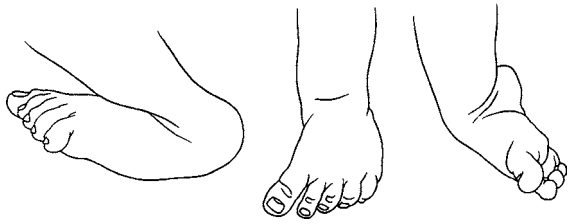


Figure 42.1 **(a)** Congenital talipes calcaneovalgus. The foot has been folded back against the front of the tibia, but there is no fixed deformity; **(b)** metatarsus adductus: **(c)** congenital talipes equinovarus (club foot): this can be postural (mobile) or structural (stiff).

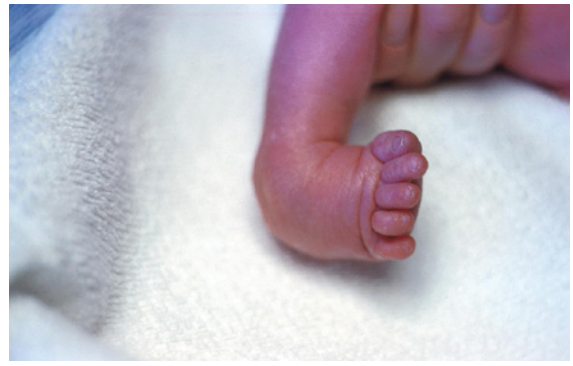


Figure 42.2 Congenital talipes equinovarus. The foot lies in a typical position and is rigid.

In metatarsus adductus, the foot curves inwards, especially the great toe, so that the sole of the foot has a 'bean' shape, but the hind foot is normal. The deformity is flexible and resolves rapidly, either with a short period of casting or spontaneously.

The foot in talipes calcaneovalgus [Fig. 42.1] has been lying along the tibia in the womb, the heel in a downward position, referred to as calcaneus, in comparison to the heel in an upward position of club foot, referred to as equinus. The deformity is flexible and corrects rapidly.

Postural club foot looks just like structural club foot, hence parental anxiety. However, the two conditions do not feel in the least like each other. Postural club feet are soft and supple. Gentle pressure from an examiner's finger can place the foot in a normal position.

By contrast, 'manufacturing defects' are structural and require surgical correction. Club foot (talipes equinovarus) is a good example and is easily recognised because of the stiffness of the deformity when the examiner attempts to place the foot in the correct position [Fig. 42.2].

The incidence of club foot is about 1 per 1000 live births in Caucasians, 0.5 per 1000 in Asians but 5 per 1000 in Polynesians. It is more common in males and is bilateral in about 40%. A family history is present, but both genetic and environmental factors are implicated. Club foot may be an isolated deformity, associated with conditions such as congenital hip dislocation, part of a syndrome or acquired because of neuromuscular disease. A revolution has occurred in the treatment of club feet in children by the 're-discovery' of the Ponseti method. This involves serial casting of the deformity starting as soon after birth as is practically possible. Sometimes simple percutaneous tenotomy of the Achilles tendon is required. Following correction in a series of four to six casts, long-term splinting using special shoes attached to a bar is required. The long-term results of this largely non-operative treatment are vastly superior to those achieved by extensive surgery. Nowadays only a few feet need surgical correction when non-operative treatment fails. However, the affected foot is usually smaller and the leg on the affected side is usually shorter in unilateral cases. These long-term issues must be carefully explained to parents whose understanding and cooperation is required to achieve good results by the Ponseti method.

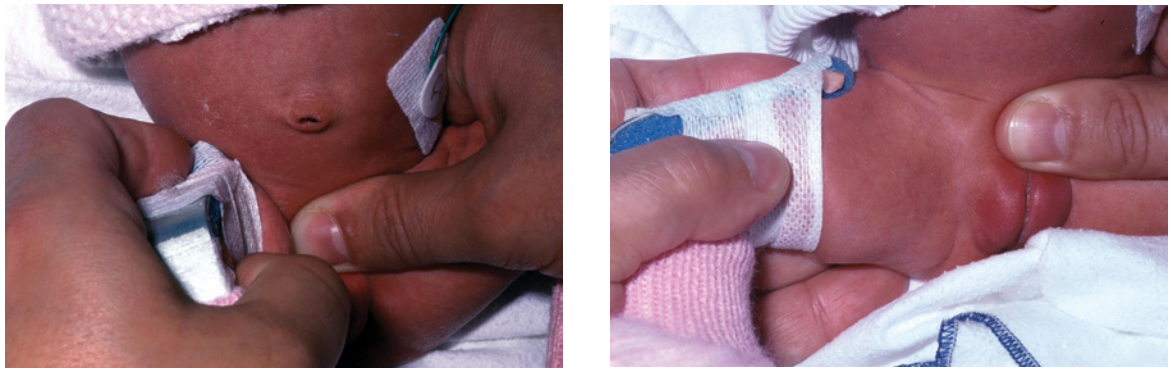
Another important 'manufacturing defect' is congenital dislocation of the hip. This rarely presents in the neonate because the parents have noted something; rather, it is found thanks to a screening programme.

Developmental dysplasia of the hip (DDH or CDH)

The preferred term is developmental dysplasia of the hip (DDH) rather than 'congenital dislocation of the hip' (CDH). This reflects two important features of the natural history of the condition:

- Not all cases are found at birth; some develop during the first year of life.
- Not all hips are dislocated; some have only a shallow or dysplastic acetabulum.

DDH covers a spectrum of hip dysplasia and instability, presenting from birth to early childhood. It is the result of both genetic and environmental factors, so that family history is a risk factor but so also is breech birth. According to diagnostic criteria, it affects 1–2 per 1000 live births and is at least five times more common in females than males. Dysplasia usually refers to a shallowness or



(a) (b)

Figure 42.3 Developmental dysplasia of the hips. Ortolani test: **(a)** the thumbs are placed over the front of the hip joints **(b)** as both are fully abducted with the knees and hips flexed.

malformation of the acetabulum, subluxation to a partial displacement of the head of the femur from the acetabulum and dislocation to displacement of the head of the femur completely outside the acetabulum.

The condition is best diagnosed soon after birth by the use of clinical tests of neonatal hip instability [Fig. 42.3], supplemented by the selective use of ultrasound examination. Hip x-rays are of little value in the neonatal period because the femoral head is cartilaginous and hence invisible until the age of 4–8 months, when the ossification centre develops.

Ideally, the examination should be performed by an experienced examiner. In practice, DDH is uncommon and not every paediatrician or GP is able to gain the necessary experience. The baby should be undressed, warm, relaxed and placed on a firm but comfortable surface. Offering a bottle, finger or dummy to suck can help.

The examiner holds the leg to be examined in the hand with the hip and knee flexed and the thumb on the inner side of the thigh over the lesser trochanter and the middle finger over the great trochanter. The right hand is used to examine the baby's left hip and the left hand to examine the right hip.

The pelvis is steadied by the other hand and the flexed thigh is abducted and adducted, carefully feeling for any 'clunk' or jerk, which may denote the hip entering or leaving the acetabulum (Ortolani test). Then, with the hip adducted, gentle downwards pressure is exerted to determine if the head of the femur is stable, or whether it may slip posteriorly out of the acetabulum (Barlow test).

Educational DVDs are now available to aid in the teaching of health practitioners who have the responsibility for performing the Ortolani and Barlow tests, including maternal and child health nurses, paediatricians, obstetricians and general practitioners.

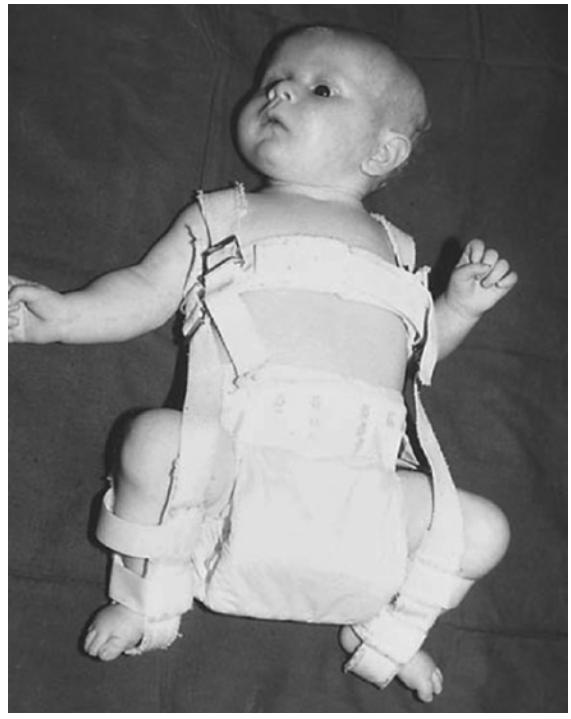


Figure 42.4 Pavlik harness. In the flexed and abducted position, the dislocated hip is held in the reduced position.

A fine 'click' is not evidence of dislocation or instability, but the baby should be examined again. A 'clunk' as the hip enters the socket from a dislocated position is the most important finding and is an indication for immediate treatment. The unstable neonatal hip tends to dislocate in

adduction and extension, and to reduce in flexion and abduction. Treatment is directed towards gently placing the hips in a position of abduction and flexion while allowing movement within a safe arc of motion. This is most efficiently and safely achieved by using a Pavlik harness [Fig. 42.4]. The progress of treatment is best monitored by careful repeated hip examinations, supplemented by ultrasound examinations, which can be performed while the harness is in place. Most hips will stabilise quickly and become normal. If the golden opportunity for early diagnosis and treatment is missed, operative treatment and a less satisfactory result are much more likely.

KEY POINTS

- Neonates hold painful limbs still, as if 'paralysed'.
- Fractures are common during difficult delivery.
- Fever and pseudoparalysis suggests osteoarticular sepsis.
- Brachial plexus traction injury at birth usually recovers (80%).
- The key to foot deformity at birth is whether it is mobile or stiff.
- Developmental dysplasia of the hip needs to be sought in high-risk infants.

CHAPTER 43
Orthopaedics in the Infant and Toddler

CASE 1

A mother brought her 12-month-old child to her general practitioner (GP) because he had just started to pull to stand, and she was worried that he had flat feet and bow legs. Examination revealed a healthy toddler with symmetric bowing of the lower limbs: the gap between the knees when standing was 4 cm. There was no medial arch in the feet in the standing position.

Q 1.1 What is the likely outcome for this child?

CASE 2

Jessica, an 18 month old, presented because she was walking with in-toeing. She had a normal birth and developmental history but walked with both feet facing inwards and sometimes tripped. Examination revealed mild bowing and medial tibial torsion.

Q 2.1 What is the natural history and management?

CASE 3

Susan presented at the age of 14 months with a limp. She was the first born to a young mother who walked with a severe limp because of an arthritic hip. Susan had been born by breech delivery and was referred for an x-ray of her hips.

Q 3.1 What is the diagnosis?

Q 3.2 Could the problem be diagnosed earlier?

CASE 4

John was brought to see his paediatrician because, at the age of 18 months, he limped on his right leg and when he ran, his right arm was held stiffly with the elbow flexed. When examined, the muscles of the right arm and leg felt stiff when compared with the left, and the deep tendon reflexes were brisk.

Q 4.1 What is the likely problem?

CASE 5

Bruce, aged 22 months, was collected from childcare by his mother who was told that he had been limping on his right leg. He was put to bed but next morning refused to walk. He had a fever and was sore when his nappy was changed. His mother took him to his GP, who arranged admission to hospital.

Q 5.1 What might be wrong?

Q 5.2 What would a bone scan show?

CASE 6

Mary, a 4 month old, presented to her GP because of persistent crying and a swollen left thigh. She was a 'difficult' baby with feeding and sleeping problems. There were several bruises and abrasions.

Q 6.1 What is the diagnosis?

Q 6.2 What would x-rays show?

CASE 7

Toby was well known at the emergency department of his local hospital. At the age of 20 months, he had been seen three times with fractures of both his upper and lower limbs. On this occasion his blue sclerae were noted.

Q 7.1 What is the diagnosis?

Limp, or abnormal gait, is the most common reason for orthopaedic referral in the infant and toddler. As children grow, there are rapid changes in the appearance and alignment of the lower limbs, such that the bow-legged toddler becomes a knock-kneed child, and eventually an adult with straight legs. Many toddlers who are referred to orthopaedic clinics are normal and have no specific disease or deformity. They are referred with a variation of normality, such as flexible flat foot, in-toeing, out-toeing, knock-knees or bowlegs. It is essential to recognise that there can be just as much parental anxiety in these circumstances as there is about a child with a definable pathological condition.

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The general principles of the management of these children are as follows:

1 These conditions are common because they are normal variations.
2 A reasonable description of 'normal' is the mean value of the measurement plus or minus two standard deviations. This is of value to doctors, but not to parents.
3 These conditions generally resolve spontaneously and there is little evidence that intervention changes the natural history.
4 Overinvestigation of these children should be resisted.
5 Overtreatment should be resisted.
6 Within the large group of normal children with physiological variants, there are a small number with specific pathology. These children should be identified, investigated, diagnosed and treated appropriately.

The toddler with a symmetrically abnormal gait

Bow legs

When the toddler first begins to walk, the appearance of bowing is very common [Fig. 43.1]. It is frequently accompanied by some degree of internal tibial torsion and the one deformity accentuates the other. Bowing seems to be pronounced.

Physiological bowing is symmetrical, not excessively severe, and improves with time. Measurement of the distance between the knees (the intercondylar separation (ICS)) in the standing child provides a simple means of follow-up to assess whether the condition is improving or not. Often this is all that parents require for reassurance. Night splints have been abandoned with the recognition that they do not influence the natural resolution of the condition.

Pathological bowing may be asymmetrical, is often more severe and deteriorates with time. Causes of pathological bowing include Blount's disease, rickets, trauma and skeletal dysplasia.

In-toeing

In-toeing is one of the commonest presenting symptoms at paediatric orthopaedic clinics because of the appearance and parental concern about long-term sequelae [Fig. 43.2]. Sometimes the concern is from the grandparents or a kindergarten teacher. There may also be a complaint that the child is clumsy or trips frequently. There is often a marked contrast between parent and child. The parent is anxious about the 'deformity', whereas the child runs around the consulting room in a carefree fashion, frequently not demonstrating nearly as much in-toeing as the parents claim is the case at home.

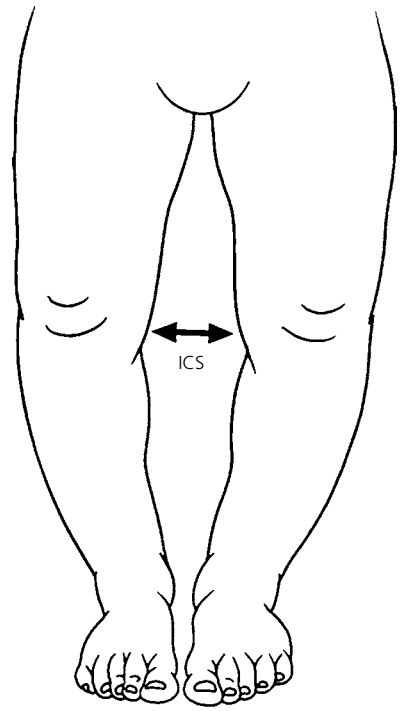

Figure 43.1 Bow legs: the tibia has an outward curve and an inward twist (internal torsion), both of which accentuate the normal 'flat' appearance of the feet. The inter-condylar separation (ICS) can be measured to monitor resolution.

Internal tibial torsion

Internal or medial tibial torsion is very common in toddlers and usually presents as in-toeing between 1 and 3 years of age. It is probably a 'packaging defect'; the result of intrauterine positioning. It frequently coexists with, and may be confused with, bowing of the tibia, physiological genu varum.

The natural history is for spontaneous resolution. A number of orthotic devices have been used; principally boots on a curved metal bar with the feet turned outwards (Denis Browne splint). This is used as a night splint and is a potent cause of disturbed sleep and family distress. It may speed resolution of the deformity, but this has never been proven. Surgery is almost never required in normal children. In pathological conditions

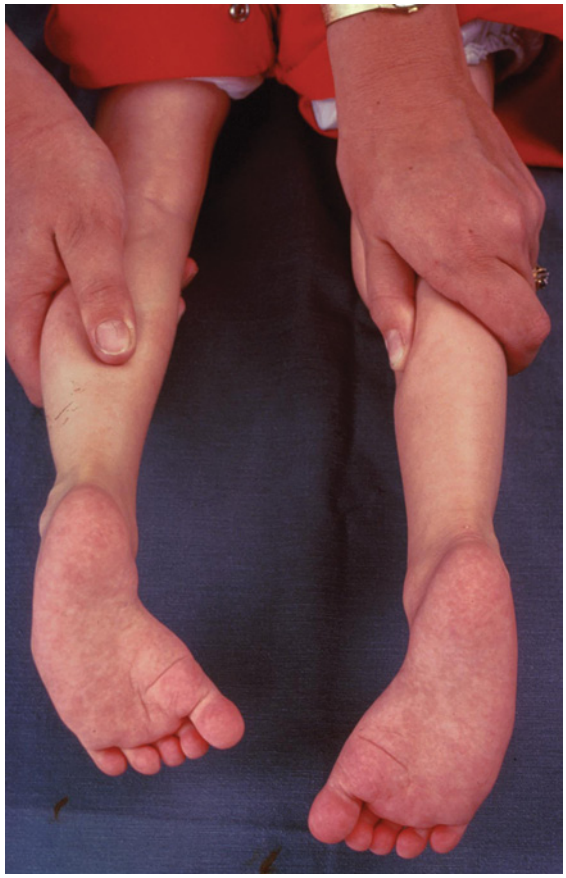

Figure 43.2 Metatarsus adductus. The midfoot and forefoot are adducted, but the heel is normal.

such as spina bifida, it can be treated by derotation tibial osteotomy at the supramalleolar level.

The toddler with a painless, chronic limp

Developmental dysplasia of the hip (DDH)

DDH may present at walking age because of an asymmetric gait [Fig. 43.3]. At this age, the hip is not painful and does not cause an undue delay in walking. Bilateral hip dislocations may present even later than a unilateral dislocation because the deformity is symmetrical. The risk factors are the first-born female child, breech delivery, with a positive family history. In addition to the usual neonatal clinical examination of the hips, an infant with many risk factors should have an ultrasound examination of the hips in the neonatal period and an x-ray of the hips at 6 months. Delayed presentation leads to a high risk of arthritis in young adults.

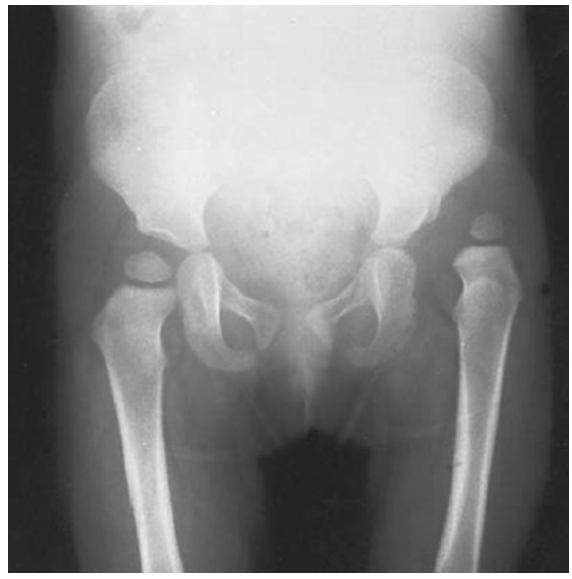

Figure 43.3 Congenital dislocation of the hip. X-ray showing permanently dislocated hip and poor acetabular development in an infant where the diagnosis was missed at birth.

Hemiplegia

The typical hemiplegic gait is a limp with a stiff, ipsilateral arm with a flexed elbow. Most children with hemiplegia have a brain lesion acquired in the perinatal period, but the mildly involved may not present until walking age. Walking and running may unmask or accentuate the posturing in the upper limb.

Cerebral palsy (CP) is the most common cause of physical disability in developed countries. It is classified according to the type of the movement disorder (spastic, athetoid, ataxic, mixed) and the distribution in the limbs (hemiplegia, diplegia, quadriplegia).

Toe-walking

When children are learning to walk, a short period of intermittent toe-walking is very common. It is then followed by a period of 'flat foot' strike, before the gait matures to the adult pattern, in which a heel strike is normal. In some children, the period of toe-walking is prolonged and pronounced, causing parental concern and referral to the orthopaedic surgeon.

Differential diagnosis

The majority of these children are otherwise normal and are called 'idiopathic toe-walkers'. Pathological causes of 'toe-walking' are diplegic CP, muscular dystrophy, Charcot-Marie-Tooth disease and spinal dysraphism. Unilateral toe-walking is almost always pathological and the most common causes are hemiplegic CP and unilateral DDH.

With the rapid increase in the number of children diagnosed with 'autism-spectrum disorders' (ASD), the number of children with unexplained toe-walking has rapidly increased. There is a definite overlap between the tendency to toe-walking and the presence of autism for reasons that have not yet been explained.

Acute onset limping in the toddler

This is a common age for presentation of acute haematogenous osteomyelitis (AHO). Over a period of 12–48 h, the child develops fever, a limp and then refuses to walk. A toddler's fracture of the tibia may present in a similar manner because the fall or injury is not observed and the fracture may be difficult to see on x-ray [Fig. 43.4].

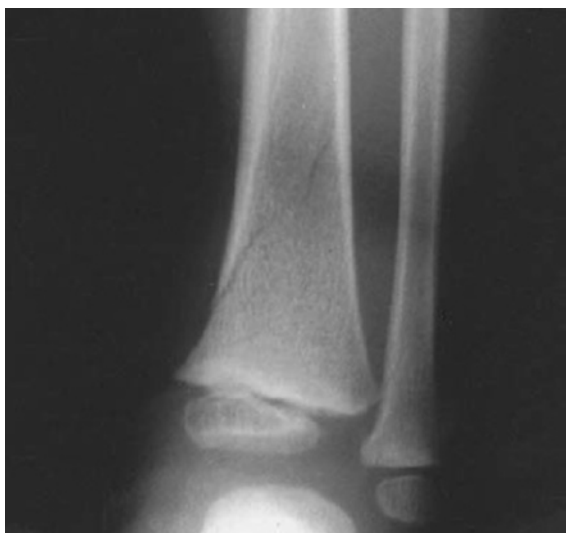

Figure 43.4 The 'toddler's fracture'. A spiral fracture of the distal tibia often presenting as a limp of unknown cause (as the fall was not witnessed), and which is difficult to see on x-ray.

Fractures in the infant and toddler

It is difficult for a normal infant to sustain a femoral fracture. Infants cannot climb, they have limited mobility and most accidental falls in this age group do not result in a fracture. The younger the child with any fracture, especially a femoral fracture, the higher the incidence of child abuse. As many as 40% of femoral fractures in the under-12-month age group are caused by child abuse. A bone scan will detect any occult fractures [Fig. 43.5].

Unfortunately the prevalence of fractures caused by parents seems to be increasing and fractures of the femur up to the age of 2–3 years may be caused by this unhappy mechanism.

Most infants and children with fractures and an unconvincing history have been abused. However, a number may have fragile bones because of osteogenesis imperfecta, and a premature diagnosis of child abuse may cause irreparable harm [Fig. 43.6].

Figure 43.5 A technetium bone scan is a sensitive means to detect occult fractures in child abuse. Note 'hot-spots' in forearm (arrow) and ribs.

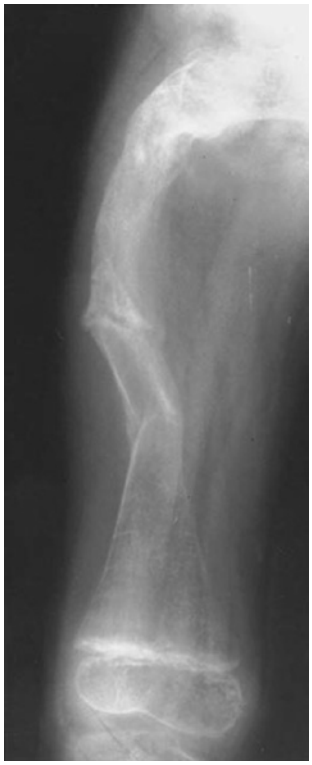

Figure 43.6 Multiple fractures and deformity of the femur in osteogenesis imperfecta.

KEY POINTS

- Lower limb shape changes with age, and rarely is pathological if symmetrical.
- Symmetrical bow legs is common between 1 and 3 years of age.
- In-toeing is common and rarely needs treatment.
- Painless limp in a toddler needs investigation for developmental dysplasia of the hip.
- Painful limp in a toddler needs investigation for fracture or osteomyelitis.
- Fractures in a toddler should arouse suspicion of child abuse.

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CHAPTER 44
Orthopaedics in the Child

CASE 1

Mary, a 4 year old, was brought to see her general practitioner (GP) because of in-toeing. Examination revealed that she walked with both feet and knees facing inwards by 20°. Her mother commented that she had been described as 'double jointed' as a child. Both had signs of generalised joint laxity.
Q 1.1 What is the diagnosis?

CASE 2

Jacky, a 5 year old, was brought to see an orthopaedic surgeon because of knock-knees. Examination revealed symmetric genu valgum with 6 cm between the ankles in the standing position.
Q 2.1 Is treatment required?

CASE 3

Sara, a 6 year old, was a keen gymnast and was noted to have 'flat feet'. Expensive orthotics were prescribed.
Q 3.1 Are these necessary?

CASE 4

A 7-year-old boy falls out of a tree on to his outstretched hand. He presents shortly after with a very swollen, painful elbow and decreased radial pulse.
Q 4.1 Why is this important?

As children become older, parental anxiety about the appearance of their feet, legs and walking continues. As in the younger age groups, the majority of these children are also normal, but a different spectrum of problems are seen from those seen in the toddler. It may be rare to see developmental dysplasia of the hip or cerebral palsy presenting for the first time in the child, but irritable hip, Perthes' disease [Fig. 44.1], osteomyelitis and septic arthritis are all seen.

As children become more adventurous in play and participate in sport, an increasing number and variety of fractures and epiphyseal injuries are seen.

Internal femoral torsion (inset hips)

This is frequently seen in children between the ages of 3 and 10 years. The in-toeing is symmetrical. Parents complain that their children look awkward and trip frequently, but the degree of disability is not great. The child often has signs of generalised joint laxity and may have associated features, such as flexible flat feet.

Examination reveals a characteristic shift of the arc of hip rotation inwards, hence the synonym 'inset hips'. A typical finding would be internal rotation of 80–90° and external rotation of 0–10°. This is the reason why the children can sit comfortably in the 'W' position [Fig. 44.2]. It is doubtful if sitting in this position causes the condition, but there is some evidence that habitually sitting in this posture slows down the natural tendency to spontaneous recovery.

In some children, correction of the in-toeing is accomplished by a compensatory tibial torsion. In these children, the feet no longer turn in, but in standing and walking the patellae are facing inwards or 'squinting'. This combination of deformities can look unattractive and gives the appearance of bowlegs.

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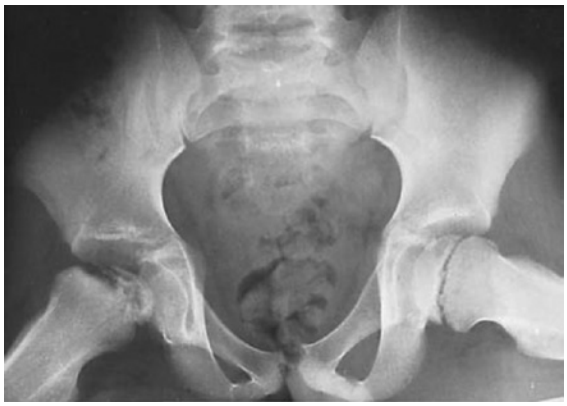


Figure 44.1 Typical x-ray appearance of Perthes' disease.

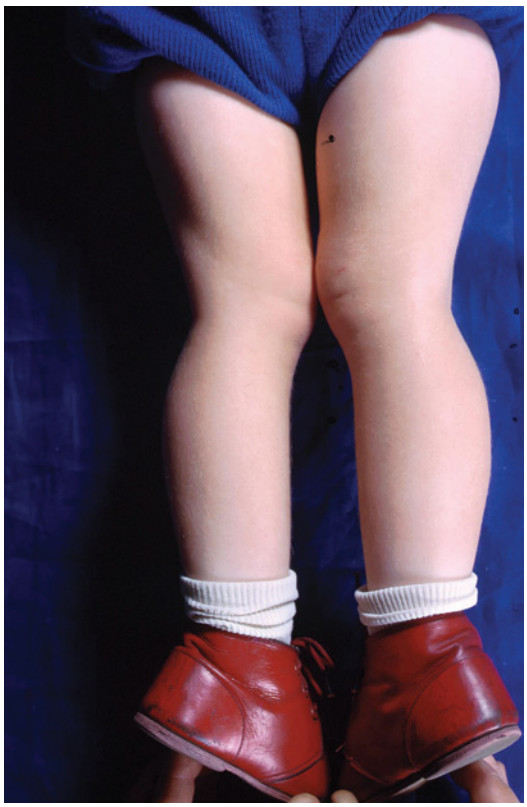


Figure 44.2 Internal femoral torsion (inset hips): the child can sit on the floor in the 'W' position.

Management

The natural history of the condition is for spontaneous resolution during the growing years. There is no evidence that any form of exercises or orthotic devices influences the resolution. The condition can be treated surgically by means of external rotation osteotomy of the femur, but the vast majority of children improve spontaneously and do not require intervention.

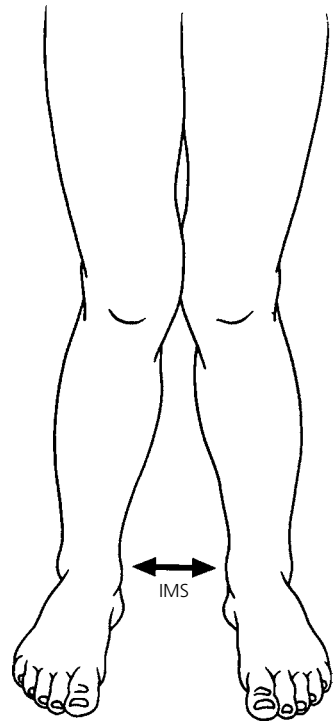


Figure 44.3 Knock-knees (genu valgum). (IMS: intermalleolar separation).

Knock knees

Physiological genu valgum, or knock-knee deformity, is often seen in children between the ages of 3 and 8 years [Fig. 44.3]. The majority of children straighten spontaneously. The deformity is symmetrical, not excessive (e.g. gap between ankles on standing <10 cm) and improves with time. Pathological genu valgum is usually more severe, asymmetrical and increases with time. Causes include trauma (proximal metaphyseal greenstick fracture of the tibia or growth plate injury) rickets, skeletal dysplasias and congenital limb deficiencies.

Management

There is no evidence that the natural history of the condition is affected by exercises, shoe inserts or night splints.

A small number of children with physiological genu valgum do not correct completely. The reasons to consider surgery are discomfort from 'knee-swishing' while running, concern about the appearance and progression of the deformity in the pathological cases. In order to assess the degree and site of deformity, a standing x-ray of the lower limb should be obtained. Correction can be achieved by restricting growth in the distal femoral or proximal tibial growth plates on the medial side of the knee using staples or screws. New, safer devices for growth plate surgery are now widely available, minimally invasive and very successful for the correction of angular deformities in children. However, by definition the children must be 'growing' and must be referred in time!

When the growth plates have already fused, unfortunately osteotomy of the distal femur or proximal tibia may be required.

Flat feet

Almost all infants have 'flat feet', and in the majority an arch will develop by the age of 6 years. The clinical findings of a flexible flat foot include absence of the medial longitudinal arch and a variable degree of hind foot valgus. When the child stands 'at ease', the only support to the medial arch is the interosseus ligaments and intrinsic muscles of the foot, which are not continuously active. When the child stands on tiptoe, the long flexor and extensor muscles are recruited into continuous activity. In the correctable flat foot, the medial longitudinal arch usually appears and the heel tilts into neutral or varus. This 'tiptoe test' can be used to explain the nature of the condition to parents and to reassure them that the internal structure of the foot is normal. In the flexible flat foot, the medial arch is also reformed on weight bearing when the hallux is passively dorsiflexed. This is referred to as the 'toe-raising test of Jack'.

Pathological causes of flat foot include hypermobility syndromes and cerebral palsy.

Management

Most of the enthusiasm for 'treating' flat foot has probably been based on the observation that with use of any of the popular forms of treatment, the majority of children are noted to get 'better'.

Although shoe modifications and inserts do not change the shape of the foot in the long term, there is some evidence that orthotics may prolong the life of the shoe by decreasing deformation and wear. If excessive shoe wear and cost of replacements are important to the parents, or pain is a problem, the Helfet or UCBL heel cup or a simple medial arch support may be helpful. Expensive, custom-made orthotics are rarely, if ever, required.

In children with normal flexible flat foot, surgery is very rarely required.

Growing pains and night cramps

About 15% of children go through a period where they waken at night, crying because of pains in their legs. The child goes to sleep after an energetic day only to waken in pain and misery, but the following day all is well. Presentation is often delayed until there have been many disturbed nights.

Clinical features

The child has no daytime pain and no limp. The pain at night is relieved by rubbing, heat and simple analgesics. Examination reveals no abnormalities.

Differential diagnosis

Night pains are a feature of osteoid osteoma, but this is always unilateral and often reasonably well localised. One cause of bilateral leg pains is leukaemia, which can be excluded in most children by a full-blood count. There are usually other features in leukaemia or an atypical story, so investigation is not necessary in all children with bilateral nocturnal leg pain.

Management

Full history-taking and thorough examination excludes pathological causes and allays parental anxiety. Reassurance is very important, and fortunately, most parents can accept the situation. There may be a role for a programme of stretching exercises.

Fractures and epiphyseal injuries in the child

As the child becomes more adventurous in play, and then active in organised sport, the incidence of musculoskeletal injuries increases dramatically. The weak link in the child's skeleton is the growth plate or physis. In

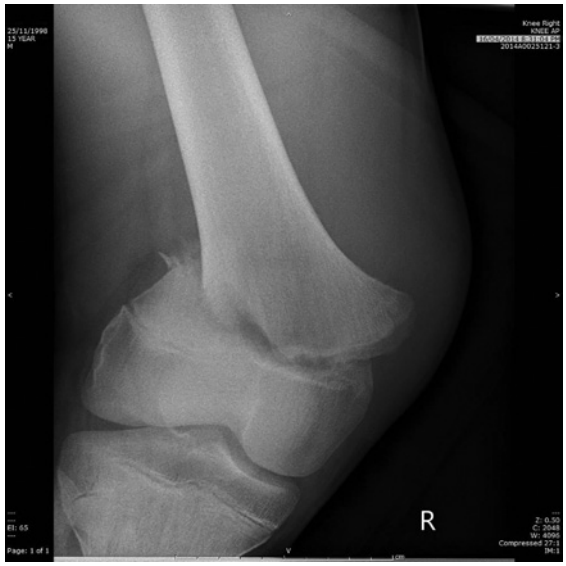


Figure 44.4 An x-ray image of valgus injury to the knee. In an adult, a tear of the medial ligament would be likely, whereas in this child the result is a Harris–Salter type 2 separation of the distal femoral plate.

children, epiphyseal separations are common, as are fractures of the long bones.

Specific soft tissue injuries, such as collateral ligament tears, are rare and the diagnosis of a 'sprain' in the child is frequently incorrect. A valgus force at the knee, which would result in a tear of the medial collateral ligament

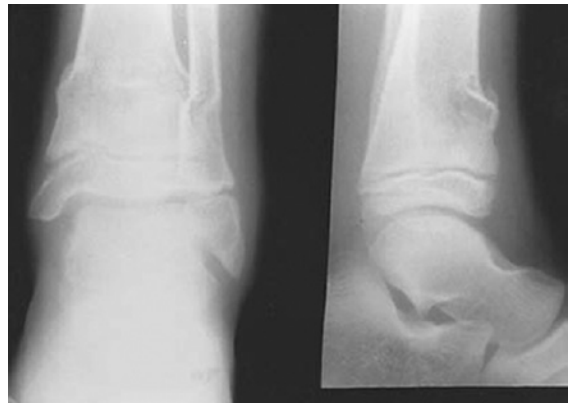
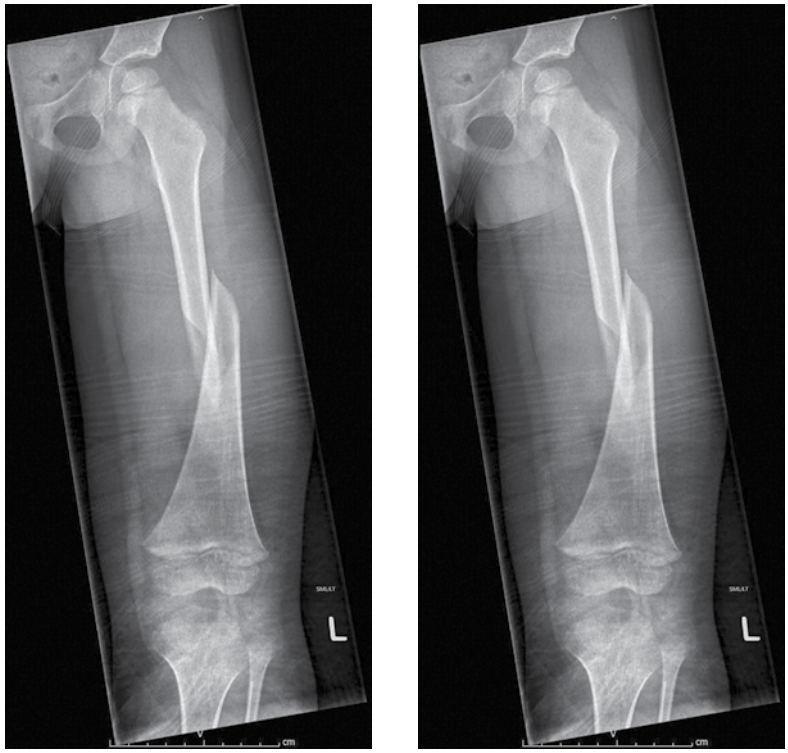


Figure 44.5 Children's bones may bend and buckle, as in this fracture of the distal tibia and fistula.



(a) (b)

Figure 44.6 (**a**) X-ray of open fracture of the femur as a result of a fall from a tree. Note the gross displacement and shortening. (**b**) X-ray after wound care, reduction and traction, the fracture is healing in good position. Up to 1 cm of overlap is acceptable because of anticipated overgrowth.

in an adult, is more likely to cause a separation of the distal femoral epiphysis in the child [Fig. 44.4]. The equivalent of an anterior cruciate tear in a child is avulsion of the tibial spine.

Non-specific, minor soft tissue injury is common in the child, including abrasions and bruising.

Fractures in children

Fractures are caused by forces applied to the skeleton which result in failure of the bone under the applied load. Because children's bones have different biomechanical qualities from adult bones, the patterns of failure are different. Children's bones may bend and buckle rather than breaking cleanly [Fig. 44.5]. Plastic bowing, buckle fractures and greenstick fractures are all incomplete fractures frequently seen in children but not in adults. Children's fractures heal more quickly than adult fractures, and recovery of function is also faster and generally more complete. Children's fractures are subject to a process of remodelling during further growth by which residual deformity may correct and function improve. Remodelling is faster and more complete in younger children and for fractures close to an active growth plate. Hence, residual angulation or displacement of distal radial fractures in younger children is well tolerated and there are few poor results in the long term. Fractures of the femur in children aged between 4 and 10 years are subject to 'overgrowth'. During the remodelling phase, which may last for more than 12 months, the hyperaemia results in faster growth of the injured limb compared with the uninjured limb. During the first year after fracture, this may amount to between 0.5 and 1.5 cm. With this in mind, femoral fractures in this age group may be allowed to heal with up to 1 cm of overlap or shortening, in the expectation that overgrowth will tend to make up the deficit and equalise the length of the lower limbs [Fig. 44.6].

In children, most fractures are isolated injuries caused by indirect forces:

- In the upper limb, a fall on the outstretched hand
- In the lower limb, a twisting injury, for example, roller skating

These are usually closed injuries, with a good prognosis.

A small percentage of injuries are caused by direct violence, usually road trauma. These injuries are more likely to be multiple, severely displaced, open or compound, and have associated injuries to the head, spinal cord or abdomen.

Fractures in children are treated in many ways, including cast immobilisation [Fig. 44.7], traction, internal fixation and external fixation. The choice of management is based on an understanding of the risks and benefits of each type of treatment, with safety, efficacy and convenience being the most important factors.

Upper limb fractures

Fractures occur most frequently at the ends of the long bones but may be seen in the midshaft. The area next to the growth plate, the metaphysis, is especially vulnerable. In the upper limb, the most common injuries are fractures of the distal radial metaphysis, the diaphyses of the radius and ulna, and fractures around the elbow. Most fractures of the radius and ulna are managed by closed reduction and cast immobilisation for about 6 weeks.

Elbow fractures in children are common; there are many types, and a variety of management strategies are required. An accurate diagnosis is required which in turn requires good quality anterior-posterior (AP) and

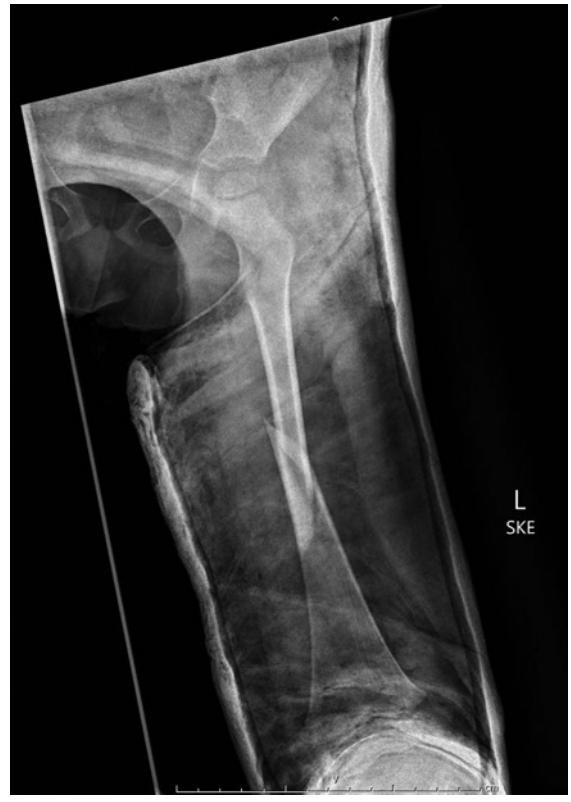
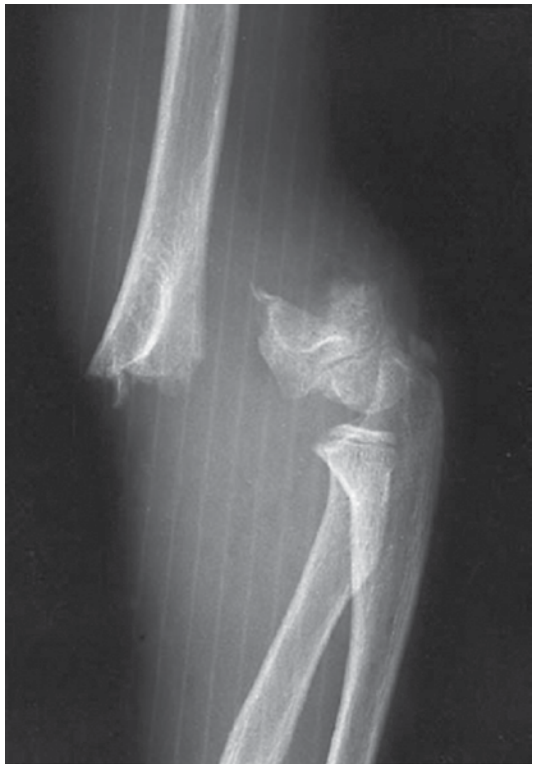
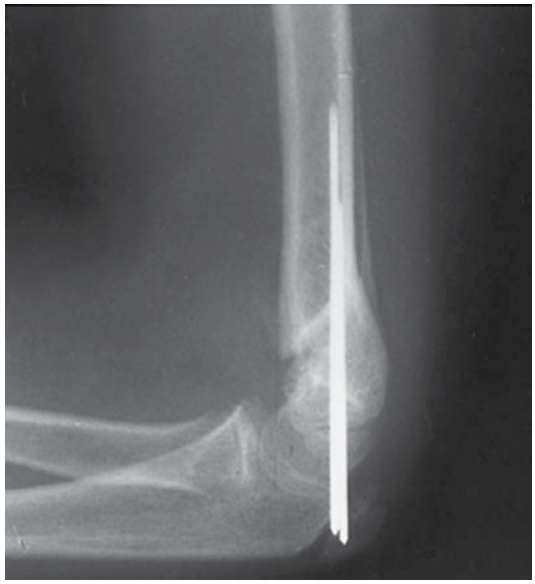


Figure 44.7 X-ray of cast immobilisation of a femoral fracture, with acceptable overlap in good position.



(a)



(b)

Figure 44.8 (**a**) Supracondylar fracture of the humerus with gross displacement: the neurovascular structures are at risk. (**b**) The appearance after closed reduction and Kirschner wire fixation.

BOX 44.1 Features of arterial ischaemia in fractured limbs

1. Pain, severe and unremitting
2. Pallor of the digits with lack of capillary return
3. Paralysis with inability to move the digits
   if full passive mobility produces pain, suspect ischaemia
4. Altered sensation

lateral x-rays, and a knowledge of the normal growth patterns of the elbow.

The most common of the more serious injuries is the supracondylar fracture of the distal humerus. This is a transverse fracture of the distal humerus, just above the growth plate, and is usually displaced backwards as the result of a fall on the outstretched hand [Fig. 44.8a]. The fracture displacement or subsequent swelling may result in vascular problems in the forearm and hand and Volkermann's ischaemia. Nerve palsies are also common. In the past, these fractures were usually managed by reduction and casting with the elbow flexed, but this increases the risk of Volkermann's ischaemia. The preferred management for displaced supracondylar fractures is now closed reduction and percutaneous fixation with Kirschner wires [Fig. 44.8b, Box 44.1].

Displaced fractures of the lateral condylar physis are Harris–Salter type 4 injuries and require open reduction and internal fixation. Fractures of the radial neck can usually be managed by closed reduction or an indirect percutaneous reduction with a Kirschner wire.

The Harris–Salter classification of growth plate injuries

There are many classifications of growth plate injuries, but that by Harris and Salter is the most popular and useful [Fig. 44.9]. The line of separation of the growth plate is identified on good quality AP and lateral x-rays, looking for both horizontal and vertical components. Most injuries can be readily classified in one of the five groups. Some complex injuries require further imaging including CT scans or MRI.

Type 1 and 2 injuries are the most common and are usually managed by closed reduction and cast immobilisation. Epiphyseal injuries heal very quickly and in type 1 and 2 injuries, the prognosis is usually good. In type 3 and 4 injuries, the growth cartilage and articular

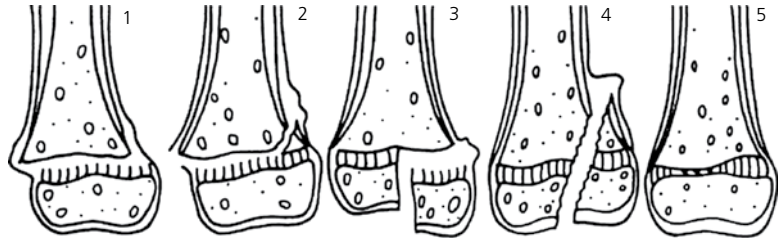


Figure 44.9 The types of growth-plate injury, as classified by Salter and Harris.

cartilage are both disrupted. Precise reduction is required (this usually means an open reduction) but growth disturbance is still a possibility. Partial growth arrest may cause a progressive angular deformity in the limb; a complete arrest results in progressive shortening.

Lower limb fractures

Fractures of the femur and tibia are common and are usually classified according to the position of the fracture in the diaphysis; for example, the upper, lower or middle third. Femoral fractures can be managed by a wide variety of methods including traction, hip spica casts, internal fixation and external fixation [Fig. 44.6]. The method is chosen according to the age of the child, the fracture type and displacement, and the experience and preference of the surgeon. Younger children tolerate traction and casts very well. Open fractures and those associated with head injuries, tibial fractures and multiple injuries are better managed by internal fixation. Flexible intramedullary nails are the most widely used fixation devices. The time to healing is closely related to age: 2–3 weeks in the first year of life, 6–8 weeks in children and 8–12 weeks in teenagers. Remodelling and overgrowth have been referred to earlier.

Tibial fractures are very common but are usually more easily managed than femoral fractures. The majority are treated by closed reduction and cast immobilisation for 6–10 weeks. Displaced diaphyseal fractures carry a risk of compartment syndrome, and neurovascular monitoring is important for 48h after injury. Tibial and femoral fractures cause a prolonged period of limping in most children because of weakness, stiffness and loss of confidence. Time and reassurance of parents is of more help than physiotherapy.

KEY POINTS

- In-toeing needs no treatment if symmetrical.
- Knock-knee deformity is common between 3 and 8 years and usually needs no treatment.
- Custom-made orthotics are unnecessary in flat foot.
- Night pains in legs need full history and physical examination to exclude rare serious pathology and reassure parents.
- Fractures and epiphyseal injuries are much more common than 'sprains' or ligamentous injuries.

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CHAPTER 45
Orthopaedics in the Teenager

CASE 1

A 12-year-old girl was on holidays with her family. When she was on the beach in her swimsuit, her mother noticed that her shoulders were uneven and when she bent forwards the ribs on the right side were prominent. Although she had no pain, she agreed to go for an x-ray of her back.

Q 1.1 What is the diagnosis?
Q 1.2 What did the x-ray show?
Q 1.3 What is the management?

CASE 2

A 14-year-old boy attended the emergency department for the third time in 6 weeks complaining of pain in his left knee, associated with limping. Symptoms were worse after basketball and relieved by rest. Blood tests and x-rays of the knee were normal and a diagnosis of sprained knee ligaments had been made. He had been prescribed anti-inflammatory medication and a knee brace. On this occasion, it was noted that his left hip lacked internal rotation and that the hip went into external rotation during flexion.

Q 2.1 What is the diagnosis?
Q 2.2 What investigations are appropriate?
Q 2.3 What is the management?

CASE 3

Sue, a 14-year-old girl, presents with painful knees. There has been pain in the right knee, then the left and currently both are sore. She has been seeing a physiotherapist for over a year with similar symptoms, including pain, giving way and clicking. Many sets of x-rays had been taken and were reported as normal.

Q 3.1 What is the diagnosis?
Q 3.2 What investigations are appropriate?
Q 3.3 What is the management?

CASE 4

Mary, a 13-year-old girl, complains of pain in her right knee for the past 8 weeks. The pain has been mild and intermittent, but has become more constant, keeping her awake at night. She had physiotherapy for a pulled muscle with some temporary benefit. She agreed to her mother's request to see her general practitioner after she noted a lump on the inner aspect of her thigh, just above the knee.

Q 4.1 What is the differential diagnosis?
Q 4.2 What investigations are needed?
Q 4.3 What is the management?

The teenage years encompass the final period of skeletal growth leading to the closure of the growth plates. The adolescent growth spurt is relatively short but intense, a time of rapid growth in the length of long bones and remodelling of the skeleton to meet the needs of the young adult.

Evaluating musculoskeletal symptoms in teenagers can be difficult. Pain may be referred to the lower limbs from the back and hip pathology frequently presents with knee pain. Teenagers may conceal symptoms and signs from parents. A scoliosis may reach an advanced degree of deformity before being noticed by parents.

Scoliosis

Scoliosis means a lateral curvature of the spine and may be classified as structural or non-structural. Non-structural curves have a cause outside the spine, the most common being a difference in leg lengths. Structural scoliosis is a complex, three-dimensional deformity of the spine, in which a rotational deformity is an important component [Fig. 45.1]. Idiopathic adolescent scoliosis is equally common in both sexes (minor curves are found in up to 4% of the population), but far more girls than boys come to surgery because their curves are more likely to progress. There is often a family history and curves

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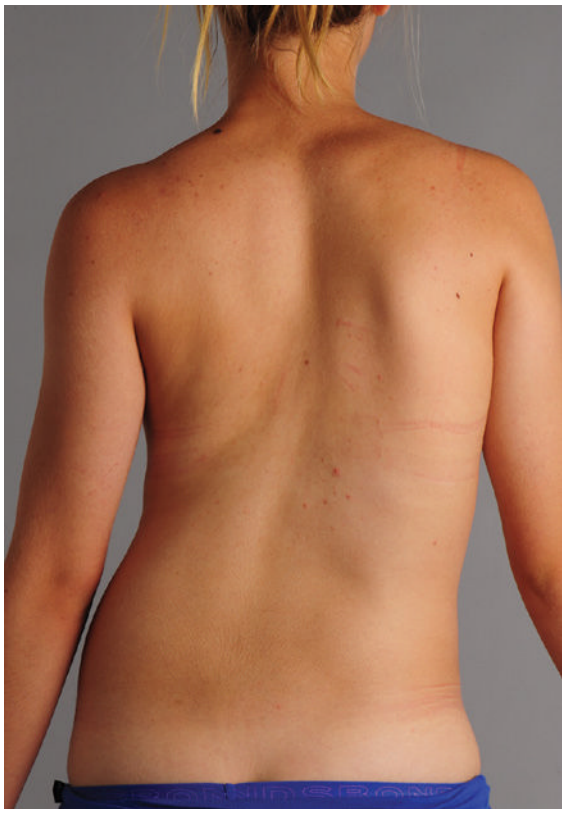

Figure 45.1 Scoliosis. There is a right thoracic scoliosis producing prominence of the right scapula and ribs due to rotation of the vertebral bodies. The left shoulder is lowered and the left waist is increased. The deformity of the spine and chest is more apparent on forward bending.

may progress rapidly during the adolescent growth spurt. Scoliosis is recognised clinically by the 'forward bend test' and confirmed on x-ray. Curve progression should be monitored clinically and by measuring directly from the x-ray. Curves of less than 20° are unlikely to progress, curves of between 20° and 40° may be controlled by bracing and curves of more than 40° may require surgical correction by spinal instrumentation and fusion.

Slipped upper femoral epiphysis

Diagnoses such as 'sprains' and 'pulled muscles' can be dangerous. They lack precision and are often incorrect, a smokescreen for fuzzy thinking, wrong diagnoses and/or incorrect treatment.

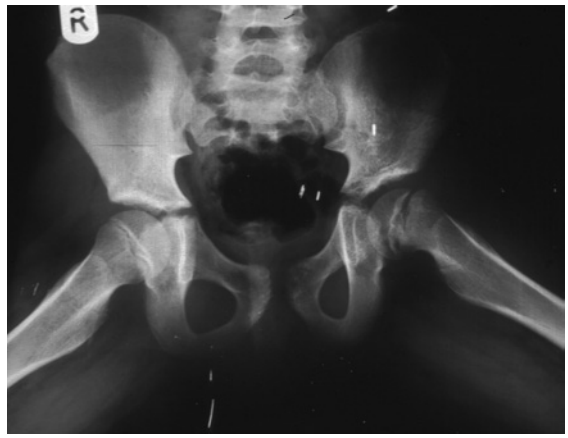

Figure 45.2 Slipped upper femoral epiphysis on x-ray.

There is only one growth plate which may fail under normal physiological loads, the proximal femoral growth plate. During the last 2 years of rapid growth that lead up to the closure of the growth plate, the upper femoral growth plate may slip, allowing posterior displacement of the femoral head in relation to the shaft [Fig. 45.2]. This process may occur in normal teenagers, but is more likely if the loads on the growth plate are increased (e.g. in obesity) or the growth plate is weakened by endocrine disorders, radiation or renal disease.

The time frame of the slipping dictates the clinical presentation. If the slip is acute (or unstable), the adolescent will have severe pain in the hip, will be unable to walk and all movements of the hip will be grossly restricted. The x-ray appearances are usually easily recognised and ultrasonography of the hip will usually show a haemarthrosis, as well as the acute slip. However, if the slipping occurs gradually over a period of many weeks, the presentation can be much more difficult to recognise. The pain, which may be mild and intermittent, is often felt mainly or only in the knee. The adolescent can walk, but there may be an intermittent limp. There is usually a good range of hip motion except internal rotation, which is lost or restricted early in the slipping process, and eventually there will be a characteristic sign whereby the hip rolls into external rotation when it is flexed. Early chronic slips are often difficult to recognise on x-ray and the lateral view is the most sensitive projection. Delay in diagnosis of chronic slips is frequent because of failure to consider that knee pain may be referred from the hip or because a lateral

hip x-ray is not done. Delay in diagnosis may result in continued slipping and a much worse prognosis. Such delays in diagnosis frequently lead to litigation.

Both types of slip require immediate operation. Chronic (stable) slips are pinned *in situ* to prevent progression and have a good outcome if the degree of slip is not severe. The management of acute (unstable) slips is more controversial and the outcome is uncertain. A major operation to openly reduce the severe unstable slip as soon as the slip is recognised is increasingly widely used but only available in specialist centres. Open reduction certainly improves the shape of the proximal femur but unfortunately does not abolish the risk of avascular necrosis.

Anterior knee pain

Knee pain is very common in adolescents and there are many causes. Most knee disorders are self-limiting and are managed by advice and reassurance. However, knee pain may be the presenting symptom of a limb- and life-threatening condition, such as an osteosarcoma. Not every patient with knee pain should have a bone scan or MRI, as the history and examination are the primary tools to distinguish the serious from the trivial.

Anterior knee pain is a common clinical syndrome in adolescents, especially girls, with up to 30% affected. Pain is usually intermittent, is located around or behind the patella and is made worse by exercise and relieved by rest and simple analgesics. It is often bilateral, but one side may be more symptomatic than the other.

Examination is usually unremarkable apart from patellar tenderness and crepitus. X-rays are normal but are done to exclude more serious pathology. Arthroscopic examination of the knee may reveal changes in the retropatellar cartilage, which are sometimes called chondromalacia. However, the relationship of these changes to pain is not clear. Some of the most painful knees are normal on arthroscopic examination, and the changes of chondromalacia may be present without pain. Anterior knee pain can be considered to be an overuse syndrome affecting the immature retropatellar cartilage, which is ultimately benign and self-limiting. It does not lead to arthritis or any other sequelae in later life.

Management is conservative: explanation, education and simple measures to control symptoms, including analgesics, restricting activities which provoke symptoms, hamstring stretching and quadriceps strengthening. Neither arthroscopic examination nor surgery should be advised routinely.

The differential diagnosis includes osteochondritis dissecans of the tibial tuberosity (Osgood–Schlatter's disease), bipartite patella, meniscal tears, plica syndrome, patellar instability and referred pain from the hip.

Bone tumours

The principal symptoms of a bone tumour are pain and the presence of a mass or lump. Slow-growing bone tumours often present as a mass, with pain as a less prominent feature. The most common bone tumour is the benign osteocartilaginous exostosis, usually abbreviated to 'exostosis' [Fig. 45.3]. Exostoses are benign bone tumours which are usually solitary and are found near

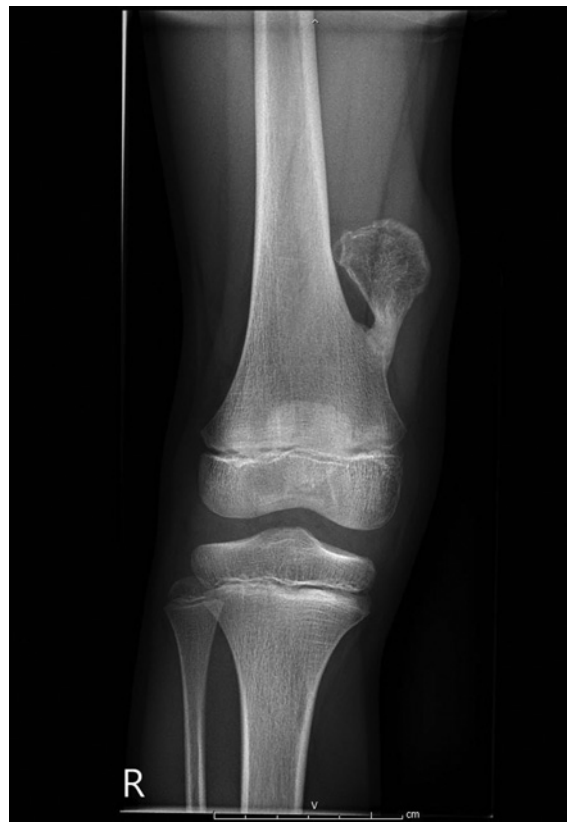

Figure 45.3 X-ray of exostosis of the distal femur.

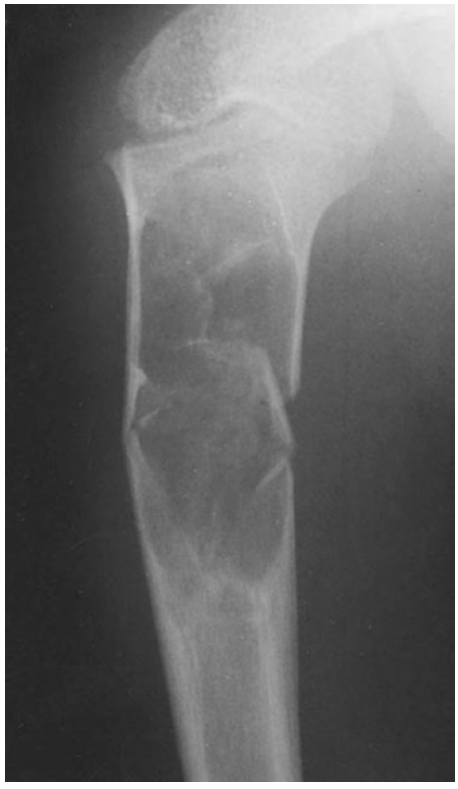

Figure 45.4 Solitary bone cyst of the humerus. Note the fractures of the thinned cortex.

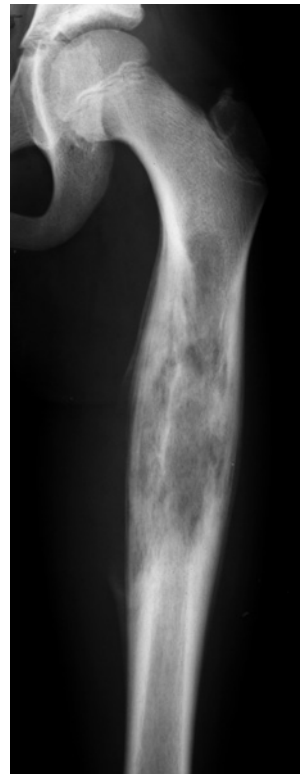

Figure 45.5 Ewing's sarcoma involving the proximal half of the diaphysis of the femur, with layers of new bone and fusiform swelling of the soft tissue.

the ends of long bones because they originate from aberrant cartilage cells from the growth plate. Some children have multiple exostoses, and this may be inherited as an autosomal-dominant condition, 'hereditary multiple exostoses'. The bony lumps are noticed incidentally or after minor trauma. They grow slowly until skeletal maturity or a little later. Clinically and radiologically they are benign in behaviour, although incomplete excision may lead to local recurrence. Excision is advised if the lumps are symptomatic or the diagnosis is in doubt.

A solitary bone cyst usually presents as a pathological fracture of the proximal humerus or femur [Fig. 45.4]. Often the fracture stimulates healing of this benign lesion.

Painful benign bone tumours include osteoid osteoma and osteoblastoma. These are small tumours which present with night pain, characteristically relieved by aspirin. Diagnosis may be delayed because the tumours are small and are not always easily demonstrated on x-ray. Excision is usually performed by CT guidance using a trocar passed percutaneously into the nidus.

Malignant bone tumours usually present with well-localised pain, which is progressive, disturbs sleep and is not easily or fully relieved by rest or analgesics. The rapidly growing ends of long bones are most likely to be affected, including around the knee (the distal femur and proximal tibia) and the proximal femur and the proximal humerus. The two most common primary malignancies of bone, osteosarcoma and Ewing's sarcoma [Fig. 45.5], are most common in childhood and adolescence.

Diagnosis and staging is achieved by imaging studies that may include plain x-rays, technetium bone scans, computed tomography (CT), MRI scans and positron emission tomography scans (PET scans) followed by biopsy, in all cases. Although there are features which suggest malignancy on x-ray (e.g. bone destruction by the lesion, a large soft tissue mass and a marked periosteal reaction), these features can be mimicked by benign bone tumours, bone dysplasias and infection.

The prognosis for saving both life and limb in adolescents with primary bone tumours has improved

dramatically with limb salvage surgery and chemotherapy. The tumour must be completely excised with a margin of healthy tissue, and the limb reconstructed, whenever possible, using a variety of techniques including bone grafts (autografts and allografts) and endoprosthetic replacement. Chemotherapy is usually commenced after the diagnosis has been established by biopsy but before the tumour excision surgery.

KEY POINTS

- Scoliosis is common (4%) in both sexes, but is often more severe in girls.
- Structural scoliosis persists when 'touching the toes' (forward bend test).
- 'Knee pain' in adolescents is commonly referred from the hip (e.g. slipped upper femoral epiphysis).
- Pain around the patella is common in girls from overuse affecting immature retropatellar cartilage.
- A mass (+pain) in the limb needs urgent investigation for bone tumour.

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CHAPTER 46
The Hand

CASE 1

A 3-year-old has a fixed flexion deformity at the interphalangeal joint of the thumb.

Q 1.1 What is the diagnosis?
Q 1.2 Is splinting indicated?
Q 1.3 Is surgery required?

CASE 2

A 4-month-old child of a diabetic mother has weakness of the right arm. Hand movement has recovered following a difficult vaginal delivery. However, elbow flexion and shoulder control have not yet appeared.

Q 2.1 What are the diagnosis, cause (aetiology, pathology and anatomy) and treatment?

Reconstructive surgery of the hand in a child has a better prognosis than for similar conditions in adults. Injuries should be repaired immediately, while malformations are often corrected in the first year of life or at least prior to starting school. The stiffness that adults experience after upper limb surgery is seldom a problem. The hand needs to be protected, immobilised and maintained in an elevated position until sufficient healing has taken place to allow the child to play and use the hand without restriction. A plaster slab or cast must be applied in such a way that the child cannot easily remove it. It must be comfortable and immobilise the hand, the wrist and often the elbow in a safe position. In most cases, the tips of the digits should be able to be inspected to ensure that vascular compromise does not occur. Elevating the arm in a well-designed sling under the clothing is the best way of ensuring that the limb is protected. In a child, a plaster should be maintained for 2 weeks after simple suturing, 3 weeks after skin grafting (e.g. syndactyly release) and almost 4 weeks following tendon surgery. Ongoing protective splinting may then be required.

Congenital anomalies

These are common and varied. A classification based on embryological aberrations is useful in describing and recording malformations [Box 46.1]. Unfortunately, many unrelated conditions, such as trigger thumb, syndactyly, clinodactyly and so on, are classified as a failure of differentiation. Anomalies may be confined to the hand or be a local manifestation of a generalised condition, such as arthrogryposis multiplex congenita. Some deformities, for example, certain forms of syndactyly, are strongly familial; others are sporadic, with no known cause. Other associated congenital conditions should be looked for. Some are potentially lethal, such as when radial club hand coexists with cardiac and haemopoietic abnormalities.

Management

A child with a hand anomaly (congenital *difference*) may have functional and/or cosmetic impairment. They will, however, learn to use it with dexterity, provided certain basic anatomical elements are present that provide

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BOX 46.1 Classification of malformations of the hand

Failure of formation
- Transverse congenital amputations
- Phocomelia
- Radial club hand
- Cleft hand

Failure of differentiation
- Syndactyly
- Clinodactyly
- Camptodactyly
- Clasp thumb

Duplication
- Polydactyly
- Triphalangeal thumb

Overgrowth
- Giantism
- Macrodactyly

Undergrowth
- Hypoplasia
- Brachydactyly
- Symbrachydactyly

Constriction ring syndrome
- Intrauterine amputations
- Congenital constriction bands
- Acrosyndactyly

Congenital
- Skeletal abnormalities (achondroplasia)

rudimentary pinch and/or grasp. The psychological effects on the parents need to be managed.

Syndactyly

This affects 1 in 2000 births and may be incomplete or complete (to the fingertip) and simple or complex (fused bones). It may be part of a syndrome, such as the Poland or Apert syndrome. Family history is positive in up to 40% of cases; inheritance is dominant but there is reduced expression and penetrance. Early surgery (in the first 6 months) is indicated when border digits, particularly the thumb, are involved, leading to growth disturbance in digits of unequal length [Fig 46.1].

Clasp thumb

This may be due to a congenital trigger thumb caused by a swelling within the flexor pollicis longus tendon impinging on the fibrous flexor sheath. Alternatively, it may represent a weakness in the extensor muscles, which can be improved by splinting initiated in the first month of life. A deficiency of skin on the palmer surface may be improved by splinting but may need correction using skin flaps or grafts.

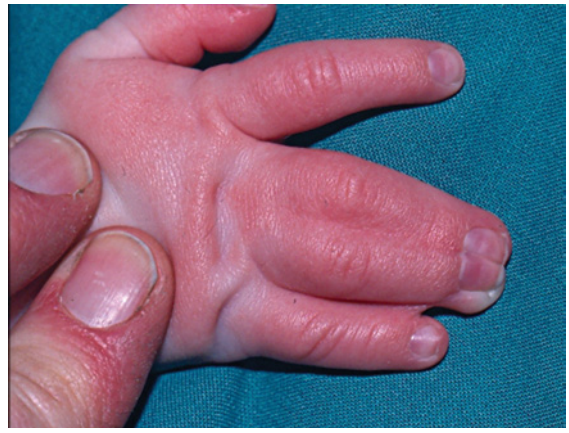

Figure 46.1 Syndactyly, complete and complex (fused) distal phalanges between ring and middle fingers, with simple and almost complete syndactyly between the ring and little fingers.

Trigger thumb

Trigger thumb is a common and often bilateral abnormality. It is sometimes identified at birth but is usually found in children up to 5 years of age. There may be a family history of this condition. The primary lesion is thickening in the flexor tendon of the thumb associated with narrowing of the tendon sheath at the metacarpophalyngeal joint [Fig 46.2]. Once the affected digit is flexed, extension is restricted by impingement of the thickened tendon proximal to the narrowing. Usually, children present with fixed flexion deformities of the interphalangeal joint. Clinically, the thickening in the tendon can be palpated at the level of the metacarpal head in the palm. Treatment involves longitudinal incision of the tendon sheath to allow full excursion of the thickened tendon through the area of narrowing. Splinting is not effective.

Distal arthrogryposis

This condition, which may be inherited, is associated with skin deficiency, underdeveloped flexure creases and weakness associated with underdevelopment of the musculature. The thumbs may be clasped and the digits ulnar deviated. An intense programme of splintage, initiated in the neonatal period, produces quite dramatic improvements.

Camptodactyly

This condition affects 1 in 100 hands and causes a flexion contracture, particularly of the proximal interphalangeal joint of the little finger, and is due to an

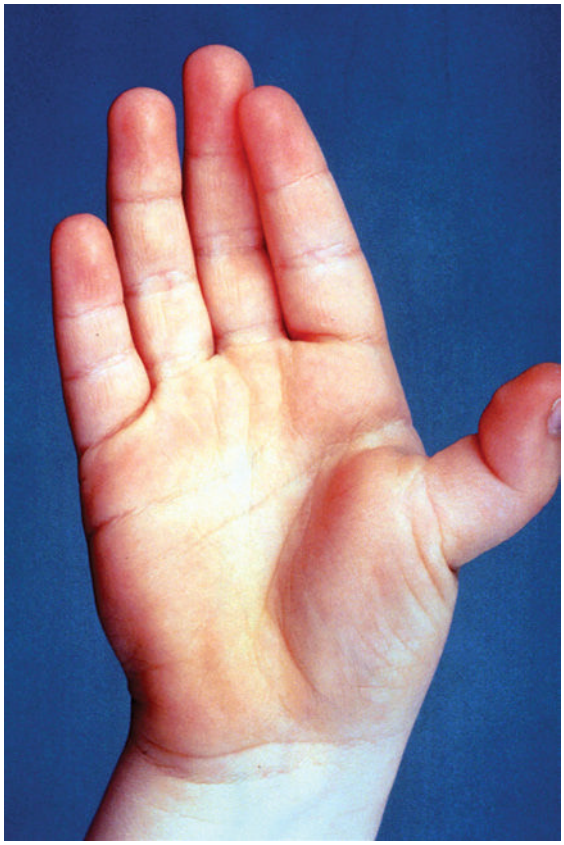

Figure 46.2 Right trigger thumb presenting as a fixed flexion deformity.

imbalance of anomalous muscles, tendons and fascial structures. In severe cases, the joint becomes deformed. The results of surgery are frequently disappointing, but early aggressive splinting can be effective.

Clinodactyly

This condition affects 1 in 100 hands. Radial deviation, particularly of the little finger, is often due to a delta-shaped middle phalanx associated with an anomalous C-shaped cartilage growth plate. Resection during childhood of the abnormal cartilage bridging the radial border of the phalanx can allow the finger to grow almost normally.

Polydactyly

Radial (preaxial) polydactyly involving the thumb occurs in 1 in 1000 births, particularly in Asia. This condition is sporadic and unilateral, except when one of the duplicated thumbs is triphalangeal in which situation the condition is often bilateral and is inherited as an autosomal dominant condition. Ulnar (post-axial) polydactyly occurs in 1 in 300 in Africans, and in 1 in 3000 in Caucasians. This condition is often bilateral and inherited in an autosomal recessive pattern and may be part of a syndrome.

Symbrachydactyly

This is a sporadic unilateral condition with hypoplasia of the digits that are short and stiff (aplasia of bones and joints). In several cases, the digits are represented by soft tissue nubbins. Microsurgical transfer of toes can significantly improve function in some patients. Others may benefit from free grafting of toe proximal phalanges into redundant soft tissues in the first 2 years of life.

Obstetric brachial plexus palsy

This is due to a traction injury associated with a difficult delivery frequently in babies weighing in excess of 4 kg. Injuries to the upper (C5, C6+7) nerve roots or the upper (+middle) trunk of the brachial plexus are known as Erb's palsy. These improve in 90% of cases. If there is no recovery after 3 months, particularly if all nerve roots are involved, then exploration and reconstruction with nerve grafts should be contemplated within the first year of life. The results of reconstruction are far better than those achieved in adults with brachial plexus trauma.

Injuries to the hand

The repair of injuries to the hand differs little from that in adults, except that the results are generally better. Absorbable skin sutures should be used. Restoration of function is more rapid and more complete, and prolonged hand therapy is rarely necessary.

Failure to diagnose the extent of the injury when the child is first seen leads to unsatisfactory results and the need for later reconstruction. Subjective tests for nerve injuries are of little value, and cooperation during a detailed examination is unlikely. Injuries to tendons and nerves may be overlooked until loss of function is noted by parents, weeks or months later.

Awareness of the likelihood of these injuries associated with lacerations of the hand is essential. Abnormal posture of the fingers with the hand at rest suggests possible

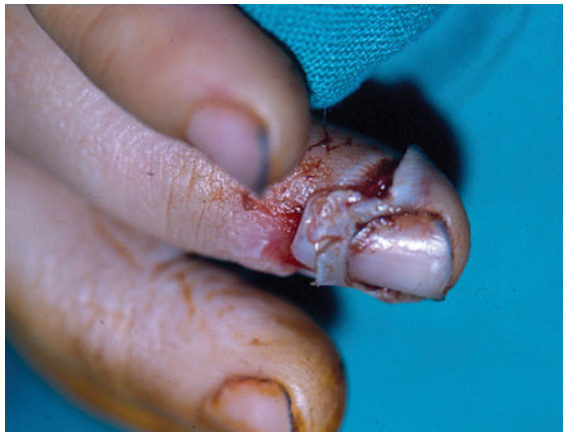

Figure 46.3 Finger crushed in a door.

tendon injury. Most injuries are cuts or crushing injuries. Any wound in a child that cannot be adequately examined, particularly if it overlies an important structure, must be explored under general anaesthesia. Any damage to tendons and nerves can be repaired primarily under ideal conditions. No matter how minor the injury is, a full plaster and sling should be used to promote uncomplicated healing.

Injuries to fingertips

Crushing and slicing injuries with loss of a part of the fingertip are common in childhood [Fig. 46.3]. In most crush injuries, the tissue remaining after judicious debridement and thorough lavage is viable. Any associated fracture of the distal phalanx should be reduced and in some cases pinned with a fine Kirschner wire. If possible, a portion of the fingertip should be left exposed so that the circulation can be observed in the early postoperative period.

In slicing injuries with skin loss, some form of primary closure with a skin graft or local flap may be indicated. In younger children, the cross-sectional area of the fingertip is small, healing is rapid, and the results of healing by second intention can be excellent. Amputations at the level of the mid-distal phalanx can often be microsurgically replanted by anastomosis of preferably two blood vessels. More distal amputations or unreplantable crushed or avulsed parts may be, after defatting, replaced as a graft with moderate success.

KEY POINTS

- Congenital hand anomalies may be complex and require sophisticated reconstruction, but prognosis for function is good.
- A toddler with a fixed flexion deformity of the thumb has *trigger thumb*, needing surgery.
- Injuries to the hand need absorbable sutures and plaster immobilisation.
- Lacerations may need exploration under general anaesthetic (GA) to exclude tendon, nerve or vessel injury.

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PART VIII
Chest

CHAPTER 47
The Breast

CASE 1

An 8-year-old girl presents with a slightly tender lump behind the left nipple, which has been present for 2 months. There has been neither a growth spurt nor menarche.
Q 1.1 What are the problem and its natural history?
Q 1.2 When is further investigation or surgery indicated?

CASE 2

A 2-week-old baby has bilateral enlargement of the breast buds with milk production. The right breast is painful, red and tender.
Q 2.1 What are the diagnosis and treatment?
Q 2.2 What is the gender of the baby?

In children, there are virtually no serious conditions involving the breast in either sex, but there are a number of minor conditions that may give rise to anxiety or inconvenience, and some of these require treatment. Malignancy is exceedingly rare.

Absent breasts and multiple nipples

The absence of the breast (amastia) is a rare, and usually unilateral, anomaly that is frequently part of a regional dysplasia, which also affects the pectoral muscles and subjacent ribs (Poland syndrome; Chapter 48). Breast tissue with the absence of a nipple (athelia) may occur in ectopic sites such as accessory breasts.

Multiple nipples, which are also rare, may occur anywhere along the embryological mammary line, which is a curved line from in front of the anterior fold of the axilla down to the groin and medial thigh. It is even rarer to have a supernumerary breast, which is usually in the axilla. This may be removed by surgery.

Neonatal enlargement

Galactorrhoea

Transplacental passage of lactogenic hormones may lead to hyperplasia and the secretion of breast milk in newborn babies of either sex. The enlargement usually lasts a week if left alone, but attempts to empty the breast by massage may actually prolong and increase milk production. Unusually overactive secretion may be stopped within 24 h by oral oestrogens, but this is rarely necessary. The engorgement predisposes to infection, which is potentially serious although not particularly common.

Neonatal mastitis

If an abscess forms, it should be aspirated or drained by a very small incision placed peripherally (at the edge of the areola) in girls to minimise disruption of the canaliculi beneath the nipple. Since the entire breast is no larger than the areola, extreme care is required to avoid unintentional mastectomy or physical damage to the breast bud. Damage to breast tissue may lead to breast deformity in adult life.

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Precocious puberty

This is very uncommon but may occur in girls as early as 12–18 months of age. Menstruation and bilateral hyperplasia of the breasts should always raise the possibility of an underlying cause, such as an ovarian (or adrenal) tumour or an intracranial lesion. However, in the constitutional type, no cause is found, and the possibility of dwarfism from excess production of oestrogens should be investigated. Early referral to a paediatric endocrinologist is advised.

Premature hyperplasia (premature thelarche)

This is probably the most common minor physiological aberration and, when unilateral, may present a diagnostic problem. The usual finding is the development of one breast in a girl, sometimes as young as 5 years, though more commonly 7–9 years of age.

The presenting feature is a firm discoid lump, 1–2 cm in diameter, situated symmetrically and concentrically beneath the nipple. It is initially symptomless and found accidentally, although it may become mildly tender, perhaps in part due to repeated palpation and to anxiety. There are no other signs of puberty, which develops at the normal time.

The clinical signs are so diagnostic that no confirmation by other means is required. This is not precocious puberty, for the menarche will occur at the normal age. The affected breast may return to normal, but the swelling commonly remains static, and the same changes frequently appear in the opposite breast within 3 to 12 months: both then remain static without further increase in size until puberty. Reassurance and explanation are all that are required. Biopsy should not be performed, for it may damage the breast bud.

Pubertal mastitis

This occurs in boys as well as girls. In girls, there are some tenderness, discomfort and a granular texture on palpation; and serous fluid may be expressed from the nipple. One breast is often affected more than the other. It is a temporary phase of development and no treatment is required.

In boys, the discoid, subareolar lesion as described in premature hyperplasia occurs in one or both breasts at about 12–14 years. No treatment is necessary.

Gynaecomastia

Gynaecomastia is defined as male breast enlargement due to benign proliferation in breast glandular tissue. This occurs in several conditions, perhaps most commonly in a spurious form in obese preadolescents, when it is formed of fat alone. Gynaecomastia affects up to 60% of boys in adolescence, but resolves spontaneously in 75% cases within 2 years. Only 5% of cases persist into adulthood.

In thin boys, the possibility of a disorder of sexual development should be considered and requires close examination of the genitalia. Chromosomal and hormonal studies, urethroscopy, biopsy of the gonads or even laparoscopy may also be indicated (Chapter 10). The combination of gynaecomastia and small testes suggests Klinefelter syndrome.

Other pathologic causes for gynaecomastia should be considered for persistent cases and/or cases arising outwith the typical neonatal and peri-puberty age groups. Gynaecomastia may arise as a side effect of various medications, most commonly oestrogen therapy for some other condition. Uncommonly, hormone-producing tumours (e.g. testis, liver, adrenal) may cause gynaecomastia.

In most cases, no cause can be found, and if the enlargement is of sufficient magnitude to cause embarrassment, simple mastectomy may be justified. The standard curved submammary incisions should not be used, for the scar may simulate the contour of a breast even after it has been removed. Preferably, a lower half-circle peri-areolar incision of the breast provides sufficient access to remove the breast tissue.

KEY POINTS

- Premature thelarche, without other signs of precocious puberty, is common and requires no treatment.
- Neonatal lactation in response to maternal hormones resolves spontaneously, but predisposes to secondary infection.
- Transient gynaecomastia is common in boys in early puberty.

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CHAPTER 48
Chest Wall Deformities

CASE 1

Bruce is a 10-year-old boy whose parents are becoming increasingly concerned about the appearance of his deformed chest, which is making him reluctant to socialise and play sport. He is embarrassed about his appearance and wants it corrected.

Q 1.1 Is the deformity significantly limiting his cardiorespiratory performance?
Q 1.2 What investigations are required?
Q 1.3 What advice would you give him and his parents?

Deformities of the chest wall often appear early in life but more commonly become apparent at the start of the pubertal growth spurt. They are classified as follows:

1 Primary depression deformities of the sternum (pectus excavatum, also known as funnel chest)
2 Primary protrusion deformities of the sternum (pectus carinatum, also known as pigeon chest)
3 Deficiency deformities of the chest wall (e.g. Poland syndrome)
4 Failure of midline sternal fusion.

Aetiology

The deformity is a primary condition, except in a few of the protrusion deformities where it may be secondary to chronic asthma, congenital heart disease, a massive lung cyst, diaphragmatic hernia, an anterior mediastinal lesion such as a teratoma or, rarely, hydatid cysts of the liver. Rarely, depression deformities may be seen in Marfan syndrome, homocystinuria and congenital laryngeal stridor with inspiratory retraction.

A hereditary factor may generally be established but is of mixed penetrance or recessive and consequently may miss one or more generations.

Depression deformity (pectus excavatum or funnel chest)

Clinical features

Pectus excavatum (funnel chest) is four times as common as pectus carinatum (pigeon chest). It is more frequent in males, and may be symmetrical or asymmetrical. Affected children are usually thin: muscular and ligamentous laxity may be marked, and posture tends to be poor. The sternal depression is typically maximal at the sterno-xiphisternal junction but may extend up the sternum to any level.

When most of the sternum is affected, the depression is shallow and the deformity may be described as the *saucer* type. A localised depression is compatible with a well-developed upper chest, and the very localised *funnel* may be referred to as the *cup* type.

In asymmetrical lesions, the sternum is rotated, usually from right to left around its longitudinal axis, producing prominence of the costal cartilages on one side and recession of those on the other.

A shallow sulcus may extend laterally on each side from the sterno-xiphisternal junction, a sulcus that corresponds to the attachment of the diaphragm to the lower costal cartilages (Harrison's sulcus). The costal

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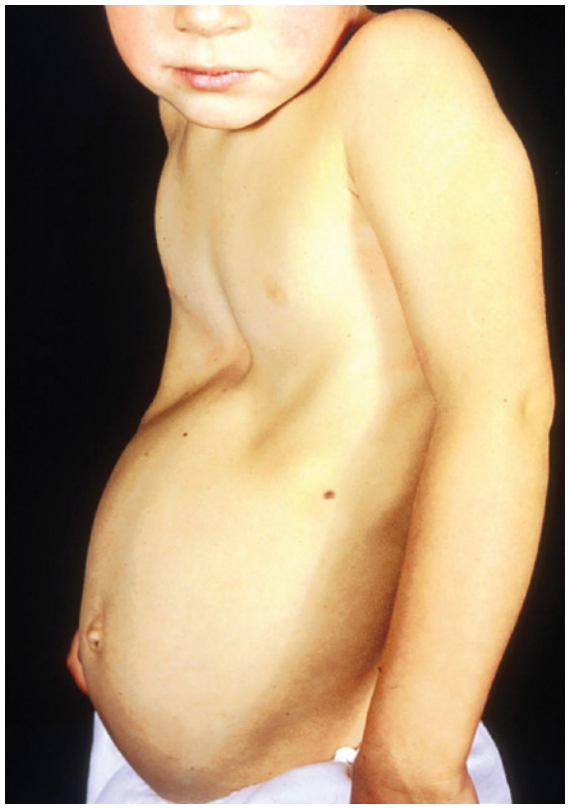

Figure 48.1 Depression deformity. An extensive deep depression centred on the xiphisternal junction. Note the lateral sulcus, in profile, on the right side.

margin on each side may become generally everted or protrude as a boss (*costal flaring*). Postural kyphosis is common, and scoliosis not infrequent.

The deformity may be of any degree of severity, from being barely detectable to one in which the lower sternum seems to almost touch the front of the vertebral column [Fig. 48.1].

A pectus excavatum may be present at birth or become apparent at any age up to 16 years. There is a group of males in which it develops rapidly at about 14 years of age, when rapid growth in height is occurring.

When the deformity is severe at birth, there is paradoxical retraction of the sternum on inspiration. This ceases at about the age of 4 years and is replaced by orthodox respiratory movement of subnormal amplitude. In the great majority of cases, there is a tendency for the deformity to progress until growth ceases, but in others, it becomes less severe.

Symptoms

These are largely related to psychological effects and occasionally to cardiac function.

Psychological features may become apparent from an early age. The child resents being *different* and, when undressed for examination, brings the arms together to try to conceal the shape of the chest. Older children may refuse to go swimming for fear of becoming an object of attention. Comments from schoolmates may be unkind. The psychological effects may completely alter the child's personality and affect social development.

Diminished exercise tolerance is uncommon and confined to those with the most severe deformities. Pain may be mentioned, but has no obvious organic basis. The deformity is not responsible for any supposedly increased tendency to respiratory infections or asthma.

The progression and severity of the condition, and its significance to the patient and his parents, may best be assessed by serial examinations at yearly intervals. Photographs recording the deformity at each visit may be useful to objectively document the degree of the pectus excavatum, as well as any change that occurs over time.

Investigations

Investigations are often not necessary in children and adolescents with pectus excavatum. This is because they are usually asymptomatic, and the indication for intervention is usually based on the clinical appearance and its associated psychological impact.

A CT scan of the chest can be undertaken to assess the deformity of the sternum and to measure the Haller Index. The Haller Index is a measurement of the internal transverse diameter of the thorax divided by the shortest anteroposterior distance, as measured from the inner aspect of the sternum to the closest vertebral body. The Index is usually around 2.5, with anything above 3 seen as a significant deformity.

Significant debate exists about the cardiopulmonary effects of pectus excavatum and its relationship to the Haller Index. A number of reports have suggested a reduction in total lung capacity, vital capacity and forced expiratory volume in 1 s with significant depression anomalies. Echocardiography has also shown a reduction in stroke volume and cardiac output in some cases as well as shows evidence of mitral valve prolapse in up to 25% of cases. In cases where there is concern about reduced exercise performance, then pulmonary function tests and echocardiography can be arranged.

In our experience though, these have usually been normal even with quite significant anomalies. If there is a clinical concern about Marfan syndrome, then referral to cardiologists, ophthalmologists and clinical geneticists can be arranged.

Treatment

The deformity cannot be improved by any form of exercises or brace, though there are some reports suggesting success with a vacuum bell applied to the anterior chest for a number of hours each day. Encouragement of participation in physical activity helps some children to become less self-conscious about lesser degrees of depression deformity. In selecting patients for operative correction of the deformity, it is important that the patient wants to have the correction.

Operative correction is often performed as the child enters puberty. Most commonly, a *Nuss* procedure is performed; this involves placing a curved metal strut behind the sternum using a minimally invasive technique. The operation causes considerable discomfort, and patients typically require a thoracic epidural for pain management in the early post-operative period.

Protrusion deformities (pectus carinatum or pigeon chest)

High protrusion

This may be associated with a deficiency deformity. The sternum is angulated forwards at the level of the third sternochondral junction, and below this point, it recedes as a depression. There may be *pinching in* of the lower costal cartilages. The deformity tends to increase with age and never regresses spontaneously.

The radiological findings are unique, for there is osseous fusion of all the synchondroses of the sternum, which probably occurs before the age of 1 year.

Low protrusion

These may be secondary to intrathoracic pathology, particularly chronic asthma. In a few of those in whom it is a primary condition, there is a tendency for spontaneous improvement, which is usually apparent by the age of 8 years; otherwise, there is a general tendency for the deformity to increase until growth ceases.

The maximal protrusion is at the sterno-xiphisternal junction or a little higher, and there is usually some pinching in of the lower costal cartilages [Fig. 48.2]. Prominence of the costal cartilages on each side near their junction with the sternum may be present, forming a median trough that, though locally depressed, is still part of the sternal protrusion. Rotation of the sternum producing an asymmetrical deformity is not uncommon.

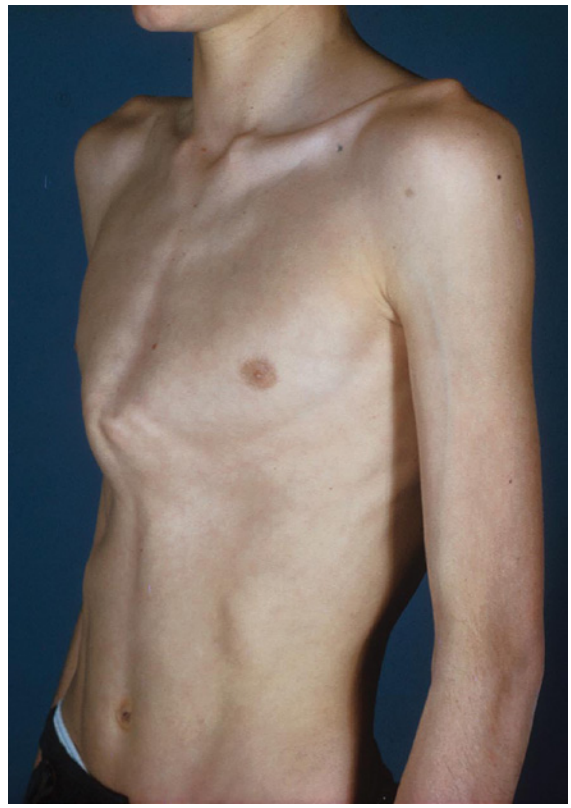

Figure 48.2 Low protrusion deformity maximal just above the sterno-xiphisternal junction.

Radiography is necessary to confirm the degree of protrusion and to exclude the presence of any intrathoracic condition that might be contributory. The symptoms are related entirely to the cosmetic defect, or to any underlying condition.

Treatment

Secondary deformities usually resolve spontaneously after elimination of the underlying cause. Occasional lesions also show a tendency for spontaneous improvement. In the first decade of life, repeated observation will indicate the trend. Many patients, however, present

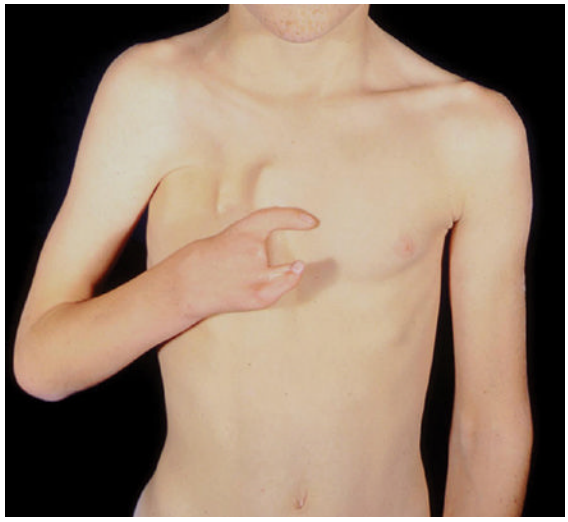

Figure 48.3 Deficiency deformity. Poland syndrome: absent nipple, areola and pectoral muscles with hypoplasia of the chest wall and syndactyly and symbrachydactyly.

for the first time after puberty, and these usually progress until growth ceases.

The management of patients with pectus carinatum has been revolutionised by the introduction of orthotic braces to provide continuous pressure to the protruding chest wall. Patients are assessed with radiology and 3D imaging to create an individualised brace that is moulded for their deformity. The brace is worn for 23 h per day, with a 1 h break to limit skin trauma and increase compliance. The initial (or corrective) phase takes a number of months and is followed by a maintenance phase that will typically last 6 months. The end results are usually excellent and operative correction is now rarely indicated.

Deficiency deformities

In the usual type, the pectoral muscles are absent on one side, and there is a variable degree of hypoplasia of the underlying ribs and costal cartilages. The third and fourth cartilages may be deficient anteriorly, with some paradoxical respiratory movement visible through the chest wall, although this is rarely of any clinical significance. All elements of the breast may be absent, but usually the nipple and areola are present. Hypoplasia of the upper limb on the affected side and syndactyly may occur (Poland syndrome; Fig. 48.3). The sternum in these patients may show a high protrusion deformity of cosmetic significance. The dominant problem, however, is the soft tissue deficiency.

Treatment

Operative intervention is only rarely indicated for filling in the bony chest wall deficiency or for correction of sternal protrusion. Muscular flaps, utilising the *latissimus dorsi*, may be used to provide soft tissue bulk, to be followed in girls by augmentation mammoplasty. It is more difficult to replace absent tissues than it is to reorganise disordered tissues; despite this, much can be done to improve the appearance in these children.

KEY POINTS

- Pectus excavatum is common in boys, usually idiopathic and maximal at the xiphisternum; symptoms are psychological.
- Pectus excavatum is treated, if required, in early adolescence by the Nuss procedure (minimally invasive insertion of struts).
- Protrusion deformities may be secondary to a thoracic disorder or deficiency deformity.

Further reading

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CHAPTER 49
Lungs, Pleura and Mediastinum

CASE 1

Jemma, a 3-year-old, became unwell with a viral upper respiratory infection. Forty-eight hours later, she became very sick with high fever, lethargy, cough and shortness of breath, eventually becoming cyanotic.

Q 1.1 What could be the problem?

Q 1.2 How is it best treated?

CASE 2

Adrian is 16 years old, athletic and tall for his age. He developed sudden severe chest pain and shortness of breath, not relieved at all by his usual asthma medication.

Q 2.1 What is the diagnosis and its management?

While the principles of diagnosis and treatment of pulmonary disease in children are similar to those of adults, there are additional aspects that need to be considered:

1. The respiratory passages are small, and encroachment upon the lumen by exudate or oedema may cause airway obstruction earlier than anticipated.
2. The cough reflex is relatively ineffective at clearing mucus or exudate in infancy, and retention of secretions may have serious consequences.
3. Obstruction may allow infection to supervene and lead to patchy or lobar collapse.
4. Hypoxia develops rapidly in neonates, and increased respiratory effort increases the consumption of oxygen; the vicious circle may culminate in respiratory failure (the most common cause of arrest in paediatrics).
5. Disturbances of intrathoracic pressure relationships are tolerated poorly, and a tension pneumothorax may cause irreversible cardiorespiratory failure, particularly if there is a pre-existing lung disease.

With few exceptions, for example, hydatid disease, operations on the lungs and pleura in childhood are of two kinds: the provision of pleural drainage and the resection of pulmonary tissue.

Resection, often involving a lobectomy, is required for a variety of developmental anomalies, including congenital lobar emphysema, hamartoma, sequestrated lobe, and rarely bronchiectasis. Metastases from osteosarcoma or Wilms tumour may also warrant resection.

Inhaled foreign bodies sometimes produce few symptoms. The final diagnosis of an intrabronchial foreign body occasionally is made after resection of a diseased segment of the lung.

Pleural drainage may be necessary in three conditions:

1. Pneumothorax, for example, neonatal pneumothorax (Chapter 4) or traumatic pneumothorax (Chapter 38)
2. Empyema (see following sections)
3. Haemothorax, an unusual condition because of the rarity of major thoracic injuries in childhood

Aspiration of the pleural cavity may be used diagnostically to obtain pleural fluid for microscopy and culture or occasionally as a preliminary step in providing immediate continuous drainage of an empyema by means of an intercostal catheter.

Staphylococcal pneumonia

This occurs usually as a complication of a viral infection of the upper respiratory tract [Fig. 49.1].

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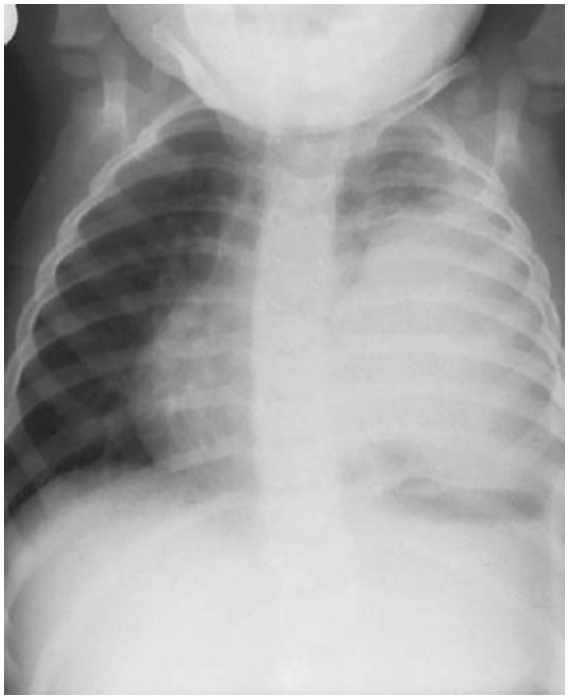

Figure 49.1 Staphylococcal pneumonia involving the left haemithorax

Diagnosis

Although chest x-rays make the diagnosis with a high degree of certainty, identifying the organism responsible is the absolute criterion. When *Staphylococcus aureus* is isolated from a throat swab or sputum, and typical changes are present in lung radiographs, the diagnosis is at least presumptive. The development of a pleural complication and culture of *Staphylococcus* from a specimen of pus from the pleural cavity confirm the diagnosis.

The diagnosis may be delayed at presentation because the clinical signs suggest upper respiratory tract infection and the pathological changes initially may be confined to this area. Progression to pneumonia may not be recognised immediately. The baby or young child may become worse rapidly for the following reasons:

1. Septicaemia and a shock-like state of collapse
2. A collection of pus:
   - Intrapleural (a parapneumonic effusion becoming an empyema, or pyopneumothorax, when both pus and air are present)
   - Intrapulmonary (single or multiple lung abscess)
3. An air leak under tension at one or more of the following three sites:
   - Intrapleural (pneumothorax)
   - Intrapulmonary (pneumatocele)
   - In the mediastinum (pneumomediastinum)
4. Obstruction of the airways

Treatment

Intravenous antibiotics are given, and until the results of culture and sensitivity tests are known, an effective combination is flucloxacillin and gentamicin. Serial x-rays are obtained because of the rapid evolution of the pathological changes on radiology.

Complications and sequelae

Suppurative pericarditis, meningitis, septicaemia and osteomyelitis may complicate staphylococcal pneumonia, but are uncommon when appropriate antibiotics are given early and in adequate dosage. Empyema is the complication that is likely to require operative intervention.

Empyema

The incidence of parapneumonic effusions and empyema appears to be increasing in many Western societies [Box 49.1]. It is more common in boys and during early childhood.

Aetiology

Pleural effusions are usually unilateral and are a complication of acute bacterial pneumonia. Initially, the effusion is clear (parapneumonic effusion), but it subsequently becomes purulent (empyema) and loculated. *Staphylococcus aureus* is the predominant organism, but other causative pathogens include:

- *Streptococcus pneumoniae*
- *Haemophilus influenzae*
- *Mycoplasma pneumoniae*
- *Pseudomonas aeruginosa*

Clinical presentation

There are two patterns of presentation:

1. This occurs in a child who presents with symptoms of pneumonia (e.g. fever, cough, breathlessness, poor appetite, abdominal pain, lethargy and malaise) but is more unwell than would be expected with simple

Box 49.1 Definitions pertaining to pleural infection

Parapneumonic effusion
Pleural fluid that collects between the visceral and parietal pleura as a result of underlying infection (pneumonia)

Empyema
The presence of pus in the pleural space

Exudative phase
The earliest phase in which the underlying pneumonia causes accumulation of clear fluid within the pleural space

Fibropurulent phase
The second phase, where deposition of fibrin in the pleural space leads to septation and loculation. The fluid thickens (*complicated parapneumonic effusion*) and contains many white cells

Organisational phase
The final phase where fibroblasts infiltrate the pleural cavity. The intrapleural membranes become thick and non-elastic (*the peel*), which may prevent lung re-expansion and impair lung function

pneumonia alone. They often have pleuritic chest pain and decreased chest expansion on the affected side. There is dullness to percussion and reduced or absent breath sounds. The effusion is obvious on a chest x-ray.

2 This occurs in a child with known pneumonia who does not respond to the usual and appropriate treatment. If a child remains pyrexial or unwell 48 h after admission and treatment with antibiotics, empyema should be suspected.

Investigations

An erect plain x-ray of the chest demonstrates obliteration of the costophrenic angle and a rim of fluid around the lung (*meniscus sign*). On a supine film, there may be a homogeneous increase in opacity over the whole lung field. A lateral chest radiograph may sometimes assist in differentiating pleural from intrapulmonary shadows.

Ultrasonography provides information on the size and distribution of the effusion, and may differentiate free (simple) from loculated (complicated) pleural fluid. It may also be used to guide chest drain insertion or thoracocentesis, if required. Chest CT scans are not performed routinely.

Blood cultures are performed in all patients with parapneumonic effusions. Similarly, sputum, if available, should be sent for bacterial culture. There is no role for routine bronchoscopy.

Treatment

All children with parapneumonic effusion or empyema should be admitted to a tertiary paediatric centre.

Supplementary oxygen is administered if the PaO_2 is less than 92%. Intravenous fluids are provided if the child is dehydrated or unable to drink, and antibiotics are given intravenously. The severity of pleuritic pain means that many of these children need regular analgesia. Physiotherapy has not been shown to influence outcome.

Many small parapneumonic effusions will respond to antibiotics without the need for further intervention. However, effusions that are enlarging and/or compromising respiratory function in a pyrexial and unwell child need drainage. This may be done by chest tube drainage with introduction of fibrinolytics or by operative drainage and debridement. Chest tube drainage alone is usually ineffective.

Intrapleural fibrinolytics (tissue plasminogen activator) administered via the chest drain have been shown to shorten hospital stay and may be used in early non-loculated disease. Dosage is typically for 3 days using 40,000 U/40 mL 0.9% saline for children aged 1 year or above or 10,000 U/10 mL 0.9% saline for children aged under 1 year.

Early referral to paediatric surgeons is advantageous. There is a trend towards early operative intervention, the main indication being where there is ongoing sepsis with a persistent or increasing pleural collection. In many centres, the initial approach is via thoracoscopy (video-assisted thoracoscopic surgery [VATS]) to debride the fibrinous pyogenic material, to break down loculations and drain pus from the pleural cavity and to irrigate the pleural space with saline. If this is unsuccessful or the disease is advanced, a mini-thoracotomy may be required to better debride the organising empyema. This usually involves a small postero-lateral thoracotomy. A chest drain is left *in situ* to allow drainage of any residual fluid and pus. An operation is more likely to be required when the empyema is long-standing or there is significant underlying lung pathology.

Outcome and prognosis

The majority of children have complete recovery and regain normal lung function but this may take many months.

Pneumothorax

Clinical features

The clinical picture is determined by the age of the patient, the size of the pneumothorax, the presence or absence of tension and the nature of any underlying pulmonary disease [Box 49.2]. Symptoms include pain in the chest and dyspnoea.

Signs include displacement of the trachea to the contralateral site and displacement of the apex beat. There is diminished movement of the chest wall, a hyper-resonant percussion note and diminished air entry on the side of the pneumothorax. In the typical *spontaneous pneumothorax* of adolescence, there is sudden onset of symptoms, usually in a tall, athletic boy with no pre-existing illness.

Box 49.2 Causes of pneumothorax

1. Spontaneous, often in teenagers, from apical bleb
2. Rupture of subpleural abscess in staphylococcal pneumonia
3. Neonatal pulmonary disease (e.g. hyaline membrane disease)
4. Rupture of subpleural emphysematous bulla (e.g. asthma, cystic fibrosis)
5. Traumatic, following chest wall injury (Chapter 38)
6. Perforation of oesophagus by ingested foreign body or during oesophagoscopy
7. Iatrogenic (e.g. after paracentesis)
8. Rupture of hydatid cyst
9. Post-operative, after thoracotomy

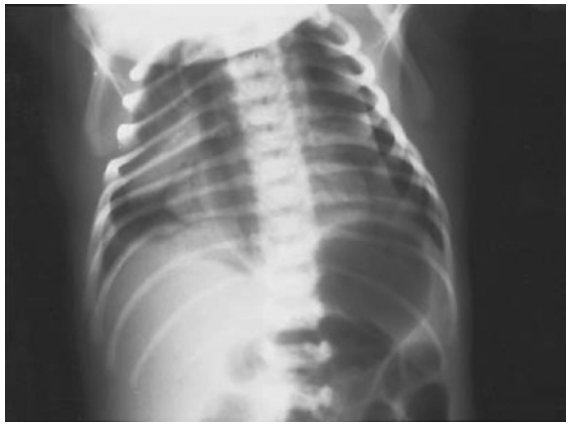

Figure 49.2 Neonatal pneumothorax. Tension pneumothorax has caused displacement of the mediastinum to the right with collapse of the right lung.

Chest x-rays are usually diagnostic [Fig. 49.2], but occasionally, a huge cyst or unusual types of diaphragmatic hernia may look similar radiologically.

Treatment

Urgent relief of tension by needle thoracocentesis or intercostal drainage with an intercostal chest tube is required. The underlying cause may require further investigation (usually a CT scan of the chest). In *spontaneous pneumothorax*, the scan will often reveal apical cysts. If this is the first episode, the patient is managed with chest drainage as required and no further intervention. If the pneumothorax persists and recurs on the same side or apical cysts are seen on imaging, most surgeons perform a thoracoscopic apical blebectomy ± an apical pleurectomy.

Pleural effusion

The accumulation of fluids (pus, serofibrinous exudate, blood, chyle or transudate) in the pleural cavity occurs in a variety of conditions [Box 49.3].

The physical signs are usually unmistakable, but radiographs are necessary and may require careful interpretation to distinguish a pleural *collection* from such conditions as hydatid cyst or neuroblastoma.

Paracentesis may provide information on the nature of the fluid and material for cytological and bacteriologic examination, and indicates the necessity for further drainage.

Chylothorax

Chylothorax may occur spontaneously or after cardiothoracic operations where the thoracic duct has been injured. Some cases require aspiration, while some patients require thoracoscopy or thoracotomy to ligate the leaking thoracic duct. Replacing the fat content of

Box 49.3 Causes of pleural effusions

1. Pulmonary infection: acute (staphylococcal pneumonia) and chronic (TB)
2. Disturbed haemodynamics: cardiac failure and hypoproteinaemia
3. Malignancy: lung, pleura and mediastinum
4. Chylothorax: damaged and/or obstructed thoracic duct

the diet with medium-chain triglycerides that are absorbed via the portal vascular system may control, or even cure, the condition, as only the long-chain fats enter the lacteals and travel via the thoracic duct.

Bronchiectasis

Recurrent or chronic bronchitis is the most common precursor of bronchiectasis. This type of bronchiectasis tends to be widespread, although the disease is usually most marked in one area.

Infection associated with permanent collapse of one or more lobes of one lung may be the cause. However, the bronchiectasis may remain *dry*, that is, structural changes may be demonstrated by bronchography, but symptoms and signs are minimal or absent if the affected area remains free of infection.

Other causes include pulmonary tuberculosis, hydatid disease, congenital weakness of the bronchial wall (bronchomalacia), cystic fibrosis and the inhalation of a foreign body. In healed tuberculosis, the bronchiectasis is usually *dry*, while in bronchomalacia and cystic fibrosis, dilatation of the bronchi is usually widespread and associated with copious sputum.

Clinical features

Physical signs may be diffuse, localised or even absent, according to the aetiology and the extent of the disease. Clinical assessment of the nature and amount of sputum, any interference with normal life and school attendance, and the frequency of acute toxic episodes of pneumonitis, should precede any investigations.

Bronchoscopy aims to confirm the diagnosis and determine the distribution and severity. A CT scan of the chest confirms the extent of the disease process. Culture of the sputum is a useful guide to antibacterial choices, particularly during exacerbations of infection.

Treatment

Conservative treatment, with physiotherapy, postural drainage and antibiotics, is usually effective.

Lobectomy and/or segmental resection is required occasionally and, in localised disease, is curative. As a general rule, resection should be deferred until late childhood, in case progressive disease in the remaining lobes of the lungs develops. When the bronchiectasis is more widespread, resection of a particularly diseased area may considerably improve the patient's well-being and reduce the amount of sputum produced, although some coughing may persist.

Congenital malformations

Congenital lobar emphysema and congenital cystic lung lesions (Chapter 4) may present in older children, and there is no typical clinical pattern. Often recurrent respiratory infections lead to an x-ray examination of the chest, with a subsequent diagnosis.

A CT scan will often provide additional information, and fluoroscopy may demonstrate *trapped air* in the lobe concerned, that is, movement of the mediastinum towards the affected side during inspiration and away from it during expiration.

Treatment typically involves removal of the affected portion of the lung.

Pulmonary sequestration

A sequestrated lobe is pulmonary tissue that does not have a normal communication with the bronchial tree, and it receives its blood supply from an anomalous systemic artery, usually from the aorta. It does not participate in the normal function of the lung and is consequently prone to infection. Two types are recognised: extralobar and intralobar.

In extralobar sequestration, there is complete anatomical and physiological separation from the normal lung, and the sequestrated portion may be above or below the diaphragm. This is sometimes seen in association with congenital diaphragmatic hernia. The arterial supply is from the aorta (above or below the diaphragm) or one of its branches (Fig. 4.3, Chapter 4).

In intralobar sequestration, the abnormal tissue is contiguous with the normal lung, which partially surrounds it. This type is almost always in the postero-lateral portion of the right or left lower lobe. The blood supply comes from large direct branches of the aorta (75%) or from other thoracic or abdominal vessels (25%), and the venous drainage is through the pulmonary veins.

The sequestrated lobe may consist of a large cyst, multiple cysts, branching bronchi without cysts, or all three of these.

Clinical features

The diagnosis is most often made on routine antenatal ultrasonography. At birth, the neonate is often asymptomatic. In children presenting beyond the neonatal period, the usual history is of repeated episodes of pulmonary infection with signs confined to one area. Although the infection commonly subsides, acute suppuration may supervene. Subsidence of the acute phase often leaves in its wake chronic suppuration with poor health, a persistent cough and sometimes low-grade pleural pain. Haemoptysis occurs occasionally.

Chest x-rays show an opacity in the posteromedial part of one of the lower lobes or cystic spaces, with or without fluid levels, in a lower lobe. In the acute phase, the opacity increases in size and may produce mediastinal displacement.

Ultrasonography and CT scanning demonstrates the anomalous arterial supply, confirms the diagnosis and is useful in planning the operative approach.

Treatment

Resection, usually by thoracoscopy, is indicated because of the susceptibility to infection. Where acute infection has occurred, it is treated with antibiotics, and the sequestration is subsequently resected during a quiescent phase.

The child with a mediastinal mass

A child may present with a mediastinal mass in one of two ways:

1. A symptomless mass demonstrated in a chest x-ray
2. Symptoms caused by compression of mediastinal structures

A symptomless mass

The thymus is large in infancy, and determining whether its appearance on chest x-ray is normal requires experience. Some radiographic techniques cause apparent enlargement.

A symptomless mediastinal mass may develop in the course of a generalised disease, for example, from enlarged hilar lymph nodes in leukaemia or Hodgkin's disease, or as metastases from a malignant disease. Paravertebral and para-aortic masses of neuroblastoma may involve the mediastinum, often as the primary site but also as metastases from tumour elsewhere, for example, in the abdomen (Chapter 25).

Incidental chest x-ray presentation

Sometimes, a mass is found incidentally on chest x-ray. Initial investigations should include:

1. Examination of the peripheral blood for evidence of leukaemia
2. Examination of the bone marrow for metastatic neuroblastoma
3. Examination of the urine to determine the excretion of methyl hydroxyl mandelic acid (MHMA) in a 24 h specimen
4. A complete radiological assessment of the mediastinal mass, including oblique views, computerised tomography and magnetic resonance imaging, as indicated

In many instances, these investigations will give a good indication of the diagnosis, but depending on discussions with the oncology service, surgical exploration may still be required to obtain material for a histological diagnosis or to determine whether the mass is removable.

Surgical excision is generally curative in conditions such as teratomas, duplication (*enterogenous cysts*), bronchogenic cysts and ganglioneuromas. In the latter, both thoracotomy and laminectomy, combined or in stages, may be required to remove both the components of a dumb-bell tumour.

Total removal of infiltrating primary neoplasms or extensive metastases in para-aortic lymph nodes is often impossible, but there is evidence that even incomplete removal may be of benefit in neuroblastoma, in which maturation to benign ganglioneuroma may occur, particularly in those arising in the posterior mediastinum.

The operative findings, the histology and the results of the preliminary examinations will determine the need for chemotherapy and radiotherapy.

Compression of mediastinal structures

In childhood, this is nearly always the result of a malignant mass, of which lymphosarcoma is the most common. Congestion of the veins of the head, neck and upper limbs from obstruction of the superior vena cava, wheezing, an unproductive or reverberating brassy cough and increasing dyspnoea are all ominous signs.

Lymphosarcomas usually arise in the anterior mediastinum in the region of the thymus, and x-rays usually show a large mass that extends laterally. The edges may be irregular, rounded or ill defined; the tumour may extend into the pleura and cause a pleural effusion. The histological diagnosis may be made from cytology of the cells in the pleural effusion.

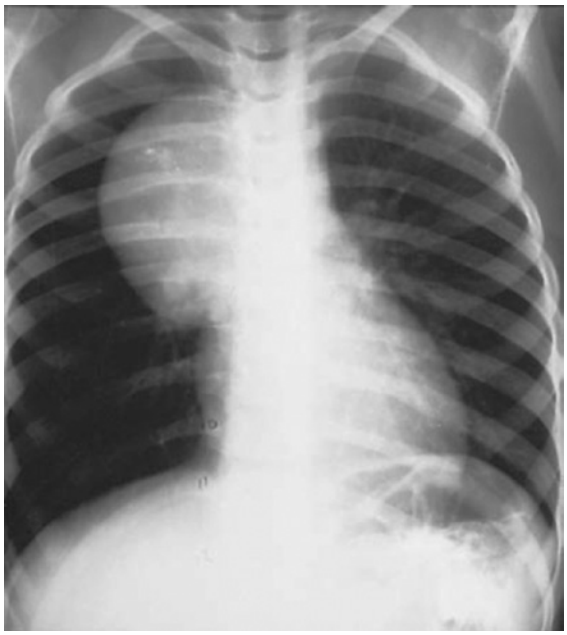

Figure 49.3 A mediastinal mass: in this case a neuroblastoma with calcification in the posterior mediastinum.

A neural crest tumour (neuroblastoma or ganglioneuroma) is the usual cause of a paravertebral mass: areas of both neuroblastoma and ganglioneuroma may be present in a single tumour [Fig. 49.3]. Erosion of the ribs and extension into a vertebral foramen causing spinal symptoms are seen with infiltrating mediastinal neuroblastomas.

A ganglioneuroma is a benign tumour arising in a paravertebral gutter. It often grows through a vertebral foramen into the spinal canal, resulting in two solid elements connected by a narrow isthmus in the intervertebral foramen, a *dumb-bell* tumour.

Thymomas are much less common and are difficult to distinguish from lymphosarcomas. They tend to grow slowly, to reach an even larger size and to have a more distinct margin in the x-rays of the chest.

Teratomas in the anterior mediastinum are very rare. They are cystic or solid, only occasionally malignant, and may extend laterally into one or other pleural cavity.

Bronchogenic cysts arise close to the trachea or the hilum of the lung; they usually contain air and many show a fluid level in x-rays. There may be a history, suggestive of intermittent partial obstruction of one of the larger bronchi, and operative excision is curative. This is now often performed through a thoracoscopic approach.

Management

Compression of the trachea and/or bronchi may cause severe respiratory distress, sometimes precipitated by a supervening virus infection. When a chest x-ray shows a mediastinal mass, tracheostomy is usually of no assistance, for the obstruction is below the level of the suprasternal notch. Nasotracheal intubation may be necessary as an emergency. This situation is most commonly caused by a lymphosarcoma, and this presumptive diagnosis should be confirmed quickly by examination of the peripheral blood, the bone marrow or an accessible enlarged lymph node (or occasionally the mass itself). As with other malignancies, the stage of the disease will determine the treatment. However, use of steroids and cytotoxic agents, even without a histological diagnosis, is justified in an emergency and brings dramatic relief. The prognosis for most types of lymphosarcoma is good.

KEY POINTS

- Staphylococcal pneumonia may rapidly lead to empyema, lung abscess and/or air leak.
- Children with parapneumonic pleural effusion should be admitted to a tertiary paediatric centre.
- Spontaneous pneumothorax is most common in adolescent boys.
- Chronic/recurrent bronchitis may lead to bronchiectasis.
- Chest x-ray should be done in recurrent respiratory infections to exclude underlying cause (congenital cystic lung, foreign body, tumour).

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PART IX
Skin and Soft Tissues

CHAPTER 50
Vascular and Pigmented Naevi

CASE 1

The mother of an 11-year-old is concerned about a few small brown flat moles that are slowly enlarging and becoming slightly nodular.

Q 1.1 Is it possible that any of these could be a melanoma?

Q 1.2 How should they be managed?

CASE 2

A 2-month-old was born with a red spot on the left upper eyelid. This has progressively grown into a large red nodular lesion, and partial ptosis is now present.

Q 2.1 What are the possible functional and cosmetic consequences?

Q 2.2 How should this be managed?

Vascular lesions

Cutaneous vascular malformations are common. Both haemangiomas and vascular malformations may occur as isolated lesions or as part of a syndrome with multisystem involvement.

Haemangiomas

Haemangiomas are the most common tumours of infancy and childhood. They have increased endothelial cell turnover and many mast cells, with relatively predictable behaviour and life cycle. They occur in 12% of all 1-year-olds.

Haemangioma of infancy (strawberry naevus)

These usually appear shortly after birth as a pale, pink or bright red spot or patch on the skin, the so-called herald spot. The hallmark of these tumours is their rapid growth in infancy, which continues for anywhere from 4 to 12 months, but typically ends around 9 months [Fig. 50.1]. Gradual involution often begins at about 1 year of age, with a grey patch appearing centrally, and is usually complete by 6–8 years. A small amount of redundant atrophic telangiectatic skin and subcutaneous tissue may remain, requiring cosmetic correction. This is especially true on the lip. Multiple skin lesions may be associated with visceral involvement.

Haemangiomas on the skin surface have the typical red, *strawberry* appearance. When haemangiomas are under the skin, they are more blue in colour, but histologically, they are the same. Apart from presenting later, their behaviour is identical. Often, haemangiomas have superficial and deep components.

Congenital haemangioma

A subgroup of haemangiomas is present fully formed at birth. The haemangiomas of this subgroup either regress rapidly in the first few months of life (rapidly involuting congenital haemangioma or RICH), or they persist into adolescence (non-involuting congenital haemangioma or NICH). Congenital haemangiomas have a blue colour and a pale periphery. They can be differentiated from infantile haemangiomas on histology as they do not stain for markers of immature endothelium, in particular Glut-1, for which haemangiomas of infancy are uniformly positive.

Management of haemangiomas consists of accurate diagnosis and careful observation. The parents need reassurance during the normal rapid growth phase of the lesion and must be warned that the haemangioma may increase significantly in size. Failure to give this warning leads to a loss of confidence in the treating doctor if alarming growth occurs. Problems of ulceration, bleeding and rarely infection occur secondary to minor trauma, especially in the *nappy* area. These are usually

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best dealt with non-operatively. Bleeding is controlled with pressure. Ulcerated haemangiomas are painful and can be very slow to heal. Excision of an ulcerated haemangioma may be indicated if the defect can be directly closed, particularly in the nappy area where recurrent ulceration may occur. Haemangiomas of the eyelid are of particular concern as pressure on the globe can distort the shape of the cornea, causing strabismus, and this, or the occlusion of the visual axis, for even a few weeks, can produce an amblyopic eye [Fig. 50.1]. Early referral to a paediatric ophthalmologist is mandatory for haemangiomas of the eyelid.

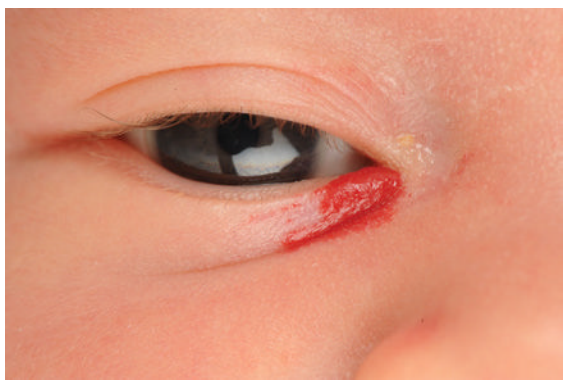
Systemic steroids used to be the first-line treatment for haemangiomas not amenable to surgery, but most centres now use oral propranolol as the first-line treatment. Response to propranolol is generally rapid and occurs in around 90% of cases. Steroids, alpha interferon and vincristine are alternative treatments. Topical Timoptol eye drops can be effective on superficial lesions.

Large haemangiomas of the liver can be associated with high-output cardiac failure and can also deplete thyroxine, causing unrecognised hypothyroidism. These lesions may be fatal.

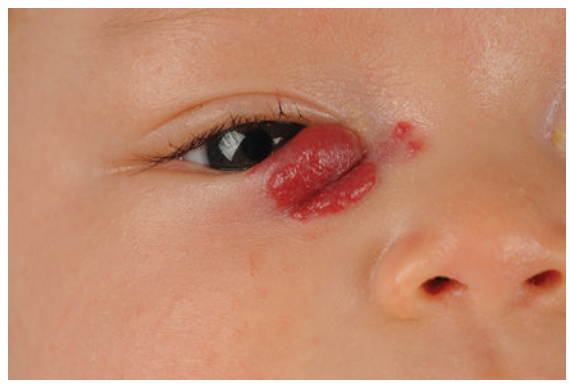
Consumptive coagulopathy (Kasabach–Merritt syndrome) with a very low platelet count is associated with two rarer lesions, kaposiform haemangioendothelioma (KHE) and tufted angioma, but is not caused by true infantile or congenital haemangioma. KHE and tufted angioma (along with congenital haemangiomas) do not respond to propranolol. Steroids and vincristine are the preferred treatments, and rapamycin is sometimes used.

Pyogenic granuloma

Pyogenic granuloma is the other common *cellularly dynamic* lesion. It may follow minor trauma to the face and grows rapidly, becomes ulcerated and friable, and bleeds readily. It is characterised by a central feeding vessel supplying a mass of new capillaries and an associated inflammatory infiltrate. The appearance is often similar to haemangioma of infancy, but the age of appearance differentiates it. Pyogenic granuloma is



(a)



(b)

(c)

Figure 50.1 a,b,c The common haemangioma of infancy (strawberry naevus) on the lower eyelid, shortly after birth **(a)**, 6 months **(b)** and then at 2–3 years of age **(c)**. The lesion required frequent assessment, as obstruction of the visual axis, for even a few weeks, will result in an amblyopic eye.

Table 50.1

	Haemangioma of infancy	Vascular malformation
Growth pattern	Appears just before or just after birth and grows rapidly	Grows in proportion with child
Involution	Starts to involute between 6 and 18 months	Does not involute
Endothelial characteristics	Plump endothelial cells showing rapid turnover	Stable flat endothelium
Pharmacological treatment	Most respond to propronolol, backup treatment of steroids, interferon or vincristine	No pharmacological treatment available at present

treated by excision, which should include the feeding vessel to prevent recurrence.

Vascular malformations

Regional vascular malformations, in contrast to haemangiomas, are composed of mature vascular elements and do not regress [Table 50.1]. They may be capillary, venous, arteriovenous, lymphatic, or a combination of these.

Capillary malformations

Naevus flammeus medialis is the *salmon patch* or *stork's beak mark* seen on the nape of the neck in infancy. This lesion does not change with time. *The port wine stain* is a cutaneous capillary malformation. There is gradual darkening and hypertrophy of the lesion over a patient's lifetime. This lesion may be part of a syndrome, for example, the Klippel–Trenaunay syndrome (limb overgrowth with lymphatic and venous anomalies) or the Sturge–Weber syndrome (lesions involving the upper face with intracerebral involvement and epilepsy). Any capillary malformation around the eye requires assessment for glaucoma.

Current therapy centres on the use of lasers, with wavelengths selective for haemoglobin and rapid, short-duration target heating times. This coagulates the lesion while producing the least scarring in the skin. The results of the treatment in children are better than those in adults.

Venous malformations

These compressible lesions occur anywhere on the body and may occasionally undergo thrombosis. The presence of phleboliths (small, smooth, hard and mobile) is diagnostic of venous malformation [Fig. 50.2]. Venous malformations tend to slowly expand over time. MRI and ultrasound are the most useful investigations: venous malformations are not visualised on conventional angiography. The most effective treatment is usually injection sclerotherapy performed by the interventional radiologist. Surgery can be useful for specific problems but complete excision is rarely possible. Compression garments and low-dose aspirin therapy may be helpful.

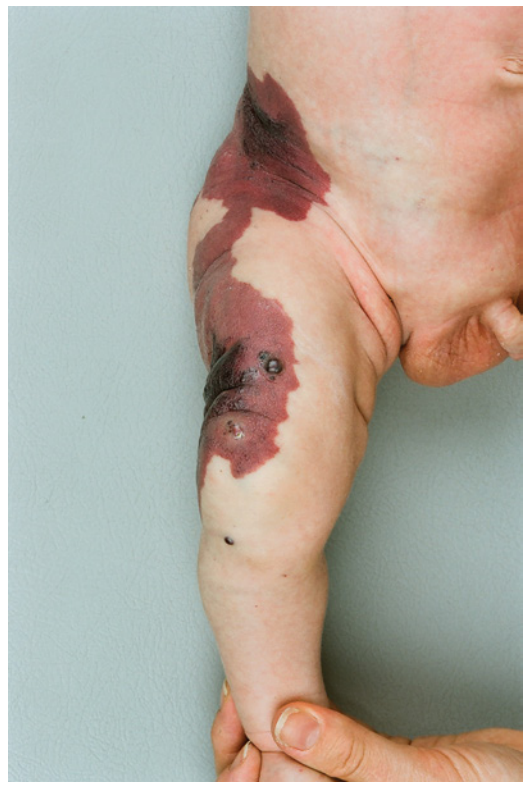


Figure 50.2 A venous malformation affecting the pelvis and thigh.

Arteriovenous malformations

Arteriovenous malformations are high-flow lesions that tend to have a more malignant course than low-flow malformations. They often have an overlying capillary *stain*. They tend to gradually increase in size over time, especially around puberty and in pregnancy, and can

ultimately develop skin ulceration and infection. Large lesions become at risk of catastrophic bleeding. Ligation of feeding vessels, either surgically or by angiographic embolisation, is not curative and complicates future treatment. Treatment is either surgical excision or intra-arterial ethanol infusion under radiological control. Recurrence is common.

Lymphatic malformations

Malformations of the lymphatic system range from the small nodular lesions of lymphangioma simplex to large cervical cystic hygromas. They can consist of large cysts (macrocystic), multiple small cysts (microcystic) and sometimes contain areas of solid fibrous tissue. Lymphatic malformations have a propensity to infection, which must be treated with prolonged antibiotic therapy. Well-defined lesions should be excised if possible, although recurrence is common and may occur in the scar. *Cystic hygromas* are usually located deep in the cervical and upper thoracic area and may be associated with inflammatory and infectious complications. Macrocystic lesions are amenable to injection sclerotherapy under radiological control, which can be performed with a variety of agents. Microcystic lesions and those not responding to injection, if causing problems, require operative treatment. Surgery is technically difficult as the lesions are not confined by tissue planes (see Fig 16.2, Chapter 16) and nerves frequently pass through the centre of the lesion.

Telangiectasia and spider naevi

Telangiectasias may also be congenital and may be isolated or appear as part of a syndrome, for example, Rendu–Osler–Weber syndrome. Spider naevi are common and are distinguished by a central feeding vessel and radial filling following careful occlusion of this vessel. Treatment is by laser.

Pigmented naevi

True pigmented naevi are melanocytic in origin.

Junctional naevi

Histologically, these naevi show clusters of melanocytes in the basal layers of the skin. They are flat brown or black spots clinically and normally persist throughout childhood. Junctional activity after puberty is a very slight risk for malignant melanoma. Malignant change is so rare that excision of these in childhood should be avoided unless they have particularly worrying features or if they are a significant cosmetic blemish.

Compound and intradermal naevi

A compound naevus has both junctional and intradermal components. Naevus cells bud off into the dermis where they proliferate and form a cluster of cells, resulting in a raised palpable lesion. In later years, the junctional activity ceases and the lesions become mature intradermal naevi. They are felt to be benign with no risk of malignant transformation. Surgical excision is for cosmetic reasons.

Spitz naevi

The juvenile or Spitz naevus is usually reddish in colour, as melanin is less prominent. There is considerable junctional activity and spindle cells are present in the dermis. The presence of mitotic figures and atypical cells sometimes leads to confusion with malignant melanoma.

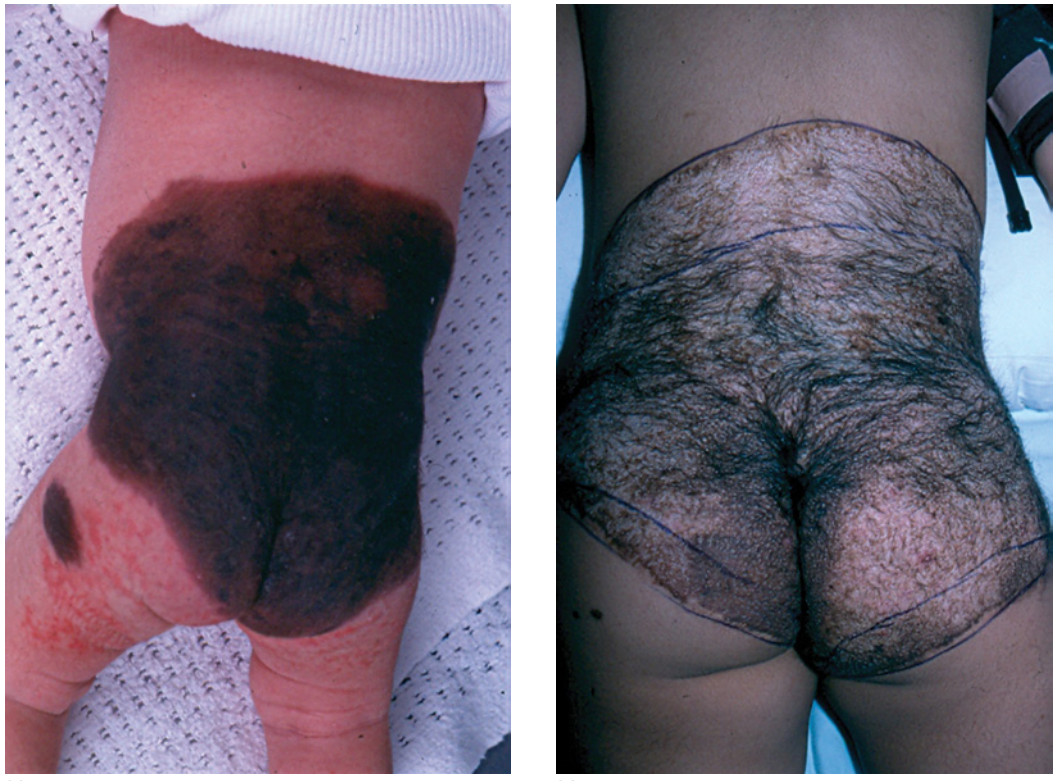
Congenital naevi

Congenital naevi are found in 1% of babies. They may be small (<1.5 cm), medium or large (>20 cm). These are histologically similar to acquired compound naevi, and the cells form nests deep within the dermis in association with hair follicles and sebaceous glands. The giant naevus occurs in 1:20,000 babies and covers a major segment of the body, for example, *bathing-trunk* naevus [Fig. 50.3a]. Multiple smaller naevi may also be present in other areas, and there may be meningeal involvement. The giant naevus is largely intradermal, but may have a junctional component. The risk of malignant melanoma, mainly after puberty, is about 4% over a lifetime in large naevi and may be higher in giant naevi. With adolescents, the lesions tend to become more nodular and hairy.

Treatment is performed mainly for cosmetic reasons by excision and direct closure where possible, or reconstruction with flaps, with or without tissue expansion. In many instances, complete removal is impossible. In giant naevi, extensive skin grafting is best avoided. Early referral is essential, as many giant naevi can be improved by curettage in the first few months of life [Fig. 50.3b]. Occasionally, melanocytic naevi respond to laser treatment, but generally, results are disappointing.

Halo naevi

A halo naevus occurs when melanocytes disappear from the periphery of a pigmented naevus. This is felt to be an immunological phenomenon, and lymphocytes



(a) (b)

Figure 50.3 Giant hairy naevus: **(a)** at 3 weeks of age, **(b)** later in childhood, following healing after removal using a sharp curette (and excision of the rim that failed to cleave off with the curette).

are seen on histological examination. The naevus may disappear completely, leaving a pale patch of skin that later re-pigments.

Ephelis

An ephelis or freckle is a localised increase in epidermal pigmentation only. The melanocytes are normal in number.

Lentigo

A lentigo is a patch of increased numbers of melanocytes occupying the basal layer of the epidermis.

Blue naevi

A blue naevus consists of spindle-shaped melanocytes with large amounts of pigment in the deeper dermal layers. The presence of the dark pigment beneath the translucent epidermis produces the characteristic blue-grey colour. The *Mongolian spot* found on the back and buttock of Asian and black infants, which fades with age, is a classic example.

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CHAPTER 51
Soft Tissue Lumps

CASE 1

A 1-year-old has a non-hair-bearing patch of yellow, slightly raised and irregular skin, measuring 2 × 3 cm on the scalp.
Q 1.1 How and when should this be treated?
Q 1.2 What may happen if it is not treated?

CASE 2

A year ago, a 13-year-old girl, whose parents are from South Asia, had her ears pierced. She now has 1 cm lumps at the sites of piercing.
Q 2.1 What is the diagnosis and how should this be managed?

There are many cutaneous and subcutaneous lesions that occur in childhood. Some of the more common are covered in this chapter. Vascular and pigmented lesions of the skin are discussed in Chapter 50.

Warts

These are small epidermal tumours produced in response to papilloma virus infections of the skin. They are contagious to a limited degree. Those on the soles of the feet can cause discomfort on weight bearing. Treatment of troublesome lesions involves destruction of the lesion by topical application of liquid nitrogen, salicylic acid or podophyllin paint or curettage.

Molluscum contagiosum

This is caused by a pox virus, which produces clusters of pink papules with a central plug which can be squeezed out. They usually resolve after 2 months or may be treated like warts.

Epidermal and pilar cysts

These are common cysts of epidermal and pilar (hair follicle = trichilemmal) origin, respectively. Although most often seen in adults, they are not uncommon in children. Treatment is by complete surgical excision.

Dermoid cysts

These are most commonly found in the head and neck. Dermoid cysts contain epithelium and adnexal structures, such as sweat glands or hair follicles of varying degrees of differentiation. Nasal dermoids may have a small pit or sinus with associated hair growth. External angular dermoids appear in the lateral eyebrow region [see Fig 16.7]. They are sometimes located under the pericranium and appear hard and fixed. Treatment is by complete surgical excision; however, for nasal dermoids, possible deep, intracranial extension must be excluded by CT scan, MRI, or both.

Pilomatrixoma

This relatively common skin appendage tumour occurs on the face, neck and upper extremities of young children. It is a hard, non-tender and irregular intradermal or subcutaneous lesion, which may appear white or yellow, or sometimes blue in colour through the skin [Fig 51.1]. They enlarge slowly. It is characterised histologically by areas of calcification and *ghost cells*. Excision

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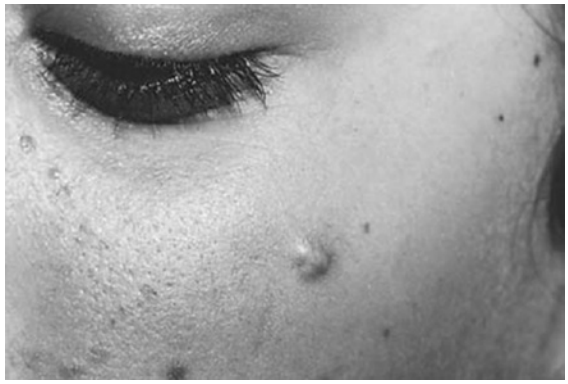

Figure 51.1 Pilomatrixoma.

is recommended. Multiple lesions should arouse suspicion of Gardner's syndrome.

Naevus sebaceous

Naevus sebaceous is present at birth and appears as a yellowish, slightly raised lesion, usually on the scalp or face. Around puberty, they become thicker and more wart-like. There is a 15–20% incidence of basal cell carcinoma developing during adulthood if it is left untreated. These lesions are best excised electively during early childhood.

Hypertrophic scars and keloids

Some children tend to produce thick red scars that take 1–2 years or more to involute to become acceptably pale and flat. All scars will become thicker and redder in the first 2–3 months before flattening, and hypertrophic scars are considered to be an exaggerated form of this normal healing behaviour. Hypertrophic scars are more common on the sternum and deltoid regions and over the extensor surface of joints. A keloid continues to grow beyond the margins of the original wound and is more like a tumour [Fig 51.2]. Keloids are more common in people of African and Asian origin but can occur in other children, particularly on the earlobe. The appearance is often very unsatisfactory, and the scars are itchy and sometimes cause contractures. Management can be difficult and involves counselling parents (about the time course and expected outcome),

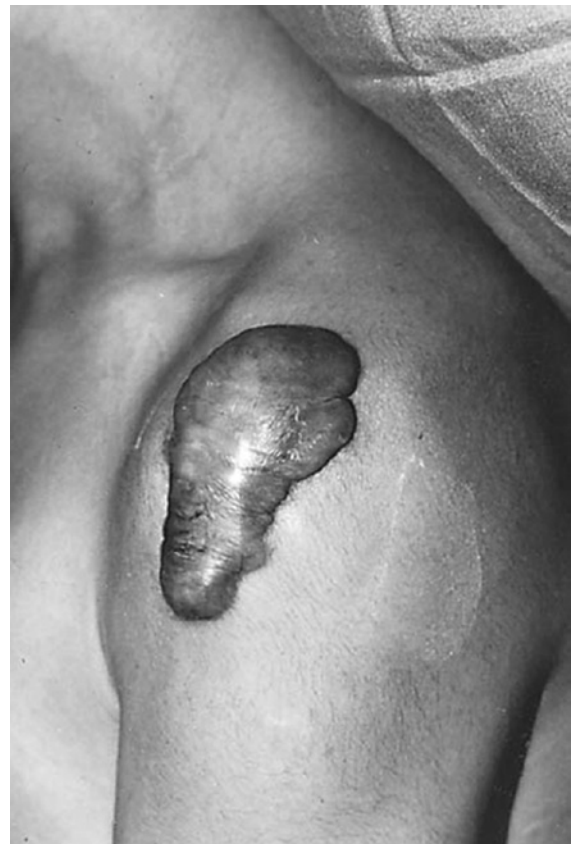

Figure 51.2 Keloid scar in an adolescent Asian male following a BCG vaccination.

intralesional corticosteroid injections and application of pressure (often applied through specially designed Lycra garments), and the use of silicone gel sheeting may be helpful with hypertrophic scars, although steroids should be used with caution as overcorrection is possible. Keloids are more difficult to treat and may not respond to even the highest dose of steroids. Excision is often followed by recurrence (except sometimes in the earlobe). Excision by a plastic surgeon with immediate steroid injection or, on occasion, immediate radiation is the most effective treatment.

Ganglia

Ganglia contain gelatinous synovial fluid and frequently arise from joints, particularly the wrist, principally from the scapholunate ligament. They also arise from tendon

sheaths. They may be uncomfortable and can be treated with aspiration and corticosteroid injections followed by splintage or by excision. If excision is not complete down to the joint of origin, recurrence is common.

Neurofibromatosis

This autosomal dominant condition affects 1 in 3000 children and may present as café-au-lait pigmented spots or soft tissue tumours. These consist of fibroblasts, Schwann cells and nerve fibres. Large plexiform lesions of the nerve trunks may be quite disfiguring. Neurofibrosarcoma occurs in approximately 5% of patients.

Fibromatosis

This is an idiopathic, self-limiting fibroblastic proliferation that infiltrates locally and tends to recur, but never metastasises. Fibromatosis often affects the digits or palm and may be aggressive (e.g. extra-abdominal desmoid).

Soft tissue sarcomas

These make up 6–8% of all malignancies in childhood, with rhabdomyosarcoma and undifferentiated sarcoma being the most common. After diagnostic biopsy and complete staging, combined treatment with chemotherapy, wide excision, and sometimes radiotherapy is planned. Sophisticated reconstruction, often with microsurgically transferred tissues, may be required.

KEY POINTS

- Unless a firm diagnosis of a benign tumour can be made, excision of a soft tissue lump to exclude malignancy is prudent.
- Naevus sebaceous is best treated by excision to prevent basal cell carcinoma in adulthood.
- Keloid scars may need local treatment.
- A ganglion may be treated by aspiration and corticosteroid injection.

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CHAPTER 52
Answers to Case Questions

1 Antenatal diagnosis: Surgical aspects

1.1 Regular ultrasounds to monitor progress and ongoing discussion with, and counselling by, paediatric surgeon.

1.2 Yes. Antenatal ultrasound allows antibiotic prophylaxis and early postnatal investigation to be initiated before kidney becomes further damaged by infection and/or obstruction.

2.1 Chromosomal analysis and search for other major anomalies.

2.2 No. Only rarely is time or mode of delivery important.

2 The care and transport of the newborn

1.1 Wrap the exposed viscera and abdomen in a plastic wrap (be careful not to twist bowel); insert IV line for 10% dextrose and 1/5 (0.18%) normal saline; nil orally; nasogastric tube with aspiration; put baby in incubator; check for other anomalies; talk to parents about surgery and prognosis (good); obtain consent for transport and surgery; and take photograph for parents.

2.1 Bronchopulmonary damage and pneumothorax.

2.2 Avoid high airway pressures during resuscitation and allow baby to breathe spontaneously. The key metabolic problems are respiratory.

3 The child in hospital

1.1 No. General anaesthetic is required to prevent fear and keep the patient still during surgery. Local anaesthetic is used for post-operative analgesia.

1.2 Removal of sutures (with blade or scissors) is frightening to children, because they assume it will be painful (which it is).

1.3 Allow regular washing and activity.

2.1 Fear of pain, staying overnight away from home and parents, unknown technology and unfamiliar people.

4 Respiratory distress in the newborn

1.1 Cystic lung disease (sequestration, cystic adenomatoid malformation, congenital tumour [e.g. teratoma])

1.2 Excision

2.1 Pneumothorax ± tension

2.2 Intercostal (IC) drainage: formal catheter (fourth IC space, mid-axillary line) or needle (second IC space, mid-clavicular line) if urgent

5 Congenital diaphragmatic hernia

1.1 Diaphragmatic hernia.

1.2 X-ray of chest and abdomen (±insertion of NG tube to determine the position of stomach).

1.3 Pulmonary hypoplasia and pulmonary hypertension are the main intrinsic factors. Iatrogenic factors include excessive ventilation with pneumothorax, rough handling or premature surgical intervention prior to stabilisation.

2.1 No.

2.2 Insert nasogastric tube and apply gentle suction to keep the bowel decompressed.

2.3 Tension pneumothorax.

6 Oesophageal atresia and tracheo-oesophageal fistula

1.1 Pass a still 10 French catheter gently via the mouth: arrest at 9–11 cm from gums confirms diagnosis of oesophageal atresia.

1.2 Yes. Look for VATER/CHARGE and others.

1.3 Standard neonatal preoperative *first aid*, plus 10 minute suction of the oesophagus.

2.1 VATER: clinical examination and imaging.

2.2 Renal and cardiac ultrasonography, as well as a preoperative workup.

7 Bowel obstruction

1.1 Hirschsprung disease, small bowel atresia, volvulus neonatorum and meconium ileus.

1.2 Call the Neonatal Emergency Transport Service (NETS).

1.3 See under 'General Treatment'.

2.1 Rectal biopsy for histochemical tests and an examination for ganglion cells.

2.2 Neonatal primary pull-through (laparotomy/transanal + laparoscopy) or colostomy above the transition zone at diagnosis and pull-through operation at 3–6 months of age.

2.3 Initial prognosis is good and surgery is life-saving, but there are some problems with long-term morbidity, especially faecal soiling, constipation and proximal gut function.

3.1 Duodenal atresia, volvulus neonatorum and high jejunal atresia.

3.2 Plain x-ray and contrast meal.

3.3 Volvulus neonatorum is one of the most urgent conditions seen by paediatric surgeons.

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8 Abdominal wall defects

1.1 Exomphalos.
1.2 Failure of correct formation of the umbilical ring with embryonic folding.
1.3 See Table 8.1.
1.4 Aim to excise sac and umbilical cord and close the defect primarily. Prognosis reasonable if no other anomalies present.
2.1 Yes, if *first aid* is good.
2.2 Heat and water losses cause rapid hypothermia and dehydration.
2.3 Yes, but not very common, except for secondary atresia of the bowel compressed at the level of defect in the abdominal wall.
3.1 Bladder exstrophy.
3.2 Staged surgical reconstruction of bladder, bladder neck, urethra and external genitalia.
3.3 Each corpus cavernosum forms separately. Sexual function should be fair, but not normal (penis bent and short), but prognosis of urinary continence is guarded.

9 Spina bifida

1.1 Level and appearance at birth, motor and sensory levels, any secondary deformities and other abnormalities (e.g. hydrocephalus).
1.2 There is a high risk of Arnold–Chiari malformation and associated hydrocephalus.
1.3 Intellect should be reasonable, as long as hydrocephalus is controlled; walking is unlikely although hip flexion and adduction may be preserved.
2.1 Incontinence ± partial outlet obstruction, infection, stone and renal failure.
2.2 Clean intermittent catheter for urine (±reconstruction procedures); diet and washouts/laxatives for faeces.

10 Disorders of sex development

1.1 Any child with (i) apparently enlarged clitoris, (ii) apparent hypospadias with bifid scrotum and/or undescended testes and (iii) truly ambiguous genitalia.
1.2 As much as possible, so that they understand and can participate in decision-making.
1.3 Genital anomaly is a medical and social emergency, requiring urgent referral to a tertiary centre.
1.4 (a) Possible adult fertility and (b) relative degree of development of male versus female organs.
2.1 No.
2.2 Bifid *scrotum* and/or bilateral or unilateral *cryptorchidism*.

11 Anorectal anomalies

1.1 Same as for Case 1, Chapter 2
1.2 Either definitive surgery if low anomaly or colostomy if high anomaly and then pull-through at about 3 months
1.3 Outcome for faecal continence poor for more severe anomalies: bowel washouts ± diet and laxative treatment required. Less severe anomalies with better outlook but soiling or constipation still common
2.1 MRI
2.2 Diet and laxatives or regular washouts (via anus or appendicostomy)

12 Head and neck

1.1 Head circumference crosses the percentiles (hydrocephalus) or abnormal shape of skull.
2.1 Headaches are frequent or *different*; there is significant child or parental anxiety; there are other signs of raised ICP or focal signs.
2.2 Some are benign (e.g. pilocytic astrocytoma, craniopharyngioma, pineocytoma) and may be cured with surgery. Germinoma and medulloblastoma are malignant, but prognosis is quite good with treatment. The rest do badly (e.g. brainstem glioma, pineoblastoma, disseminated medulloblastoma or ependymoma).
3.1 Not always. Neurosurgical referral is required.
4.1 Surgery is possible, but prognosis varies. Occipital lesions are worse than sincipital (because of blindness/ataxia/hydrocephalus). Genetic counselling is needed.
5.1 Plagiocephaly with *sticky* lambdoid suture. Often improves, but strip craniectomy is needed sometimes. Definite synostosis does need surgery.

13 The eye

1.1 Amblyopia may develop if squint ± poor vision is untreated.
1.2 Eye examination, including cover test and check of vision.
2.1 Yes, there is nystagmus, which may be secondary to poor vision.
2.2 Vision and eye examination, referral to ophthalmologist.
3.1 The cause is nasolacrimal duct obstruction, which resolves in 95% by 1 year. Probing (under GA) reserved for persisting symptoms only.
3.2 Antibiotic drops only needed if secondary infection with conjunctivitis occurs.
4.1 History and examination should reveal cause, for example, foreign body.

14 The ear, nose and throat

1.1 A pale, opaque membrane occurs with fluid behind the membrane and/or inflammatory thickening.
1.2 Amoxicillin 40 mg/kg/day in × 3 doses for 5 days.
1.3 The infant has otitis media with effusion (± acute suppuration). The fluid in the middle ear reduces hearing.
2.1 Tonsillectomy plus adenoidectomy is recommended if attacks of tonsillitis are very frequent.
2.2 Whether or not the symptoms of obstructive sleep apnoea are present (see box 14.2).
3.1 Physical examination to determine the site of stridor plus degree of airway obstruction, followed by flexible laryngoscopy.
3.2 Nose blocked, stenosis and choanal atresia; glossoptosis; laryngomalacia; vocal cord lesions; subglottic stenosis; and tracheomalacia.

15 Cleft lip, palate and craniofacial anomalies

1.1 Pierre Robin sequence with mandibular hypoplasia and secondary cleft palate.
1.2 Nurse prone ± airway for first few days. Long-term spontaneous resolution with growth.
2.1 4% if no parents affected.

2.2 16% if parent affected.
3.1 Apert syndrome.
3.2 Craniofacial surgery for synostosis, repair of fingers and genetic counselling.
3.3 Mental deficiency is present in a significant number of patients.

16 Abnormalities of the neck and face

1.1 Cervical lymphadenitis, abscess or sialectasis
1.2 Once proven/suspected to contain pus: incision and drainage
2.1 External angular dermoid: excision
3.1 Thyroglossal cyst ± infection, ectopic thyroid, dermoid cyst and submental lymph node (goitre)
3.2 Sistrunk operation (removal of cyst and track including middle one-third of hyoid bone)

17 The umbilicus

1.1 No, spontaneous closure in >90% by 2 years of age.
1.2 If not closed by 2–3 years, it is because of increasing risk of incarceration.
1.3 Normal gap in abdominal wall of fetus for placental vessels (umbilical arteries × 2, umbilical vein × 1).
2.1 Residual necrotic tissue from cord stump colonised by skin organisms. Granulation tissue (*granuloma*) is produced in response to subacute inflammation. Silver nitrate cauterisation or excision.
3.1 Extraperitoneal fat.
3.2 No.
3.3 Yes, if symptoms are troublesome.

18 Vomiting in the first months of life

1.1 Observe visible peristalsis and palpate pyloric tumour.
1.2 Refer to paediatric surgeon.
1.3 Serum electrolytes and acid–base looking for metabolic alkalosis.
2.1 Malrotation with volvulus.
2.2 Contrast meal.
2.3 Ladd's operation (laparotomy/laparoscopy and detorsion of bowel) – extremely urgent.
3.1 Gastro-oesophageal reflux.
3.2 Reduce handling after feeds and thicken feeds.

19 Intussusception

1.1 Intussusception
1.2 Rectal examination, plain abdominal x-ray and abdominal ultrasound or gas enema
1.3 7% risk
2.1 Bowel obstruction secondary to intussusception (or other causes)
2.2 Resuscitation with IV fluids, nasogastric tube decompression and cross-matched blood
2.3 Operative reduction ± resection

20 Abdominal pain: Appendicitis

1.1 Mesenteric adenitis but should exclude urinary infection
1.2 Urine microscopy and culture and rectal examination
2.1 Appendicitis (retrocaecal/retroileal)
2.2 None required, as you have already made a diagnosis

21 Recurrent abdominal pain

1.1 Parents often worry that pains are caused by cancer, because of similar symptoms in an elderly relative.
1.2 Frequent short-lived pains, referred to periumbilical region, perhaps associated with stress in the child's life. Physical examination rarely shows anything other than constipation.
1.3 Surgeons' role is to exclude serious disease after careful history and physical examination and then reassure.

22 Constipation

1.1 Needs diet (fluid and fibre increase) and laxative treatment. Rule out organic causes.
2.1 Needs exclusion of organic causes, especially Hirschsprung disease and slow-transit constipation. Initial management includes enema ± faecal disimpaction (under GA).

23 Bleeding from the alimentary canal

1.1 See Table 23.3.
1.2 Anal fissure, secondary to constipation.
1.3 Correct the constipation with diet and laxatives.
2.1 Meckel's diverticulum, duplication, peptic ulcer and varices.

24 Inflammatory bowel disease

1.1 Ulcerative colitis: contrast enema, colonoscopy with biopsy, full blood examination and bacteriological cultures
2.1 *Top-and-tail* endoscopy with biopsies, contrast meal and follow-through, full blood examination, liver function tests and stool cultures
2.2 Extent of disease: extra-intestinal signs and symptoms and histology

25 The child with an abdominal mass

1.1 Hydronephrosis.
1.2 Renal ultrasound, nuclear renal scan ± cystoscopy with retrograde pyelogram to distinguish pelvi-ureteric junction obstruction from vesico-ureteric junction obstruction.
2.1 Wilms tumour or neuroblastoma, once ultrasound confirms lesion is solid.
2.2 Biopsy/excision and chemotherapy ± radiotherapy.
3.1 Catecholamine measurements (VMA, HMA) in urine, CT scan, bone scan, bone marrow and FBE; neuroblastoma.
3.2 Prognosis is poor, but some are still curable with chemotherapy and surgery.

26 Spleen, pancreas and biliary tract

1.1 Trauma (handlebar, child abuse, MCA), bile duct stone, idiopathic.
1.2 Serum lipase level, epigastric peritonism and ileus and ultrasound/CT scan that shows swollen gland.
1.3 A cystic cavity secondary to leakage of enzymes from the pancreatic duct.

2.1 Liver function tests, ultrasound and HIDA scan.
2.2 Stone is removed by laparotomy, laparoscopy or ERCP. Biliary atresia requires Kasai portoenterostomy.

27 Anus, perineum and female genitalia

1.1 Perianal abscess with fistula *in ano*: incision of abscess and tract
2.1 Labial adhesions
2.2 Separation with thermometer, paper clip or gentle pressure (± anaesthetic jelly). Regular application of Vaseline and general measures to prevent nappy rash

28 Undescended testes and varicocele

1.1 Risk of subsequent ascent at 5–10 years of age.
1.2 If ascent occurs, orchidopexy is recommended once the testis no longer can reside spontaneously in the scrotum.
2.1 Disorders of sex development, bilateral intra-abdominal testes and bilateral perinatal torsion/agenesis.
2.2 Hormone/chromosome tests: laparoscopy + orchidopexy/orchidectomy.
3.1 6–12 months.
3.2 Not known for certain, but expected to be normal or near normal in most boys with early treatment.

29 Inguinal region and acute scrotum

1.1 Indirect inguinal hernia.
1.2 Inguinal herniotomy – some surgeons would do bilateral operation.
2.1 Torsion of testis or its appendages, epididymitis and idiopathic scrotal oedema.
2.2 No – this affects testis after puberty.
2.3 Exploration of scrotum, R/o appendage or detorsion and fixation of (both) testes.

30 The penis

1.1 No, not necessary; normal washing (externally) is sufficient.
1.2 After 5 years in the majority of boys; partial adherence is still normal up until adolescence.
2.1 Urethral meatus and glans visible in normal child on retraction or traction.
2.2 Topical corticosteroid cream or circumcision.
3.1 Neonatal circumcision not necessary for hygiene, and procedure is dangerous.
3.2 Acceptable standards are general anaesthetic (after 6 months), full surgical technique and adequate post-operative analgesia.
3.3 Bleeding, infection, penile deformity, glanular/meatal ulceration and acute retention (pain).
4.1 6–12 months.
4.2 Fix chordee and put urethra on top of glans; satisfactory cosmetic appearance and minimum complications.
4.3 Extensive mobilisation to fix chordee, foreskin advanced to ventral surface, neourethra constructed, ± post-operative stenting/urinary diversion, one or two stages.

31 Urinary tract infection

1.1 Urine microscopy and culture to confirm UTI and then renal ultrasound. Further tests only if ultrasound scan abnormal
1.2 Low risk of anomaly but still possible
1.3 Perineal contamination secondary to poor perineal hygiene ± constipation with soiling
2.1 Suprapubic aspirate for urine specimen
2.2 Renal and bladder ultrasound scan, MCU ± MAG3 or DMSA scan

32 Vesico-ureteric reflux

1.1 Renal ultrasound and MCU ± DMSA scan
1.2 Unpleasant and invasive but gold standard for vesico-ureteric reflux (VUR) and urethral and bladder anomalies
1.3 Indirect MCU
2.1 Both
2.2 Should not (although scars become visible very slowly and may appear to be occurring later)
2.3 Failed medical treatment, anatomical anomalies and persisting VUR in prepubertal girls

33 Urinary tract dilatation

1.1 Spontaneous resolution is common, especially in the less severe cases. Severe dilatation suggests progressive obstruction requiring treatment.
1.2 Pelvi-ureteric junction obstruction, vesico-ureteric junction obstruction, vesico-ureteric reflux and urethral obstruction.
1.3 Antibiotic prophylaxis, early ultrasound and MCU (<1–2 weeks) and MAG3/DTPA scan (2–6 weeks).
2.1 Vesico-ureteric obstruction, vesico-ureteric reflux and urethral obstruction.
2.2 MCU and renal ultrasound, especially in infants and preschool children. MCU less useful in older children.

34 The child with wetting

1.1 Each predisposes to the other.
1.2 Renal and bladder ultrasound, x-ray ± MCU, ± cystoscopy to document bladder anatomy and cystometry for bladder function.
1.3 Consider surgery for VU reflux if still present, bladder training and antibiotics ± anticholinergics.
2.1 Ectopic ureter in perineum.
2.2 Patch test (wet patch on sheet within 10 min), renal ultrasound and DTPA scan + MRI.
2.3 Heminephroureterectomy
3.1 Occult spina bifida: hairy/pigmented patch, lipoma, sinus and palpable defect in vertebral spines. Absent sacral segments.
3.2 Spinal ultrasound can identify the spinal cord and roots in babies, but later, an MRI is required.
3.3 Clean intermittent catheterisation is the mainstay of treatment.

35 The child with haematuria

1.1 Meatal ulcer
1.2 Soak off scabs, apply Vaseline, leave off nappy and treat nappy rash.

2.1 Ruptured kidney, Wilms tumour and hydronephrosis.
2.2 Renal ultrasound: if minor renal trauma, then treat conservatively unless significant urine leak. Tumour and hydronephrosis: treat condition.

36 Trauma in childhood

1.1 Airway, breathing and circulation; resuscitation; secondary (head-to-toe) survey; continued resuscitation; and definitive care.
1.2 Detailed examination of all body systems to identify all injuries.
1.3 Airway obstruction (from tongue or blood) may depress conscious state.
2.1 Surgical examination and debridement under anaesthetic.
2.2 Tetanus management and excision of devitalised tissue, foreign bodies and blood clot.

37 Head injuries

1.1 See Box 37.1.
1.2 See Box 37.3.
2.1 See Box 37.2.
3.1 Acute left extradural haemorrhage.
3.2 Extremely urgent.
3.3 Determine the GCS, urgent resuscitation and X-match, CT and then craniotomy and removal of collection.
4.1 See Table 37.5.
5.1 Transient/prolonged amnesia after head injuries (HI). No pathology with minor concussion; severe concussion with diffuse axonal injury (swelling on CT with haemorrhage in corpus callosum/dorsal brainstem) may cause a persisting vegetative state.

38 Abdominal and thoracic injuries

1.1 Ruptured spleen.
1.2 ABC, correction of hypovolaemia, CT or U/S to confirm rupture and conservative treatment (unless uncontrollable haemorrhage, which is very rare).
2.1 The girl has a fractured femur and possible intra-abdominal bleeding (liver or splenic rupture). The leg needs an air splint and traction/fixation. After airway control and oxygen, the girl needs resuscitation with fluids and urgent transfusion. CT scan will demonstrate the injury; however, conservative treatment is likely unless there is bowel perforation.

39 Foreign bodies

1.1 No
1.2 No
2.1 Under GA with mosquito forcep inserted via a small incision guided with image intensifier

40 The ingestion of corrosives

1.1 Immediate copious irrigation of face and mouth with water and drinking of copious water/milk.
1.2 Observation to see if drooling/dysphagia is present, and if so, oesophagoscopy to diagnose full-thickness oesophageal burn, which needs treatment. The aim is to prevent oesophageal stricture.

41 Burns

1.1 Remove clothing and immerse affected parts in cold water for 10–20 min. Commence IV (Hartmann's solution). Give IV morphine 0.05–0.1 mg/kg over 5 min. Wrap child in a clean sheet and blanket. Take swabs of nose, throat, faeces and burn. Insert urinary catheter. Give maintenance fluids orally. Start IV rate at 3–4 mL/kg/1% burn area and give the half of this volume over the first 8 h, as Hartmann's solution.
1.2 Yes: Call burns unit for advice and to arrange transfer.
2.1 Possible child abuse by deliberate immersion.
3.1 Possible inhalation injury with airway burn.

42 Neonatal orthopaedics

1.1 Callus of healing fracture of humerus secondary to birth injury.
1.2 Spontaneous healing with full recovery.
2.1 Dislocated hip with soft tissue oedema around capsule. Aspiration of joint revealed pus, consistent with the diagnosis of septic dislocation of hip. Even with drainage, IV antibiotics and abduction bracing, there is a risk of proximal femoral growth arrest and short limb.
3.1 Birth injury to brachial plexus.
3.2 Spontaneous recovery with elbow movements at 6 weeks and shoulder movements normal by 3 months. Failure of recovery by 3 months suggests microsurgical repair is needed.
4.1 *Postural* club foot.
4.2 Normal function after a few weeks of stretching exercises.
5.1 Talipes calcaneovalgus may be associated with developmental dysplasia of hip.
6.1 Serial plaster casts commencing immediately. Surgical release of contracted tendons may be required at 6 months if there is residual deformity.

43 Orthopaedics in the infant and toddler

1.1 The child has bow legs, which resolves over 6–12 months. Monitoring the deformity in the standing position with serial photographs is useful.
2.1 Internal tibial torsion is common with mild bowing. Six-monthly reviews should demonstrate resolution by 3 years of age.
3.1 Developmental dysplasia of the hip.
3.2 Ortolani test with confirmation by hip ultrasonography should have been done at birth, as she had all the risk factors for DDH.
4.1 Cerebral palsy with hemiparesis.
5.1 Osteomyelitis of proximal femur.
5.2 A hot area in the right upper femur.
6.1 Child abuse.
6.2 Fracture of femoral shaft and a strong likelihood of other healing fractures, such as rib, skull and collarbone.
7.1 Osteogenesis imperfecta (type 1): a skull x-ray would show multiple Wormian bones.

44 Orthopaedics in the child

1.1 Internal femoral torsion.
2.1 The gap is not excessive and spontaneous resolution is expected without surgery.

3.1 Simple orthotics may prolong shoe life but make little difference to the condition itself.
4.1 This is likely to be a supracondylar fracture with brachial artery compromise. There is a high risk of Volkmann's ischaemia unless treatment is rapid and appropriate.

45 Orthopaedics in the teenager

1.1 Scoliosis.
1.2 Likely to show an intrinsic curvature ± rotational deformity.
1.3 Less than 20°, observe; 20°–40°, brace; and greater than 40°, spinal fusion.
2.1 Slipped upper femoral epiphysis.
2.2 AP and lateral x-ray; ultrasonography may help to diagnose secondary haemarthrosis.
2.3 Pinning of epiphysis *in situ* to prevent further chronic slippage.
3.1 Overuse syndrome affecting immature retropatellar cartilage.
3.2 No further tests needed if x-ray is normal. Arthroscopy is indicated occasionally.
3.3 Conservative; analgesics, restriction of provocative activities and gentle exercises.
4.1 Osteosarcoma or Ewing's tumour.
4.2 Imaging (plain x-ray, bone scan, CT/MRI) and biopsy.
4.3 Chemotherapy followed by tumour excision ± limb salvage.

46 The hand

1.1 Trigger thumb
1.2 No
1.3 Yes
2.1 Erb's palsy; traction on brachial plexus during delivery and surgery if not resolved after 3 months

47 The breast

1.1 Premature thelarche: remains static or resolves over 6–12 months.
1.2 Only if mastitis occurs: surgery for drainage of abscess (rare at this age).
2.1 Neonatal hypertrophy: clean skin with an antiseptic, and do not squeeze breast (milk production ceases spontaneously). Right breast may have been secondarily infected: needs antibiotics + drainage (carefully).
2.2 Either male or female (female hormones from mother).

48 Chest wall deformities

1.1 Chest wall deformities only cause *subtle* changes in cardiorespiratory function, except in severe deformity.
1.2 A chest x-ray, especially if there is a low protrusion.
1.3 Reassure that there is no major functional limitation, and cosmetic surgery can be done if required (often delayed until adolescence).

49 Lungs, pleura and mediastinum

1.1 Staphylococcus pneumonia and may be developing Staphylococcus empyema.
1.2 Flucloxacillin and gentamicin IV until cultures confirm sensitivity, regular x-ray review, ± surgical drainage of empyema.
2.1 Spontaneous pneumothorax: confirm with x-ray; needle or intercostal catheter if under tension; if no tension, observe in ward; diagnose and treat cause.

50 Vascular and pigmented naevi

1.1 No risk of melanoma.
1.2 Cosmetic treatment if needed.
2.1 Amblyopia of eye, deformity of face throughout preschool years and some residual scar.
2.2 Propranolol orally as first-line management + surgical excision or laser treatment. Occasionally need intralesional steroid injection or interferon treatment.

51 Soft tissue lumps

1.1 This is a naevus sebaceous that needs excision in early childhood.
1.2 15–20% risk of basal cell carcinoma in adulthood, if untreated.
2.1 Keloid: counselling, intralesional steroid injections, pressure via Lycra or silicone gel sheeting.

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Note: Page references in *italics* refer to Figures; those in **bold** refer to Tables and Boxes

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