Handbook Series of
Consent in Pediatric Surgical Subspecialities

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Martin A. Koyle *Editors*

Consent in Pediatric Urology

Springer

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Foreword

This book is one of a handbook series of consent in pediatric surgical specialties. The subject matter is extensive and includes virtually every aspect of open and minimally invasive surgery. The editors have selected experienced and very knowledgeable authors for all of their chapters.

The first chapter "Why and How Is Consent Obtained?" is a powerful introduction to the book. It is written by R. A. Wheeler and has important sections dealing with who should disclose the information, how should consent be recorded, who should provide consent and consent dealing with young people (16 and 17 years of age), children under 16 who can demonstrate their capacity, and children under the age of 16 who lack competence.

Diagrams of the anatomy and surgical procedures are present in many of the chapters. In some, both the open and endoscopic techniques are described. For example, the chapter "Anti-Reflux Surgery" has a lay description of ureteral reimplantation, describing open and endoscopic techniques. In this particular chapter, there are sections "Intended Benefit" and "What Happens Before Surgery". The operative techniques with photographs are well described for open and endoscopic surgery. There is a section "Expected Post-operative Course" with follow-up and expected outcome. Complications of both open and endoscopic surgery are discussed.

Many chapters have sections dealing with the intended benefits as well as general and specific risks and alternatives to the operative procedures.

There are discussions, when appropriate, dealing with assessment of kidney anatomy, conditions, and function using various radiologic modalities.

There is an excellent section "Renal Impairment Surgery" consisting of chapters "Hemodialysis and Peritoneal Dialysis" and "Kidney Transplantation".

In the chapter on Wilm's tumor and other renal neoplasms, there is an extensive section on surgical techniques for radical nephrectomy, partial nephrectomy, and also nephrectomy with minimally invasive techniques.

In the chapter on trauma, not only are complications described, but the incidence of each potential complication is mentioned.

There are excellent discussions regarding urinary continent diversion as well as surgery for fecal incontinence. There is even an interesting section in the chapter "General Laparoscopy" on risks to the operating surgeon and in the chapter "Hydrocele and Hernia" there is another very unique section on iatrogenic injury.

Not only is this a book on *Consent in Pediatric Urology* but in many ways it is a comprehensive discussion of many of the surgical conditions we deal with and their overall management. This is a book which should be read by every urologic surgeon caring for infants and children.

Boston, MA, USA Alan B. Retik

Preface

With advances in evidenced-based medicine and the ability to research medical topics on the Internet, families and patients are increasingly coming to clinic with a thorough understanding of their condition. Consequently, there has been an increasing emphasis on shared decision making in clinical management. The aim of this book is to impart information, often in lay language, that will enable the reader to inform patients and families about the management options and importantly to discuss relevant complications and the likelihood of them occurring.

We are very grateful to all the authors who were kind enough to put a lot of time and effort into writing their chapters. They have skillfully compiled chapters that are informative, up to date, and succinct. As well as being timely.

We would also like to thank Andre Tournois and Melissa Morton at Springer for the advice with this project which has been invaluable to ensure the successful and almost on time completion.

Lastly, and most importantly, we would like to thank our families for supporting us and allowing us the precious time to complete this project.

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Part I
Introduction

Chapter 1
Why and How Is Consent Obtained?

Robert A. Wheeler

'Choice' has achieved high priority in many developed countries, reflecting the fundamental role of autonomy. That is, the right of every citizen to influence their own destiny. In English health care terms, this has two important consequences, and these are shared in many jurisdictions.

Firstly, that there must be agreement with the patient or the parent before any clinical intervention can ensue.

Secondly, that there must be similar agreement before any of the confidential information revealed during clinical management can be further disclosed.

These two agreements can occur only after a formal conversation, during which is disclosed information about the matter itself, its benefits, risks and alternatives. The parent or child must have the mental capacity to engage in the conversation, and be able to make the relevant decision. The final agreement to treatment (or information sharing) is known as consent.

Consent is the legal key that makes both physical intervention and sharing of information lawful. The standard of clinical management that patients receive is judged against several yardsticks.

(i) The most straightforward of these is that clinicians should not touch patients without their consent.

The need for consent for the otherwise unwanted touch is self evident. Unwelcome attentions from another person, who tries to touch you against your wishes, are repellent. There are times when such touches are unavoidable…packed like sardines on the London Underground or jostling fellow shoppers (and their baggage) in the supermarket during shopping frenzies, there is little choice but to resign yourself to being touched, irrespective of your lack of consent.

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But in less frenetic circumstances, there is an absolute understanding that we are entitled to choose who touches us, and when. The patient who is lying on her hospital bed, when suddenly confronted with a surgeon who puts his hand on her abdomen without first asking if he may do so, would justifiably complain that her treatment fell below the reasonable standard she was entitled to expect. Such behaviour is simple rudeness, irrespective of the legal context. However, the legal context is suddenly placed into stark relief when a patient complains that an intimate examination was performed without consent; and further still when such an examination was irrelevant to her clinical presentation.

These latter actions move the lack of consent into the arena of professional disciplinary regulation, as well as civil litigation and potentially criminal prosecution. The medical defence organisations' case reports are a testament to this frequent (and potentially devastating) error of judgement.

(ii) A second standard that clinicians must attain is that they should supply the patient with enough information to make an informed decision as to whether they wish to undergo the proposed intervention. This allows the patient to judge whether allowing themselves to be subjected to an otherwise unwanted touch is worthwhile.

Doctors often fail to appreciate how little patients understand about the consequences of intervention. How many parents or patients appreciate that surgery on the spine may lead them to urinary and faecal incontinence, or failure to move their legs? Or that misdirection of a subclavian needle may lead to a thoracotomy, to arrest the hemorrhage thus caused? Or that a difficult inguinal hernia repair could lead to loss of the ipsilateral testicle?

Trained surgeons are acutely aware of these hazards, but our patients are not. The process of consent, with disclosure of risks and side effects, is designed to allow patients an insight into the risks that they and the surgeon jointly face. Furthermore, patients cannot be expected to anticipate the limitations of treatment, or the possible alternatives. A 14 year old facing the excision of his pulmonary metastases from osteosarcoma may be blissfully unaware that cure is unlikely. If he had known, he might have chosen to accept a different route to palliation, and avoided thoracic intervention. A 15 year old preparing to be a bridesmaid at her mother's wedding tomorrow morning might just elect, tonight, to have her acute appendicitis treated with antibiotics, at least for 24 h….and accept any consequent risk. Disclosing alternatives to treatment may be very important to a patient who is otherwise unaware that they had a choice.

It is very unusual for cases where patients claim that they were not provided with valid disclosure (and thus their consent was invalid) to reach court. However, when this happens, English courts do not rely on expert witnesses to set the standard for what the appropriate disclosure should have been. This is in contradistinction to the great majority of clinical negligence cases, where a medical expert will be asked to set the standard of care,[1] against which the defendant doctor will be judged.

[1] Bolam v, Friern Barnet Hospital Management Committee [1957] 1 WLR 582.

This gives an insight into the importance that the judiciary set on disclosure for consent. Judges put themselves in the shoes of the ‘reasonable’ patient…and enquire what such a person would want to know, before giving consent in the particular set of clinical circumstances. This position was articulated in 1999, and reinforced by the United Kingdom’s Supreme Court in 2015[2], providing the standard for disclosure of risk:

> …Ensure that the patient is aware of any material risks involved in any recommended treatment, and of any reasonable alternative or variant treatments. The test of materiality is whether, in the circumstances of the particular case, a reasonable person in the patient’s position would be likely to attach significance to the risk, or the doctor is or should reasonably be aware that the particular patient would be likely to attach significance to it.[3]

Seeking consent from a person who is fully aware of the clinical risks and benefits should concentrate the mind of both surgeon and patient. If the process has failed to dispel doubts in either mind that intervention is the right thing to do, abandon the procedure, reconsider the situation, and do something different.

Statistics are a valuable when articulating risk to patients. The risk may be tiny, but of great importance when deciding whether or not to have surgery, particularly in the elective setting. In a recent case, the court confirmed the importance of comparative statistics when setting out the therapeutic alternatives that a patient might want to consider in deciding which intervention she should consent for. Faced with a choice between a catheter cerebral angiography and an MR angiogram, the patient was not informed of the comparative risks of stroke.[4] The court held that the patient, as a result, could not provide properly informed consent for the cerebral catheter whilst remaining oblivious to the excess risk of stroke that this entailed.

The most commonly asked question relating to disclosure refers to the importance, or otherwise, of the numeric threshold for risk; how common does a foreseeable risk have to be before we must disclose it to the patient? Doubtless, surgeons are comfortable with ubiquitous numeric thresholds to guide their interventions, and depend upon on plasma levels, physiological or radiological measurements to carry a patient across a threshold from non-treatment to treatment. Perhaps this explains the existence of a common but mistaken supposition by English surgeons that there exists a numeric threshold of improbability beyond which there is no need to disclose; leading them to the awkward but inevitable question of where should such a threshold line be drawn?

One problem with a threshold is that the numerical risk of most complications of therapy is usually low, and may not be caught by a realistic threshold. Is it right that such a threshold should (inadvertently) conceal relevant matters from the putative patient’s consideration?

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[2] Montgomery v Lanarkshire HB (Scotland) [2015] UKSC 11.

[3] Montgomery v Lanarkshire HB (Scotland) [2015] UKSC 11 para 87.

[4] Birch v University College London Hospital NHSFT [2008] EWHC 2237.

Internationally, courts have explored the notion of a numeric threshold. In the 1980, a Canadian court[5] held that a 10 % risk should automatically be disclosed when obtaining consent; in this case, to disclose the possibility of a stroke following surgery. This built on the American concept of a material risk, where a reasonable person in the patient's position is likely to attach significance to the risk.

But since then, courts have steadily distanced themselves from a numeric threshold. An American[6] case determined that a 200/1 complication rate would not equate to a material risk. A 'landmark' English consent case[7] held that Mrs Sidaway, who had suffered spinal cord damage after surgery, failed to prove that a prudent patient would regard a <1 % complication rate as constituting a significant risk.

In 1997, it was held that there was no certainty that an unqualified duty to disclose a risk of around 1 % existed, in the context of a family who were not told that permanent neurological damage could flow from cardiac transplantation surgery.[8] An Australian court [9] found that the failure to warn of the 14,000/1 risk of blindness following ophthalmic surgery fell below the reasonable standard of care.

From the legal perspective, this was the death knell of the numeric threshold. To disclose all risks of this frequency would be impractical. The court was demanding that significant risks should be disclosed, irrespective of the likelihood of occurrence. The UK courts followed this lead in 1995,[10] holding that failure to disclose the risk of spontaneous vasectomy reversal (2,300/1) equated to substandard care.

The explicit switch from a quantitative to a qualitative approach came in a maternity case,[11] when a patient lost her baby. She had reluctantly agreed to the deferral of her delivery, in the absence of full disclosure of the possible consequences of so doing. Lord Woolf, giving the leading judgement, held that it was not necessarily inappropriate to fail to disclose a risk in the order of 0.1–0.2 %; but that the correct standard was to disclose '…. A(ny) significant risk which would affect the judgement of the reasonable patient'.

As in the general rule for disclosure in the United Kingdom, the most recent Supreme Court decision in Montgomery makes it clear that the assessment of whether a risk is significant cannot be reduced to percentages. The significance of any given numerical risk is '…likely to reflect a variety of factors besides its magnitude, including the nature of the risk, the effect which its occurrence would have upon the life of the patient; the importance to the patient of the benefits sought to be achieved by the treatment, together with the alternatives available, and the risks involved in those alternatives'.[12] The assessment thus becomes fact sensitive, rather than founded on a numeric threshold.

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[5] Reibl v Hughes. *DLR* Canada 1980; 114: 11.

[6] F v R. *South Australian Supreme Court* 1983;33: 189.

[7] Sidaway v Board of Governors of the Bethlem Royal Hospital. *All England Reports*, House of Lords 1985; 1: 643.

[8] Poynter v Hillingdon Health Authority. *Butterworths Medical Law Reports* 1997; 37: 192.

[9] Rogers v Whittaker. *CLR HC Australia* 1993;175: 479.

[10] Newell v Goldenberg. Medical Law Reports 1995;6: 371.

[11] Pearce v United Bristol Healthcare Trust. *Butterworths Medical Law Reports* 1999;48:118.

[12] Montgomery v Lanarkshire HB (Scotland) [2015] UKSC 11 para 89.

Who Should Disclose the Information?

There is no clear rule in England. However, since it is very clear that significant risks should be disclosed, perhaps a surgeon familiar with the procedure is best placed to provide the disclosure. Some surgeons prefer to delegate this duty to trainees or nurses who are unable to perform the intervention, but who are armed with an information sheet; and have been 'trained' in consenting patients. One wonders whether they would wish their own families to be treated in this way. It is recommended that consent, and the disclosure that makes it valid, should be taken by the person who is about to perform, or at least who is capable of performing the procedure.

How Should Consent Be Recorded, and for What Procedures?

Consent is necessary for any intervention, but the form of consent, and it's mode of recording, differ widely in surgical practice. Individual hospitals often have their own view of what type of procedures merit written consent, and it is prudent to adhere to local rules. However, from a broader perspective, there is no doubt that oral consent is good consent. There is no English statutory requirement to obtain written consent for surgery, although written consent is required for fertility treatment.[13]

But the reality remains; that the existence of oral consent is very hard to prove in retrospect, and this difficulty is proportionate to the time that has elapsed since the intervention in question took place. For this reason, our Department of Health advocates written consent for all forms of surgery.[14] However, neither the form of the written consent, nor the definition of 'surgery' is stated.

Which interventions are 'so serious' that formal consent form must be completed? It may be difficult, months later, confidently to recall that you obtained consent orally, and what you disclosed in obtaining it. If in doubt, make a note in the case notes. This will usually have been decided for you, codified by the local Trust's consent policy. Commonly, any surgical or interventional radiology procedure will require a consent form, as will any procedure which requires local or general anaesthesia. If in doubt, it is prudent to use a form, but please ensure that it is properly filled in. The disclosure that leads to valid consent is infinitely more important than the signature.

Indeed, errors such as; failure to ensure that the patient has capacity; inadequate disclosure of information; or failing to ensure that the patient provides consent voluntarily; all these invalidate the consent, irrespective of a signature: 'Consent expressed "in form only" is no consent at all'.[15]

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[13] Human Fertilisation and Embryology Act 1990 Sch3.

[14] DH, Reference guide to consent for examination or treatment, Second edition, 2009.

[15] Chatterton v Gerson [1981] 1 All ER 257 @ 265.

For this reason, providing evidence by a handwritten entry in the case notes becomes potent evidence of a diligent approach to consent. This is also a prudent move when you have completed a consent form with a patient, but still remain anxious that the form does not wholeheartedly reflect the conversation, or the inherent uncertainties of the procedure. You may have spent an hour with some parents, weighing up the risks and benefits of excising their infant's thoracic neuroblastoma. The local consent form gives precious little space for a full account of your deliberations, or a relevant diagram which better describes the surgical dilemma. Putting your thoughts on paper will allow you to state your position, beyond doubt; and remember it years later.

Oral consent is valid, but writing provides a record. Get consent for any intervention, but take a proportionate approach as to what form of documentation is necessary.

Who Should Provide Consent?

From the English perspective, a child is a citizen who has not yet reached 18 years of age. Legal synonyms include 'minor' and 'infant'. The latter is instructive, since it is derived from the Latin: *Infans*, unable to speak. This reflects the legal rules which prevent children from speaking for themselves in court, although this impediment has been at least partly addressed over the last two decades. Nevertheless, it begs a fundamental question, as to whether children can provide their own consent, or whether they must depend upon their parents to provide it for them.

People under 18 years are considered in three broad groups in England and Wales. Bearing in mind that the law is different even in Scotland, a country nevertheless within the United Kingdom, it is hoped that readers outside these small islands will forgive a parochial approach, since a comprehensive international review of child law is not feasible here.

Young People: 16 and 17 Years of Age

Citizens of 16 and 17 years of age are described as young people, presumed to have the capacity to provide consent for surgical, medical and dental treatment. (*Capacity* rather than *competence* is used to describe this attribute, a convention only necessary to acknowledge the words of the Mental Capacity Act 2005, which applies to those of 16 years and above).

The presumption of capacity in young people was made possible by a law enacted in 1969,[16] which recognised that the 'lifestyle' decisions that teenagers were taking, irrespective of the law, contrasted sharply with the age of majority (21 years) at the

[16] Family Law Reform Act 1969s8.

time. The new law reduced the age of majority to 18 years, and introduced the presumption of capacity for 16 and 17 years olds.

The new law did not extend this right to consent for research, or to interventions that do not potentially provide direct health benefit to the individual concerned. However, if competent along ‘Gillick’ lines, a young person may be able to provide consent for these activities.

Young people of 16 and 17 are thus able to provide consent for treatment in absence of their parents. However, the parental right to provide consent for treatment prevails until the end of childhood. This has the effect of providing a ‘safety net’; allowing a 16/17 year old the opportunity of consent for herself; or deferring to her parents, if she sees fit. Once the young person reaches adulthood on her 18th birthday, her parents’ rights evaporate. For the rest of her life, she alone can provide consent, either directly, in person; or in some circumstances by a proxy method.

If parents and a young person disagree over consent for surgery, it is wise to exercise caution. In the situation where a 16/17 year old wishes to exercise his right to consent, whilst his parents oppose his decision, then you would be entitled to rely on his consent. However, it would be important to understand the basis for their disagreement. For instance, if you suspected that the young person lacked capacity, you should challenge the presumption that he is able to provide consent for himself. This can simply be done by establishing whether he understands the relevant information; can retain the information, believe it, weigh it up, use it….and communicate his decision. If he can, then he has capacity. But it is still wise to tease out where the problem lies, since this is an unusual situation, and it would be in the young person’s interests to resolve the issue before surgery, if that is feasible.

The problem, reversed, is of a young person who refuses treatment, but who is accompanied by a parent who provides consent. Valid parental consent will make the procedure ‘legal’, but as with the situation of consent withdrawal, you will have to make a clinical judgement as to whether proceeding with the treatment against the young person’s wishes is both practicable, and in her interests. In summary, it is recommended that an elective procedure should be abandoned until the dispute is resolved. If emergency treatment is required, but could be administered in a different way which was still consistent with her best interests, that alternative should be explored. If her life or limb is threatened, and there is no choice but to provide a definitive operation, then reluctantly, you may feel the need to restrain and proceed. A supracondylar fracture of the humerus that has resulted in an ischaemic hand could be an example of this situation. It should be noted that in reality, the amount of resistance that a child of any age puts up is usually inversely proportional to their malaise and discomfort. In the gravely ill, refusal is rare.

There are those who are gravely ill, but need urgent rather than emergency treatment. If a 16/17 year old in this category refuses treatment for the preservation of her life, such as the transfusion of blood,[17] or feeding[18] (in anorexia), English courts have invariably chosen to override the young person’s autonomy, providing an order

[17] Re P (Medical Treatment: Best Interests) [2004] 2 All ER 1117.

[18] Re W (A Minor) (Medical Treatment: Court’s Jurisdiction) [1992] 3 WLR 758.

which allows lawful provision of the treatment against the child's wishes. This either upholds the parental wishes for treatment, or overrides parental refusal. These cases are rare, but the timescale within which the clinical treatment needs to be provided usually allows sufficient time (perhaps measured in hours) for the court to be contacted, providing the surgeon with the necessary authority.

Children Under 16 Who Can Demonstrate Their Capacity

Depending on their maturity and the intervention that is proposed, children from a young age may be able to provide independent consent. A 4 year old may be able to consent to a pulse measurement; a 7 year old to a venepuncture; a 10 year old to the removal of an early stage appendicitis. It is not suggested that the parents should be excluded from this process. Rather, it is for the family as a whole to decide what part the child's potential capacity should play in the consenting process. But the involvement of children in this process will strengthen the therapeutic relationship, and is to be encouraged.

A child's previous experience is of great importance in this context. It is submitted that following the very recent diagnosis of leukaemia, a 13 year old, who has been healthy up to this point, will be so horrified by the dissolution of his comfortable and well organised life as to be incoherent, incapable of consenting for the necessary tunnelled central venous catheter (CVC). Contrast this child with a 10 year old on the same ward; suffering relapsed leukaemia. She has already undergone three line insertions and two removals. She knows (effectively) everything there is to know about CVC placement, complications and disadvantages. Now facing her fourth insertion, she may be competent to provide independent consent.

Therefore, it is important objectively to determine whether a child of 15 years or younger has capacity to provide independent consent for the proposed intervention.

For this assessment, the Gillick test is used, derived from a landmark case where it was established that a child who is competent to provide consent can do so, independently of her parents. The test requires that the child has sufficient understanding and intelligence to enable them to understand fully what is involved in a proposed intervention.[19] Thus, if a child can understand:

- That a choice exists
- The nature and purpose of the procedure
- The risks and side effects
- The alternatives to the procedure; and is able:

- To retain the information long enough…
- To weigh the information…..
- To arrive at a decision
- *And* to be free from undue pressure

[19] Gillick v West Norfolk & Wisbech AHA [1986] AC 112.

Then she would be deemed competent for the proposed intervention. It will be seen that competence rests on intelligence, maturity and experience. Not on age.

During the Gillick case, an additional set of guidelines were suggested by Lord Fraser, specifically for doctors who assist with reproductive decision-making by children under 16. It should be noted that these do not replace the Gillick test, nor are they synonymous with it.[20]

Gillick provides a high threshold for consent, consistent with public policy. It would be highly undesirable to allow incompetent children to provide consent for interventions which they could not fully understand. The fact that a child has to 'prove' their competence places a barrier to children that is not immediately faced by adults, whose capacity is presumed. One can only speculate how many adults would 'pass' the test in *Gillick*.

The Gillick competent child does not enjoy an equal right to refuse treatment. Only those cases in which the refusal of life- saving treatments in these children is at issue have reached the court. But given this opportunity, courts have resolutely denied the (otherwise) competent minor the right to choose death. A 15 year old girl[21] refusing her consent for a life-saving heart transplant had her refusal overridden by the courts. M's reason was that she 'would rather die than have the transplant and have someone else's heart…I would feel different with someone else's heart…that's a good enough reason not to have a heart transplant, even if it saved my life…. The court authorised the operation, as being in her best interests. In another case,[22] a 14 year old girl with serious scalding required a blood transfusion. She was a Jehovah's Witness, and refused the treatment. The court found that even if she had been Gillick competent, her grave condition would have led the court to authorise the transfusion. As it was, the girl was unaware of the manner of death from anaemia, and was basing her views of on those of her congregation, rather than on her own experiences. For these reasons, she was judged incompetent to make this decision for herself.

It must be remembered that the vast majority of Gillick competent children who refuse treatment are refusing relatively trivial procedures. You would be entitled to rely upon their parent's consent if necessary, but it is a matter for clinical judgement whether the procedure could be deferred, to allow the child further time to consider, and become reconciled with what is likely to be an inevitable outcome. The problem of refusal in Gillick competent children is dealt with in the same way as for the 16 and 17 year age group, above.

Children Under the Age of 16 Who Lack Competence

For children lacking competence, a person with parental responsibility has the right to provide consent for treatment.

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[20] Wheeler RA. Gillick or Fraser? A plea for consistency over competence in children BMJ 2006 332 807.

[21] Re M (Medical Treatment: Consent) [1999] 2 FLR 1097.

[22] Re L (Medical Treatment: Gillick Competency) [1998] 2 FLR 810.

Who Can Provide Consent in England and Wales?

The child's mother (the woman who gave birth to the baby, rather than the person who provided the egg from which the baby was conceived, if different) has parental responsibility automatically. The child's father gains parental responsibility automatically if married at the time of the birth registration. Since 2003, unmarried fathers achieve parental responsibility by their registration of the birth. Alternatively, parental responsibility can be acquired by the unmarried father; either with the agreement of the child's mother, or by application to a court, or by subsequent marriage to the mother.

Parental responsibility is passed to adoptive parents on legal adoption. It may be shared with guardians appointed by parents; with local authorities; and is linked to various legal orders.[23]

The person with parental responsibility who provides consent for a child's surgery must act in the child's best interests in so doing. These are usually self evident, and the agreement between parents and surgeon is reached after full disclosure of the relevant information.

This agreement is not invariable. In a case[24] concerning a child with biliary atresia, the clinicians wished to perform a liver transplant, and considered the prospects of success to be good. The parents refused their consent, on the grounds that the surgery was not in the child's best interests. The Court of Appeal held that the assessment of the child's best interests went wider than the narrower medical best interests, and that T's connection with his family held great weight in this regard. Accordingly, the court refused to enforce the hospital's request that the mother would bring T in for surgery. The judgement could be criticised, in failing to differentiate between the interests of the child and those of his mother. However, the case provides an example of the balancing act performed by courts, a common activity when there are disputes over the best interests of the child.

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[23] For a full account see Bainham A, *Children*: *The Modern Law*' 2005 Family Law, Jordan Publishing, Bristol.

[24] Re T (Wardship: Medical Treatment) [1997] 1 FLR 502.

Part II
Open Surgery of the Upper Urinary Tract

Chapter 2
Open Nephrectomy

Rachael Clark, Matthew Boal, and Mark Woodward

Key Points

- With the advent of laparoscopic surgery, simple open nephrectomy is rarely performed [1]
- The retroperitoneal approach is preferable to the transabdominal approach
- Conversion of laparoscopic to open nephrectomy is usually achieved by joining or extending port site incisions

Specific risks of the procedure:

1. Hemorrhage
2. Bowel or pancreatic injury
3. Pneumothorax
4. Ileus
5. Wound infection
6. Wound bulge or pain

Lay Description

Open Nephrectomy (ON) is an operation to remove the kidney via a single larger cut in the abdomen.

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Intended Benefits

The benefits of ON include prevention of urinary tract infection, a reduction in the risk of developing high blood pressure, removal of a potential malignancy and in the case of emergency conversion from a laparoscopic nephrectomy, to stop bleeding.

Technique

An ON is performed in the operating room under general anaesthesia and there are a number of different approaches and incisions. The retroperitoneal approach is preferable as avoidance of the peritoneal cavity reduces the risk of formation of intraperitoneal adhesions and recovery is generally quicker.

Retroperitoneal Approach

Open access to the retroperitoneum can be achieved through loin, lumbotomy, and anterior incisions. A loin (flank) approach has advantages of good access to renal parenchyma and collecting system but exposure of the renal pedicle may be difficult [2].

A dorsal lumbotomy approach allows rapid access without cutting muscle and is useful for bilateral procedures without patient repositioning [3]. An anterior subcostal/transverse approach gives the surgeon good access to the renal pedicle.

Transabdominal Approach

These approaches are generally used in suspected malignancy or trauma. Transverse, subcostal or midline incisions are most frequently employed, although occasionally a thoraco-abdominal incision may be used e.g. for a large tumour [4].

Approaches in Specific Circumstances

Converting from laparoscopic to open nephrectomy involves making an incision between port sites in a subcostal loin incision. An anterior approach with laparoscopy can either be converted to a midline laparotomy, or again at the port site with best access.

A pelvic kidney requires an extraperitoneal iliac fossa approach. An anterior subcostal extraperitoneal incision can be used for hemi-nephrectomy in a horseshoe kidney, or transverse supraumbilical transabdominal approach for bilateral access [5].

How Is Open Nephrectomy Performed?

Gerota's fascia is opened and the perinephric fat dissected to expose the vascular pedicle and ureter. The vessels are isolated, ligated and divided. The kidney is then fully mobilised and the ureter followed distally as far as possible prior to ligation and division.

How Is the Wound Closed?

Following hemostasis, the wound is closed in layers. A subcuticular layer is used for the skin. Wound drains are not generally required.

Postoperative Expected Course

Most children will have local anaesthetic infiltration of the wound to reduce initial postoperative pain and may have a morphine infusion via nurse/patient-controlled analgesia (NCA/PCA) for 24–48 h depending upon the incision used. A urinary catheter is not usually necessary. With a retroperitoneal approach, oral intake can recommence immediately; with a transperitoneal approach, oral intake may need to be delayed for 24–48 h if there is an ileus. Post-operative antibiotics may be required in certain situations. Discharge usually occurs 48–96 h following surgery.

Follow Up

Follow up is dependent on the indication for ON, but patients will be usually be assessed in clinic 3 months after surgery.

Risks of Procedure

Complications are uncommon in simple ON in children. There are no recent series reporting complication data relating to open nephrectomy in children, but extrapolating from laparoscopic nephrectomy, complication rates would be expected to be <1 % for all listed complications [6–10].

Intra-operative

Hemorrhage

Intra-operative hemorrhage is very unusual in ON in children, and blood is usually only cross-matched if the surgeon anticipates difficulties, e.g. XGP nephrectomy.

Bowel Injury

The duodenum is particularly at risk in right nephrectomy and the colon can be damaged on either side. If bowel is injured, it should be debrided and sutured directly.

Pancreatic Injury

If recognized intra-operatively, injury to the tail of pancreas is best managed with partial amputation to avoid pancreatic fistula.

Pneumothorax

The pleura may be breached during ON. The defect is closed, and a chest drain left if required.

Early Post-operative Complications (<30 days)

Pancreatic Fistula

This can produce fluid discharge from the wound, is diagnosed on ultrasound or CT, and percutaneous drainage may be required. The majority of fistulae close spontaneously.

Ileus

Ileus is more commonly encountered following transabdominal incisions.

Wound Infection and Dehiscence

Superficial or deep wound infections can occur. Wound dehiscence is rarely encountered.

Secondary Hemorrhage and Hematoma

The presentation depends on the severity of the bleed. Treatment can be conservative or by radiological or surgical drainage depending on the extent.

Late Post-operative Complications (>30 days)

Wound Bulge

Loin incisions are frequently accompanied by postoperative wound bulge, especially in infants, and this usually resolves spontaneously.

Pain

Although extremely rare in children, chronic wound pain can be encountered after any incision but is more common after loin approaches.

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Chapter 3
Partial Nephrectomy

Courtney Shepard and John Park

Name of Procedure

Partial Nephrectomy.

Lay Description

An operation to remove part of one kidney and preserving the rest.

Intended Benefit

Partial nephrectomy removes the non-functioning or abnormal portion of a kidney (caused by either blockage, reflux, blood vessel anomaly, or inflammation) and thus decreases the risk of urinary tract infections, high blood pressure, or pain. It is sometimes used to remove a kidney mass that could be malignant [5].

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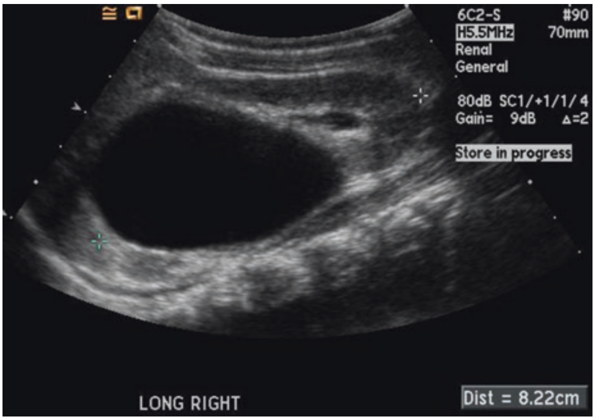
Technique

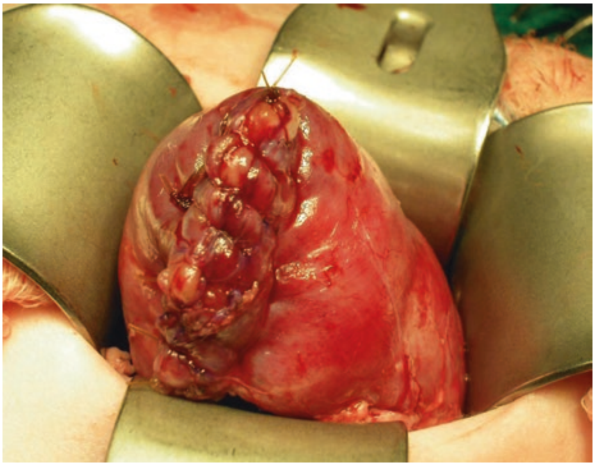
The surgery is performed under general anesthesia. If open approach is used, there is a transverse incision on the side at the level of the lower ribs. If laparoscopic approach is used, there are typically three to four small incisions, usually not much longer than an inch, either on the front of the abdomen or in the back. Once the entire kidney is exposed and visualized, the abnormal portion to be removed is carefully identified from the normal portion to be preserved. Only the blood vessels feeding the abnormal portion are carefully divided, controlling the bleeding. During the removal of the abnormal portion, surgeons sometimes will temporarily occlude the blood supply to the entire kidney to minimize the blood loss during the kidney division. Once the abnormal portion is removed, the remaining kidney is carefully repaired to prevent bleeding and urine leak. In many cases, the abnormal kidney portion has its own separate urine drainage tube (called ureter) that needs to be divided toward the bladder. Often numbing medicine is injected around the surgical incisions to help with pain after waking up, or the anesthesiologist may offer other methods of pain control depending on your child's age and anatomy. After surgery, your child may have a small drain near the incision (positioned near the partial nephrectomy location to the outside) as well as a catheter draining the bladder.

Two Illustrations

1. Renal ultrasound demonstrating an abnormal upper pole of duplex kidney (has two separate ureters) caused by congenital blockage down below. This segment has no function and is to be removed.



2. Surgical picture of remaining lower half of the kidney after removing the upper pole segment.



Postoperative Expected Course

Your child is admitted to the hospital after the surgery. On the night of the surgery, your child may start drinking fluids and possibly eat solid food, although many children are too tired and may be nauseated from anesthetics to eat much the night of surgery. The morning after surgery, your child may eat and drink as usual, although you may notice a decreased appetite for few days, which is normal. Your child may start getting up and move around on the morning after surgery. Narcotic pain medication and acetaminophen are usually given as needed for pain. The bladder catheter is often removed on either the day after surgery or the following day. The drain is removed after that if the output remains low. Usually the hospital stay is 2–5 days.

Follow Up

You will see your surgeon to make sure the incision is healing well within 2–6 weeks after surgery. You may have other follow-ups after that, often with a kidney ultrasound or other imaging test, depending on the reason for the surgery and any other abnormalities of the kidneys, ureter, or bladder.

Risks of Procedure

General

Up to 20 % of patients undergoing this surgery can have complication, although they are usually minor and do not require any further surgeries [10].

Specific

– Conversion to open: If this surgery is done laparoscopically, there is approximately 20 % chance that the surgeon may have to make a larger incision to perform a traditional open partial nephrectomy. This is usually due to unusual anatomy of the kidney that makes it difficult or unsafe to perform laparoscopically. It will not change long-term outcomes [1–6, 7–8, 11].
– Urine Leak: Occasionally the part of the kidney that is left behind may leak some urine into the abdomen after the surgery, and this is manifested by increase in drain output. If this happens, usually the drain and bladder catheter are left for a longer period, allowing it to heal. Rarely is a small additional procedure required (such as placement of a temporary stent that goes from the kidney, through the ureter, and into the bladder). Urinomas (collection of urine around the kidney) occur in 0–10 % of cases [5, 6, 9].
– Injury to surrounding structures (such as the bowel, spleen, liver, the remaining portion of the kidney, or the ureter): These injuries are rare and usually can be fixed during surgery. Rarely does the entire kidney need to be removed. These injuries occur in 0–12 % of patients [5, 9].
– Bleeding: The kidney has a large blood supply, and so there is always risk of bleeding requiring a blood transfusion or another operation. This is rare, occurring in 0–5 % of patients [5, 9].
– Urine infection: Because the kidney is operated on, along with multiple tubes used (drain and bladder catheter), there is a risk of developing a urine infection. Antibiotic is used in the operating room and for a short period after the surgery to minimize the risk. There is 0–8 % risk of developing a urinary tract infection [5, 6, 9].
– Incisional Hernia or muscle bulging: There is a small risk of developing a weakness at the sites of surgery. This is rare, occurring in 0–7 % of patients [5].

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Chapter 4
Ureteropelvic Junction Obstruction

Amy Hou

Name of Procedure

Pyeloplasty.

Lay Description

Ureteropelvic junction obstruction is the most common cause of hydronephrosis in infants. It occurs in 1 in 500–1250 live births. It accounts for almost 50 % of neonatal hydronephrosis cases. It is a narrowing of the outlet from the renal pelvis, whether intrinsic or extrinsic, which prevents urine from emptying out of the kidney efficiently. The diagnosis is may be found on workup of prenatal diagnosis. In older children, symptoms may be what bring the diagnosis to light. Older children may present with episodic flank or abdominal pain and/or cyclical vomiting. Twenty-five percent of children may have hematuria. Some patient will present with hypertension, thought to be a renin-mediated hypertension [1].

A diuretic renogram is used to confirm the diagnosis of obstruction in almost all patients. In these cases, the patient is sometimes followed by serial ultrasounds to monitor for changes in the degree of hydronephrosis, to determine if findings on imaging are a clinically significant obstruction and if surgery is required. Surgery is performed in the setting of pain, infection, loss of renal function or worsening hydronephrosis.

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Intended Benefit

The goal of surgery is to remove the area of abnormal narrowing and improve drainage of urine out of the kidney. The ultimate goal is to avoid deterioration of renal function in the obstructed kidney. The success rate of correcting the obstruction, with pyeloplasty, is over 90 %, and as high as 98 %. Adult series have demonstrated persistent success of the surgery in 5–15 year follow-up studies.

Technique

There are multiple approaches that can be used to access the kidney and ureter. Once the abdomen is entered, the kidney is mobilized so that the renal pelvis and ureteropelvic junction is exposed. Care must be taken to look for a crossing lower pole vessel, which may be the cause of the obstruction, rather than an intrinsic narrowing of the ureter itself. The most commonly performed repair is a dismembered pyeloplasty. In this repair, the renal pelvis is opened and the apex of the future ureteropelvic junction marked with a stitch, which can also be used for traction. The pelvis is examined to ensure the new ureteropelvic junction will be in a dependent position, for optimal drainage of urine. The proximal ureter is then mobilized and the dysplastic segment excised sharply. The normal appearing ureter is then spatulated, to allow for a larger funnel when reconstructing the new ureteropelvic junction. The ureter and renal pelvis is then reapproximated in a running fashion or interrupted fashion with absorbable suture. Prior to completion of the closure, a ureteral stent of some form is placed, per surgeon preference. Sometimes a drain is placed around the ureteropelvic junction repair rather than in the repair. Again, this is by surgeon preference.

Comparisons comparing the safety and advantages of different approaches have been studied extensively. Varda et al. [2] examined outcomes and cost of open vs laparoscopic vs robotic approaches in pediatric pyeloplasties and found the difference in complication rates between the different surgical approaches was not statistically significant. There are differences in length of stay, need for analgesics postoperatively in the different modalities, with an advantage in the laparoscopic and robotic group. A 2011 meta-analysis [3] also demonstrated no difference in success rates, but confirmed an advantage of shorter hospital pain, decreased pain medication requirement with laparoscopic and robotic surgical approaches. Operative times have been shown to be lower with robot-assisted pyeloplasties when compared to standard laparoscopic pyeloplasties [4].

The use of internal vs external stenting has been reviewed extensively, without demonstration of superiority of one method over the other. Multiple studies have shown no difference in long-term outcome [5]. Lee et al. [6] demonstrated equal outcomes when using an internalized double-J stent and externalized nephroureteral drains. A 70 % higher rate of bacterial colonization with indwelling stents has been reported, with an associated increase in the risk of urinary tract. In unstented/perinephric drain group, a higher rate of urine leak and ileus was seen, although this did not quite reach statistical significance [7].

Postoperative Expected Course

The patient is typically admitted overnight, with an indwelling urethral Foley catheter, to maximize drainage of urine downstream. Pain management for the first night often involves intravenous medications, along with additional medications given for control of bladder spasms, from the Foley catheter and the ureteral stent, if present. If the patient is tolerating enteral nutrition and abdomen is not abnormally distended, the Foley catheter is removed the following day and the patient is instructed to urinate on a regular, scheduled basis. Medications to minimize bladder and ureteral spasms are continued, sometimes on a scheduled basis. The patient can often be discharged home the day after surgery, with a perinephric drain or ureteral stent/drain in place.

Follow Up

Depending upon the type of drainage placed during surgery, the patient will either follow-up in clinic or in the operating room. If there is a perinephric drain or a nephroureteral drain, both of which would come out through the skin near the incision, the patient would return to clinic in 7–14 days, per surgeon preference, for removal of the drain. An indwelling stent is often left in place for a longer period of time, often 4–6 weeks. Indwelling stents require another visit to the operating room, for cystoscopy and removal of the stent from inside of the bladder.

Once the drain is removed, the patient will follow-up in the next 6–12 weeks with a renal ultrasound, to assess for hydronephrosis. It is not unexpected for some degree of persistent hydronephrosis postoperatively. The patient will have repeat renal ultrasounds to monitor for improvement of hydronephrosis over time, and also to monitor for worsening of hydronephrosis, which may signify failure of the repair and recurrent ureteropelvic junction obstruction. Some surgeons will repeat a nuclear renogram to functionally evaluate the drainage from the kidney after the surgical repair, to confirm success or failure of the surgery.

Risks and Complications

Risks of surgery include urinary tract infection, urine leak, stent or drain migration, and failure of the repair leading to recurrent ureteropelvic junction obstruction. The majority of complications present themselves in the first year after surgery [8].

Early Complications

The most common early post op complications are: urinary tract infections that occur approximately 3 % of the time, wound infections that can occur in about 2 % of patients even when prophylactic antibiotics are used and finally urinary leaks which are seen in 3–8 % of patients, typically more common in patients who do not have a ureteric stent placed [8]. The majority of the urinary leaks can be managed with observation and urinary drainage either with a bladder catheter or a wound drain. In rare occasions it is necessary to place a ureteric stent or a nephrostomy to help with drainage. A quick diagnosis is imperative as a urinary leak if left can lead to fibrosis and consequently a higher rate of surgical failure with restenosis.

Recurrent Stricture/Obstruction 0.8–3 %

When recurrence is examined at 1 year postop, the rate has been reported at 0.8–1 % [9]. The rate of recurrent obstruction increases as follow-up is extended but remains low [10]. Presentation is most often worsening hydronephrosis on follow-up. However patient may present with pyelonephritis or abdominal pain.

One retrospective review with 14-year follow-up found that the long-term risk of recurrent obstruction after an initial postoperative normal diuretic renal scan was very small. Young age at initial surgery, prolonged urinary diversion and missed diagnoses at time of initial surgery (crossing vessel) have all been found to be significant risk factors contributing to failure of the first pyeloplasty [8]. Several studies have noted a slightly increased complication rate with the dorsal lumbotomy approach, which was often statistically significant [11].

Hypertension

A Korean study [12] with post-pubertal follow-up of patients who underwent pyeloplasty found a 12.7 % rate of hypertension. The mean time to diagnosis of hypertension was 15.7 years with the number of cases diagnosed peaking between 15 and 20 years of age. In this study, cases of hypertension were defined as requiring hypertension, with baseline elevated renin aldosterone levels, consistent with a hypertension of renovascular origin.

Management of Recurrent Ureteropelvic Junction Obstruction

Endopyelotomy

Success rates of retrograde endopyelotomy have been reported to be between 20 and 60 % [13]. Faeber et al. [14] had initially reported a success rate of 80 %, with an antegrade approach, attributed to their use of the cold knife for incising the stricture, rather than cautery. The Toronto group demonstrated a lower success rate, of 13–52 % [15]. They found that length of stricture was important in success with this approach. Strictures with a mean length of 5.8 mm were successful. Mean ureteral segment length of 10.1 mm was associated with failure with endopyelotomy. One single institution review found three out of ten patient who underwent secondary endopyelotomy ultimately required redo pyeloplasty [16]. The success rate with a redo pyeloplasty is undisputedly better. However endopyelotomy is a safe option and may be desired by some families as a first step, as it is less invasive.

Redo Pyeloplasty

Published studies on open redo pyeloplasties report success rates of 75–100 %. Review of outcomes in a single institution for failed pyeloplasties demonstrated much greater success with redo pyeloplasty (92 %) or ureterocalicostomy (100 %) compared to endopyelotomy (13–52 %) [15, 17]. There had been a 2.5–4.5 year interval between the initial pyeloplasty and the secondary procedure. Piaggio et al noted success with redo pyeloplasty using a laparoscopic approach [18]. Leung et al. [19] also found that a redo pyeloplasty was superior to endoscopic interventions. They performed the redo procedure open, laparoscopically and robotically. There were redo pyeloplasties that required a third pyeloplasty in this study, resulting in a lower success rate of 50 % in their pyeloplasty group. They had only a 25 % success rate with endoscopic intervention for a failed initial pyeloplasty.

When counseling families, it will be necessary to advise them of the difference in outcome with secondary procedures, if complications arise and further intervention is needed.

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Chapter 5
Antireflux Surgery

Angela M. Arlen, Anthony A. Caldamone, and Andrew J. Kirsch

Name of Procedures

1. **Ureteral reimplantation**
2. **Endoscopic correction of vesicoureteral reflux**

Lay Description Ureteral Reimplantation

The kidneys drain urine into the bladder via the ureters. Ureteral reimplantation is an operation to correct vesicoureteral reflux (VUR), or abnormal back flow of urine from the bladder into the kidneys. The procedure involves re-tunneling the ureter(s) into the bladder to prevent urinary reflux.

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Lay Description Endoscopic Injection

Endoscopic injection is performed by placing a cystoscope into the bladder through the urethra (urine passage tube), and there is no incision. The procedure corrects reflux by injection of a bulking substance where the ureter enters into the bladder; this prevents flow of urine back into the kidney.

Intended Benefit

There is a natural tendency for reflux to improve or resolve over time; however, there remains a subset of children with persistent VUR or with associated kidney infections who are at risk for potential harm secondary to kidney infection and injury. VUR treatment goals include prevention of febrile urinary tract infections, prevention of kidney injury, and minimizing morbidity of treatment and follow-up [1]. The key focus in selecting patients for surgery is identifying those children unlikely to outgrow VUR and those at greatest risk for recurrent kidney infections [2].

What Happens Before Surgery?

Voiding cystourethrogram (VCUG) is the mainstay of VUR diagnosis, and is most often performed after a child has a febrile urinary tract infection or is found to have hydronephrosis (dilation of the kidney) on ultrasound. Children with bladder or bowel dysfunction (such as infrequent urination, urinary incontinence or constipation) should have those symptoms addressed prior to surgery, as they can lower the likelihood of a successful operation. A urine culture should be obtained before surgery. If this is not possible, a urinalysis the morning of surgery will alert the surgeon to the possibility of bladder infection.

Ureteral Reimplantation

Operative Technique

Open ureteral reimplantation is performed under general anesthesia, and typically takes between 1 and 3 h to complete depending on type of repair, whether it is one or both ureters, and patient age. Various open reimplantation techniques have been described including intravesical (which involves opening the bladder)

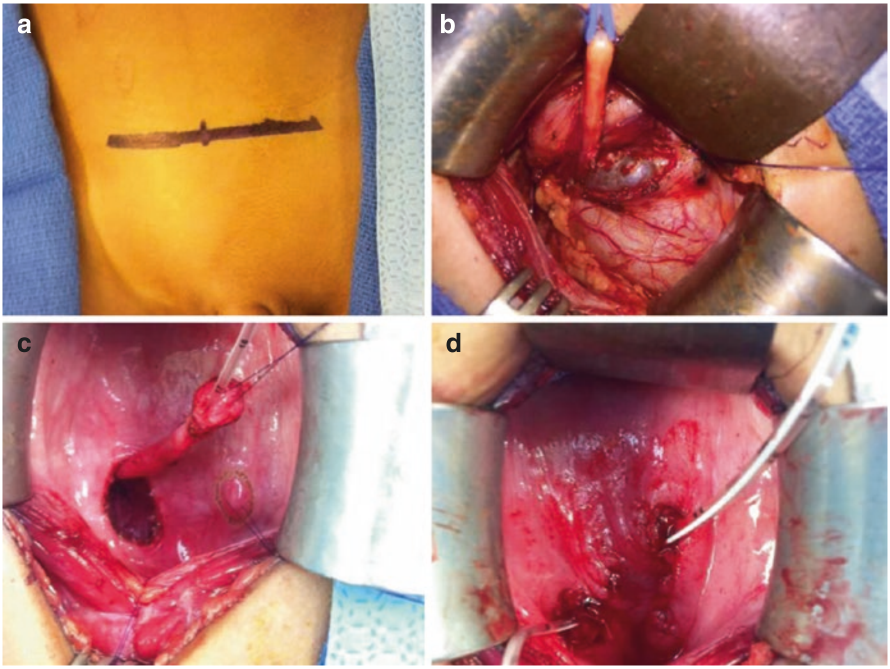

Fig. 5.1 Open ureteral reimplantation. Both intravesical and extravesical approach are via a Pfannenstiel incision (**a**). With the extravesical approach, the refluxing ureter is *encircled* with a vessel loop. A detrusor trough has been created by incising down to the mucosa, which has not been violated (**b**). With a Cohen cross-trigonal, the bladder is opened anteriorly and the ureter is mobilized to ensure the tunnel length is five times the ureteral diameter (**c**). After creation of tunnels, the ureters the approximated to the neo-orifices bilaterally, and feeding tubes temporarily passed to ensure there is no obstruction (**d**)

and extravesical (tunneling the ureter without opening the bladder) approaches, and are performed through a Pfannenstiel incision, which is a horizontal incision on the lower abdomen just above the pubic bone (Fig. 5.1a). Children receive IV antibiotics and the abdomen is cleansed with an antiseptic to prevent skin infection. After making the incision, the abdominal muscles are split to expose the bladder. The ureters can be re-tunneled into the bladder either by creating muscle flaps to place over the ureter (Fig. 5.1b) or by opening the bladder and creating tunnels from the inside (Fig. 5.1c). Feeding tubes may be temporarily placed to ensure there is no blockage of the ureter after reimplantation (Fig. 5.1d), and sometimes a stent is left indwelling in the ureter for 4–6 weeks to allow proper healing in more extensive repairs. A Foley catheter may be left to drain the bladder overnight, and is removed within a couple of days after surgery.

Expected Postoperative Course

Children are admitted following surgery, and typically have an indwelling catheter until discharge from the hospital. Diet is advanced as tolerated. Patients are often discharged within 48 h after surgery with narcotic pain medication (with or without medicine to prevent painful bladder spasms), and should be kept on prophylactic antibiotics until appropriate postoperative studies are obtained. Children typically miss 3–5 days of school. If an indwelling stent in the ureter is placed at the time of ureteral reimplantation, it is removed via the urethra 4–6 weeks later under general anesthesia.

Follow-Up

Renal-bladder ultrasound should be obtained 1–3 months postoperatively to assess for dilation of the kidney(s). Due to the high success rates of open repairs, postoperative VCUG can be limited to high-risk cases or those with postoperative kidney infection. Children also typically undergo sonography at 1 year postoperatively to assess kidney growth and detect kidney swelling. Routine urologic follow-up with imaging beyond 1 year for uncomplicated ureteral reimplantation in the absence of kidney damage is not warranted [3], however monitoring for late occurring complications of VUR should be performed yearly. Monitoring includes measurement of blood pressure, selective renal ultrasound, and a urinalysis, to access proteinuria, renal growth, and infection.

Outcome

Success following any antireflux surgery may be based on radiographic (absence of VUR) and clinical findings (absence or decrease in febrile urinary tract infections). Bladder infections after ureteral reimplantation may occur and are of no concern regarding the success of the surgery nor kidney damage. The success rate of ureteral reimplantation is >95 % in cases of primary reflux [4]. The most severe form of reflux (grade 5), however, has a lower success rate.

Endoscopic Injection

Operative Technique

Endoscopic injection is also performed under general anesthesia, and often requires approximately 30 min to complete. The child is placed in stirrups, and a small cystoscope is placed into the bladder via the urethra. The injection method

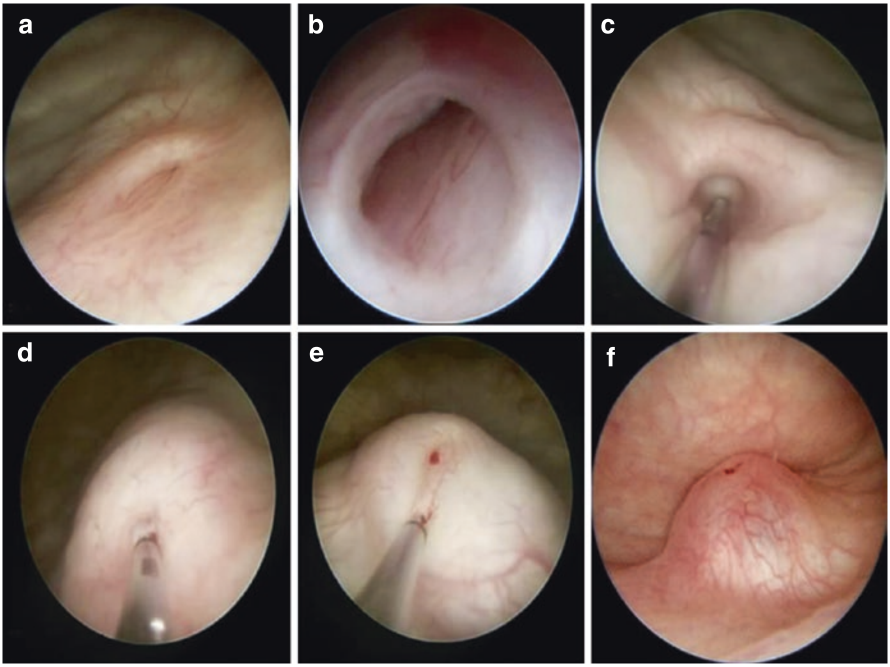

Fig. 5.2 Double HIT Method. Bladder emptied and the ureteral orifice is visualized (**a**), followed by hydrodistention (**b**). Proximal HIT is then performed with needle inserted into the mid-ureteral tunnel at the 6 o'clock position (**c**) and sufficient bulking agent is injected to produce a bulge which collapses the tunnel (**d**). Distal HIT (**e**) leads to coaptation of the ureteral orifice (**f**)

achieving the highest rates of success is the Double HIT [5]. The ureteral orifice (where urine from the kidneys drains into the bladder) is visualized (Fig. 5.2a). Refluxing ureters often hydrodistend, or "pop open" when the flow of saline is directed at them (Fig. 5.2b). A needle is inserted at the mid-ureteral tunnel at the 6 o'clock position (Fig. 5.2c), and bulking agent injected to produce a bulge which collapses the ureteral tunnel (Fig. 5.2d). A second implant is injected within the most distal intramural tunnel (Fig. 5.2e), which leads to coaptation of the ureteral orifice and prevents backflow of urine from the bladder into the kidney (Fig. 5.2f).

Expected Postoperative Course

Children are allowed to go home the day of surgery, after a 1–3 h stay in the post-anesthesia recovery unit. Patients are usually discharged without any narcotic medications, and should be kept on prophylactic antibiotics until appropriate post-operative studies are obtained. Children can return to school and full activities after 24 h.

Follow-Up

As with ureteral reimplantation, renal-bladder ultrasound should be obtained 1–3 months postoperatively to assess for dilation of the kidney(s). While a postoperative VCUG has also been recommended [1], there remains wide variability in success dependent upon the individual patient and the surgeon's clinical experience. Children also typically undergo sonography at 1 year postoperatively. Urologic follow-up with imaging beyond 1 year is patient specific, and not routinely performed in children who are doing well clinically without infections. As above, children with a history of VUR should undergo yearly blood pressure check and urinalysis by their primary care provider. Patients with recurrent febrile urinary tract infection after successful endoscopic treatment of VUR should be evaluated for bladder-bowel dysfunction and recurrent reflux.

Outcome

Endoscopic injection is effective for the treatment of most VUR, however children with higher grades of reflux and those with bladder anomaly or dysfunction may have increased risk of failure. Reported initial success rates, while up to 94 % with the Double HIT method, are known to vary widely among surgeons and techniques [4, 5].

Complications

Complications may occur in the immediate postoperative period or may manifest month to years later. Early complications occur within the first few days following surgical intervention and are typically transient. They include low urine output, blood in the urine (hematuria), bladder spasm, voiding dysfunction and infection [6]. Hematuria and bladder spasms are frequent complications of intravesical ureteral reimplantation and usually resolve within 2 weeks of surgery; while these symptoms can be distressing, reassurance and selective use of anticholinergics are typically all that is necessary.

Ureteral obstruction, or inability of urine from the kidney to drain freely into the bladder, is the most serious surgical complication of reimplantation or endoscopic VUR repair. If low urine output persists beyond 24–48 h after surgery, an ultrasound is obtained to rule out obstruction. Complete obstruction requires placement of a ureteral tube or nephrostomy tube to allow kidney drainage. The risk of obstruction is uncommon (<1 %) after both open and endoscopic repairs [1].

Postoperative reflux may be due to persistent reflux in the reimplanted/injected ureter(s) or new onset reflux into a ureter that was not operated on (contralateral reflux). For open bladder surgery the incidence of persistent VUR is up to 2 %.

In almost all cases observation is the preferred treatment as spontaneous resolution occurs over time. Treatment failure following endoscopic therapy ranges from 7 to 50 %, and is dependent on technique, VUR grade and surgeon experience [4]. New onset of contralateral VUR has been observed in up to 19 % of open unilateral reimplants, with similar rates reported in endoscopic cases [6]. As mentioned, new onset VUR is generally self-limited.

Conclusion

Ureteral reimplantation is a safe and effective intervention for management of VUR. The majority of potential complications are temporary, and managed conservatively during the immediate postoperative period. Endoscopic injection of bulking agents has emerged as a successful, minimally invasive alternative to open reimplantation, with minimal morbidity. Progressive ureteral obstruction is a serious complication of either surgery, and although it occurs in less than 1 % of children, it requires intervention with ureteral stenting or nephrostomy tube placement to achieve renal drainage. Families should be counseled about all management options, and treatment should be individualized. When surgical intervention is considered, the consent process needs to include a thorough discussion of the potential risks and benefits of each modality, and an informed decision made based on patient age, health, risk of subsequent renal injury, clinical course, renal function and parental preference.

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Chapter 6
Open Ureteral Surgery-Ureteroureterostomy and Transureteroureterostomy

Michael E. Chua and Martin A. Koyle

Name of Procedures

Open Ureteroureterostomy (U-U) and Transureteroureterostomy (TUU).

Lay Description

An open operation to cut one blocked or inappropriate insertion of an ureter and connect to another unblocked ureter that connects into the bladder. Connection may be done either to the same side ureter (ureteroureterostomy) or to opposite side ureter (Transureteroureterostomy).

Informed Consent

Informed consent follows the "PARQ" acronym for **P**rocedure, **A**lternatives, **R**isks and benefits, **Q**uestions (and answers provided).

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Intended Benefits of Procedure

To address blockage or abnormal insertion of one ureter and the kidney above it, and divert the urine flow to another ureter where the urine can drain more freely into the bladder. This will reduce the risk to the kidney from the increased pressure associated with any blockage, and/or reroute the urine flow from an abnormal location, other than that within the bladder (vagina, urethra, skin) where urine leakage (incontinence) and/or recurrent infection can occur. This may protect the kidney from damage, reduce the risk of infection, and eliminate incontinence.

Risks of Procedure

General [1–4]

Overall complications after upper tract urinary tract reconstruction are relatively low (4–13 %) and consistent with other similar ureteral procedures. The majority of complication are considered minor Clavien-Dindo Grade I or II (conservative or pharmacologic management), rarely grade III (additional procedure under anesthesia). All patients undergoing any surgery are advised of risks of: Anesthesia, and generic risks of surgery such infection, bleeding, pain and poor healing.

Specific Risks [1–8]

1. Prolonged drain output due to urine leak from anastomosis site (3–13 %), which is usually managed conservatively by maintaining drain until output decreases.
2. Urine reflux into the donor stump (12 %), this usually occur if patient had condition of vesicoureteral reflux to the ectopic upper moiety ureter (urine back flow from bladder to the ureter).
3. Urinoma/urine accumulation in abdomen space (1–5 %), which is a consequence of urine leak from anastomosis (#1), where increased urine collects in the surgical space or exudes from the wound. This can be managed by percutaneous drainage or conservative management, depending to volume of urine and presence of absence of infection.
4. Vesico-ureteral reflux (9–11 %), which is urine back flow from the bladder to both ureters from the supposedly normal ureter in the bladder. This condition is related to the ureter draining the lower ureter in a U-U or the recipient contralateral ureter in a TUU due to shorted intramuscular insertion of the ureter into the bladder.
5. Wound infection or urinary tract infection (1–12 %), although the procedure itself is considered as a clean contaminated system, involving urinary tract. Hence, antibiotics are usually given perioperatively to prevent this occurrence.

In cases where an infection occurs, the usual management is by IV or oral antibiotics, although some wounds may require opening and drainage.

6. Temporary ureteral obstruction (1–2 %) may occur in the initial phase where the anastomotic site of the ureteral segments develops edema or swelling, which temporarily blocks the urine flow and may even cause leakage.
7. Rarely a ureteral narrowing needs revision (1–3 %), which may happen in long-term due to excessive fibrosis occurring at the anastomosis site of the ureter. Management involves additional procedure under anesthesia by endoscopic incision or open revision of the anastomosis; depending on the severity of the condition.
8. Ileus or dilated bowels (1–2 %), may occur when patient had increased dose of opioid derivatives pain medication or when the peritoneal cavity is irritated by things like a urine leak or after extensive dissection.
9. Upper respiratory infection including pneumonia (1 %) rarely occurs.
10. Rarely, severe bleeding requiring transfusion, and even less commonly, reoperation (1 %) may occur. Bleeding is most often not excessive, does not induce symptoms and can be managed conservatively. When significant bleeding does occur, a previously undiagnosed, preexisting bleeding condition might need to be excluded.

Alternatives

This operation can also be performed using laparoscopic or robotic techniques alone, or in combination with open surgery. The alternatives to open U-U or TUU may include, but are not limited to: common or single ureter reimplant, pyelo-ureterostomy, partial nephrectomy, or ongoing observation.

What Happens Before the Operation?

Assessment of the kidney condition, anatomy and function using ultrasound and a nuclear scan (DMSA or MAG3 with Furosemide) will have been done. In other cases an Xray test of the bladder, called a voiding cystourethrogram (VCUG) will also be indicated. In some complex situations, an MRI scan allows an opportunity to identify anatomy more clearly. Once surgery has been suggested as an option and other alternatives, including the risks and benefits of each have been discussed in detail with the family and their questions answered, a standard consent (attached) form is signed. Teaching by the nurse then occurs and the scheduling office books the surgery, usually as an elective procedure. Preoperative anesthesia evaluation will be made prior to the scheduled procedure and any ancillary tests (blood and urine) ordered. Patients are allowed to have full diet until 8 h prior, formula milk until 6 h prior and clear liquid until 4 h prior to scheduled surgery time, depending on the patient age.

Operative Technique

Patients receive general anesthesia with appropriate adjuvant regional anesthesia, depending on incision location and age of patient. Pre-operative IV antibiotics are administered. The patient is then positioned for cystoscopy or placing a scope into the bladder. Surveillance and identification of the ureteral opening(s) is (are) made. A J stent or open-ended ureteral stent (plastic tube) is placed into the non-obstructed normal, or recipient ureter and left indwelling. We often attach this to a string so that it can be removed with the child awake 7–10 days later. A Foley catheter is placed into the bladder to help drain urine. An inguinal (modified Gibson) or midline low abdominal smile face (Pfannenstiel) incision is made, then abdominal muscles are split in order to gain access into the retroperitoneal area, where the ureters are located. The peritoneum or space where the intestines are located is not opened. The abnormal or dilated/bocked ureter is identified and carefully cut with the blood supplies being preserved. Then the upper segment ureter is connected to the non-obstructed ureter by sewing the two ureters using end to side technique (Fig. 6.1). The wound is closed in layers using dissolvable sutures that are all under the skin. Rarely, a drain (penrose or Jackson-Pratt) might be left in place, which later can be removed with the child

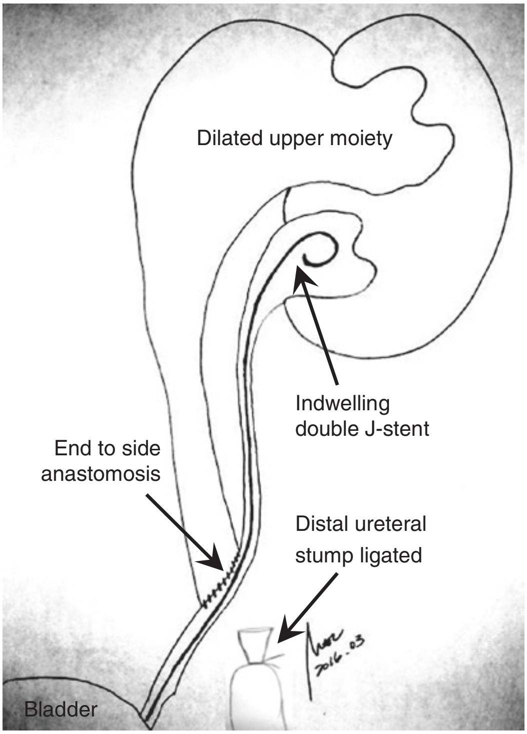

Fig. 6.1 End to side connection of ureters (uretero-ureterostomy) in a double (duplex) left kidney. The lower part of the J-stent has a suture attached that exits the urethra so that it can be removed 7–10 days after surgery

awake. The actual “cut to close” surgical time is usually 45 min or so with additional time for cystoscopy and anesthesia and room preparation and turnover (total 75–90 min).

Postoperative Expected Course

The patient may eat and drink as soon as they awaken. Pain medication: Acetaminophen/Paracetamol alternating with Ibuprofen every 4 h while awake for 48 h, then PRN; if more severe pain, Ketorolac or a narcotic as needed is offered while in hospital only. The patient may receive additional IV or oral antibiotics depending on how extensive the surgery is. Foley catheter will be removed later that day if the child is otherwise doing well. Discharge occurs that evening, or the next morning depending on pain control, normal voiding, and tolerating diet. If a drain has been left, it may be removed within 2–5 days depending on amount of drained fluid. The J stent is typically removed in 7–10 days and often by the family or local doctor if they live a substantial distance from the hospital.

Follow Up

Three months after stent removal, patient will be followed-up with repeat ultrasound of the kidney and bladder. Then routine follow-up proceeds according to the individual patient. We recommend that every patient who has had kidney surgery have their blood pressure measured at least annually.

Outcome

Success rate ranges 94–100 % for long term for uretero-ureterostomy and transureterostomy. Less than 4 % will need an additional procedure to address any problem such as urine leak or recurrent car/blockage (stenosis).

Conclusion

Ureteroureterostomy and transureteroureterostomy are considered fairly straight forward procedures with relatively low risk involved and short hospital stays. Because this operation is irreversible; the patient and family should be counselled extensively in the preoperative period about the procedure and treatment plan, and they must understand reasonable alternatives to the proposed intervention (such as monitoring or heminephrectomy) or the same procedure being performed using other minimally invasive techniques. Likewise, discussion must be made on relevant risk, benefits and uncertainties involved with each treatment options. Most important in the consent process is to assess the understanding of the patient and family by asking their concerns and answering their possible questions (PARQ).

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Part III
Surgery of the Bladder

Chapter 7
Consent on Epispadias and Bladder Exstrophy

M. İrfan Dönmez and Duncan Wilcox

Name of Procedure

Epispadias and Bladder Exstrophy Repair.

Lay Description

Epispadias and bladder exstrophy are currently some of the most challenging diseases managed in a pediatric urology practice. Epispadias is a congenital problem defined by the urethral meatus, through which urine passes, being on the front or base of the penis, rather than on the head of the penis. Bladder exstrophy is the term that refers to the bladder being exposed through the abdominal wall, so that it appears on the abdomen. These are congenital problems, where frequently the exact cause is not known. Those two problems are generally presented together and called 'Epispadias – exstrophy complex' (EEC) which are associated with other problems, such as an open pelvis and inguinal hernia. The incidence of EEC is 1 in every 10,000–50,000 births [1]. Other less common types of EEC are cloacal exstrophy and exstrophy variants in which there can be bowel, kidney, spine and neurological problems.

The only way of correcting EEC is through surgery. The surgery, is either single staged or multi-staged, this includes radical mobilization of bladder, bladder neck reconstruction to create a control mechanism, closure of bladder mucosa, closure of abdominal wall, correction of skeletal anomalies if needed, reconstruction of external genitalia and bringing urethral meatus to the correct position.

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Intended Benefit

The aims of surgery on epispadias and bladder exstrophy are to have a cosmetically acceptable appearance of both lower abdomen and external genitalia, gain a normal voiding function (i.e. urinary continence and unobstructed voiding) and furthermore have a normal sexual life. Closing of the bladder is important since the growth of the bladder is dependent on the cyclic filling. When untreated, all patients will be incontinent of urine and, bladder exstrophy has a 17.5 % risk of bladder neoplasia after 20 years with a high mortality rate [2].

Surgery

Initial surgery plays a key role in the treatment of EEC. There are different options regarding the type (single/staged), timing (initial, within 28 days/delayed) and the content (with or without osteotomy) of the surgery. EEC is very uncommon and so many authors believe that evaluation and surgical treatment should be done by experienced centers with high surgical volumes. There is however, considerable controversy about whom, when and how is the best way to achieve good results in the treatment of children with bladder exstrophy.

An important fact about bladder exstrophy closure is the improved bladder growth in patients operated under 1 year of age [3]. Also, surgery performed as early as the first 2 days of life might not require osteotomies as the pelvis is more pliable. Furthermore, it must be remembered that classic bladder exstrophy patients undergo a median of five operations/interventions throughout their lives [4].

In the complete single staged approach all of the required steps are handled in one session. However, the modern staged approach indicates early bladder closure followed by genital reconstruction at 6 months and finally bladder neck reconstruction with or without correction of the vesicoureteral reflux, the timing of this varies according to the size of the bladder.

A preoperative urinary ultrasonography of the urinary tract will determine any abnormalities regarding the kidneys and the ureters while obtaining a baseline for future imaging studies.

Anesthesia

All patients should undergo preoperative anesthesia evaluation which includes physical examination, complete blood count, coagulation panel, kidney function tests. Appropriate consultations may be requested when necessary.

Generally, significant blood loss is not anticipated however, it is usually prudent to ensure blood products are available before commencing. In addition, central venous catheters can be placed before the surgery in order to facilitate the precise control of per operative hydration status, although this is not universally performed. The operation is performed under general anesthesia, with additional local anesthetic blockage. This is frequently performed using an epidural or caudal block. Mean operation time can vary due to the content of the surgery (staged, with or without osteotomy etc.) however it takes around 4–6 h for the patient to leave the operation room. In most cases, patient is transferred to the recovery room first and then the patient room where he/she will stay until discharge. However, a need for a short intensive care unit stay may be required in rare cases.

Neuroaxial blocks can be applied to reduce the early postoperative pain. Postoperative pain medication is adjusted for the needs of the child and the route of the administration will be intravenous in the first days and it will be converted to oral route as soon as it is convenient. Acetaminophen and non-steroidal anti-inflammatory drugs are the mostly used to reduce pain in the clinical setting however opioid drugs such as tramadol and morphine can be used in limited situations.

Surgical Technique

Regardless of the technique, successive primary bladder closure is important to have a chance of obtaining normal postoperative bladder function [5].

Appropriate prophylactic antibiotic are given prior to the surgery. During the surgery, after the incision along the two sides of the bladder into the umbilicus, radical mobilization of the vesicourethral unit is obtained. The steps to the procedure are:

1. Mobilization and closure of the bladder – often in two layers
2. Surgical correction of the vesicoureteric reflux
3. Creation of a bladder neck to provide resistance to the bladder, and ultimately urinary control
4. Surgical reconstruction of the penis
5. Closure of the inguinal hernia
6. In some cases, bilateral anterior transverse innominate or vertical posterior iliac osteotomies may be needed to facilitate approximation of the pubic bones and to decrease the tension of fascia. As a result the pelvis needs to be stabilized. This is frequently achieved by: Bucks traction, a spica cast or by fixator pins and external fixators. Pelvic stabilization is usually kept in place for up to 4–6 weeks in patients with osteotomy. Some centers are now advocating keeping the child paralyzed for 3–4 days after surgery to allow healing and avoid pelvic traction.

This can be performed in a single stage or in multiple stages. In the modern staged approach it is normal to initially close the bladder and pelvis, then the penis and around 4–5 years of age the bladder neck reconstruction.

Diversions and Drains

Patients leave the room with a urinary catheter (type, diameter and material of the catheter can vary between departments and surgeons) via urethral and/or abdominal route. Also both ureters are catheterized using specific ureteric catheters to prevent the bladder over distending. The drains and urinary catheters are removed according to surgeon preference. It is utmost important that all of the catheters and drains should be kept in situ since premature accidental removal of any of them may necessitate additional interventions and/or prolonged hospitalization.

Postoperative Course

Patient will generally be able to gradually drink and eat 4–6 h after exiting the operation room. Antibiotic prophylaxis may be continued for up to 24 h. The mean hospitalization period will be between 4 and 6 weeks.

Complications

Complications can be classified into two; early and late complications. Early complications include fascial and/or bladder dehiscence which can be observed in up to 10 % of patients. Fascial dehiscence needs urgent exploration however bladder dehiscence should be re-operated at least 6 month after first operation. Urethrocutaneous fistulas are the most common complications after neonatal bladder exstrophy closures with a prevalence of 14–35 % [4, 6]. The rates are even higher (26–52 %) in delayed surgeries [7]. However spontaneous closure of those fistulas can be seen between 25 and 100 % of patients. With regard to epispadias correction, atrophy of corporal bodies, glans and urethra can be observed, as well as persistent chordee [8]. In some cases there may be a transient reduction in bowel movements, which usually resolve with proper medication and rarely require a surgical intervention. Also, there are more general postoperative complications such as pulmonary atelectasis, bleeding, deep venous thrombosis and surgical wound infections.

Bladder outlet obstruction, vesicoureteric reflux, renal deterioration, urinary incontinence, and malignancy are some of the problems that an extstrophy patient can face in the long term. Bladder outlet obstruction can develop around 7.5 % of the patients after initial closure [6, 9]. Cystoscopy might be necessary to rule out obstruction. It should be remembered that all of the EEC patients have vesicoureteric reflux (VUR) and treatment needs to be individualized to each patient. Spontaneous resolution rates are low and the rate of febrile urinary tract infection (UTI) may be up to 22 % [4]. Therefore treatment of VUR should be considered in patients with febrile UTIs. Up to a quarter of patients will show transient hydronephrosis following primary closure. However, only 3 % of the patients will result with renal impairment [6]. The bladder may not have sufficient volume and/or it may not empty adequately consequently: Clean intermittent catheterization, use of anticholinergics, intra detrusor botulinum injections and/or augmentation cystoplasty may be needed in patients with poor bladder compliance and high detrusor pressures. Continence can be assessed around 5 years of age. The definition of urinary continence varies greatly between authors; however, urinary continence after single staged operations has been reported between 19 and 73 % in the literature [10, 11]. Urinary continence rates after modern staged repair has been shown to be between 45 and 72 % [12, 13]. Up to 66 % of the boys with bladder exstrophy will have sexual problems in the future, typically persistent chordee of the penis and ejaculatory problems [14]. On the other hand, 56 % of the women with bladder exstrophy can maintain an pregnancy and delivery [15]. Since the psychological impact of the disease and the surgeries on the individual are overwhelming, psychological support is recommended in these patients throughout their life beginning as early as possible [16].

Follow-Up

Patients born with bladder exstrophy and epispadias require life long follow-up. In the first year of life this is normally every 3 months and may become less frequent as the child's situation becomes more stable. Follow-up will need to consist of regular urinary tract assessment with renal ultrasound scans, urodynamic investigations and renal blood profile. Orthopedic involvement is essential to ensure correct positioning of the pelvis and normal walking. Most centers advocate the regular input of a clinical psychologist and this is usually aided by support groups. After puberty the urologist will also need to evaluate sexual function. Due to lateral displacement of pubic bones and corporeal bodies, boys with EEC will have a shorter final penile size when they reached adulthood. Finally, as has been mentioned, many of these patients will develop malignancy and so long term surveillance is required.

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Part IV
Endoscopic Surgery of the Urinary Tract

Chapter 8
Cystoscopy and Cystoscopic Interventions

Navroop Johal and Divyesh Desai

Name of Procedure

Cystoscopy and Cystoscopic interventions.

Lay Description

Paediatric cystoscopy (PC) allows visualisation of the lower urinary tract and includes assessment of the urethra and the bladder. In addition to its role in diagnostic evaluation endoscopy enables therapeutic interventions to be performed. This includes the treatment of bladder outflow obstruction, endoscopic treatment of ureterocoeles, treatment of vesico-ureteric reflux, injection of Botulinum toxin, biopsy of suspicious lesions and the placement and removal of ureteral stents.

Intended Benefits

The aims of PC and cystoscopic interventions are twofold: (1) to evaluate the anatomy of the lower urinary tract (2) endoscopic treatment of: (a) posterior urethral valves (b) anterior urethral valves or syringocoele (c) urethral stricture (d) ureterocoeles (e) ureteric stenting (f) Botulinum toxin injection for neurogenic bladder dysfunction.

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Which Conditions Which Can Be Treated with Cystoscopic Interventions?

Posterior Urethral Valves

Posterior urethral valves remain the most common cause of lower urinary tract outflow obstruction in male infants with an estimated incidence of 1:5000 live births. Videoendoscopy using an 8.5 Fr (5° lens) to 10 F (0° lens) resectoscope and a cold knife hook allow safe and effective ablation of urethral lesions [1].

Syringocoele

A syringocoele is thought to arise due to a cystic dilatation of the main bulbourethral glands as described by Cowper in 1705. Clinical presentation is variable and includes prenatal hydronephrosis, penile swelling, urinary tract infections and voiding symptoms like poor urinary stream and dribbling. Transurethral incision or fulgration of the valve is the procedure of choice in the majority. Bagli et al. in their series of 17 cases found this technique successful in all 6 cases treated with a transurethral incision of the valve leaflet [2].

Urethral Stricture

Paediatric urethral strictures are uncommon and their aetiology can be divided into "inflammatory," traumatic, and idiopathic. The time-honored method of treatment is urethral dilatation. An alternative treatment with an equally long history is an optical urethrotomy. If instrumentation is required more frequently or is complicated then a urethroplasty is the only curative option.

Ureterocoele

The aim in ureterocoele management is prevention of renal damage secondary to obstruction. The treatment should at the same time maintain continence and minimize surgical morbidity. Endoscopic puncture is minimally invasive, can achieve definitive decompression or act as a temporizing procedure that reduces the risk of infection and has the potential to allow recovery of renal function.

Ureteric Stenting

JJ stent insertion across the vesico-ureteric junction can allow effective drainage in primary obstructive megaureters in carefully selected cases and in a proportion (up to 50 %) is curative [3].

Botulinum Toxin Injection for Neurogenic Bladder Dysfunction

Preliminary results suggest endoscopic injection of Botulinum-A toxin to be a safe alternative in the management of neurogenic bladder dysfunction and the improvements demonstrated in urodynamic parameters and continence are encouraging [4].

Technique

Cystourethroscopy is performed in the operating theatre under general anaesthesia. The duration of the operation varies depending on whether it is diagnostic or therapeutic procedure. Intravenous antibiotics are given at induction.

How Is the Patient Positioned?

Older children are placed in lithotomy with their legs secured in stirrups and pressure points are checked. Smaller infants have their legs carefully secured in a frog leg position at the end of the operating table.

Are There Different Sizes of Cystoscopes Available?

There are different sizes of cystoscopes available. Neonatal scopes are available in 6- to 8- Fr and 9.5-, 11- or 14-Fr paediatric cystoscopes are available (see Fig. 8.1). The optical views obtainable include a straight 0°, 5° or 30°. The scope is connected to the light source and the endoscopic camera, which in turn is connected to the endoscopic stack. The fluid is connected to the irrigation channel.

How Is Cystourethroscopy Performed?

In males the penis is held in a vertical position and the cystoscope in inserted into the urethra from an upright position and advanced to inspect the whole urethra including the verumontanum before entering the bladder. Care is taken to ensure the lumen of the urethra is in the middle of the field of view and the tip of the scope is maintained as steady as possible. The urethra is delicate and forced or over sized instrumentation will inevitably result in narrowing. Additionally one must be careful to not over distend the bladder as anatomy can be distorted and the risk of perforation can increase if performing a therapeutic procedure for the treatment of vesicoureteric reflux.

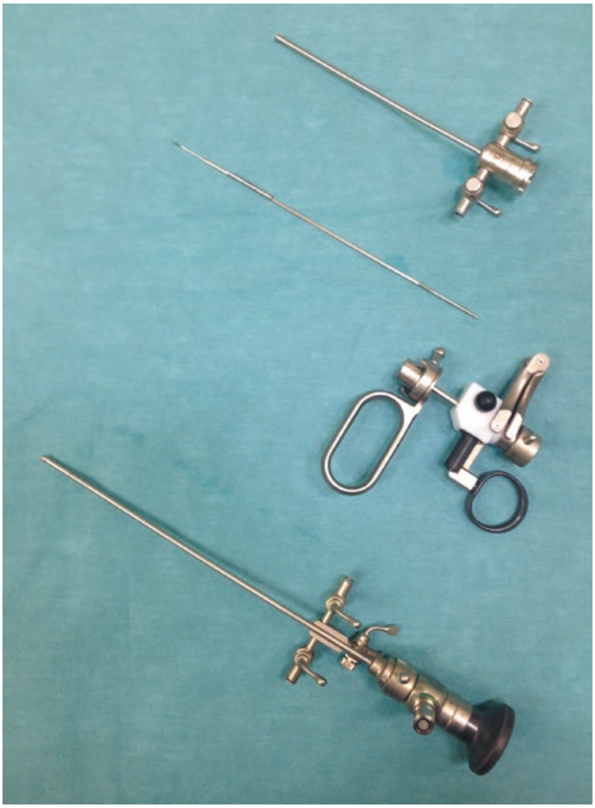

Fig. 8.1 An 11Fr cystoscope with a 0 degree lens is displayed. The upper images show a dismantled 10.5Fr resectoscope and a cold knife blade

The bladder is inspected in a systemic fashion commencing at the bladder neck and the trigone before viewing the ureteric orifices and all quadrants of the bladder in stepwise manner. The bladder is emptied before the posterior urethra is inspected as the scope exits the bladder.

In females the labia is separated to allow visualisation of the urethra. The cystoscope is inserted and the rest of the examination is completed as for males.

Postoperative Expected Course

The majority of patients will be discharged on the same postoperative day unless there are co-morbidities requiring admission to the ward.

If patients are catheterised postoperatively the catheter should be removed in the morning on the following day.

Follow Up

Most patients will commonly be discharged on the same day of the procedure. Patients are followed up in the clinic at 6–8 weeks following the procedure.

In the case of posterior urethral valves a repeat check cystourethroscopy is performed at 3 months. A review of the last 100 valve resections at the author's institution using a bugbee/insulated wire or the cold knife resection technique shows a 21 % incidence of re-resection at planned follow-up check cystoscopy [1].

Risks of Procedure

With modern instrumentation and good surgical technique, complications directly related to endoscopic manipulation are rare.

Most patients experience a degree of burning or discomfort on micturition but providing patients maintain a good fluid intake this usually resolves after a few hours. Other complications include urinary tract infection and hematuria.

Urinary Tract Infection (UTI)

Intravenous antibiotics are given at induction routinely to reduce the risk of postoperative urinary infection. If a urine culture have been sent preoperatively an appropriate antibiotic is administered based on the urine sensitivity. Routine prophylactic antibiotic minimizes the incidence of infections in the neonatal period. Postoperatively all patients are encouraged to have a good fluid intake. If a child presents with a fever postoperatively, urine should be checked and sent for culture and sensitivity and appropriate intravenous antibiotics commenced expediently.

Patients whom have impaired renal function or those whom have recurrent UTIs are particularly vulnerable to developing urine infection following urethral instrumentation and should be prescribed a course of treatment antibiotics on discharge.

Hematuria

Some patients do experience some hematuria following the procedure and most cases are self-limiting.

Urinary Retention

This is a very rare complication and most children will void providing adequate analgesia is given.

Urethral Strictures

This is an extremely rare complication and if diagnosed early they may be amenable to dilatation or visual internal urethrotomy.

Conclusions

In conclusion, PC is a safe procedure that if performed carefully has a low risk of complications.

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Chapter 9
Minimally Invasive Interventions for Stone Disease

Bernardita Troncoso Solar and Naima Smeulders

Introduction

The incidence of stones in children varies enormously geographically and is approximately 5–20 per 100,000 children aged less than 18 years [1]. A key difference between adult and paediatric stone disease is its recurrent nature. Identifying any underlying metabolic abnormality, reported in 33–93 % in the literature, is therefore essential [2]. Nevertheless, it is important to note that such assessment must not delay the treatment of the stones. At our institution, a metabolic abnormality is found in just under half the children presenting with stones, comprising hypercalciuria (57 %), cystinuria (23 %), hyperoxaluria (17 %) and hyperuricosuria (2 %) [3]. Infection is the second most common aetiology.

In children, ultrasound can give exquisitely detailed information on the size, location and extent of stones. Abdominal X-ray (AXR) increases the detection rate particularly of ureteric stones. Today, ultra-low dose non-contrast CT (Computerised Tomography) has reduced the radiation burden so significantly that this imaging modality is suitable for children. IVU (Intra-Venous Urogram) is rarely needed [4]. Urine microscopy and culture is mandatory before any treatment.

Minimally invasive treatment of paediatric stones has advanced rapidly over the last few decades with open surgery now rarely used [5]. The aim of any intervention is to render the child stone-free, while minimising the impact of the procedure(s) on the child. Treatment options include ESWL (Extracorporeal Shock Wave Lithotripsy), URS (Uretero -Reno-Scopy, if extending into the pelvi-calyceal system also referred to as RIRS – Retrograde Intra-Renal Surgery), PCNL (Per-Cutaneous Nephro-Lithotomy), PCCL (Per-Cutaneous Cysto-Lithotomy), laparoscopic surgery

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and open surgery [6–8]. Optimal treatment modality depends on stone burden, composition, location and size, symptoms and complications, as well as patient factors.

Complications relate mostly to infection, bleeding, iatrogenic injury, urine leak, pain, obstruction due to stone or stricture, residual fragments/recurrence of stones and loss of renal function. Infection is one of the most important complications of any form of stone treatment and can lead to overwhelming sepsis or even death [9]. Meticulous pre-operative checking of the urine for bacteria and appropriate timely antibiotic treatment will help reduce this potentially lethal complication. UTI may be the cause or the effect of the stone. Importantly, in the presence of infection, even small residual stone fragments can act as the nidus for recurrent stone formation. Indeed, one third of small residual fragments will regrow over a period of 1–4 years [10].

ESWL

Key Points

Treatment of choice for single renal pelvic stones up to 20 mm, upper/inter/lower pole stones up to 10 mm and proximal ureteric stones.

Specific risks of the procedure:

1. *Hematuria*
2. *Pain/colic*
3. *Infection*
4. *Steinsstrasse (stone street)*
5. *Bruising/Ecchymosis and hematoma*
6. *Injury to other viscera*

Name of Procedure

Extracorporeal Shock Wave Lithotripsy (ESWL)

Lay Description

Pressure waves are generated outside the body and travel through the skin onto the stone to shatter the stone into small fragments, which can pass from the body in the urine. ESWL has a success rate of approximately 80 %, although more than one treatment session may be required [11–13, 17].

Intended Benefit

By breaking the stones into small pieces, these can be expelled with the urine.

Technique

Shock waves are produced by electrical discharge, rapid vibration of piezo-electric crystals or electro-magnetic diaphragms and then focused into the shape of a cone. The shock waves are transmitted through the skin ("coupling") and body tissues to release their energy onto the stone. Continuous ultrasound allows the shockwaves to be focused onto the stone and the effect of the treatment to be monitored [14]. As the depth of the kidney from the skin is typically only a few centimetres in children, the shockwaves enter through an area of skin the size of a coin, making it much more painful than in adults, where the shocks enter through a much larger area of skin as the kidney and stone are much deeper in the body. The child needs to lie still for the duration of the treatment, around 30 min, to enable the shock waves to be focused consistently onto the stone avoiding neighbouring lung and intestine. While older children can tolerate ESWL under the cover of simple analgesia or sedation, young children usually require general anaesthesia or deep sedation.

Illustrations (Figs. 9.1, 9.2, and 9.3)

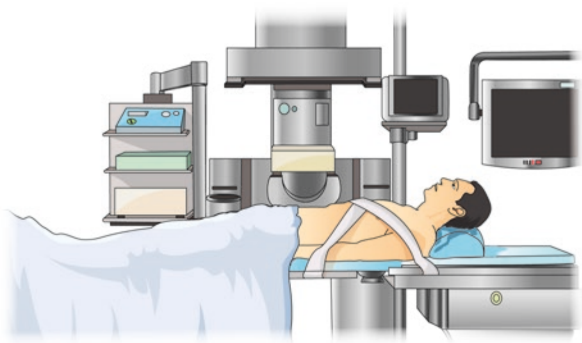


Fig. 9.1 Lithotripsy machine [14]

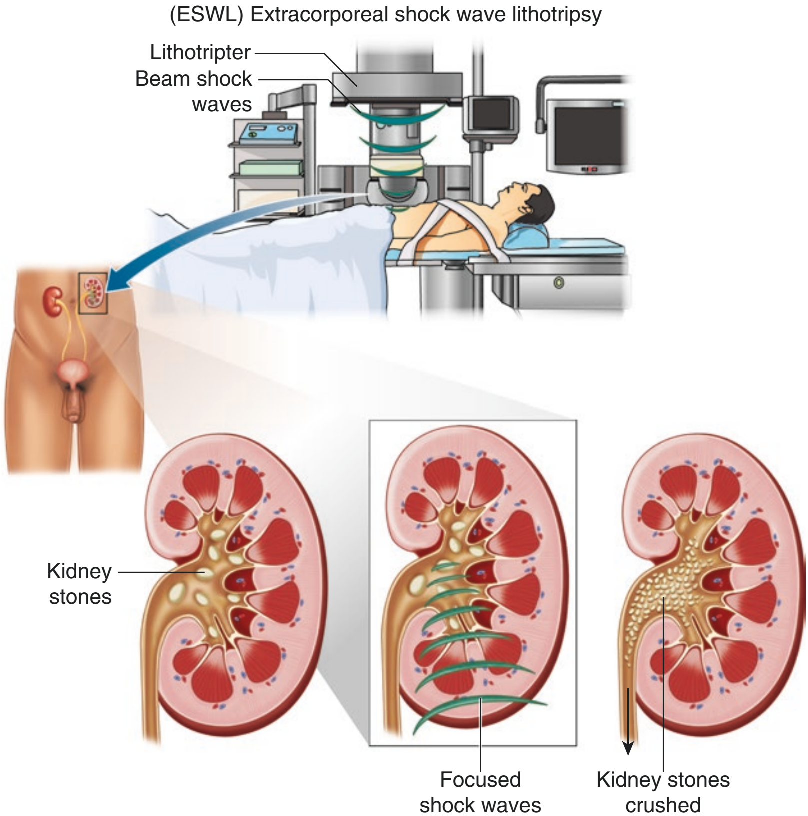


Fig. 9.2 Stone fragmentation achieved with schock waves delivered from outside the body

Post-operative Expected Course

- Patients can eat and drink as soon as recovered.
- Hematuria is expected to clear within 24–48 h.
- Ecchymosis in the skin at points of shock wave entry will resolve spontaneously.
- Fluid intake should be plentiful and physical activity encouraged to facilitate clearance of the stone fragments.
- If there are no immediate complications, patients can be discharged the same day on antibiotic therapy as indicated by the pre-operative urine culture.
- Analgesia is usually only required for the first few days after the procedure and increasing pain should prompt urgent re-assessment for complications.

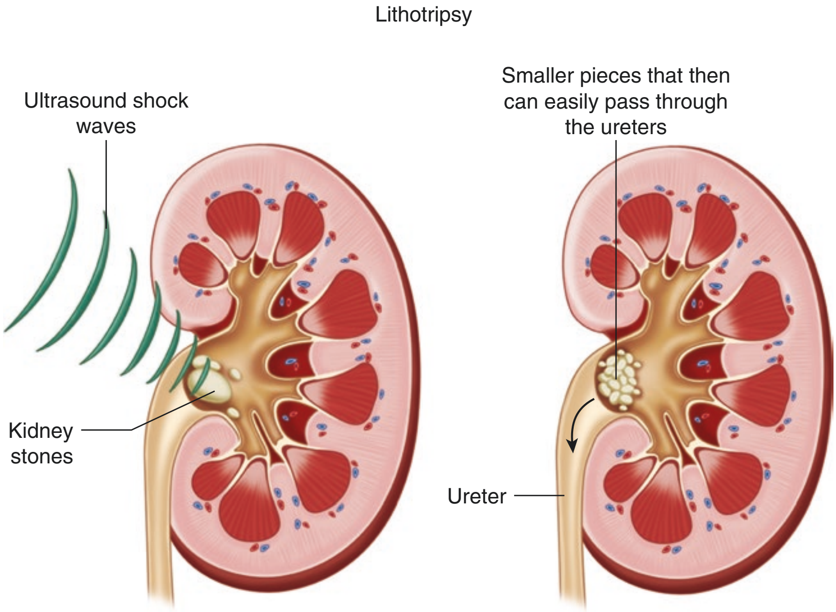


Fig. 9.3 Stone fragmentation achieved with schock waves delivered from outside the body

Follow Up

– One to 2 weeks after the procedure, the patient should be reviewed with an USS and AXR to assess for complications, in particular stone fragment ureteric obstruction (Steinstrasse), which occurs in around 1 in 20. Stone fragments should be sent for stone analysis if available.
– If there is a residual stone a further session of ESWL can be considered at 4–8 weeks after the first one.
– Once stone clearance has been achieved the stone metabolic assessments are completed and the patient is monitored for recurrence.

Risks of Procedure

General

Complications can occur in about 5–18 % patients [15–17].
Re-admission rate is up to 15 %.

Specific

Hematuria (8–44 %) [15–17]

This is the most common complication and is usually transient and self-resolving. It may be due to the effect of the shock waves on the kidney or due to the passage of fragments subsequently.

Pain/Colic (3–20 %) [15–17]

Some pain may occur in a high proportion of patients although colic is reported in less than 20 % of them. It can also be associated with nausea and vomiting. In most cases, pain control is achieved by oral analgesia and anti-spasmodics. Persistent pain should trigger further investigations to exclude Steinstrasse ureteric obstruction.

Infection (1–2 %) [15–17]

Urine should be tested pre-operatively and bacteruria appropriately treated prior to ESWL. Stone fragmentation will release bacteria where these are contained within the stone. Particularly in those with a history of UTIs, antibiotic cover for ESWL is advised. Post-operative fever (7–12 %) should be investigated and appropriate antibiotics commenced based on previous sensitivities and local protocols.

Steinstrasse (Stone Street) (1–22 %) [15, 18]

This complication results from a number of stone fragments accumulating in the ureter causing obstruction. It is largely dependent on the size of the stone that has been treated. JJ stents may be inserted prior to treatment in cases of large stones to prevent this complication, although stone clearance is reduced by presence of the stent. This complication is managed by JJ stenting to relief the obstruction and ureteroscopy to retrieve the fragments. In cases of obstruction and sepsis, an emergency nephrostomy may be needed. As Steinstrasse obstruction in children may be asymptomatic, it is prudent to screen for this complication using ultrasound and a Abdominal X-ray following ESWL.

Bruising/Ecchymosis and Hematoma [15–17]

Due to the proximity of the kidney to the skin, the area for the shock wave entry is dramatically smaller in children, and ecchymosis occurs in a large proportion but does not usually need any treatment. Occasionally an intra-renal, subcapsular or peri-renal hematoma may occur. This is usually self-resolving without any lasting effects.

Injury to Other Viscera [15–17]

Serious side effects are rare, including lung contusion, enteric hematoma and perforation. The energy of the shock wave is released at points of change in density from water (body tissues are 90 % water) to stone but also from water to air. In infants, the lungs may need to be shielded from the shock wave to prevent lung contusion, pulmonary hemorrhage and pneumothorax.

Uretero(Reno)Scopy and Retrograde Intrarenal Surgery

Key Points

First line treatment for ureteric stones and an alternative treatment modality for small renal stones in children.

Specific risks of the procedure:

1. *Hematuria*
2. *Infection*
3. *Failure of access*
4. *Ureteric injury/extravasation*
5. *Stricture*
6. *VUR*
7. *Stone retropulsion*
8. *Stone trapped in a basket*
9. *Stent migration*
10. *Forgotten stent*

Name of Procedure

Uretero(Reno)Scopy (URS) or Retrograde Intra-Renal Surgery (RIRS)

Lay Description

A small telescope is passed through the natural urinary orifice via the bladder into the ureter (and kidney). Tiny instruments can be used through the telescope to break the stone into smaller pieces and to remove these. Often an internal soft plastic tube (JJ stent) is placed along the ureter from the kidney to the bladder before and/or after this procedure for a few weeks. The procedure is performed under X-ray guidance. Approximately 90 % stone-free rates at first treatment can be achieved [19, 20, 23].

Intended Benefit

If the stone is impacted in the ureter causing obstruction, the objective is to disengage the stone and insert a stent to release the obstruction. For stone clearance, stones are fragmented using laser or mechanical energy, and the fragments are extracted by special retrieval devices called baskets. The fragments should be sent for stone analysis. Alternatively, the stone can be pulverised to such small fragments that these will clear spontaneously along a JJ stent.

Technique

Intravenous antibiotics are given during induction of general anaesthesia. Under general anaesthesia, first a cystoscope is inserted into the bladder through the urethra. Under X-ray guidance, a guidewire is inserted into the ureter on the affected side. It is recommended that a safety wire is used at all times so that if an intra-operative complication occurs the patient can be stented safely and the procedure halted. Placement of a second wire can help “open the ureter” by allowing the ureteroscope to pass between the two wires. A longer telescope (rigid or flexible) is then inserted over/along one of the wires and passed up the ureter to the location of the stone(s). The stone(s) are disintegrated using a mechanical probe or laser and the fragments extracted by a stone basket. Holmium-YAG laser energy is the preferred modality for stone fragmentation in view of the small size of probes allowing their passage through the smallest of ureteroscopes [21]. A ureteric stent is left in place in the majority of cases, together with a bladder catheter, which is inserted at the end of the procedure.

The use of a ureteric access sheath can reduce trauma to the urethra and vesicoureteric junction from repeated passage of the ureteroscope, but may risk ureteral injury [24, 25].

Illustrations (Figs. 9.4 and 9.5)

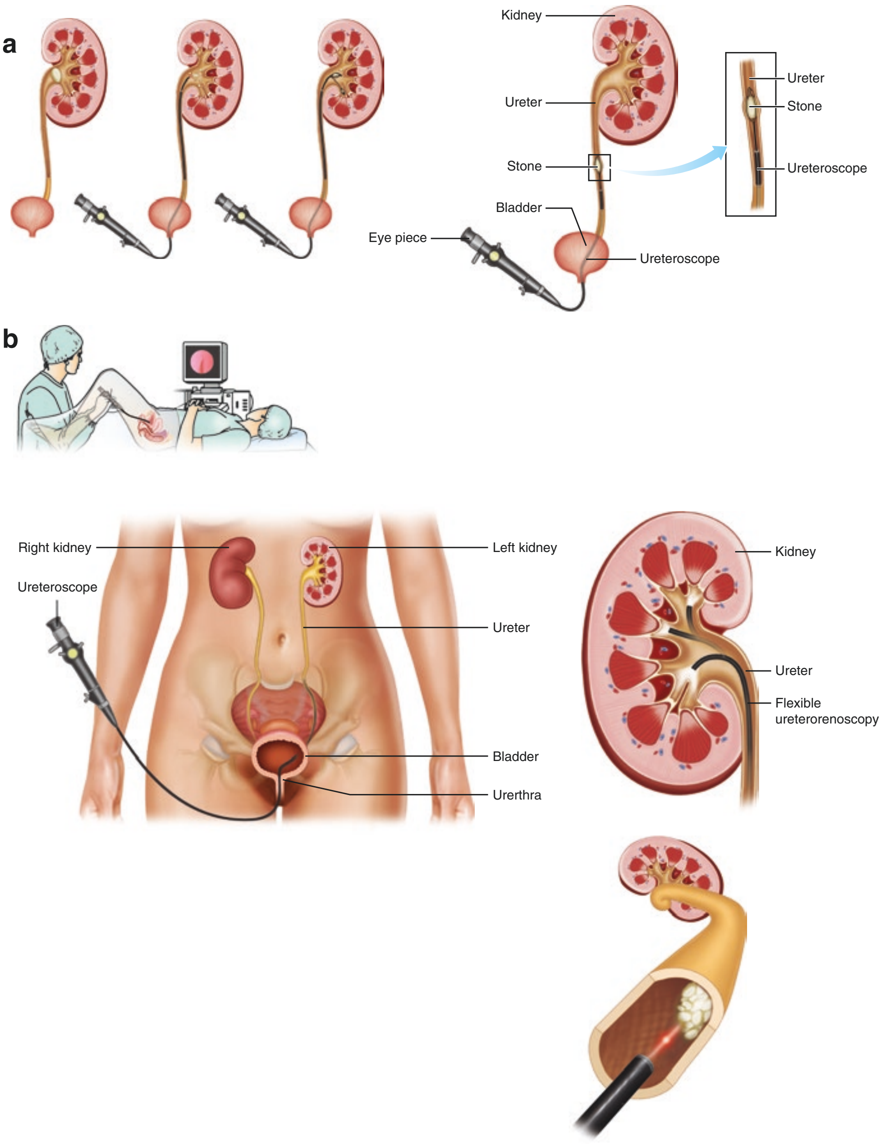


Fig. 9.4 (**a**) Flexible uretreo-reoscopy via the urethra, bladder and ureter, affording access to a renal stone which is then fragmented by laser and fragments removed using a basket. (**b**) Rigid ureteroscopy providing access via the urethra and bladder to a stone within the mid ureter

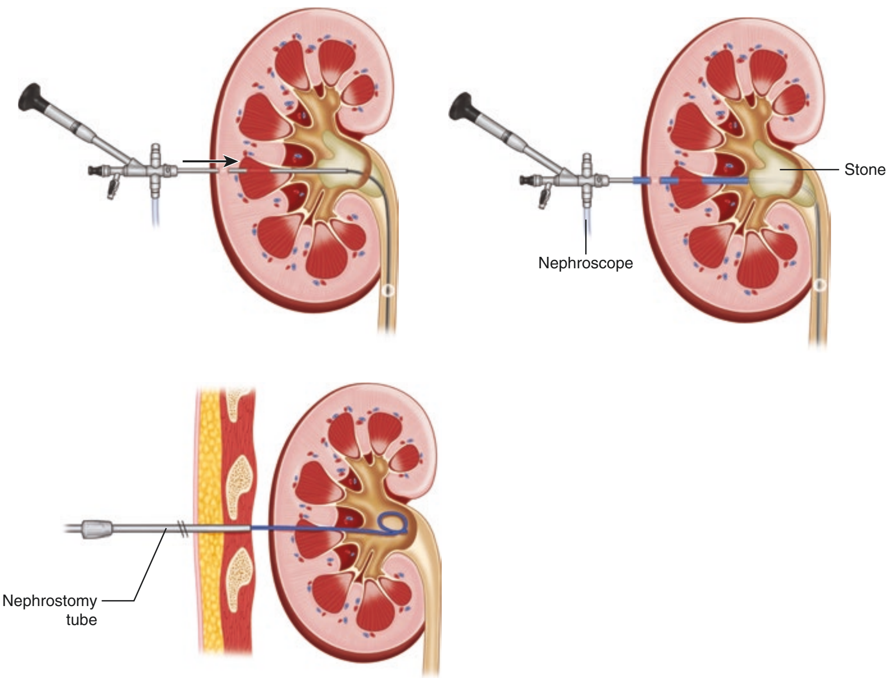


Fig. 9.5 Access tract into kidney, nephroscope is introduced through the tract to treat the stones and nephrostomy tube is left at the end of procedure

Post-operative Expected Course

– Patients can eat and drink as soon as recovered.
– Intravenous fluids help clear any hematuria and stone dust.
– Antibiotics intravenously followed by orally are administered to treat urinary infection.
– If a bladder catheter was inserted, this can usually be removed 1–2 days after surgery.
– The child may experience pain for the first 24–72 h and may require opioid analgesia for the first 24 h.
– Children are discharged home once they are passing urine normally. The average hospital stay is 2 days.

Follow Up

If a JJ stent was inserted, this will require cystoscopic removal under general anaesthetics 2–6 weeks later.

Two to three months after the procedure, the patient is reviewed with USS and AXR to assess for complications and residual stones and to complete metabolic stone assessments.

Risks of Procedure

General [22, 25–29, 32]

Complications occur in approximately 8 % of cases. Most are low-grade and self-limiting.

Specific

Hematuria (27 %) [22, 25–29, 32]

Post-operative hematuria is common but usually resolves in a matter of hours with conservative management. Profuse hematuria is rare (<1 %) and requires prompt evaluation.

Infection (3–19 %) [22, 25–29, 32]

As for any stone intervention, pre-operative checking for and treatment of bacteruria is mandatory. Despite this and the use of antibiotic prophylaxis at induction, UTI, pyelonephritis or septicaemia do occur following ureteroscopy in 3–19 %.

Failure of Access (2–10 %) [22, 25–29, 32]

Where a stone has been impacted in the ureter for some time, gaining access to the stone and the ureter above, or even the placement of a guide-wire, can be challenging. The greatest of care is required to avert ureteric injury. Hydro-distension of the ureter below the stone via extremely prudent ureteroscopy may cause the stone to be pushed back into the dilated ureter above. Alternatively judicious laser-fragmentation of the stone may allow a wire to be passed for JJ stent placement. In the presence of sepsis, insertion of a nephrostomy should be considered. Once the sepsis and ureteric oedema have improved, a second ureteroscopy allows the stone to be treated.

Ureteric Injury/Extravasation (0–10 %) [24]

Injury to the ureter can be the consequence of the use of laser or lithoclast too close to the mucosa, placing undue pressure onto the stone against the ureteric wall, high-pressure irrigation, or result from direct trauma from the guidewire or ureteroscope, dilatation of the ureteric orifice or placement of an access sheath. The procedure may be completed cautiously in the presence of a superficial mucosal abrasion before placement of a stent. Otherwise, the procedure must be terminated and a JJ stent placed. The procedure can then be completed by a second ureteroscopy 2–3 weeks later. Displacement of a stone or fragment into or through the ureteric wall is uncommon but requires close follow-up for stricture formation.

Stricture (0–2 %) [22, 25–29, 32]

Stricture formation is rare and may be a consequence of stone impaction, its trans-ureteric erosion or displacement, or iatrogenic injury. It has been postulated that this relates to active dilatation of the uretero-vesical orifice and can be avoided by a period of “passive” dilation using a JJ stent.

VUR (0–15 %) [22, 25–29, 32]

This is usually related to the use of 8.5/9.5/11.5 Fr ureteroscopes. But it is transient, low-grade and clinically insignificant. Post-operative cystography is not required.

Stone Retropulsion (1–2 %) [30]

Laser lithotripsy (especially when using a long-wave setting on the latest lasers) is associated with less retropulsion than pneumatic or electrohydraulic lithotripsy. In addition, various devices, such as for instance stone cones and parachutes, specialised baskets, and other strategies, for example reduced irrigation, head-up positioning, and gels placed proximal to the stone, have been used in an attempt to reduce the incidence of retropulsion. If stone fragments are lost into the kidney, or indeed the whole stone, then flexible URS may be utilised along with laser during the same procedure, alternatively ESWL can be used subsequently.

Stone Trapped in a Basket

This may occur if fragments are caught in the basket and the stones cannot be disengaged. The basket should be dismantled and a laser fibre passed alongside the stone and the fragments fragmented using the laser fibre. Once the trapped stones are made into smaller fragments, the basket may disengage or the stones may fall out of the basket and the basket can be withdrawn. If the vision is obscured by bleeding and a safety wire is in place then a stent may be placed and the procedure halted.

Stent Migration (1 %) [22, 25–29]

The stent may migrate into the ureter/renal pelvis or indeed fall into the bladder and be voided out per urethra. This usually relates to inappropriate size of stent, inaccurate placement, or a markedly dilated ureter.

Forgotten Stent [31]

Timely removal of a stent is paramount, and a register of patients with a JJ stent may help avert disaster. Forgotten stents may be the cause of severe encrustation, recurrent stone formation and UTIs, renal deterioration and litigation. In the most severe cases, accurate delineation of the stent stone burden may require a CT scan. A combination of interventions may be needed, including ESWL, PCNL and cysto-uretero-renoscopy, to remove encrustation, so as to enable safe stent extraction and to prevent ureteric avulsion.

Percutaneous Nephrolithotomy

Key Points
First-line treatment modality for Staghorn stones, renal stones >20 mm, lower pole stones >10 mm, cystine or struvite stones.
Specific risks of the procedure:

1. *Hemorrhage*
2. *Fever*
3. *Infection and sepsis*
4. *Hypothermia*
5. *TUR syndrome*
6. *Renal pelvic laceration, extravastion of fluid or Urine leak (urinoma)*
7. *Hydrothorax and pleural injury*
8. *Intestinal Injury*
9. *Steinstrasse*
10. *Pelvi-calyceal scarring and stricture*

Name of Procedure

Per-Cutaneous Nephro-Lithotomy (PCNL)

Lay Description

Keyhole access into the kidney allows a telescope to be passed directly into the kidney for removal of the stone(s) with or without their fragmentation. The procedure includes USS and X-ray screening for guidance. Stone clearance is achieved in over 80 %, as a single procedure for the vast majority.

Intended Benefit

Although PCNL is an invasive treatment, it achieves stone-free rates ranging from 86.9 to 98.5 %. It can offer complete stone clearance in a single hospital stay with minimal morbidity. Where residual stones do persist after PCNL, these can be cleared subsequently by ESWL, second-look PCNL and/or uretero-renoscopy. The "sandwich" treatment combines PCNL-ESWL-PCNL, allowing residual stones to be fragmented and then accumulate in fewer calyces for a second look PCNL [33–35, 45–46].

Technique

Urine infection should be treated before surgery. The child may be admitted the day before for intravenous fluids and antibiotics. Blood should be checked for clotting disorders and grouped prior to PCNL. Intravenous antibiotics are given during induction of general anaesthesia. A telescope (cystoscope) is inserted into the bladder through the urethra. Under X-ray guidance, a ureteric catheter is inserted into the ureter to just below the pelvi-ureteric junction to allow retrograde ureteropyelography and flushing to prevent stone migration down the ureter during the procedure. The ureteric catheter can be secured to a urethral balloon catheter, and both are then secured to the skin.

The patient is then repositioned, generally to a prone position. The anatomy of the collecting system and the position of the stones are assessed by either ultrasound or retrograde pyelography, in order to plan the optimal percutaneous access, including the number and location of punctures. Needle puncture under ultrasound guidance enables the shortest tract to the desired calyx and avoids visceral injury. Access is confirmed by return of clear fluid through the needle. Under fluoroscopy, a guidewire is passed via the pelvi-calyceal system into the ureter. The tract is dilated over the wire to the chosen Amplatz sheath, through which the nephroscope (12–20 Fr) is passed [35]. Under direct vision, pneumatic lithoclast or ultrasound probes, or holmium:yttrium aluminum garnet (Ho:YAG) laser fibres can be used for stone fragmentation. Stone pieces can be removed by vortex, stone graspers or wire baskets and are sent for stone analysis. Use of the combined suction and ultrasound probe can ensure complete clearance of even the smallest of stone chips. A nephrostomy drainage tube is placed via the tract in most cases and enables clots to be flushed. A JJ stent may be inserted if deemed necessary by the operating surgeon [41–46].

Illustrations *(Figs. 9.6 and 9.7)*

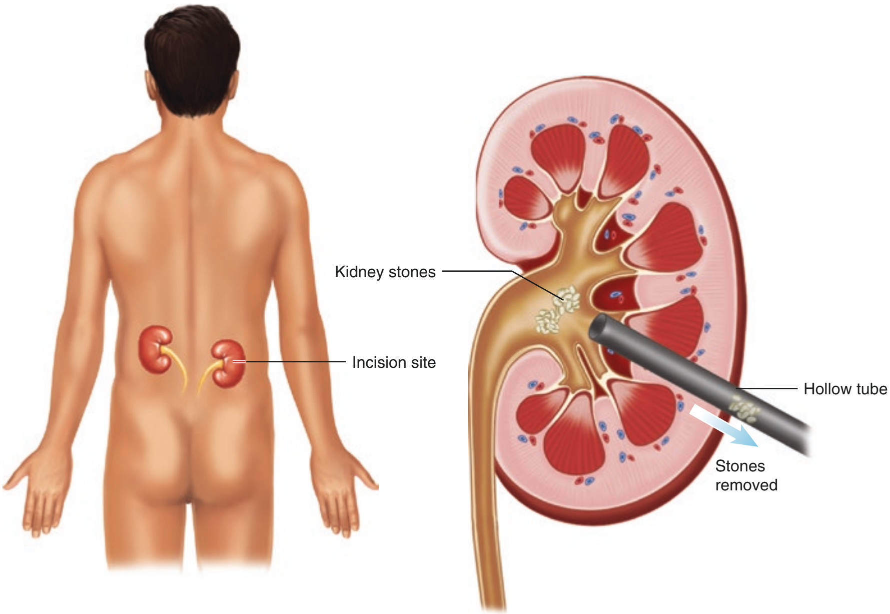


Fig. 9.6 Prone position and incision in the loin for percutaneous access into the pelvicalyceal system of the kidney

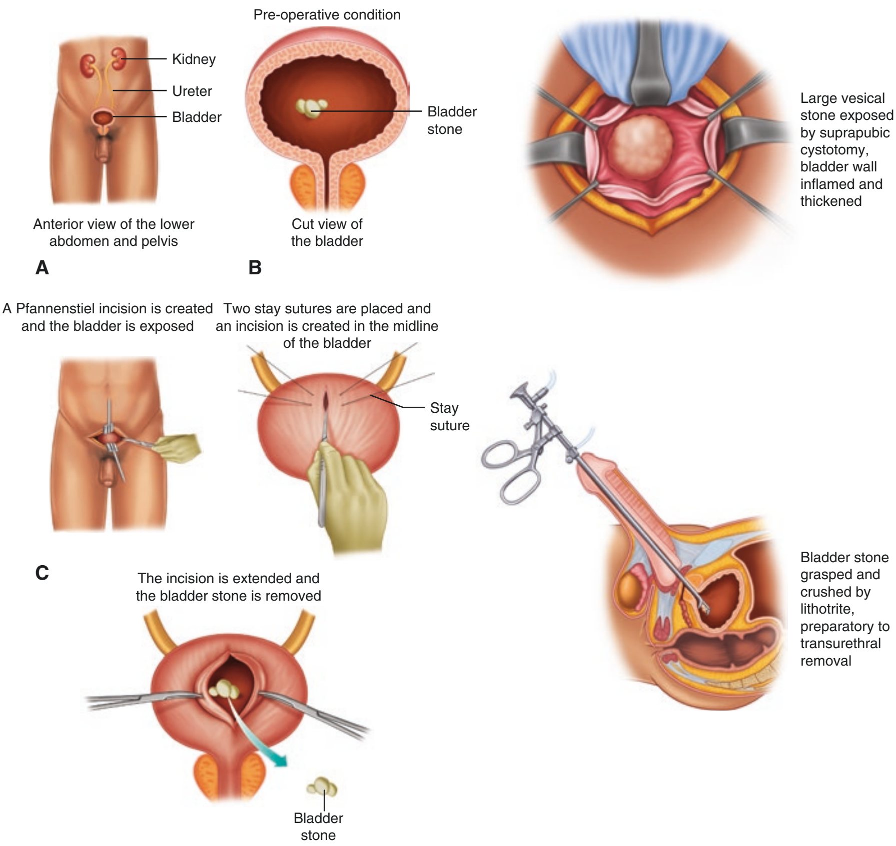


Fig. 9.7 Open cystolithotomy and transurethral endoscopic removal of a bladder stone [52]

Post-operative Expected Course

- Patients can eat and drink as soon as recovered. Additional intravenous fluids help clear hematuria.
- Intravenous antibiotics are recommended for 24–48 h in those with a history of urinary infection, followed by a treatment course and then prophylactic oral antibiotics.
- The child will experience pain over the first 24–72 h, initially requiring opioid analgesia.
- The nephrostomy tube is clamped 24–72 h after surgery and removed later that day, followed by removal of the urethral catheter. Anticholinergic medication can help combat bladder spasms during this time.
- Children are discharged home once they are passing urine normally. The usual hospital stay is 3–5 days.

Follow Up

If a JJ stent was inserted, this will require cystoscopic removal 2–6 weeks after the procedure under general anaesthesia.

Two to 3 months after the PCNL, the patient is reviewed in the outpatient clinic an ultrasound, AXR and functional imaging, and the stone metabolic evaluation is completed.

Risks of Procedure

General [36–40, 48]

Complications occur in approximately 10–28 % of cases.

Specific

Hemorrhage (9–11 %) [39, 40, 42, 43, 45]

Bleeding requiring transfusion is reported in 0.4–24 %, and it can result from the tract, or from undue manipulation of the sheath and scope. If bleeding occurs from the tract, the Amplatz should be advanced without angulation to tamponade the bleeding. A large nephrostomy tube should be placed through the tract and the procedure discontinued. If bleeding persists, a large balloon catheter can be placed into the calyx, the balloon inflated, and the parenchyma compressed by gentle continuous traction on the balloon catheter for pressure hemostasis.

Late bleeding after PCNL, typically after 1 week, may indicate a vascular injury, such as intra-renal pseudo-aneurysm or arterio-venous fistula. This rarely develops after PCNL and is managed by angiography and selective embolisation [47].

Fever (11–30 %) [39, 42]

Transient fever following PCNL is common and often resolves spontaneously. However, it may also indicate life-threatening urosepsis or dislodgment of the nephrostomy tube with a peri-nephric collection [48].

Infection (2–7 %) [39, 48]

Upto 65 % of patients requiring PCNL have positive urine cultures. Treatment of positive urine cultures prior to PCNL and antibiotic prophylaxis peri-operatively is strongly advised. Despite this, post-operative UTI/urosepsis may still occur in at least 2 % of patients, the management required ranging from parenteral antibiotics to intensive care support.

Hypothermia [36, 37]

This is a significant risk in paediatric PCNL. All measures to prevent it should be rigorously adhered to, such as, warmed operating theatres, use of irrigation and intravenous fluid actively-warmed to body-temperature, and avoidance of pooling of fluid on or under the patient by use of water-proof sticky drapes. The length of procedure time (operative and anaesthetic induction) is also relevant, with no incidence of hypothermia in shorter procedures.

TUR Syndrome

The risk of absorption of irrigation fluid causing hyponatremia and electrolyte imbalance can be minimised by using a low pressure irrigation system and ensuring that warm isotonic solution is used. The procedure should be completed in the shortest time possible and electrolytes checked post-operatively.

Renal Pelvic Laceration, Extravastion of Fluid or Urine Leak (Urinoma) (1–5 %) [39, 42]

This may present as post-operative fever or loin pain. They usually resolve with conservative management but may need percutaneous drainage if large, especially if infected.

Hydrothorax and Pleural Injury (1–2 %) [42]

Punctures above the 11th rib significantly increase the risk of intra-thoracic complications. Here a post-operative chest radiograph is advised to look for complications. Most cases of small hydrothorax are asymptomatic and resolve spontaneously; if the hydrothorax is large, a chest drain is likely to be required.

Intestinal Injury (0.3 %) [39]

This complication is rare, particularly in the prone position and if ultrasound is used to guide the puncture, as the colon is readily visualised and easily avoided. If an intra-peritoneal bowel injury is suspected intra-operatively, diagnostic laparoscopy may help delineate the injury before the onset of peritonitis.

Steinstrasse (Stone Street) [48]

Stone fragments migrating down the ureter are a well-recognised risk during PCNL. A retrograde ureteric catheter is therefore placed just below the pelvi-ureteric junction at the start of the procedure to enable any stone fragment threatening to do so to be flushed back into the renal pelvis for extraction. If stone fragment migration is unrecognised, these fragments may accumulate in the distal ureter, potentially causing obstruction. Ureteroscopy and stenting may be required and subsequent treatment by ESWL.

Pelvi-calyceal Scarring and Stricture (0.2 %) [39, 44]

Persistent urine leak via the tract can be an indicator. However, scarring of the renal pelvis or pelvi-calyceal junction can occur after the immediate post-operative period. Many advocate antegrade pyelography to exclude obstruction prior to removal of the post-procedure nephrostomy/pig-tail catheter. Placement of a JJ stent at the time of the PCNL does not always protect against this complication. Vigilance is required for its detection to avert renal loss. Endo-urological approaches can be considered for a short stricture, however extensive or persistent stricturing will require open uretero- or pelvi-calyceal anastomosis.

Minimally Invasive Cystolithotomy

Key Points

Bladder stones can be dealt with in a variety of ways, including open cystolithotomy or by minimally invasive techniques either per urethra or percutaneously.

Specific risks of the procedure:

1. *Infection*
2. *Hematuria*
3. *Extravastion and urine leak*
4. *Urethral stricture*

Name of Procedure

Open cystolithotomy (open bladder surgery)
Endoscopic cystolithotripsy (optical mechanical cystolithotripsy)
PerCutaneous CystoLithotomy (PCCL)

Lay Description

Most bladder stones can be dealt with through a small telescope passed either through the natural urinary orifice or keyhole into the bladder. If stones are very large (>4 cm), an open operation may be recommended [49–51].

Intended Benefit

The aim is to achieve a stone-free state in the shortest possible time and with minimal complications. Endo-urological procedures afford a reduced length of stay and similar procedural time as compared to open cystolithotomy.

Technique

All procedures are performed under general anaesthesia and antibiotics are given on induction [52–54].

1. Open cystolithotomy: An incision is made in the lower abdomen to access the bladder. The bladder is opened and the stone removed. The bladder is repaired with absorbable sutures, and a catheter is inserted into the bladder via the urethra.
2. Endoscopic cystolithotripsy: a telescope (cystoscope) is inserted into the bladder through the urethra. A combination of pneumatic/ultrasonic lithotripsy devices can be used, with aspiration of the stone fragments. Stone-free rates of 90 % can be achieved with reduced operative time and postoperative stay as compared to open surgery.
3. PCCL: After suprapubic needle puncture and dilatation of the tract over a guidewire, an Amplatz sheath is placed directly into the bladder, allowing telescopic visualization, fragmentation and removal of the stone(s). PCCL avoids prolonged instrumentation of the urethra and decreases the risk of iatrogenic urethral stricture. The large working sheath facilitates the extraction of larger stone fragments and thereby reduces procedure time. After PCCL, a suprapubic catheter is left in situ, as well as a urethral catheter to enable irrigation if required.
4. In the presence of a Mitrofanoff channel, this can be serially dilated to an 18Fr Amplatz for access onto a bladder stone.

Illustrations (Fig. 9.8)

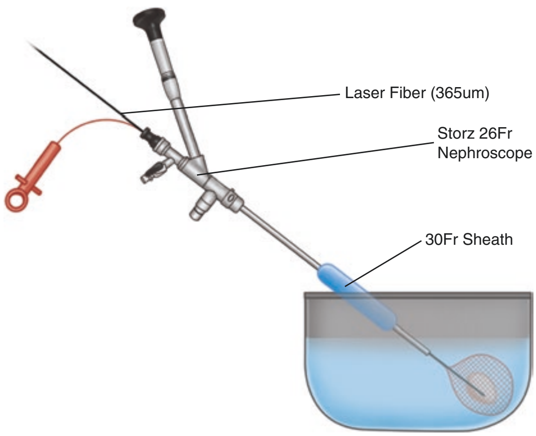


Fig. 9.8 Schematic view of laser lithotripsy during PCCL

Post-operative Expected Course

- Patients can eat and drink as soon as recovered. Intravenous fluids will be given to clear any hematuria.
- Intravenous antibiotics are recommended for 24–48 h in those with a history of urinary infection, followed by a treatment course and then prophylactic oral antibiotics.

  1. Open cystolithotomy: Most patients will stay 2–3 days after the operation. The catheter will remain in place for 7–14 days, depending on the size of the incision in the bladder.
  2. Minimally invasive techniques: The average hospital stay is 2 days. The catheters are removed after 24–48 h. After a PCCL, removal of the suprapubic catheter at 1 or 2 days after the surgery is followed by that of the urethral catheter 24 h later, to afford sufficient time for the percutaneous tract to close.

Follow Up

The patient is followed up with an ultrasound and AXR 2–3 months after the procedure.

Risks of Procedure

General [54–56]

Complications rates range from <1 to 10 % and are more common after endo-urological approaches.

Specific

Infection (11 %) [56]

Mainly associated with PCCL and is managed by parental antibiotics for 2–3 days.

Hematuria (4 %) [56]

This is rarely a significant issue. Injury to bladder mucosa with the laser or due to inadvertent crushing within the jaws of the stone punch can result in bleeding which may necessitate bladder wash-outs/irrigation. It usually resolves spontaneously in 24 h.

Extravasation and Leak

Percutaneous suprapubic puncture should be performed with a moderately full bladder so as to avoid a trans-peritoneal course and intestinal injury [56]. Extravasation is associated with large working tracts and high irrigating pressure and vesical overdistension. A recurrent or persistent suprapubic leak generally settles over a matter of days by placement of an appropriately sized urethral catheter. Intra-peritoneal leaks may require operative repair [54, 55].

Urethral Stricture (2–4 %)

Repeated instrumentation of the small calibre paediatric urethra, especially where the stone size is large, can lead to stricture formation. Optical urethrotomy and intermittent dilatation have been described [54], but if the stricture persists a formal repair is indicated.

Paralytic Ileus and Abdominal Distension (10 %) [54–56]

These are reported with large working tracts and high irrigating pressure.

Laparoscopy and Robotic Surgery

Key Points

It is primarily indicated for nephrectomy of non-functioning moieties, concomitant correction of pelvi-ureteric junction obstruction, for stones in abnormal locations (e.g. *in a calyceal diverticulum*) *or anomalous urinary tracts, such as ectopic kidneys, or after failed endourological procedures as an alternative to open surgery.*

Specific risks of the procedure:

1. *Coversion to open*
2. *Stone migration*
3. *Hemorrhage*
4. *Extravastion and urine leak*
5. *Urethral stricture*

Name of Procedure

Laparoscopic/robotic nephrolithotomy, pyelolithotomy or ureterolithotomy

Lay Description

Laparoscopy and robotic-assisted laparoscopy are current treatment options for urinary stones in adults, but have a limited role in children [62]. Keyhole surgery allows access to the urinary system and the stones. Post-operative recovery and hospital stay are shorter than with open surgery.

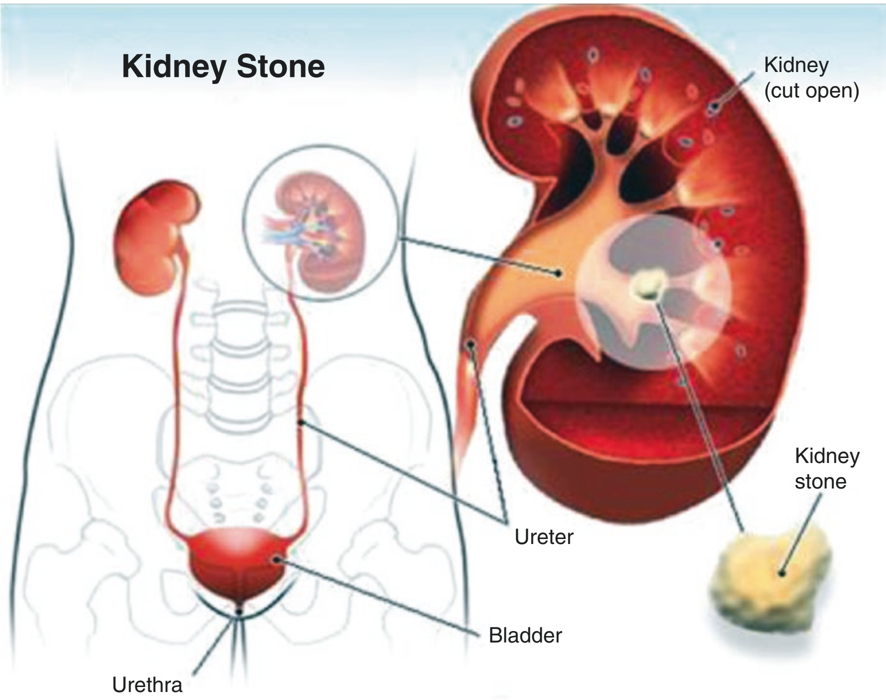
Intended Benefit

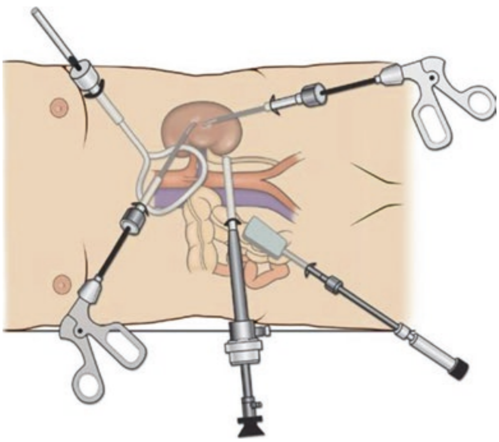
It is primarily indicated where concomitant pyeloplasty is required, for large stones resistant to extra or intracorporeal lithotripsy, for stones in abnormal locations (e.g. in a calyceal diverticulum) or anomalous urinary tracts, for cystine stones unsuitable for PCNL, or in cases of failed access or failed endourological procedures as an alternative to open surgery. Laparoscopic pyelolithotomy takes a longer time to perform, and requires considerable skills, although it carries similar hospital stay and stone-free rates (70–100 %) to PCNL [57–59, 64, 65].

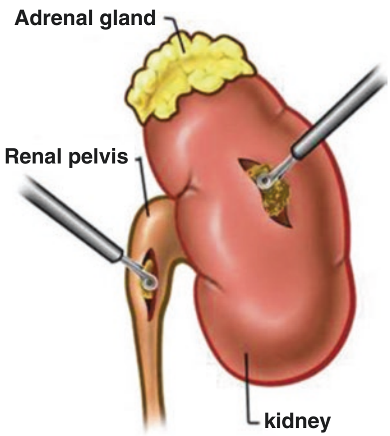
Technique

Laparoscopic nephrolithotomy and pyelolithotomy are performed under general anaesthetic using either a transperitoneal or retroperitoneal approach. In the transperitoneal approach, the abdomen is insufflated with carbon dioxide. In the retroperitoneal approach, a small incision is made in the back and a dissecting balloon is inserted to create space: carbon dioxide pneumoretroperitoneum is then established. Two or three additional small incisions are made [60]. In a nephrolithotomy, once the kidney has been mobilised, the stone is located by X-ray, laparoscopic ultrasound or by evidence of a bulge or depression secondary to scarring. The renal capsule and parenchyma are incised and the stone(s) is removed. A JJ stent may be inserted through the kidney. In a pyelolithotomy, the stone is accessed through an incision in the renal pelvis (pyelotomy). Once the stone is removed, the pyelotomy is usually closed with sutures, with or without a stent. A perinephric drain may be left in situ after closure of the renal pelvis [61–63].

Illustrations







Post-operative Expected Course

– Patients can eat and drink as soon as recovered. Additional intravenous fluids help clear hematuria.
– Intravenous antibiotics are recommended for 24–48 h in those with a history of urinary infection, followed by a treatment course and then prophylactic oral antibiotics.
– Where a perinephric drain has been placed, this can usually be removed after 24 h.
– Hospital stay averages 2–3 days.

Follow Up

If a JJ stent was inserted, this will require cystoscopic removal under general anaesthetics 2–6 weeks later.

Two to 3 months after the procedure, the patient is reviewed with USS and AXR to assess for complications and residual stones and to complete metabolic stone assessments.

Risks of Procedure

General

Complications rates range from 5 to 10 % [62–66].

Specific

Fever (3 %) [63–66]

Is treated by parental antibiotics.

Conversion to Open Surgery (10–16 %) [62, 64, 65]

Can be necessitated by dense peri-renal/ureteric inflammation/fibrosis, bleeding or stone migration.

Stone Migration [62, 65]

Laparoscopic ultrasound may help locate a stone that has migrated into the calyces. In this situation, irrigation, laparoscopic graspers or flexible nephroscopy can help retrieve a stone. Care must be taken not to lose the stone in the peritoneal cavity on extraction.

Hemorrhage

Blood loss appears to be similar to PCNL [64, 65].

Leak (7–12.5 %) [62]

Urine leak is primarily reported after laparoscopic ureterolithotomy; this is managed by JJ stenting with or without placement of a drain. The retroperitoneoscopic approach has the advantage that a urinary collection is limited to retroperitonium and usually resolves spontaneously.

Ureteric Stricture [63, 66]

Strictures may result from stone impaction in the ureter or as a complication of the procedure. A persistent stricture may require resection and uretero-ureterostomy.

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Chapter 10
General Laparoscopy

Nathalie Webb and Chris Kimber

Key Points

- Laparoscopy is safe in trained surgical hands
- Attention to equipment set-up and ergonomics improves task performance and reduces risks to surgeon and patient
- Energy sources must be fully understood and used judiciously to prevent inadvertent injury

Specific risks of the procedure:

1. Inadvertent vascular or organ injury during port insertion or surgical manipulation
2. Insufflation risks: barotrauma and gas embolism
3. Risk of conversion to open access
4. Risks associated with tissue retrieval
5. Risks to operating surgeon: musculoskeletal and neural injury

Name of Procedure

General Laparoscopy.

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Lay Description

"Key-hole surgery" for the purposes of diagnosis, removal or pathology or reconstruction. It involves insertion of a thin telescope through a small cut in the abdominal wall or back under general anaesthesia. Gas is gently pumped into the abdominal cavity to create workspace for the telescope and special surgical instruments, allowing surgery to occur through very small incisions.

Intended Benefit

Undertake diagnostic evaluation, address pathology or reconstruct anatomic abnormality via minimally-invasive route to reduce post-operative pain, recovery time and external scarring.

Technique

- General anaesthesia with muscle relaxation
- Insertion of ports (transperitoneal or retroperitoneal)
- Insufflation
- Procedural steps specific to the purpose of the operation, including use of catheters, stents and drains
- Port site closure

Illustrations

Expected Postoperative Course

- Eat and drink as soon as recovered
- Regular oral analgesia
- Antibiotics as indicated by specific procedure
- Discharge same day or next day as determined by specific procedure

Follow Up

Determined by specific procedure

Risks of Procedure

General

There is little data from which to draw reliable conclusions as to the rate of complications associated with laparoscopy in children specifically, however the overall rate of complications associated with a laparoscopic approach appears low, in the range of 1–3 % [1–4]. Although laparoscopy is usually safe, a small minority of patients experience life-threatening complications, including injuries to nearby organs or blood vessels. Most complications are related to access and port insertion. Systematic literature review of laparoscopic entry technique [5] found no significant difference between closed-entry and open-entry techniques with respect to vascular and visceral injury. Open access appears safer in children, with 1.2 % significant complication rate compared with 2.6 % for Veress needle technique [4]. Overall rate of complications reduces with surgeon and unit experience [4, 6].

Positioning Risks

Depending on the specific procedure being performed laparoscopically, patient positioning can pose risk of neuromuscular injury. The rate of this is unknown, but surveys of adult populations has found a rate of 2.8 % [1].

Vascular Injury

Injury to major vessels associated with laparoscopy alone is rare: 0.01–5 per 1,000, but associated mortality is high (8–17 %) [2, 7].

Organ Injury

Inadvertent injury to other organs most commonly involves damage to the bowel and this risk is likewise very low: 0.04–6 per 1,000 [8]. Most can be repaired easily if recognised at the time of the procedure, however even small injuries can have catastrophic sequelae if unrecognised and not addressed [1].

Insufflation Risks

Increased intra-abdominal pressure from pneumoperitoneum can cause hemodynamic and respiratory consequences, especially in small children and infants. Minimal pressure must therefore be employed to minimise this risk.

Gas embolism has been reported, but relates more to direct intravascular insufflation (closed-entry technique) than pneumoperitoneum [9].

Conversion to Open

Failed access risk is higher with closed-entry techniques (31 per 1,000 vs 1–20 per 1,000 for open-entry techniques) [5].

Rates of conversion from laparoscopic to open procedure vary widely with procedural indication and surgeon experience. Risks associated with surgeon fatigue rise after 2 hours of advanced endoscopic operating. Emergency conversion for significant bleeding or visceral injury is associated with poorer outcomes in adult series [10].

Tissue Retrieval Risks

Peritoneal and port site implantation and metastasis has been demonstrated after tumour spillage [11]. Use of specimen bags and port site enlargement can help avoid this.

Post-operative Risks

Severe Pain

Shoulder pain is a common complaint after laparoscopy. This is occasionally severe in children, especially after long procedures (1–2 per 1,000) [12].

Port Site Prolapse and Hernia

Omental prolapse may occur after laparoscopy in adults (1.5 %) [13]. It seems less common in infants and children (0.15 %) [4], but can occur through 3 mm port sites. It is usually identified in the early postoperative period and will require return to the operating theatre for omental ligation and port site closure.

Later hernia at port site is less common (0.14 % in adult series); related to port size, closure technique and other patient factors [14].

Risks to Operating Surgeon

Radial nerve injury, shoulder strain and rotator cuff injury, anterior osteophyte and spinal degeneration have all been recognised as risks to the operating surgeon. Attention to ergonomics and additional surgeon-mentors can help reduce this [12].

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Chapter 11
Laparoscopic Surgery of the Upper Urinary Tract

Jessica M. Ming and Walid A. Farhat

Name of Procedure

Laparoscopic Nephrectomy/Hemi-nephrectomy/Pyeloplasty.

Lay Description

Laparoscopic surgery of the upper urinary tract involves an operation using multiple small incisions to either [1]:

1. Remove a part of or entire kidney
2. Repair a kidney/ureteral abnormality.

Intended Benefit

To remove a portion of (partial, heminephrectomy) or a whole kidney (nephrectomy) if a patient is having issues with recurrent urine infections, pain, high blood pressure, tumors or compression of surrounding structures. A kidney may become blocked due to abnormal anatomy (ureteropelvic junction obstruction, UPJ-O) and require reconstruction (pyeloplasty) to resolve pain or kidney swelling.

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Why Use the Laparoscopic Approach?

Goals of laparoscopic surgery are to decrease the amount of scar tissue in the abdomen, improve cosmetic result with small incisions and decrease pain after the operation.

What Happens Before the Operation?

The hospital will contact the family on the time to arrive in the preoperative holding area the day of surgery. All regular medications, if any, should be taken before these surgeries. Children will need to stop eating and drinking for a specific amount of time before surgery, depending on the hospital policy.

Technique

Where Are the Incisions?

Depending on the side of the operation, multiple 1–2 cm incisions (cuts) are made on the abdomen or back for instruments and camera access (Fig. 11.1).

How Is the Surgery Performed?

Through the small incisions, laparoscopic ports are placed for access to the abdomen. The abdomen is filled with CO_2 gas for working space to clearly see the inside structures. Using a camera and TV screens, the kidney is identified. The kidney is either removed or reconstructed inside of the abdomen using laparoscopic instruments (Fig. 11.2a). Decision to repair or remove is dependent on the primary pathology.

The incisions are all closed with stitches that dissolve and covered with a surgical glue or bandages that fall off in few days. If a drain is left, a stitch is passed through the skin to keep it in place.

Postoperative Expected Course

It is expected that there will be minimal pain after the operation and children may require tylenol or small amounts of narcotic medications (such as morphine).

Fig. 11.1 Sites for right sided kidney surgery. One cut is made in the belly button for cosmesis and two other incisions are inserted for dissection

All children are allowed to have clear liquids immediately after surgery and can advance to solid foods as long as they feel well.

A Foley catheter is placed through the urethra (where urine comes out) into the opening of the bladder while the child is asleep. This is kept in place overnight to drain the bladder.

A stent will be placed in the kidney and down the ureter (the tube that drains the urine from the kidney to the bladder) after pyeloplasty surgeries (repair of blocked kidneys) (Fig. 11.2b). This stent is removed 2–6 weeks after the operation. Occasionally, a drain is left in place after the operation and stitched in place. This drain is removed after 5–7 days in the clinic depending on the amount of fluid drainage.

Most patients will be discharged from the hospital within 24–72 h. Some surgeons may prescribe antibiotics after surgery, especially if drains are left in place.

Follow Up

Patients will return to clinic or have a procedure while asleep to remove the stent for a pyeloplasty or any other drains.

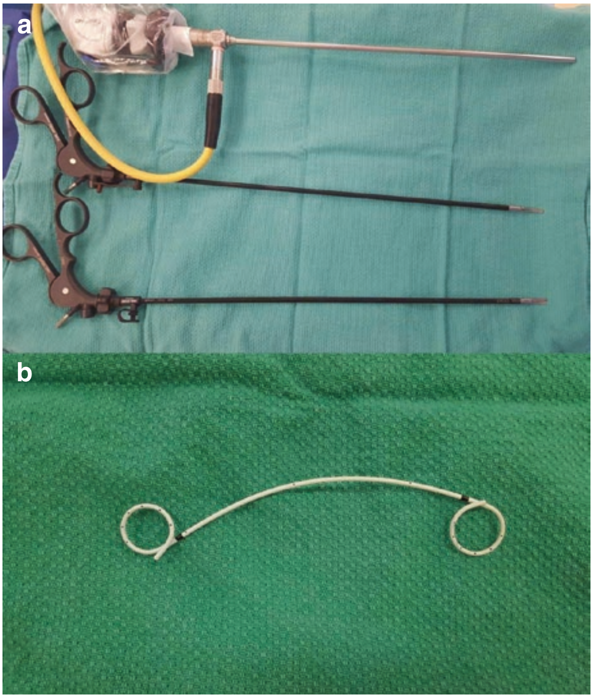

Fig. 11.2 (**a**) Sample laparoscopic instruments and a camera. The camera is connected to a TV screen to see inside the abdomen. The instruments are used to work around the kidney. (**b**) Sample of the ureteral stent, tube to drain the urine from the kidney to the bladder

Follow up for all kidney surgery is 3 months after the operation with a kidney/bladder ultrasound to evaluate the results. After nephrectomy or heminephrectomy, it is important to follow up with primary care physicians for yearly blood pressure and kidney function evaluation.

If a kidney is removed for a tumor or cancer, follow up with the oncologist (cancer doctor) is important for continued long term evaluation based on the type of disease.

Success rates of laparoscopic pyeloplasty are 95–98 % [2].

Risk of Procedure/Complications

Though the surgical, nuring and anesthesia teams optimize every patient pre-operatively, every surgery still carries some risk [3].

Intraoperative

Laparoscopic surgery, when compared to open, can take a longer amount of time to complete.

During the surgery, other abdominal organs can be injured while operating on the kidney. Inside the abdomen, the kidney is surrounded by other organs (intestine and liver/spleen) that sometimes need to be moved to gain access to the kidney. If damage occurs, every effort is made to repair the injury. When noticed in the surgery, the injuries are repaired during the same operation.

If a lot of bleeding is encountered, the surgery may require a larger incision and access to the abdomen with the traditional open technique [4].

Early

The kidney is made up of a solid portion (parenchyma) that makes urine from the body and a hollow portion (collecting system) that is used to collect and drain the urine. When kidney reconstruction is being performed and reconnecting the collecting system is being done then urine may leak from between the suture line (less than 2 % of cases) [5]. Patients can develop pain, fevers, or nausea and vomiting as warning signs of such an occurrence. If this complication happens, a stent in the ureter or drain into the kidney (nephrostomy tube) is used to prevent urine from collecting inside the abdomen. Occasionally and prior to any major intervention such as a stent or tube in the kidney, a foley catheter is placed back into the bladder.

Late

Return of UPJ obstruction can occur in up to 7 % of pyeloplasty cases due to scar formation [6]. We do not usually identify a specific reason for scarring but we think that urine leak around the suture line may increase the chances of scarring and blockage recurrence. If recurrence of the blockage occurs, patients can return to clinic with swelling of the kidney or symptoms of continued pain, similar to those experienced before the surgery.

The incisions may form abnormal scars leaving an unattractive appearance. These may require a fix with a small operation.

After removal of the kidney, it will be important to follow up with your family doctor to check regular blood pressure and kidney function.

Conclusions

Laparoscopic kidney and upper urinary tract surgery is a reasonable option to open surgery. It has the benefits of small scars and less post-operative pain, however can be a longer operation. Parents and patients should be evaluated by the surgeon to discuss all the available options to make an informed decision. Every patient should also be evaluated by all involved members of the medical team to ensure the child receives the best possible care.

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Chapter 12
Robot-Assisted Laparoscopic Ureteral Reimplantation

Andrew C. Strine and Paul H. Noh

Name of Procedure

Robot-assisted laparoscopic (RAL) ureteral reimplantation.

Lay Description

A minimally invasive surgery through a few small incisions to re-tunnel the ureter in the wall of the bladder and thereby correct vesicoureteral reflux (VUR).

Risks

Bleeding, urinary tract infection (UTI), surgical site infection, injury to intra-abdominal organs, ileus, migration or encrustation of stent (if placed), urinary leakage, persistent or recurrent VUR, de novo contralateral VUR (if unilateral surgery), ureteral obstruction, acute kidney injury, chronic kidney disease, lower urinary tract symptoms, urinary retention, open conversion, neuropathy, rhabdomyolysis, and anesthetic complication.

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Benefits

Correction of VUR, prevention of pyelonephritis, preservation of renal function, less postoperative pain and shorter convalescence than open surgery, discontinuation of antibiotic prophylaxis, and less frequent follow-up for imaging studies.

Technique

Cystoscopy may be performed before surgery to evaluate for the presence of UTI and suitability of the ureter for an extravesical approach at the discretion of the surgeon. Either a double-J ureteral stent or externalized ureteral catheter may also be placed at that time.

RAL ureteral reimplantation may be performed through either an intravesical or extravesical approach. Only a few small, single-center series have reported their outcomes with an intravesical approach. An extravesical approach seems to be preferred by most pediatric urologists due to its relative ease and reproducibility when compared to the more technically challenging intravesical approach [1, 2].

The extravesical approach replicates the Lich-Gregoir technique for open ureteral reimplantation. The patient is placed under general endotracheal anesthesia. The patient may be placed in a supine or modified dorsal lithotomy position. A Foley catheter is placed on the sterile filed to allow for intraoperative manipulation of bladder filling. A three-trocar configuration is typically used with one trocar for the endoscope through the umbilicus and two working trocars on each side of the midline. An assistant trocar may be used at the discretion of the surgeon. The ureter is mobilized from the pelvic brim to the bladder after opening the overlying peritoneum. The vas deferens or uterine artery is preserved. A detrusorotomy of adequate length is created in line with the ureterovesical junction (UVJ). The presumed dorsomedial location of the neurovascular bundles is avoided by dissecting in close proximity to the ureter and avoiding any circumferential dissection around the UVJ. A detrusorraphy is performed with either a running absorbable suture or simple interrupted absorbable sutures. Additional considerations to maintain the length of the submucosal tunnel include an advancement suture at its distal aspect and incorporation of the ureteral adventitia along the detrusorotomy. During the detrusorotomy and detrusorraphy, a transabdominal suture through the bladder and/or around the ureter is particularly helpful to allow for adequate exposure and retraction (Fig. 12.1). The fascia and skin are closed at each trocar site. A drain is not routinely placed.

Postoperative Course

The patient is typically admitted to the hospital, although select patients may be a candidate for outpatient surgery without a Foley catheter in our experience. The diet is advanced as tolerated on the day of surgery. Our preference is to start a regimen of scheduled and alternating intravenous acetaminophen and ketorolac for postoperative

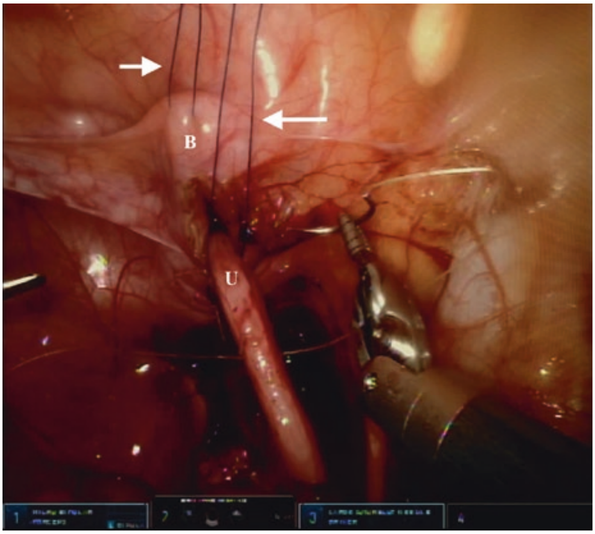

Fig. 12.1 Transabdominal suture through the bladder (*B*, *short arrow*) and around the ureter (*U*, *long arrow*) during detrusorraphy

analgesia, which may be transitioned to oral acetaminophen and ibuprofen prior to discharge. Intravenous narcotics may be administered as needed but are not routinely required in our experience. A first generation cephalosporin is administered for perioperative antibiotic prophylaxis. Early ambulation is encouraged. The Foley catheter is removed for a voiding trial on postoperative day #1. A vast majority of patients are discharged to home on postoperative day #1.

Follow-Up

The patient returns for a postoperative evaluation with a renal and bladder ultrasound in 1–3 months.

Evidence

The success rate after RAL extravesical ureteral reimplantation is variable in the literature, ranging from 72 to 99 % [3–13]. This variability may be attributed to the differing severity of VUR, treatment of contralateral non-refluxing ureters, and definition of success in these series. Some series routinely obtained a postoperative voiding cystourethrogram, while others only obtained them as clinically indicated or not at all. With the largest multi-institutional series to date, Grimsby et al. observed a fairly low success rate of 72 % in 93 ureters by robotically experienced surgeons [12]. Nevertheless, the success rate has been improving over time. Gundeti et al. demonstrated an improvement in their success rate from 67 to 87 % with specific technical modifications [13].

The overall complication rate is also variable, ranging from 0 to 30 %. Specific complications include urinary leak (0–10 %), ureteral obstruction (0–5 %), and ileus (0–4 %) [3–13]. Only one case of open conversion has been reported [13]. The incidence of de novo VUR for unilateral surgery has been inconsistent reported but observed to be as high as 22 % of patients in one series [8]. Transient urinary retention is a complication that is unique to an extravesical approach, particularly when performed bilaterally. An overly aggressive dissection around the UVJ is thought to disrupt the neurovascular bundles from the pelvic plexus and contribute to this complication. Its incidence is quite low but has been demonstrated in up to 10–12 % of patients in several small series [3–13]. Kasturi et al. reported the large single-center series of 150 patients undergoing RAL extravesical ureteral reimplantation with follow-up for at least 2 years. All patients were toilet trained before surgery and evaluated with a pre- and postoperative voiding diary, uroflowmetry, measurement of postvoid residual volumes, and validated questionnaire. They did not observe any de novo lower urinary tract symptoms or urinary retention after surgery. They argued that the magnified three-dimensional visualization with a robotic platform facilitates the careful dissection of tissues around the UVJ and preservation of the pelvic plexus [7].

Several studies have performed a retrospective comparative analysis between open and RAL extravesical ureteral reimplantation. A comparison was made to either an open intravesical or extravesical approach in two studies each. All studies demonstrated similar success and complication rates between open and RAL extravesical ureteral reimplantation. They also observed a decreased postoperative narcotic requirement in patients undergoing RAL extravesical ureteral reimplantation but conflicting results for operative time and length of hospitalization [4, 5, 9, 14].

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Part V
Genitalia

Chapter 13
Hydrocele and Hernia

David Chalmers and Emily Serrell

Name of Procedure

Hydrocelectomy or herniorrhaphy.

Lay Description

A hydrocele is a fluid-filled sac that accumulates around the testicle. It may be an enclosed pocket (noncommunicating) or along a patent path through the inguinal canal into the abdomen (communicating). If contents from the abdomen are able to enter the tract, it is called an indirect inguinal hernia.

A hydrocelectomy involves a small scrotal incision to remove the fluid around the testicle. Stitches may be used to reinforce or close any communication between the abdomen and scrotum.

A hernia repair involves a small groin incision to locate and close the opening between the abdomen and scrotum. Sometimes this procedure is done laparoscopically with a small camera and thin instruments inserted through small abdominal incisions to view and close the opening between the abdomen and scrotum.

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Intended Benefit

Simple hydroceles are a benign condition that are rarely symptomatic and do not negatively affect the testicle or future fertility. The majority resolve spontaneously by 2 years of age. Persistent simple or communicating hydroceles may be repaired in order to reduce parental anxiety, decrease the size of the hemi-scrotum, and avoid enlargement or progression to an inguinal hernia. The latter occurs in <5 % of hydroceles [1].

Communicating hydroceles share a common etiology with the indirect inguinal hernia due to a persistent patency of the processus vaginalis, which typically closes after the testis passes through. Inguinal hernias should be repaired in order to avoid complications of incarceration (non-reducible) or strangulation (vascular compromise) of bowel and intraabdominal contents. Incarceration occurs in a minority of hernias, most commonly in infants [1], but the incidence increases with duration of time [2]. In very rare cases, incarceration can lead to ischemic bowel necessitating resection or in compression of spermatic cord contents resulting in gonadal atrophy or infarction [3].

Technique

Hydrocelectomy is typically performed under general anesthesia, although spinal anesthesia may be considered in special circumstances. The patient is placed in the supine position. For a simple hydrocele, a small scrotal skin-incision is used to access the hydrocele sac and testicle. The sac is incised, drained and then may be either plicated with sutures or partially excised and reapproximated. The former approach has the benefit of less dissection, intra-operative bleeding, and need for electro-cautery hemostasis, though the complication rates are similar [4]. The testis is replaced into the subdartos tissue, and the scrotal skin is closed with absorbable sutures.

The communicating hydrocele and indirect inguinal hernia may be repaired either as an open or laparoscopic approach. Communicating hydroceles may be repaired via a scrotal approach, and the patent processus vaginalis is ligated as cranially as possible towards the internal ring. This is particularly appropriate for children >12 years of age [5]. In an open inguinal hernia repair, an inguinal crease incision is made to mobilize and then separate the hernia sac from the surrounding cord structures. The sac is then ligated high in the inguinal canal. Laparoscopic repair can be accomplished with the patient in supine or slight Trendelenburg position. Recent studies have examined the efficacy of one, two, or three-port laparoscopy. The typical transperitoneal approach involves placement of camera and trocars beyond the peritoneum, and the internal ring is closed with either absorbable or non-absorbable suture, traditionally in a purse string fashion. Percutaneous approaches have also been described in an attempt to minimize additional trocar incisions. This involves placement of an endoscope at the umbilicus to visualize a suture passing extracorporeally, around the internal ring, and then passed extracorporally so the ring is closed. The, peritoneum then fascia are reapproximated with running, absorbable sutures, by some authors. Skin is closed in a subcuticular fashion and then either sealed with tissue adhesive, skin strips, or gauze and transparent film.

Postoperative Expected Course

Hydrocelectomy and hernia repair surgeries are minimally invasive and relatively low risk. The majority of procedures are performed on either an outpatient or day-surgery basis with patients discharged the same day as surgery. Overnight stay is rare and indicated only for complications or inguinal hernia repair in premature or very young infants. Parents should be educated on post-operative expectations for recovery. For example, some discomfort, redness, crusting, bruising, and swelling is normal, but worsening of these features or fever >101.3 °F requires follow-up with the surgeon or other healthcare provider. Scrotal swelling may persist for several weeks.

The use of perioperative antibiotics is controversial. The trans-scrotal, laparoscopic and open inguinal approaches are all classified as class II, clean-contaminated surgeries with <10 % risk of infection due to the risk of exposure from the genitourinary or gastrointestinal systems [6]. In addition, post-operative pediatric wounds may be exposed to colonic flora in the environment of the diaper. As such, perioperative antibiotic prophylaxis to prevent infection with *S. aureus* and enteric gram-negative bacilli contamination is optional.

Full recovery to normal activity is expected within a few days to weeks, and children will self-regulate their activity according to pain and energy. Typical recommendations include limiting rigorous activity and straddle activities for 2 weeks. Pain management is initiated before surgery is complete with injection of local anesthetic at the surgical incisions. Most children's pain will be managed well by alternating Acetaminophen (Tylenol) and Ibuprofen (Motrin) for the first 24–48 h. Narcotic medication such as liquid codeine or oxycodone may be appropriate for older children.

Diet following surgery should be normal as tolerated. Post-anesthetic nausea and vomiting occurs in 6 % of pediatric patients and should be controlled with serotonin receptor blockers such as ondansetron [7]. Sutures are absorbable and gauze bandage may be used over the incision sites for inguinal hernia repair or laparoscopic incisions. Children should avoid full water submersion for at least 48 h but then have no restriction on bathing or showering.

Follow Up

Follow-up timing, and even necessity, should be per surgeon's discretion. If there are signs of infection as described above within 2 weeks of surgery, the patient should be seen in the office as soon as possible. If it will provide reassurance to the family, follow up in 2–4 weeks. Otherwise, office staff may call for updates on patient recovery and well-healing wounds.

Risks and Complications

The complications following hydrocelectomy or herniorrhaphy can be classified as early—iatrogenic injury, scrotal edema, hematoma, wound infections, sensory impairment—or as late—hydrocele or hernia recurrence, testicular atrophy, chronic pain, and testicular ascent. It should be noted that the rates of these complications may be underreported, as late complications may present after the patient has aged out of follow-up of hernia or hydrocele surgery and go unrecognized by study authors.

Early

Infection: <1 %

Infection following hydrocelectomy or herniorrhaphy can be expected to be <1 % due to the classification of both surgeries as clean-contaminated [8]. Because of this, many surgeons opt to avoid perioperative antibiotics. In general, wound infection is higher in open than laporoscopic approach. Three meta-analyses reviewing 70 studies report infection rates of 0–2 % with the majority of cases reporting close to zero infections [9–11].

Bleeding and Hematoma: ≤1 %

Bleeding is a risk of any procedure that breaks the dermal barrier of the skin. Following hydrodcelectomy or herniorrhaphy bleeding may occur due to inadequate hemostasis of the superficial fascia, damage to the pampiniform plexus, or from the edges of the excised distal hernia sac. In the later scenario, blood can travel via the processus vaginalis and result in scrotal hematoma. The incidence of reported hematoma as a complication is <1 %, and these resolved spontaneously over the course of several weeks. A study in the Netherlands reported 0.9–1.6 % incidence of hematoma or seroma as well as 0.1–0.5 % incidence of bleeding necessitating reoperation [12]. There appears to be little effect of the surgical approach on subsequent hemorrhage or hematoma.

Scrotal Edema: 5 %

Scrotal edema and induration, as defined by postoperative swelling without indication of recurrent hydrocele or hernia, is a common complication of surgery. Up to 5 % of patients may experience significant scrotal swelling, particularly following scrotal hernia repair. However, in all cases this subsided spontaneously within 2 weeks [13].

Iatrogenic Injury: ≤1 %

The highest incidence of iatrogenic injury is damage to the seminal pathway structures during hydrocele or inguinal hernia repair. In addition, the hernia sac may contain abdominal viscera, intestine, bladder, ovary, or uterus. Gentle mobilization will aid in protecting sac contents, though unintentional cystectomy has been reported as a complication of routine herniorrhaphy [14].

Damage to the testicle is possible in the transcrotal hydrocele approach. Injury of the vas deferens or epididymis can occur with excessive electrocautery use or excision of the hernia sac or hydrocele, particularly at the tail of the epididymis [15]. The inguinal sac may contain embryonal cells of the spermatic cord contents as well as outright portions of the vas deferens and epididymis. In samples analyzed by pathology, true vas deferens and epididymis were identified in 0.33 % [16]. A similar pathologic analysis identified either vas or epididymis remnants in 0.53 % of hernia sac samples [17]. Damage to either of these structures along the spermatic pathway may affect future fertility either through direct obstruction or by damage-induced sperm-agglutinating antibodies [15, 18].

Sensory Change: 2–5 %

The genitofemoral nerve passes through the internal ring and travels with the spermatic cord, and the ilioinguinal nerve passes lateral to the internal ring. These nerves are at particular risk during laparoscopic and open hernia repairs, respectively. Trauma can occur during inguinal ring dissection, via heat-transfer from electro-cautery, or due to compression due to postoperative swelling or scarring and produce sensory neuropathy. Traditional pelvic nerve distribution attributes sensation from the genitofemoral nerve over the femoral triangle, ilioinguinal nerve over the groin, and anterior femoral cutaneous nerve over the anterior thigh. However, this is highly variable. Absence of the cremasteric reflex may be another indicator of sensorineural damage, particularly the genitofemoral nerve. Following laparoscopic surgery, 2–5 % of children may report numbness in the thigh or groin that presents zero to 10 days following surgery and resolves in 92 % of cases by 8 months [19]. Chronic pain following hydrocele or hernia repair is infrequently reported but may occur >3 months in 2 % of patients [12]. A small 50-year follow-up survey of adult patients who had undergone childhood inguinal hernia repair found that 3 % of adults reported chronic groin pain [20].

Late

Reactive Hydrocele: ≤1 %

Postoperative hydrocele should be differentiated from scrotal edema, though the entities may be confused. Following hydrocele or hernia repair, the distal sac will continue to produce fluid. In the case of very large hernias, this fluid may be produced faster than it may be resorbed and accumulate as a postoperative hydrocele. In Ein's series of 6,361 patients, only 2 persisted [21]. Incidence is increased in infants <3 kg, with one small study reporting ipsilateral hydrocele in 11 % of herniotomies [22].

Testicular Atrophy: ≤0.5 %

Testicular atrophy (necrosis) is an exceedingly rare but potentially devastating outcome of inguinal surgery. Laparoscopic repair of inguinal hernia in and of itself does not impair gonadal perfusion or size [23]. However, ligation of the testicular artery results in testicular atrophy in 20–40 % of cases. The 60–80 % of persistent testes have adequate collateral blood flow from the deferential and cremasteric arteries [24]. The majority of reviewed studies report zero incidence of testicular atrophy in simple hydrocele or hernia repairs [3, 25]. In two case series of 6,361 and 1,565 patients, there were reported rates of 0.3 % and 0 % atrophic testes respectively [21, 26]. The risk of testicular atrophy increases in premature infants [22] and in children with incarcerated inguinal hernias [27]. This is likely due to compression of gonadal vessels by intra-abdominal contents in the inguinal canal.

Testicular Ascent/Iatrogenic Cryporchidism: ≤1 %

Testicular ascent, or iatrogenic undescended testis, is a rare complication. It can result from either mechanical tethering of the testis or post-operate scarring that prevents the spermatic cord's growth with the rest of the body. Studies vary in incidence, with most series reporting no cases; indeed, two meta-analyses of 29 studies did not mention this as a complication [10, 11]. An assessment of 11,272 inguinal hernia repairs in China reported five cases of tethered high testis [28]. The incidence of iatrogenic cryptorchidism does increase to up to in premature infants [29]. This risk may be decreased by confirming proper testis positioning in the scrotum and, if necessary, anchoring the testis in the scrotum prior to closure [30].

Recurrence: <5 %

Recurrence of hydrocele or hernia is the most common long-term consequence of surgery. Recurrence rates are approximately 0.5–1 % in uncomplicated open repairs, 2 % in premature infants, and 3–6 % following repair of an incarcerated hernia [24]. Esposito et al. performed a 20 case meta-analysis reflecting a 0–5.5% rate of recurrence, with most studies trending toward the lower end of that spectrum [9]. This rate reflects a number of other retrospective and prospective studies [21, 31, 32]. The long-term recurrence risk into adulthood is not well-described, although one small study reports a rate of up to 8.4 % repeat groin surgery [20].

The likelihood of recurrence depends on a variety of surgical and patient factors. A meta-analysis of ten studies comparing open and laparoscopic inguinal repair demonstrated recurrence of hernia in 2–4 % of laparoscopic cases and 0–2 % of open cases [33]. A similar meta-analysis of seven studies reflected no statistically significant difference in the rate of recurrence based on surgical approach [11].

When recurrences did occur, they were more likely right-sided (75 %), direct, and discovered at 6 months (50 %) to 5 years (96 %) of age [31]. Recurrent hernias were more likely to occur with patient presenting with incarceration, prematurity, postoperative complications [34] as well as comorbidities like growth failure/malnutrition, increased intraabdominal pressure, genitourinary tract abnormalities. Surgical associations with recurrence included low ligation of the hernia sac, damage to the floor, peritoneal inclusion with spermatic cord, missed direct hernia, and less surgical experience [31, 34, 35].

Though few well-powered studies have been published regarding trans-scrotal approach for hydrocelectomy or herniotomy, the reported recurrence rate is low (0–2 %) [13, 32, 36, 37]. It should also be noted that rare cases of late abscess following primary repair, most likely to occur with non-absorbable silk suture, may be mistaken for recurrence and should be considered on the differential [38].

Contralateral Hernia: 7 %

The presence of metachronous contralateral hernias following unilateral hernia repairs is well-recognized, with an estimated incidence of 7 % by meta-analysis of over 15,000 patients with a variety of surgical approaches [39]. It is unclear whether the contralateral hernias were not identified at time of initial surgery, if surgery affected development, or whether the hernias developed independent of surgery. However, even following negative laparoscopic evaluation during surgery, metachronous inguinal hernias presented in 1.3 % of patients. These new hernias were more likely to be right-sided and associated with laparoscopic inguinal approach vs. periumbilical, small angle laparoscope, and high pneumoperitoneum pressure [40, 41]. As such, parents should be educated that the natural history of the inguinal ring is unpredictable and unilateral inguinal hernia repair will not prevent contralateral hernia.

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Chapter 14
Orchidopexy and Orchidectomy

Kim A.R. Hutton

Orchidopexy

Name of Procedure

Open orchidopexy.

Lay Description

An open operation that relocates a testis, that is sited in an abnormal position, into the scrotum. Usually the undescended testis is positioned in the groin or occasionally within the tummy.

Intended Benefit

To provide improved cosmesis so the patient has two normal appearing testes within the base of the scrotum, to maximize future fertility, to allow self-examination in adult life which is important because of a small increased risk of malignancy in undescended testes, and to reduce the risk of torsion or trauma which is increased in testes not within the scrotum. The undescended testis may be associated with an inguinal hernia sac and this can be transfixed and divided at the same time as the orchidopexy.

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What Happens Before the Operation?

All patients are managed as a day-case. No preoperative blood tests are required. Patients with a palpable testis are operated with open orchidopexy and where the testis is impalpable the child is scheduled for an examination under anesthesia, open orchidopexy if the testis can be felt, and laparoscopy if still impalpable. Preoperative ultrasound or magnetic resonance imaging is not routinely required. Starvation requirements for general anesthesia are 4 h for breast milk and 6 h for formula milk and solids. Clear fluids are allowed until 2 h before the procedure.

Technique

An open orchidopexy is performed under general anesthesia within an operating room environment under usual strict aseptic conditions. The procedure takes 30–75 min depending on complexity. According to surgeon preference and the position of the testis there may be two incisions – one in a horizontal skin crease in the groin around 2–3 cm in length and another in the scrotum about 1–1.5 cm long, or just one 1.5–2 cm incision sited at the junction of the scrotal and groin skin – a scrotal orchidopexy.

For an intraabdominal testis an open procedure may be performed with or without division of the testicular vessels (a Fowler-Stephens procedure) in one or two stages (operations 6 months apart) or a microvascular transfer, although contemporary management of these high testes is usually via keyhole surgery (laparoscopic orchidopexy).

How Is an Orchidopexy Performed?

The surgeon deepens the incision, locates the testis and starts to mobilize it. All tethering tissues are carefully separated from the testis whilst preserving the important structures going to and coming from the organ – the blood supply including an artery and several veins and the sperm tube (doctors call this the vas deferens). Once enough length has been obtained, so the testis can sit in the scrotum without tension, it is routed into its new position and placed in a small pocket the surgeon creates just underneath the scrotal skin. The wound is closed with buried dissolvable sutures and some surgeons prefer tissue glue to close the skin. A wound dressing is optional depending on surgeon preference.

Postoperative Expected Course

The child can eat and drink as soon as recovered from the anesthetic. The anesthetist will often have performed a caudal nerve block in theatre to provide early postoperative pain relief. Otherwise the surgeon as an alternative performs an intraoperative nerve block and tissue infiltration with local anesthetic. Oral analgesics are prescribed, usually Paracetamol and Ibuprofen, and these are advised regularly in the first 48–72 h. No antibiotics are required before, during or after the procedure and as soon as the boy is mobile, pain-free, and has passed urine he is allowed home. Discharge is usual within 2–4 h of the procedure.

Follow Up

In infants there are no specific precautions apart from a need to keep the wound/dressing area clean and dry. Any wound dressing can be removed after 5–7 days and if Steristrips have been used these can be allowed to fall off on their own. Older patients are advised to refrain from sporting activities, swimming, riding a bike etc. for up to 6 weeks post procedure. A follow up clinic visit is normally arranged at 6–8 weeks post surgery to assess testis position and size.

Risks of Procedure

The success rates for open orchidopexy depend on the initial position of the undescended testis, with testes within the abdomen having higher complication rates of loss of the testis (atrophy), the testis remaining in an unsatisfactory position, or re-ascending into an abnormal location.

General

These are the risks of any anesthetic and of any operation including cardiorespiratory problems, anaphylaxis, pain, bleeding, hematoma, swelling of the scrotal area, wound separation and wound infection. Wound related complications occur in 2.5 % of patients – 2 % related to infection and 0.5 % dehiscence and bleeding [1].

Specific

These are the risks related to orchidopexy itself and the majority fall into two groups:

1. Intraoperative: Failure to achieve enough length on the cord structures to position the testis comfortable within the scrotum, difficulty with securing closure of a flimsy hernia sac, inadvertent damage to the vas deferens or testicular vessels (very rarely the blood vessels to the testis can snap during traction on the cord and dissection), unrecognized torsion of the testis pedicle during routing to the scro-

tum which may affect testis blood supply, and damage to one of the nerves in the groin during dissection that can leave an area of long term numbness in the upper inner thigh (the nerve is called the ilio-inguinal nerve). Intraoperative complications and problems are reported in 1.5 % of orchidopexy operations. Most are related to the vas deferens (1.14 % of cases) with a finding of no vas, the vas not connected to the testis, or the vas inadvertently cut (0.07 %). The need for orchidectomy is 0.36 % [1].

2. Long term: These are related to persistent testis malposition or re-ascent, a smaller than normal testis or a testis that shrinks completely (atrophy) where the blood supply has been severely compromised, possible reduced fertility and a small increased risk of testicular cancer. If the testis re-ascends and is still of adequate size a re-operation is appropriate. A small testis may require excision depending on its location. A testicular prosthesis can be inserted later in childhood if a testis is lost due to atrophy or excision. Success for single stage orchidopexy can be as high as 99 % [2], although reported atrophy rates of 5 % in straightforward orchidopexy and 9 % for high testes highlights the difficulties with some of these surgeries [1]. Although semen parameters may be abnormal in men with unilateral undescended testis corrected in childhood, paternity is close to normal at 90 %, compared to men without a history of undescended testis (94 %). In men with previous bilateral undescended testis paternity rates are reduced to 35–53 % [3]. Boys with isolated undescended testis are three times more likely to develop testicular cancer later in life than those without testis maldescent [4].

Orchidectomy

Name of procedure

Open orchidectomy.

Lay Description

An open operation performed to remove a testis.

Intended Benefit

Removing a small testicular remnant found during surgery for an impalpable undescended testis or at redo orchidopexy is performed because of a small increased risk of testis cancer. Removing a non-viable testis at exploration for acute torsion prevents complications of retained necrotic tissue – pain, wound discharge and breakdown.

What Happens Before the Operation?

Surgery for dysplastic/atrophic testes is carried out as a day-case. No preoperative labs are required, standard starvation guidelines are followed and no antibiotics prophylaxis is necessary. In cases of testis torsion emergency surgery is performed without delay. In all cases the surgeon marks the side preoperatively. In patients with acute torsion the surgeon obtains consent for contralateral testis fixation. In other pathologies the need to fix a solitary testis is unclear and a discussion between the parents, patient and surgeon, with the formulation of a definite management plan is advised [5].

Technique

An open orchidectomy is performed under general anesthesia. In cases where laparoscopy has been performed for impalpable testis and a small dysplastic gonad is found intraabdominally, the orchidectomy is completed laparoscopically. When the hypoplastic testis or nubbin is beyond the deep inguinal ring an inguinal skin crease or scrotal incision is used for excision [6]. Exploration for torsion is through a midline raphe or transverse scrotal incision. The vessels to the testis are ligated and divided separate to the sperm tube and the testis removed. The wound is closed in layers using absorbable sutures, with buried (subcuticular) skin sutures providing optimal cosmesis. No wound drains are required.

Postoperative Expected Course

The child may resume oral intake once recovered from anesthesia. A caudal nerve block performed in theatre prior to surgical incision is often used to provide pain relief in the first several hours postoperatively. Otherwise the surgeon as an alternative performs local tissue infiltration with Bupivacaine. Oral analgesics are prescribed, usually Paracetamol and Ibuprofen, and these are advised regularly in the first 48–72 h. As soon as the child is mobile, pain-free, and has passed urine he is allowed home. Discharge is usual within 2–4 h of the procedure, although cases requiring emergency surgery may stay a little longer.

Follow Up

Most children are back to normal within a couple of days and if of school age can return within a week. Wound dressings if used can be removed after 7 days. Avoidance of sporting activities and riding a bike is recommended for 6 weeks.

Outpatient review is performed 6–8 weeks post surgery to assess the results, discuss pathology reports and answer any ongoing concerns the family may have.

Risks of Procedure

General

There are risks related to the general anesthetic, which are increased in patients with preexisting cardiorespiratory disease. However, modern day anesthesia is extremely safe. Surgical risks include bleeding, infection and wound dehiscence. Wound related complications occur in 1.9 % of patients [7].

Specific

These risks fall into three groups:

1. Risk of torsion in remaining solitary testis: There is limited scientific evidence to guide the need for contralateral testis fixation in cases undergoing orchidectomy, except in patients with testis torsion [5]. When performed in non-torsion cases a sutureless dartos pouch technique is preferred.
2. Cosmetic concerns: Some boys have body image issues following orchidectomy and request surgery for a testicular prosthesis. Implants are saline-filled with a silicone shell or a silicone elastomer envelope filled with a highly resilient silicone gel [8]. Whilst important cosmetic and psychological benefits can be gained from surgery not all patients are satisfied with the results – 23 % claiming the prosthesis is too small, 38 % feeling it is too heavy and 38 % stating it's position is too high in the scrotum [8, 9].
3. Future fertility: Paternity in adult life is close to normal for boys undergoing orchidectomy for a dysplastic testis and normal contralateral testis [3]. Testicular torsion in teenagers and young adults seriously affects spermiogenesis in about half the patients and on long term follow up only 5–50 % have normal semen analysis [10]. Although robust data are lacking prepubertal boys may fair better with regard to future paternity [11].

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Chapter 15
Laparoscopic Orchidopexy

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Name of Procedure

Laparoscopic orchidopexy.

Lay Description of Procedure

A minimally invasive approach to identify and relocate intraabdominal testes into the scrotal position

Intended Benefit

Quick diagnostic tool to determine location and presence of non-palpable testes. If an intraabdominal testicle is found, then laparoscopic orchidopexy can be performed to move testis to the scrotum (Fig. 15.1). The goals of procedure include improving fertility, decreasing potential for malignancy, and permitting easier examination of the scrotal testis. Laparoscopic technique provides a minimally invasive approach that is well tolerated and has reduced morbidity as compared to the open approach.

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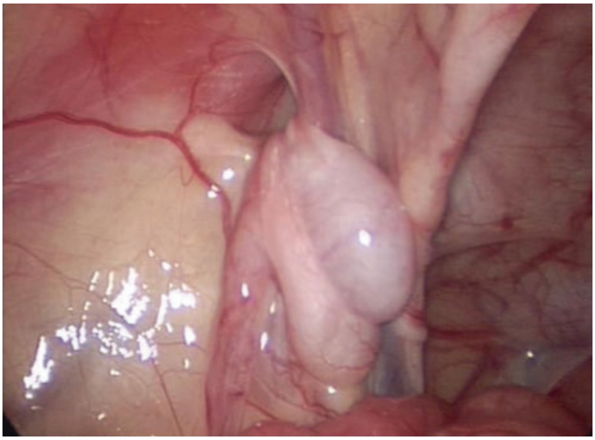

Fig. 15.1 Depicts left intraabdominal testes located at the level of the internal ring. The vas deferens can be seen behind the testicle coursing medially

Technique

Laparoscopic orchidopexy can be performed either as vessel-sparing (primary) or non-vessel sparing approach (Fowler-Stephens). Fowler-Stephens orchidopexy can be single stage or two-stages. The former requires clipping of the spermatic cord vessels and enough mobilization of the testicle and vas deferens to allow sufficient relocation into the scrotal position. If testicle is too high or maximal mobilization cannot be achieved then a two stage repair can be performed.

General anesthesia with paralysis is recommended. Preoperative intravenous antibiotics are indicated. Patient is usually positioned supine, adequately padded, strapped/taped and placed in Trendelenburg position.

The patient will have three port site incisions on abdomen, and a scrotal incision. Fascia underlying abdominal incisions are usually closed if 5 mm ports were used. The skin is usually closed with monofilament absorbable suture. If testicle is on some tension or cannot be brought to the most dependent part of the scrotum, then an external button device is utilized and removed usually after 7 days in clinic.

If performing a primary orchidopexy, the spermatic vessels are mobilized first by incising the peritoneum laterally. The gubernaculum is then incised thus freeing the caudal attachments. The testicle, vessels and vas are gently dissected away from the external iliac vessels and peritoneal attachments to provide length. Peritoneum overlying the vessels is sometimes incised to provide more length. The peritoneal triangle between the vas deferens and vessels are generally preserved.

The neohiatus to scrotum is created and can be done antegrade (intraabdominal to scrotum) or retrograde (from scrotum to abdomen). Understanding the anatomy is key in prevention of injury during neohiatus creation (Fig. 15.2). Whichever approach, the neohiatus should be medial to medial umbilical ligaments and lateral to bladder. Once the neohiatus is created the testicle is brought down to scrotum and fixated in the scrotum by creating a dartos pouch and/or applying a fixation stitch.

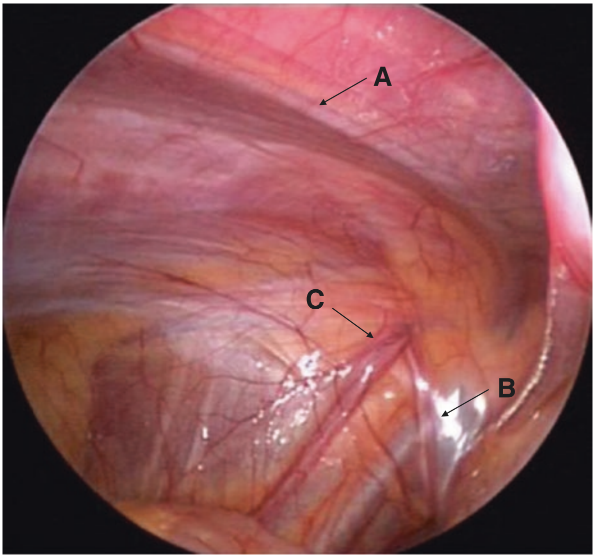

Fig. 15.2 Important landmarks during laparoscopic orchidopexy. (*A*) Epigastric vessels, (*B*) Vas deferens entering internal ring, (*C*) Testicular vessels

Post-operative Course

Typically, this is an outpatient procedure and patients go home the same day. Patients can eat/drink as soon as they have recovered from anesthesia. Narcotics can be used postoperatively if patients are older (typically >12 months, or 10 kg), but should be used sparingly. Acetaminophen or NSAIDs should suffice for postoperative pain control. Typically, local anesthetic is injected into the port sites at the end of case.

Postoperative wound care is minimal. Restrictions of activity are identical to open procedure and are surgeon specific.

Follow-Up

If staged procedure, then patient will return to operating room in 6 months for second stage. If single stage procedure, follow up is usually in 4–6 weeks with just a scrotal exam to evaluate size and position of testicle.

Timing of Surgery

Spontaneous descent of cryptorchid testes may occur within the first 4–6 months of life [1]. The generally accepted age to perform orchidopexy is between 6 and 18 months. At 6 months age, general anesthesia is consider safe.

Outcome

With improved laparoscopic techniques, laparoscopic orchidopexy has been shown to have high success rates as compared to an open approach for management of non-palpable testes (intraabdominal testes). Several single institutional and multi-institutional studies have been performed to evaluate the outcomes of laparoscopic orchidopexy. A successful outcome was defined as intrascrotal position of testis after surgery without evidence of atrophy on clinical exam. A multi-institutional study performed across ten centers (Baker et al.) revealed success rate of 97.2 % for primary orchidopexy. One and two-stage Fowler-Stephens orchidopexy were successful in 74.1 % and 87.9 % of cases, respectively [2].

Chang et al. reported their single institutional outcomes of laparoscopic orchidopexy with an overall success rate of 91 % at 6 months follow-up, with an overall atrophy rate of 4 %. For primary laparoscopic, one-stage Fowler-Stephens, two-stage Fowler-Stephens, the success rates were 94 %, 84 % and 86 %, respectively [3]. Samadi et al. published their series with a 95 % overall success rate, with a 97 % success rate in primary laparoscopic approach versus 90 % success rate in Fowler-Stephens technique [4]. Radmayr et al. also reported a high overall success rate at 97 %, 100 % with primary orchidopexy and 93 % with Fowler-Stephens [5].

Not many studies have been done directly comparing open versus laparoscopic approach. A single prospective, randomized study performed by Abolyosr et al. comparing open with laparoscopic approach revealed 100 % success rate for primary orchidopexy using both technique. Open Fowler-Stephens approach had an 85 % success rate as compared to 90.5 % success rate with laparoscopic Fowler-Stephens approach. The laparoscopic technique resulted in decreased morbidity with regards to resuming diet, length of hospital stay and resumption to normal activities [6].

Risks of Procedure

Rates of complications from laparoscopic orchidopexy are quite similar to that of an open approach; the difference being that in laparoscopy, surgeon experience is a strong predictor of complications [7]. Complications that have been reported in literature include testicular atrophy, vascular injury, bladder injury, bowel injury, ileus, transection of vas deferens, wound dehiscence/infection, and testicular vessel avulsion leading to one-stage Fowler-Stephens orchidopexy. In a large multi-institutional review, Baker et al. reported a major complication rate of 3.0 % and minor complication rate of 2.0 % [2]. Peters et al. performed a survey of pediatric urologists in which a complication rate of 5.4 % in 5,400 cases was reported [7].

The remainder of this section will highlight several complications seen intraoperatively and post-operatively.

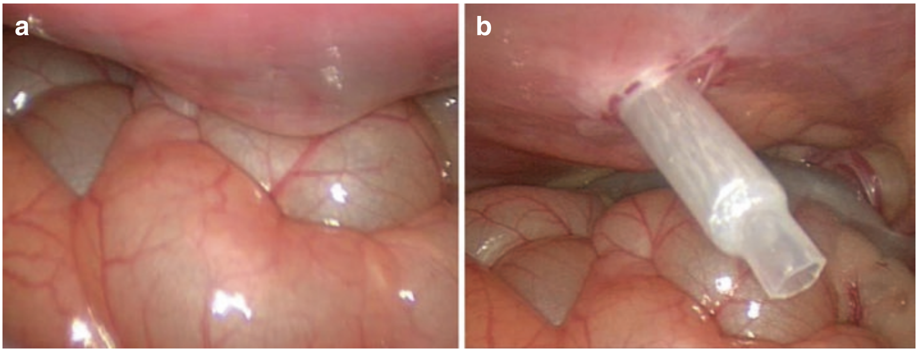

Fig. 15.3 (**a**) Abdominal wall is compliant, as you can see trocar is tenting anterior abdominal wall. (**b**) Epigastric vessels and bowel are in close proximity to port site entrance

At the start of the case, proper positioning and padding are important to reduce neuromuscular injuries. Patients should be adequately padded and secured with either tape or straps to prevent any shifting or positional related injuries when table is moved to Trendelenburg position.

Access related complications are common and often related to technique of entry into the abdomen. Two standard laparoscopic access techniques are open Hasson and Veress needle. Open peritoneal access has been associated with slightly fewer complication than with Veress needle access [8]. The pediatric abdomen is very compliant with limited space, and epigastric vessels, iliac vessels and bowel come in close proximity to the access devices (Fig. 15.3), thus open technique allows for direct visualization of intraabdominal compartments. In their series, Passerotti et al. report a 0.4 % epigastric and iliac vessel injury rate during access. Small bowel injury during access is around 0.1 % [8].

Bowel injury is a concern during laparoscopy, specifically during trocar placement, instrument passage and use of cautery. Serosal tears or isolated small enterotomies can be repaired laparoscopically by an experienced surgeon. More extensive injuries may require laparotomy and possible intraoperative consultation with pediatric surgery [3]. Omental or bowel herniation through a port site is a rare complication but has been reported, with rates approximately 0.15 % in pediatric literature [7, 9]. As a result most surgeons typically close 5 mm port sites. On the contrary, most 3 mm port sites are not closed, but there has been a reported omental herniation [10].

Injury to iliac vessels or femoral vessels can be potentially catastrophic. Iliac vessels can be lacerated during testicular mobilization and there is concern for femoral vessel injury during passage of instruments through neohiatus. Prompt recognition, vascular control and conversion to open, if necessary, are keys to management [11].

Bladder injury typically occurs at time of creation of neohiatus with instrument passage to/from scrotum. There is little published data regarding bladder injury rates, of that what is published the rates are small, between 0.03 and 0.17 % [7, 12].

In their series, Hsieh et al. report that 3 out of 93 patients undergoing laparoscopic orchidopexy had bladder injuries, and all 3 patients were bilateral and 2 of the 3 had previous inguinal surgery creating scar tissue [12]. Steps to prevent injury would include placing a urethral catheter during the case to empty bladder and dissection lateral to bladder to identify its edge. Surgeon must be familiar with the pelvic landmarks when creating neohiatus to prevent bladder or vessel injury.

Inadvertent avulsion of testicular vessels is another intraoperative complication that is strictly due to technique and tissue handling. If this happens, conversion to single stage Fowler-Stephens is necessary. Although orchidopexy can still be accomplished, there is unfortunately a higher chance of testicular atrophy [13, 14]. Transection of looping vas deferens through the internal ring is a potential complication. A surgeon must always be cognizant of the possibility that a looping vas may exist. Often the looping vas can be pulled into the abdomen laparoscopically, but if this unsuccessful and small inguinal incision to help deliver vas through internal ring can be made with high success [15, 16]. Aggressive mobilization and dissection of the vas deferens can lead to vassal injury, obstruction or testicular atrophy.

Testicular atrophy is the most common long-term complication associated with laparoscopic orchidopexy, and a marker of failure of the procedure. Single stage Fowler-Stephens have highest rates of atrophy, ranging from 3 to 25 %. Vessel sparing single stage orchidopexy have the lowest rate of atrophy with ranges from 0 to 5 %. Lastly, two stage Fowler-Stephens results in a 0–15 % testicular atrophy rate [2, 3, 5, 13, 17–20]. Although the ranges of atrophy varies, the reported rates are not inconsequential and surgeons should be cognizant of this complication when discussing the procedure with patients and families. Intraoperatively, surgeons must be careful handling testicular and cord structures and limiting use of cautery close to the vessels. Preservation of vassal blood supply is important especially is spermatic vessels are taken during a Fowler-Stephens approach. Deferential artery is known to provide collateral blood flow to testicle.

Recurrent cryptorchidism is yet another post-operative complication one must consider. Reported rates range from 0 to 19 % in the literature and is usually due to insufficient mobilization of testis or suboptimal fixation of testicle in scrotum [2, 21, 22]. This may require a second procedure, usually open, to relocate testicle into the most dependent portion of the scrotum.

Conclusion

Laparoscopic orchidopexy is a safe, effective approach for non-palpable intraabdominal testes that has high success rates and decreased morbidity. As with any type of surgery there are certain inherent risks to the procedure, but the overall complication rate is <5 % and minimized with increased surgeon experience. Overall indications, success/failure rates, and complication rates have been published in the literature and should be discussed with patients and their families prior to surgery.

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Chapter 16
Varicocelectomy

Brian Caldwell

Lay Description

Varicoceles, described simply, are varicose veins of the testicle. These dependent veins become dilated because blood is unable to return to the heart against the force of gravity. This occurs because of a combination of an obstruction to flow and incompetent valves within the veins. The majority of varicoceles are on the left side, since the blood flow returns through the left renal vein rather than directly into the vena cava like the right.

The testicle remains outside the body in the scrotum in order to maintain lower temperature that is ideal for testicular function and production of sperm. Varicoceles keep blood that is body temperature surrounding the testicle, thus increasing the resting temperature. This increase is thought to interfere with proper growth and functioning of the testicle and larger varicoceles even raise the temperature of both testes. This alteration in temperature is postulated to contribute to infertility.

Varicoceles are classified by their presentation on physical exam. The classic description is that of a "bag of worms" on scrotal examination. If there is no palpable varicocele or can only be detected on color doppler ultrasound, it is considered a grade 0. Those only palpable while bearing down or valsalva are grade 1, while grade 2 varicoceles are palpable at rest while standing. Grade 3 is the most severe and is visible to the naked eye while standing.

Intervention for varicoceles centers around ligation or division of the dilated spermatic veins, thus forcing blood return via the non-dilated, more competent veins that surround the vas deferens.

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Intended Benefit

Controversy continues to surround the decision for intervention on varicoceles. Approximately 15 % of all men have a clinically noticeable varicocele, and puberty appears to play a role. This fits with the 6 and 16 % rates of varicocele noted in prepubertal and adolescent patients, respectively. Since varicoceles are not thought to resolve spontaneously, the ones that present in childhood are likely the same that are seen in adulthood. The rate of varicocele is up to 40 % in men who present to infertility clinics, thus producing concern for varicocele contribution to infertility. Therefore, proponents of intervention extrapolate that early repair should have a positive effect on fertility. On the other hand, 85 % of men with varicoceles have fathered children, a percentage not significantly different than the paternity rate of "normal" men.

Reasons for intervention in the adolescent population often focus on testicular asymmetry with testicular growth arrest on the side with a varicocele. The smaller size (2 ml or 20 % size disparity) is often used as a surrogate for impaired testicular function, with preservation of testicular tissue proposed as an indication to intervene. In support of this concept, catch-up growth has been seen in some studies of patients who undergo intervention. However observational studies have revealed similar growth with conservative management. Semen analysis data also shows some improvement in total motile sperm with varicocele repair, but longer-term studies on paternity are lacking.

Techniques

Numerous approaches are available for treatment of varicoceles: inguinal or subinguinal, retroperitoneal or laparoscopic approaches, and percutaneous sclerotherapy or embolotherapy. The approaches are normally evaluated based on rates of success and the most common complication, hydrocele. These rates have wide variance depending on the study, partly due to a lack of long-term outcomes in the published literature. Modifications of the original approaches such as artery and/or lymphatic sparing procedures, make the decision process even more convoluted.

For adolescents, the techniques have been adapted from and are the same as their adult counterparts. One exception would be that almost all children undergoing a varicocelectomy will require general anesthesia, while adults undergoing subinguinal surgery or percutaneous sclerotherapy often do so with only local anesthetic. Regardless, all procedures in the adolescent population are outpatient or day procedures without the need for overnight hospitalization. Patients are positioned supine for each procedure.

Inguinal/Subinguinal

In inguinal and subinguinal procedures, a transverse incision is made in an inguinal skin fold above the level of the external inguinal ring and over the external inguinal ring, respectively. The subcutaneous tissues are divided as is the external oblique fascia for the inguinal approach, however a subinguinal approach will encounter the spermatic cord without this step. The spermatic vein at this point has multiple branches that must be ligated, however it allows for the ligation of accessory external spermatic veins that may contribute to recurrence or persistence of the varicocele in some cases. Each vein branch is identified and ligated individually, and outcomes have significantly improved with institution of microscopic magnification over no or loupe magnification [1, 2]. While the procedure yields excellent results in terms of success and complications, it is often lengthy, requires microscopic skills and in adolescents, general anesthesia.

Retroperitoneal/Laparoscopic

The retroperitoneal approach was first described by Palomo and encounters the spermatic vein at a point where it has not significantly branched. An incision is made superior to the external inguinal ring and medial to the anterior superior iliac spine. The incision is carried down into the retroperitoneal space without entering the peritoneum. The spermatic vessels are identified, then both artery and vein are ligated en bloc. This approach is expedient and separated from the vas deferens, so there is little concern for injury to the testicle's alternative blood supply.

For laparoscopy, the abdomen is accessed via either open or veress needle approach at the umbilicus, where a camera is introduced. Two working ports are placed in a standard triangulation pattern which varies by surgeon preference and whether unilateral or bilateral. The patient is arranged in Trendelenberg position with head down, allowing the abdominal contents to shift away from the internal inguinal rings. The posterior peritoneum is incised adjacent to the spermatic vessels approximately 2–3 cm cranial to the internal inguinal ring. This position assures protection of the vas deferens as well. The vessels are elevated and ligated with a choice of ties and cold division or use of a variety of energy devices as vessel sealants. The laparoscopic variant is adapted from the Palomo procedure and the vessels are generally ligated en bloc.

Artery/Lymphatic Sparing

Modifications to the procedures above are well described with most focusing on selective dissection and ligation of the spermatic vein. Sparing the spermatic artery theoretically preserves the blood supply to the testicle, however, testicular atrophy has rarely been described as individual cases and only with the subinguinal approach. This likely occurs due to injury of the vasal blood supply during dissection in patients where the testicular artery is ligated with the vein. Microscopic subinguinal dissection helps to combat this by accurate identification. With hydrocele being the most common complication of varicocelectomy, an effort for reduction utilizes lymphatic sparing procedures, described in both inguinal/subinguinal and laparoscopic approaches. With any procedure that spares the artery or lymphatic, the risk of missing a patent vein branch increases the chance of persistence or recurrence of the varicocele.

Percutaneous Sclerotherapy or Embolotherapy

Sclerotherapy or embolotherapy are minimally invasive alternatives to surgical varicocelectomy. These procedures are usually done by an interventional radiologist and can be completed under general or local anesthesia depending on the patient's comfort level. An antegrade approach is well described, with venous access at the level of the femoral vein or through small scrotal incision and direct access. The spermatic vein is selectively embolized with coils and/or foam of 3 % sodium tetradecyl sulfate or polidocanol. This too, is a day procedure and averages about 15 min in length with hospital time of 4 h in several studies. There are also some descriptions of retrograde injection and combination with surgical ligation.

Post-operative Expected Course

Individual procedures for varicocelectomy and sclerotherapy are almost uniformly day procedures with overnight stays rare and a result of complications not directly related to the varicocele itself. The procedures are generally minimally invasive, low risk and well tolerated. Incisions are closed with absorbable suture and are dressed by surgeon preference, often with skin sealant or gauze covering. Patients can resume their regular pre-procedure diet after clearance of general anesthesia. Activity will be autoregulated by the patient based on discomfort and most are back to normal activities within a few days. Depending on surgeon preference, a limitation is often placed on submersion of incisions in bath or pool for 48 h to 1 week. In the inguinal and scrotal approaches, some scrotal swelling can be expected that is self-resolving by a few weeks time. Otherwise, hydrocele formation is the most common complication of the procedures and can be seen in up to 30 % of patients depending on the specific procedure. Hydroceles are self-limiting in most cases, but in some series required intervention.

Follow-Up

Follow-up varies by provider, but it would be reasonable to expect a post-operative clinic visit within a few months of the procedure. Ongoing clinical physical exam with orchidometer or ultrasound to assess testicular size and catch-up growth has been advocated. With hydrocele presentation at up to 2 years in some studies, patients can be actively followed over that time or instructed to return if a hydrocele develops. Follow-up semen analysis at 18 years of age has been advocated by some, however others will only obtain semen analysis if fertility issues arise in the future.

Risks and Complications

Complications can be classified into categories of early and late. Early complications are generally associated with the intervention itself while late complications focus around success of the operation and longer-term issues. Varicocelectomy has an overall low complication rate with very low rate of major complications.

Early Complications

Bleeding

All surgical interventions where incision is made will have some risk of associated bleeding. The risks are not well studied in varicocelectomy but can be extrapolated from similar procedures such as inguinal hernia repair and general laparoscopic access. Quoted rates of bleeding risk from inguinal surgery and laparoscopic procedures are <1 %. Scrotal hematomas have been described in subinguinal varicocelectomy, but seem to be largely associated with procedures where the testicle is delivered into the incision with a rate of 0.6 % in a small series [3].

Percutaneous sclerotherapy and embolization holds unique risks due to vascular access. If femoral access is gained, an overall major complication rate of <1 % has been recorded [4]. Information on pediatric patients is limited to those that are critically ill and do not generalize well to the varicocele population. Antegrade scrotal sclerotherapy, which approaches the spermatic vein through a small scrotal incision, displays a scrotal hematoma rate of approximately 1–1.5 % [5, 6].

Infection

Very few studies on varicocelectomy report the incidence of wound infection, however, all approaches including open inguinal, subinguinal, retroperitoneal as well as laparoscopic and percutaneous interventions would be considered a clean procedure by National Nosocomial Infections Surveillance System (NNIS) wound classification, correlating with a 2–5 % rate of surgical site infection (SSI) [7].

General recommendations in this wound class would be to proceed to surgical intervention without antibiotic prophylaxis [8]. In support of this, a large pediatric surgery study on inguinal surgery exhibited an overall infection rate of 1.2 % [9]. Similarly, two studies of antegrade scrotal sclerotherapy revealed scrotal wound infection rates of approximately 2 % [5, 6]. In a review of all surgical cases of wound class 1 and 2 (clean and clean/contaminated) for pediatric urologists at a single institution, the rate of SSI was 0.34 % and 2.28 %, respectively. There was no difference in SSI in wound class 1 between patients who received antibiotic prophylaxis versus none [10].

Iatrogenic Injury

Iatrogenic injury in subinguinal or inguinal approaches are centered around damage to more than one of the arterial supplies to the testis (testicular, vasal or cremasteric) compromising vascular inflow and thus testicular necrosis and atrophy; or injury to the path of sperm efflux, almost exclusively by disruption of the vas deferens. These are very rare occurrences with data published only about testicular atrophy. In a large single center study of 522 varicocelectomies using all approaches, the rate of testicular atrophy was 0.2 % overall and 0.6 % of the open cases. No incidents of testicular atrophy were reported in the laparoscopic series [11].

While complications directly related to laparoscopic varicocelectomy such as injury to the vas deferens and testicular atrophy have not been reported, laparoscopy holds other risks mostly associated with abdominal access, especially first trocar placement. A survey assessment of pediatric urologists and their experience with laparoscopy revealed an overall rate of complications as 5.6 %, however, excluding self-resolving preperitoneal insufflation and cutaneous emphysema, the complication rate was a reasonable 1.18 % [12]. In a follow-up evaluation of 806 laparoscopic cases in a single institution, a 1.6 % rate of abdominal access injury was noted. This was higher in the Veress needle entry versus open entry to the abdomen. Types of injury were preperitoneal insufflation, vessel injury, small bowel injury, bleeding, and bladder perforation. Surgeons completing greater than 12 laparoscopic cases per year had a significantly lower rate of complication [13].

Late Complications

Hydrocele

Hydrocele after varicocelectomy is the most common complication overall and varies significantly with the surgical approach that is implemented.

– Inguinal/subinguinal – Open inguinal and subinguinal approaches have evolved over time and with that evolution has brought improvements in outcomes. In an adult study comparing high ligation and microsurgical high inguinal varicocelectomy, the hydrocele rate was 9.09 % and 0.69 %, respectively [14]. Others have shown similar rates of up to 2 % for hydrocele after microscopic varicocelectomy [15]. Efforts to decrease the rate of hydrocele led to lymphatic sparing procedures, with initial decreases in rate of hydrocele to 0 %, but this compromised success rate [16]. Overall improvement in both success rate and hydrocele rate (0 %) was noted with microscopic magnification over either loupe magnification (2.9 %) or no magnification (5.9 %); therefore microscopic subinguinal varicocelectomy with artery and lymphatic sparing has become a favored option in treatment, especially in adults [2].

- Retroperitoneal/laparoscopic – The retroperitoneal and laparoscopic techniques are shown on meta-analysis to have similar rates of success and hydrocele [17], likely because the spermatic vessels are ligated in mass, incorporating the lymphatics in the ligation. This leads to a higher rate of hydrocele than more selective procedures. A wide range of results are published with hydrocele rate ranging from 4.9 to 43 % [16, 18–20]. A recent multicenter database study evaluated the age at varicocelectomy and found a significantly higher rate with younger age. In fact, there was a 14 % decreased rate of hydrocele with each year older [21]. Modification of the technique similarly progressed toward lymphatic sparing because of the high rates of hydrocele and inherent magnification with laparoscopy. This change led to a significant reduction in hydrocele from 15.1 to 8.4 % in one publication [22] and from 43.3 to 4.5 % in another [20], with reports as low as 0 % [23]. Additionally, when lymphatic dye is injected in the scrotum therefore highlighting the lymphatic channels, the hydrocele rate has been shown to drop to 0 % [24].
- Percutaneous sclerotherapy or embolotherapy – Through several outcome studies, selective embolization with antegrade approach via groin or scrotal access have displayed no evidence of hydrocele following the procedures [5, 25]. In a multicenter database study, a hydrocele rate of 5 % was found in patients who underwent percutaneous embolotherapy [21].

Persistence/Recurrence

- Inguinal/subinguinal – In similar fashion to outcomes in hydrocele, evolution of the procedure significantly impacted the success rates of inguinal and subinguinal varicocelectomy. Recurrence rates fell from 15.5 to 2.11 % with the introduction of microscopic high ligation [14]. Initial attempts at arterial and lymphatic preservation resulted in at much higher rate of recurrence (14 %) [16]. In another study comparing magnification levels, using a microscope in an arterial and lymphatic sparing subinguinal varicocelectomy led to 0 % recurrences in comparison to 2.9 % using loupes and 8.8 % without magnification. A 10 year review at a single institution supports these excellent outcomes with the same success rates while using arterial and lymphatic sparing as without and a 0 % rate of hydrocele as well [19].

- Retroperitoneal/laparoscopic – Retroperitoneal and laparoscopic approaches are efficient procedures due to ligation of vessels in mass, without needing individual dissection of multiply branched veins. The most common procedure used by pediatric urologists is the laparoscopic approach. Because of the more cranial ligation, much less venous branching has occurred leading to a generally high success rate with laparoscopy. Recurrences range from 0 to 3.9 % across several long-term studies [18, 19, 26, 27]. Introduction of arterial and lymphatic sparing laparoscopic varicocelectomy showed an increased recurrence rate to 6 %, however, modification to only lymphatic sparing improved the rate to 0–1.3 % while maintaining low hydrocele rates [20, 23].
- Percutaneous sclerotherapy or embolotherapy – Regardless of approach, rates of persistent varicocele after sclerotherapy or embolotherapy range from 5.9 to 9 % [5, 25, 28, 29]. A multicenter database review revealed an even higher rate of retreatment at 9.9 % with percutaneous embolotherapy [21].

Testicular Atrophy

As noted previously, testicular atrophy is a theoretical risk when multiple testicular arterial supplies are compromised. This can occur during inguinal or subinguinal procedures where both the vasal and testicular arteries can readily receive injury. Similarly, there is concern for arterial compromise with prior inguinal surgery, especially hernia repair and orchiopexy. The rarity of testicular atrophy makes the rate of occurrence difficult to calculate, but one study of 522 varicocelectomies reported an overall rate of 0.2 %. None occurred in the laparoscopic cohort, therefore making the rate in the open surgery cohort 0.6 % [11]. Since this is the only study reporting rates of testicular atrophy, it is hard to imagine that the incidence is even that high.

Pain and Parasthesia

Orchalgia was recorded in one study of microsurgical subinguinal varicocelectomy with artery and lymphatic sparing at a rate of 1 % [15]. Due to the rarity of presentation in the literature the overall rate of orchalia is likely significantly less than 1 %.

Nerve injury can occur and is usually associated with damage to the genitofemoral nerve. This causes transient numbness of the ipsilateral anterior thigh that resolved within 8 months of the procedure in two studies. Nerve injuries in both studies occurred during laparoscopic intervention, with an overall rate of 4.9 % [30]. There was a distinct difference in the type of ligation leading to nerve injury: 0 % in those ligated with clip and sharp division versus those ligated with ultrasonic shears (17 %) [31]. There is a theoretical risk of injury to the ilioinguinal nerve with inguinal approach or genital branch of the genitofemoral nerve with either inguinal or subinguinal procedures due to the intimate association with the spermatic cord, but published data does not directly assess this risk.

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Chapter 17
Circumcision

Andrew Freedman and Richard S. Hurwitz

Name of Procedure: Circumcision

Lay Description

Circumcision is an operation to remove the foreskin of the penis. Circumcision is most commonly done in the newborn period for cultural preferences or religious reasons. It is also performed for medical indications after the neonatal period, usually related to non-retractability or infections of the foreskin.

Intended Benefits

Neonatal circumcision is a non-therapeutic procedure in that it is not treating any active disease. For many families there is a cultural or religious reason for wanting a circumcision. Many families however request circumcision due to a belief that there are health benefits. It is generally accepted in the American medical community that there are potential health benefits from circumcision, however the benefits are quite modest and are not sufficient to recommend routine neonatal circumcision. The main benefits are reported to (1) decreased urinary tract

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infections, (2) decreased risk of HIV transmission and other Sexually Transmitted Infections, and (3) decreased risk of penile cancer. For any individual these health risks are quite small. There are indeed risks as well associated with circumcision. The relative risk to benefit ratio is a matter of great debate. One should be willing to acknowledge that the primary benefit for most families is satisfying their desire based on their culture, religion, family identity and esthetic preference, not for medical benefit. Basically circumcision is a "tribal" custom however one may choose to define their "tribe."

Risks

Neonatal circumcision does entail inherent risks. The magnitude of risk, especially for non-acute complications has not been fully elucidated.

Acute Risks

In the setting of a trained medical provider in a hygienic environment and with a stable healthy newborn the most common acute risks include:

Bleeding- the risk of perioperative bleeding requiring a significant acute intervention is approximately 1/200. The risk of any bleeding beyond the expected, requiring any treatment has been reported to be as high as 3 %.

Infection- the risk of a significant post-operative wound infection is 1/200.

Injury of the glans/urethra- the incidence is unknown though is believed to be a very rare event. However this can be a devastating complication.

Removal of too much or too little skin- the true incidence is unknown and the subjective nature of this complication makes it difficult to estimate. However it is clear that there are numerous revision circumcisions preformed every year for this complication.

Non-acute Risks

Adhesions/skin bridge- post circumcision adhesions of the sub-coronal collar to the glans are so common in infants that it should be considered a phase that resolves spontaneously rather than a complication. Post circumcision adhesions of the circumcision incision to the glans that form a true scar (skin bridge) are a common complication though accurate incidence data is unknown.

Trapped penis- a trapped penis is caused by migration of the circumcision cicatrix distal to the glans causing a secondary phimosis. The true incidence of this complication is unknown.

Meatal stenosis- meatal stenosis has been associated with neonatal circumcision. The incidence has been reported as high as 7 % though the methodology of the studies was not definitive.

Techniques

How is the neonatal circumcision performed?

The minimum requirements to proceeding to circumcision include:

A stable healthy baby- as the procedure is elective and non-therapeutic it is critical that the risks be at a minimum. The baby should be healthy and stable. Premature babies in particular should be cleared by the pediatrician or neonatologist responsible for their care.

Normal penile anatomy- the penis and foreskin should be normal. A prior circumcision can compromise the later repair of a penile abnormality.

The newborn should have been documented to have voided.

There should be no family history of a bleeding disorder.

Contraindications- the most common contraindication to newborn circumcision is a penile anomaly. Common anomalies that should postpone circumcision include:

Hypospadias
Chordee
Buried penis
Penoscrotal fusion
Ambiguous genitalia

Procedure- Neonatal circumcision is typically preformed with the aid of a specially designed clamp or device. The most common techniques are the Gomco Clamp, Mogen Clamp or Plastibell ring. All efforts should be focused on trying to preform the safest procedure with the least amount of discomfort. Proper pain management technique should be routine employed.

- Following medical clearance and prior to the procedure the phallus can be dressed with a topical anesthetic ointment (EMLA or LMX4)
- The infant is gently restrained with securing the legs and swaddling the upper body
- The penis is cleansed with antibacterial prep
- Effective pain control measures should be instituted including

    Local anesthetic injection

        Penile block and/or ring block

    Sucrose nipple

- Begin with gentle separation of the adhesions- great care needs to be used at the 6 o'clock position due to the proximity of the frenular vessels

- The amount of foreskin to be removed is estimated
- Gomco clamp, Mogen clamp, Plastibell can be applied according to operator's preference
- In the case of the Gomco and Plastibell a dorsal slit incision is made and the prepuce fully retracted and all adhesions taken down
- The devices provide a crushing together of the inner and outer foreskin to create hemostasis
- The desired amount of skin is excised and device removed
- The phallus is dressed with Vaseline impregnated gauze
- The infant is release from restraint and the wound is reexamined for bleeding in 10–15 min, prior to the child being released from care.

How is the circumcision performed in the non-newborn?

Circumcision in the non-newborn is a very different operation and experience. The procedure is preformed in the operating room under general anesthesia.

The procedure similarly begins with an antibacterial prep and a formal sterile drape. The preputial adhesions are taken down and two parallel circumferential incisions are made creating a sleeve of foreskin that is removed. Any bleeding in the bed of the excision is controlled by electro-cauterization. The edges of the inner and outer foreskin are sewn together with absorbable sutures. Surgical glue is also often used to bind the edges together. The phallus is dressed per surgeon preference. A penile block or caudal block is given at the start or end of the procedure.

Postoperative Expected Course

Following the procedure the newborn does experience pain. Pain should be controlled with oral Tylenol. Usually the pain lasts just overnight and most infants have returned to normal behaviors in 24–48 h.

Proper and attentive care is an important factor in preventing many of the non-acute complications that can occur. The Vaseline gauze dressing will typically fall off or can be removed within 24 h. Once the dressing is removed it is very important that with each diaper change the penile shaft skin is gently retracted to expose the entire glans and circumcision line and that the glans and incision is liberally coated with a barrier ointment such Vaseline or Aquaphor. It is very important that the caregiver not be afraid to touch the penis or pull back the skin. If untreated the incision can attach to the glans causing a skin bridge adhesion or migrate over the glans resulting in a trapped penis. The family should be informed that there will be a thick layer of white or yellow discharge covering the raw areas. The discharge is not purulent but rather scab. The discharge does not need to be forcefully removed but recovery benefits from keeping the scab soft by the liberal use of the neutral ointment until the underlying skin is healed.

Follow Up

The child is typically seen in 10–14 days for a follow-up check. It is important to assess for a trapped penis or skin bridge. The complications can be corrected most easily if identified early at this postoperative visit.

Adhesions- One of the most common sources of parental anxiety is post-circumcision adhesions. Adhesion between the sub-coronal collar and glans, not including the circumcision incision, will resolve spontaneous over time and should not be considered a true complication. Betamethasone ointment 0.05 % used twice a day for 6 weeks can speed up the natural separation and reassure families. Manual lysis is very painful and unnecessary. Adhesions between the incision and the glans are true scars and would benefit from lysis. In the immediate post-operative period this can often be performed with just topical anesthesia and without the need for suturing.

Concealed penis- the penis may be concealed due to the migration of the circumcision incision distal to the glans causing a secondary phimosis and trapped penis. If recognized early this can often be ameliorated with Betamethasone ointment and avoid a secondary procedure. The penis may also appear concealed due to a large suprapubic fat pad. This will generally resolve over time with thinning and growth and typically requires only reassurance and time.

Unhappy cosmetic outcome- All too often parents are unhappy with the final appearance due to excess residual prepuce or asymmetry of the penile skin. Rarely is there a functional issue. Treatment options are reassurance with acceptance or formal revision. One should be cautious about recommending revision if there is significant concealment due to a large fat pad.

Revision circumcision- a circumcision revision at times is warranted due to a poor cosmetic outcome. The technique is typically similar to that of a non-newborn circumcision and an excellent cosmetic result should be a very obtainable goal.

Conclusions

Newborn circumcision is a non-therapeutic, elective procedure done primarily for esthetic, cultural or religious reasons. Therefore it is particularly incumbent on the provider to provide as safe, comfortable and successful procedure as possible. Complications unfortunately do occur. Acute complications can include bleeding, infection or injury to glans or urethra. Careful attention to technique, having a well trained or well supervised provider, and thoughtful patient selection should keep complications at a very minimal rate. Non-acute complications such as penile adhesions and trapped penis can be prevented by careful parent education. Recognizing that complications occur and providing timely evaluation and treatment is the best antidote to prevent the most severe sequela.

Chapter 18
Genitalia

Jason P. Van Batavia, Douglas A. Canning, and Mark R. Zaontz

Name of Procedure

Hypospadias Repair.

Lay Description

Hypospadias is a very common anomaly in boys (0.3 % of male population). Hypospadias occurs when the opening of the urethra (i.e., meatus) is placed more proximal along the ventrum of the penile shaft [1]. The meatus can be anywhere from nearly at the normal location just below the tip of the glans all the way to a position between the scrotum and anus (Fig. 18.1). Other findings commonly associated with hypospadias include abnormal penile curvature or bending (i.e., chordee), incomplete foreskin development [2] and difficulty urinating with a deflected downward stream while standing. In rare cases the scrotum may be in an abnormal position, placed superiorly to the penis or surrounding the penis rather than below the penis. Common associated anomalies (10–15 %) include undescended testes, hernia or hydrocele,

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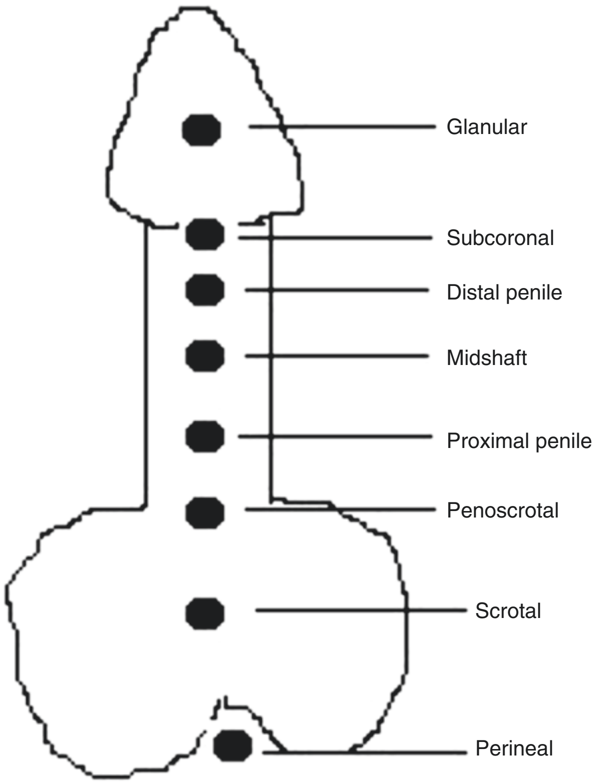

Fig. 18.1 Locations of penile opening in various types of hypospadias

which if present, may be addressed surgically at the same time as the hypospadias surgery.

Intended Benefit

Single Stage Procedure

The goals of hypospadias repair are to: (1) straighten the penis (i.e., orthoplasty), and (2) bring the opening of the urethra close to its normal location on the head of the penis to allow for urinating in the upright standing position. thereby allowing for normal sexual activity and ejaculation

Two Stage Technique

In severe cases where the opening in far away from the glans and close to the scrotum or even underneath the scrotum (i.e., proximal or penoscrotal; Fig. 18.1), the surgeon may plan on correcting the hypospadias in two stages (i.e., two separate surgeries). In these cases, the goal of the first surgery is to straighten the penis and bring the foreskin from its abnormal dorsal position to the front of the penis. In this way, the foreskin can be used during the second surgery. The goal of the second surgery is to create a new urethra and bring the opening of the urethra to a normal or near normal position at or near the glans. If the scrotum is not in the normal location, surgery can address this additional defect at the first or second stage.

Technique

The preferred timing for surgery is between 6 months and 1 year with preference to complete all surgery by 18 months if possible to minimize issues with parenteral separation and psychosocial concerns. Most surgeons perform hypospadias repair in the operating room under general anesthesia as an outpatient procedure. In addition to general anesthesia, local anesthesia is usually injected either directly into the penis or via a caudal regional nerve block to prevent pain during and immediately after the surgery. Your anesthesiologist will discuss the risks and benefits of caudal blocks. It may take several hours for your child to regain strength and mobility of his or her legs after the caudal block. The total length of the hypospadias repair varies from 1 to 4 h depending on the severity and complexity of the hypospadias. Surgery most commonly occurs under magnification provided by special glasses (loupes) worn by the surgeon or under microscopic guidance.

Where Is the Incision?

The incision will be on the shaft and surrounding the glans or head of the penis. After the surgery, if a single stage repair is planned the penis will look circumcised with no or little foreskin present since the foreskin is used in the surgical repair. Occasionally if the penile curvature is severe the surgeon may use additional tissue from the either the scrotum (no additional incision needed), the groin (a separate incision will be made in the groin crease), or the inside lip or cheek. If a two stage approach is planned, very little or no skin is removed at the first stage. After the second stage your child will look circumcised

Urethral Diversion: Use of Urethral Stents/Catheters

Depending on the specific type of surgery performed and surgeon's preference, a urethral catheter or stent may be placed to help drain the bladder. This small tube us usually left to drain into the diaper and helps allow healing of the new urethra. The urethral stent usually is stitched into place and left for 3 days to 2 weeks In certain cases, the surgeon may place a suprapubic tube (i.e., a tube through the lower abdominal wall that goes directly into the bladder to drain it in addition to the urethral stent, but this is uncommon except in older boys or in reoperative cases.

Bandages/Dressing

Surgical bandages/dressing can range from surgical glue with no bandage to clear adhesive film bandages to gauze and compression dressings that hold the penis to the lower abdomen. The type of dressing is mainly determined by surgical preference. There have been no convincing studies to show improved benefits or outcomes based on type of bandage/dressing utilized [2].

Post-operative Expected Course

Patients should expect to return home the day of surgery after recovering for a few hours in the hospital or surgicenter. After surgery, patients start by drinking liquids and should be able to eat solid foods after returning home. Pain control with oral medications and may include acetaminophen, nonsteroidal anti inflammatory drugs such as ibuprofen and/or narcotics depending on the boy's age. If the boy has a urethral stent, he may also go home on antibiotics (to take while stent is in place) and a medication that will help minimize pain from bladder spasms and irritation from the catheter/stent. Bladder spasms are suspected if the boy arches his back or suddenly cries Spasms may even occur during sleep. You may see urine squirting around or through his stent in those instances. This is not harmful. If a bandage or dressing is placed, it is usually removed from 1 to 14 days after surgery depending on surgeon's preference. Full tub bathing is usually restricted in the first couple of days. Instead surgeons recommend to sponge bathing for the first 2 or 3 days. After the dressing is removed plenty of petroleum jelly or ointment should be placed over the incisions with each diaper change or three to four times per day in potty trained boys to protect the healing tissues. After the initial dressings are removed, boys with urethral stents in place can be bathed without any restrictions.

If a stent is used, the boy should be diapered with penis up against the abdominal wall to prevent undo pressure on the surgically repaired site until the stent is removed. Once the stent is out, the penis may be diapered in any position.

Blood spotting in the diaper is common for the first few days. This is not concerning unless persistent or copious.

If your boy has a messy bowel movement that penetrates the penile dressing, one may use a bulb tip syringe to gently rinse the area with warm water.

Follow Up

Follow up varies by type of surgical repair and surgeon preference but usually varies between 3 days and 2 weeks post surgery for removal of the urethral stent After the first stage of a placed two stage correction, follow up should continue every 3–6 months until the skin is considered healed and pliable enough to allow for a second stage procedure to be scheduled. This is usually between 6 months and 1 year.

After surgery for single stage procedures, parents should note any observed erections to assess straightness of the penis and any voids to assess urinary stream quality. After the first stage of a planned two-stage correction, assessment of the penis during erections by the parents may help the surgeon determine the success of the first stage repair and plan for the second stage. If persistent curvature is present, the curvature must be corrected before creating the new urethra.

Following hypospadias repair, all boys after hypospadias repair should be followed through potty training and seen again after puberty to inspect urinary flow patterns, ensure a single strong straight urinary stream, and to confirm straight erections.

Risks of Procedure

General Outcomes

Overall outcomes and success rate depend on the specific surgical technique and on the severity of the hypospadias. Long term follow up data on most of the commonly used procedures are incomplete. Complications may not become evident until many years after surgery. In general, the further the opening of the meatus is from the tip of the penis (i.e., the more severe or proximal the hypospadias) the higher the complication rates. For more distal hypospadias repaired in a single stage, short term complication rates averages 9 % [3–5]. Single stage repairs for more severe or proximal hypospadias have higher complication rates of approximately 15–60 % [5, 6]. Similarly, with long term follow up, two stage repairs for proximal hypospadias have been shown to have a complication rate as high as 53 % with a reoperation rate of 49 % [6].

Complications

Early: Early complications can happen within the first few weeks of surgery and include bleeding, wound infections, distal separation of the repair near the tip of the penis (i.e., glans dehiscence), and loss of skin tissue used for the repair. Skin loss occurs when there is poor blood supply to parts of skin rearranged during hypospadias surgery to provide extra support and coverage to the front of the penis.

Late: Late complications can occur a month after surgery or more. Urethrocutaneous fistula or leaks along the shaft or further out on the penis can occur within weeks or months of surgery but may not occur or be diagnosed until years later when the child is potty training or even later. Other intermediate to late complications include narrowing of the urethral opening (i.e., meatal stenosis), narrowing of the urethra (i.e., stricture), recurrence of penile curvature and ballooning or dilation of the new urethra (i.e., diverticulum formation).

Specific Risks

Urethrocutaneous Fistulas

Fistulas are extra openings between the urethra and the skin. These are most often first noted during urination. If a fistula is present, a variable amount of the urine leaks out the extra hole. The extra stream may be just a mild drip, or a vigorous second stream. Fistulas may occur anywhere along the length of the repaired urethra alone or in combination with other complications such as meatal stenosis, urethral stricture, or diverticulum. Rates of fistula formation vary with type of hypospadias repair. Depending on the location and size of the fistula, repair will require one or more additional surgeries.

Meatal Stenosis

Narrowing of the new urethral opening can obstruct urinary flow. Signs and symptoms of meatal stenosis include difficulty or straining with voiding, urinary tract infections, or fistula development. Rates of meatal stenosis range from 0 % to 10 %

for distal hypospadias and 0–13 % for proximal hypospadias [2, 6]. Surgical correction is nearly always required when stenosis is diagnosed.

Urethral Stricture

Narrowing of the new urethra is an uncommon (0–3 % for distal and 0–13 % for proximal) but significant complication when it occurs [2, 5, 6]. Most strictures are diagnosed when symptoms such as urinary tract infection, urinary straining, or inability to empty the bladder completely occur. As is the case for meatal stenosis, strictures require surgical correction, often in two stage repairs, once diagnosed.

Glans Dehiscence

Separation of the repair near the tip of the penis results in an abnormal appearance to the penis as well as recurrent hypospadias as the opening regresses below the coronal margin to the distal shaft of the penis. Glans dehiscence is more common with proximal hypospadias repairs (11–14 %) compared with distal hypospadias repairs (0–12 %). Treatment requires surgical correction.

Recurrent Penile Curvature

True rates of recurrent penile curvature are unknown and likely higher than reported since most studies have short follow up and, only by following boys through puberty will the true incidence of recurrent curvature be known. Recurrent curvature rates are dependent on the type of hypospadias repair and on the specific technique used to correct the curvature. In general, recurrent curvature is less common with distal hypospadias repair (0–10 %) and usually milder in this group (<30°) while recurrent curvature is more common (2–17 %) and more severe in the proximal hypospadias repairs [2, 5, 6]. Correction of recurrent curvature if significant requires one or more surgeries.

Diverticulum

Ballooning of the new urethra during voiding with dribbling of urine after voiding may indicate a urethral diverticulum. If a diverticulum is present, evaluation for distal obstruction (i.e., stricture or meatal stenosis) is important since this may be the reason for the diverticulum formation. Diverticula occur up to 10 % of the time depending on the type of surgical technique used. All require surgical correction with one or more surgeries [2, 6].

Conclusions

There is a range of hypospadias severity based up the location of the urethral opening, curvature of the penis, and quality of the tissue and foreskin. Based upon the type of hypospadias there are a variety of surgical repairs available including single stage and two stage operations. While each surgical technique carries similar risks, the complication rates are significantly higher the more severe the hypospadias. Most complications become evident within months to a year of hypospadias repair although these boys should be followed through potty training and seen again during puberty to check for straightness of the penis during erections and to confirm a single strong urinary stream.

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Chapter 19
Surgery for Correction of Disorders of Sex Development (DSD)

Joao Luiz Pippi Salle and Rodrigo Romao

Name of Procedure

Feminizing genitoplasty (Clitoroplasty, Vaginoplasty and vulvoplasty).

Lay Description

Feminizing genitoplasty consists of clitoroplasty, vaginoplasty and vulvoplasty. **Clitoroplasty** is performed for an enlarged clitoris. The surgery is aimed at reducing its size, while keeping sensitive function intact. **Vaginoplasty** is a general term used for surgeries to bring the vaginal opening to the exterior. Preference is given to using native vaginal tissue for this purpose, but sometimes replacement with other structures such as intestine, oral mucosa, skin, peritoneum (lining of abdominal cavity) or even other tissues made in laboratory may be required. **Vulvoplasty** is the reconfiguration of the skin into labia minora and majora in situations where the initial appearance resembles a scrotum in order to accomplish a typical feminine perineum.

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Intended Benefit

Feminizing genitoplasty is an operation performed with the goal of achieving two objectives: (1) to modify the severely masculinized appearance of the external genitalia in a female while maintaining sensitive function; (2) to create a vaginal opening that is appropriate for menstruating and intercourse. Furthermore, the operation is also planned in the context of achieving long-term social adaptation for the patient and family as well as enhancing self-esteem and quality of life [1–3].

Risk of Procedure

All patients are counselled with regards to generic risks of surgery, such as those associated with general anesthesia, bleeding, infection and poor healing. Risks and complications specific to the feminizing genitoplasty can be divided into early and late.

Early risks: The most significant risk associated with clitoroplasty is disruption of the blood supply to the clitoris with resulting necrosis and clitoral loss or atrophy. Although it has not been reported in contemporary series of this operation using the same technique employed by the authors, it should be discussed with patients and families since historically it has been reported in 2–3 % of cases [4].

In the early postoperative stage, swelling, discoloration and sometimes bleeding and hematoma formation are common issues. A pressure dressing is used for a few days to prevent those problems. Wound infections and dehiscence can occur in 1–5 %. Labial dehiscence (2–5 %) requiring reoperation is very rare [3, 4].

Long-Term: Clitoroplasty: Decrease in clitoral sensitivity, undesirable cosmetic result (particularly in patients with poor endocrine control where clitoral hypertrophy can recur).

Vaginoplasty: Vaginal stenosis (narrowing) in various degrees (most are mild) preventing adequate sexual intercourse are common, requiring re-intervention after puberty; re-intervention ranges from simple dilatation to need for more extensive surgical reconstruction [4].

Alternatives

Not performing any surgery in infancy is an option that should be discussed thoroughly with parents in the multidisciplinary setting. In patients with severe virilization and a very high vagina, it is possible to perform only clitoroplasty and leave the vaginal reconstruction for the post-pubertal period, although from a technical standpoint there are benefits in doing the procedures concomitantly [3, 4]. Finally, a surgical technique where the erectile tissue from the clitoris (corporal bodies) are not removed but rather concealed underneath the labial tissue could theoretically preserve the option for future female-to-male conversion surgery, although that has not been yet been performed in clinical practice [5].

What Happens Before the Operation?

The process to reach the decision to perform a feminizing genitoplasty or not is as or more important than the actual operation. Once a decision to operate has been reached by the multidisciplinary team working with the patient and family, educational session(s) instructing the pre and postoperative steps are key.

Most patient will undergo a test called genitogram, where contrast is injected through the opening at the bottom to delineate the internal anatomy of the vagina and urethra. Adequate preoperative endocrine control is mandatory for patients with Congenital Adrenal Hyperplasia.

In babies and pre-pubertal patients the mothers are instructed to apply estrogen cream in the perineal area three times per day, 1 month before the surgery. Patient is admitted the day before the surgery and given a small volume enema. If congenital adrenal hyperplasia is the diagnosis, bloodwork and administration of steroids in the perioperative period is planned with endocrinology and anesthesia. Patients are allowed to have full diet until 8 h prior, formula milk until 6 h prior and clear liquid until 4 h prior to scheduled surgery time, depending on the patient age and fasting guidelines followed by the hospital.

Techniques

Patients receive general anesthesia and preoperative IV antibiotics. The procedure starts with a cystoscopy and vaginoscopy (inserting a camera through the opening in the perineum to delineate the anatomy). This is a key step to, in conjunction with the preoperative genitogram, establish the optimal approach for the vaginoplasty.

Clitoroplasty and Labioplasty

The procedure can take 3–5 h, depending on the complexity. It consists of careful separation of all clitoral structures, reshaping them in a typical female appearance while preserving function (by purposefully sparing blood vessels and nerves to the clitoris during the dissection). In some cases the tissues that engorge (corpora) are removed and sometimes they are maintained, depending on the surgeon and family/patient preferences. The skin of the labia is also reconfigured in order to modify the scrotal appearance for a more typical female one [5].

Vaginoplasty

The technique for the vaginoplasty will be contingent upon the information gathered by the genitogram and endoscopy at the start of the case. In cases where the vagina is low (i.e. close to the exterior/perineum), it can usually be mobilized and externalized easily. Sometimes urethra and vagina (urogenital sinus) can be mobilized as a unit and secured to the perineum in the appropriate location with absorbable sutures. A urinary catheter is left in place for a few days.

In cases where the vaginal open is high (i.e. distant from the exterior), an incision through the anus and rectum may be necessary in order to reach the vagina, mobilize it and bring it down to the perineum [6]. In exceptional cases the vagina is so small (or even absent) that there could be a need to enlarge it using non-genital tissue such as intestine, peritoneum, oral mucosa and skin grafts or flaps.

Postoperatıve Expected Course

Pain control is achieved with scheduled Paracetamol and Ketorolac postoperatively. Many children will be given a caudal or epidural nerve block at the end of the procedure to minimize pain after surgery. If a catheter is left in situ for analgesia, it usually remains in place for about 2 days. Pain is usually well-controlled with this strategy, but narcotics might be required in the first few days postoperatively.

The patient is kept in the hospital for 3–4 days and then discharged with a urinary catheter that is removed 1 week after surgery.

A pressure dressing is applied in the OR and left for 2 or 3 days post-op. Once it is removed estrogen cream is applied to the perineum for 1 week. The area usually remains swollen and the skin might be discolored for a few weeks.

Follow Up

In some cases the surgeon may want to exam the operated area and calibrate the vagina under general anesthesia 3–4 weeks postop. All patients who underwent vaginoplasty are examined under general anesthesia before puberty, around age 10, to evaluate the patency of the vagina for menstrual flow.

All patients undergoing feminizing genitoplasty for DSD should be followed by a multidisplinary team that typically includes pediatric endocrinology, pediatric urology and gynecology, genetics, psychology/psychiatry, social work. Psychological counselling and support throughout the developmental stages is essential for patients and families. Patients with CAH must comply to lifelong steroid replacement therapy.

Outcomes

The appearance of the external genitalia after the swelling has subsided (usually within 10–14 days postoperatively) is the first outcome to be assessed. A successful cosmetic outcome, as defined by the surgeon and family, is commonly observed with this procedure. In most cases of feminizing procedures the patients are young to provide valuable feedback. Obviously, functional outcomes can only be assessed when patients are going through puberty and/or ready to start sexual activity. Many patients that undergo vaginoplasty develop an area of circular fibrosis that needs re-intervention after puberty, ideally when they are ready to collaborate with vaginal dilations post-op. Unfortunately a small percentage of patients resent having surgery performed and consented by the parents. The exact percentage of such dissatisfied patients is unknown but underscores the importance of maintaining all patients on close surveillance and on psychological follow-up. Around 5 % of CAH severely virilized patients can develop gender dysphoria.

Conclusions

In conclusion, the nuances associated with feminizing genitoplasty transcend its technical execution. The procedure should only be performed by experienced surgeons in a multidisciplinary setting with the expectation of long-term follow-up and possible need for further intervention.

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Chapter 20
Surgery for Persistent Cloaca

Prasad Godbole and Govind Murthi

Name of Procedures

Posterior sagittal anorectal vaginourethroplasty (PSARVUP) and Total Urogenital Sinus Mobilisation (TUM) +/− abdomino-perineal pull-through of rectum.

Lay Description

Persistent cloaca is a severe malformation exclusively in girls where a separate rectum, urethra and vagina fail to develop normally and drain via a single common channel onto the perineum. It may be associated with a number of other anomalies including urinary tract, vertebral, genital, cardiovascular, respiratory, neurological and gastro-intestinal. A colostomy is formed in the newborn period to divert bowel content and further investigations undertaken prior to definitive surgery. Sometimes it may be necessary to drain the urine either from the bladder or the vagina by a vesicostomy or vaginostomy respectively.

The aim of definitive surgery is the achievement of bowel and bladder control and normal sexual function in adult life. The surgery involves separating the three structures to place the rectum in its normal intra-sphincteric location and the urethra and vagina onto the perineum. The operation is normally conducted from the posterior sagittal approach although may require a combined abdominal and posterior sagittal approach. More recently the anorectoplasty is combined with a total

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urogenital sinus mobilization (TUM) where the urogenital sinus (UGS) is mobilized en bloc down to the perineum.

Informed Consent

Informed consent follows the "PARQ" acronym for **P**rocedure, **A**lternatives, **R**isks and benefits, **Q**uestions (and answers provided).

Intended Benefits of Procedure

The intended benefits of the procedure are to separate the three components namely the rectum, urethra and vagina and relocate them onto the perineum in the appropriate location to enable normal bowel, bladder and sexual function.

Risks of Procedure

General

All patients undergoing any surgery are advised of risks of: Anesthesia, and generic risks of surgery such as infection, bleeding, pain and poor healing.

Postoperative Procedural Related Risks

The reported percentages of total postoperative complications are between 0 and 57 % in various series. Versteegh et al. in a systematic review showed from pooled data that postoperative complications were seen in 30 % of patients. The complication rate was 40 % in the PSARVUP group and 30 % in the TUM group.

The most commonly reported complications were:

Recurrent or persistent fistula (urethrovaginal, persistent UGS, rectovaginal, vesicovaginal) – 10 %
Rectal prolapse – 10 %
Vaginal complications such as stenosis, stricture, or occlusion – 9 %
Wound dehiscence may occur especially where a TUM under tension is performed for the longer common channels but this has been poorly reported
In the series where indications for reoperation were reported, 65 % required further surgical intervention for their complications.
Other risks are: injury to ureter(s), vaginal retraction, urinary retention or failure to catheterize the urethra.

Alternatives

This operation can also be performed using laparoscopic approach for the anorectal aspect of the PSARVUP, or in combination with open surgery. However this requires technical expertise in the procedure.

What Happens Before the Operation?

Girls would have had an initial colostomy in the newborn period and a cystoscopy to assess the anatomy and length of the common channel. Depending on how well the vagina and bladder are decompressed, in some instances they may also have had a vaginostomy or vesicostomy although most girls can be managed with a colostomy alone or in conjunction with intermittent catheterization of the common channel. Assessment of the kidney condition, anatomy and function using ultrasound and a nuclear scan (DMSA or MAG3 with Furosemide) will have been done. In other cases an X-ray test of the bladder, called a voiding cystourethrogram (VCUG) and a contrast study of the bowel (distal loopogram) will also have been carried out at the same time. In some complex situations, an MRI scan allows an opportunity to identify anatomy more clearly. Whilst the above tests help the surgeons to plan the operation, often the complete anatomy is revealed only at operation and the reconstruction is performed accordingly. Hence it may not be possible to predict the exact nature and extent of the operation that is necessary for each patient. The surgery is performed as an elective procedure. Preoperative anesthesia evaluation will be made prior to the scheduled procedure and any ancillary tests (blood and urine) ordered. Patients are allowed to have full diet as per local anesthetic protocols for milk and clear fluids.

Operative Technique

Patients receive general anesthesia with appropriate adjuvant regional anesthesia, Preparing the patient involves the whole torso, perineum and lower limbs which are wrapped in sterile cotton wool and drapes. This allows for the child to be turned from prone to supine position and vice versa. Pre-operative IV antibiotics are administered. The patient is then positioned in the prone position with a roll to elevate the hips. The legs are well supported and taped in position and adequate support for the chest and abdomen is ensured.

The rectum is approached by the posterior sagittal route and is dissected off the vagina. The vagina is separated from the urinary tract by careful dissection. Pena described the technique of TUM where the UGSis mobilized en bloc and brought down to the perineum and is the procedure of choice for most urologists along with the anorectoplasty.

The rectum is placed within the sphincter mechanism and sutured in place with absorbable sutures after closing the pelvic floor musculature. The anus is calibrated to a 10–12 Hegar dilator. The mobilized UG sinus has enough length to connect the vaginal edges and urethra to the perineum.

A catheter is left in the bladder for 7 days. Sometimes a supra-pubic catheter is also left in place to help with assessment of bladder emptying in the post-operative period.

Postoperative Expected Course

The patient may eat and drink as soon as the colostomy functions, usually on the same postoperative day assuming there has been no abdominal mobilisation. Pain relief is usually initially by nurse controlled analgesia (NCA) and then substituted with oral analgesia. The patient may receive additional IV or oral antibiotics depending on how extensive the surgery is. Foley catheter will be removed after 7 days and the child is observed for spontaneous voiding and if not possible, intermittent catheterization commenced. Sometimes it is not possible to access the bladder via the urethra (failed catheterization); in such instances the supra-pubic catheter remains in place for medium to long term drainage of the bladder. Discharge occurs as soon as full feeds are established and the patient returns for removal of the Foley catheter after 7 days.

Follow Up

Two weeks after the procedure (1 week after removal of the Foley catheter), the child returns to the consulting suite for anal calibration and graduated anal dilatation. This is performed by the parents under specialist nurse supervision. Once dilatation achieves the desired size, colostomy closure is scheduled, usually about 2 months after the procedure. A renal ultrasound is performed 3 months after the surgery.

Outcome

The specific risks relate to the main aims of the procedure. However there are a number of associated co-existent morbidities that need to be taken into consideration.

Renal Function

Renal function may be impaired as a result of structural abnormalities of the kidney in up to 60 % of girls with cloaca. A high incidence of renal failure is observed in cloaca patients. Hence careful postoperative fluid and electrolyte management and nephrology input is essential.

Bladder Dysfunction

Bladder dysfunction can be overlooked as the surgery is performed at an age where the child is not expected to be continent. Early detection and evaluation of bladder dysfunction is important to prevent progression of renal failure. Girls can suffer recurrent urinary tract infections as a result of poor emptying of the system – due to a combination of structural and functional abnormalities.

Urinary Continence

Urinary continence is achievable in between 54 and 95 % in various series but it is difficult to ascertain whether this is from the original surgery alone where the child can void spontaneously or whether this is as a result of further additional surgery. It is important to warn the family that urinary continence with the PSARVUP alone may not be achieved without further intervention. The Great Ormond Series quote 22 % of children voiding spontaneously and are dry while a further 12 % required intermittent catheterization. Overall 46 % in their series required major reconstructive surgery. This view is supported by other series.

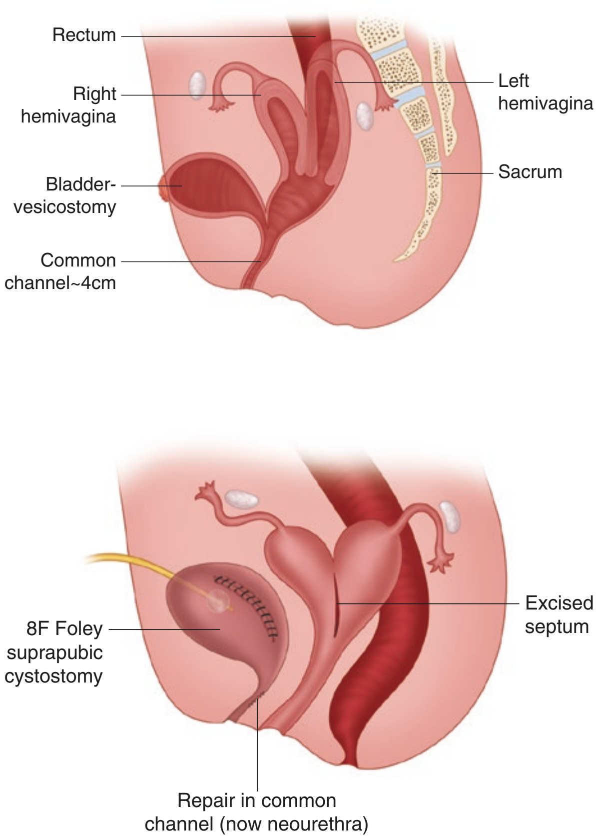
Following surgery, change in bladder dynamics may occur and may require intermittent catheterization or urinary diversion. One series has demonstrated change in bladder function in 50 % of patients, all of whom needed either IC or diversion.

Fecal Continence

Long term reviews have demonstrated fecal continence in 60 % but only 28 % of these achieve this by spontaneous bowel action and sphincteric control. The rest require an aggressive bowel management program such as rectal washouts or antegrade enemas to achieve social continence.

Gynecological aspects

Up to 60 % of girls with cloaca may have inherent structural abnormalities of the Mullerian system and vaginal abnormalities including a partially septate vagina to complete duplication of the vagina, cervix and uterus. Thirty six to 41 % may present in adult life with hematometra/hematocolpos due to stenosis of the UG sinus or of the vaginal reconstruction. Often girls with condition have a double vagina; this is corrected by dividing a septum between the two vaginas. This procedure is done either at the time of the main operation or later, usually before puberty. Due to the complex nature of the genito-urinary abnormalities and the complex surgical reconstruction undertaken, often with multiple operations, in future, a Caesarean section is the recommended method for delivery.



Conclusion

Social urinary and fecal continence can be achieved by the majority of cloaca patients although many require multiple interventions to achieve this. Co-morbidities are significant and need to be assessed and managed lifelong. Bladder dysfunction and renal failure are apparent in many children with cloaca and need careful monitoring. All girls should have a gynaecological assessment in the peri-pubertal age to assess the urogenital anatomy. It is recommended that these girls undergo a Caesarean section for delivery of babies in the future.

Part VI
Renal Impairment Surgery

Chapter 21
Hemodialysis and Peritoneal Dialysis

Alun Williams

Key Points
1. Catheter-related infection, occlusion or migration occur in hemodialysis and peritoneal dialysis and may need catheter removal or revision
2. Central venous catheters for hemodialysis can cause central venous stenosis which may then interfere with vascular access later in life
3. The exit sites of dialysis catheters should be planned to minimise mechanical complications and preserve the incision site for later transplantation
4. Dialysis in infants and small children carries a higher incidence of complications and need for catheter revision than in old children and adults

Lay Description

Dialysis refers to the process of removal of fluid and certain toxins in patients whose kidneys have failed. It is a form of renal replacement therapy.

Broadly speaking there are two routes for dialysis: hemodialysis (via the bloodstream) and peritoneal dialysis (PD – via the lining of the peritoneal cavity in the abdomen). Both of these require surgery to create dialysis access, either by a tube into a large blood vessel or a tube into the peritoneal cavity.

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Intended Benefit

When kidney function falls below 20–30 %, symptoms may develop. When function falls below 10–15 %, build up of toxins (measured by urea and creatinine levels in blood tests), salts – particularly potassium, and fluid overload, can cause symptoms and sometimes be dangerous. Dialysis aims to clear toxins, control potassium, and offload fluid to improve symptoms and avoid complications.

Generally, hemodialysis is a good means of emergency renal replacement therapy, although is very effective in the longer term. Its disadvantage is that it commonly requires very frequent visits to hospital (several times a week) and can sometimes cause difficulties in maintaining and regulating blood pressure during dialysis sessions because it involves removing and replacing blood from the circulation. PD is generally more suited to home dialysis.

Hemodialysis

The majority of children undergo hemodialysis via a central venous catheter (CVC). In adults, and occasionally in older children, a preferable route for hemodialysis is via a high flow conduit (arteriovenous fistula) which can be created by direct arteriovenous anastomosis or by placement of a prosthetic graft. This has the advantage of durability and lower infection rates than CVC [1]. The disadvantage of this is that it requires the placement of needles at each dialysis session. This section will consider CVC access as it is by far the commonest modality in children.

Technique

The upper body circulation is preferable, as it allows easy catheter access on a day-to-day basis, but also crucially preserves lower body vessels for subsequent transplantation. The internal jugular veins are the easiest for surgical access, right more so than left because of its straighter course. Subclavian vein access is associated with a higher rate of thrombosis and venous stenosis than jugular.

The jugular vein is accessed most commonly via needle puncture in the neck and passage of a guide wire under X ray control [2]. Alternatively, the vein can be mobilised through an incision in the neck. This may be required for revision access.

If the period of dialysis is predictably short (relatively few weeks) then direct placement of the line with its exit site through the neck is acceptable. However for the placement of a chronic line, it is better to tunnel from the venotomy onto the chest wall. The line should have a gentle curve to prevent kinking, and the approximate exit site should be determined so that the retaining cuff is placed conveniently in the subcutaneous tissue (Fig. 21.1).

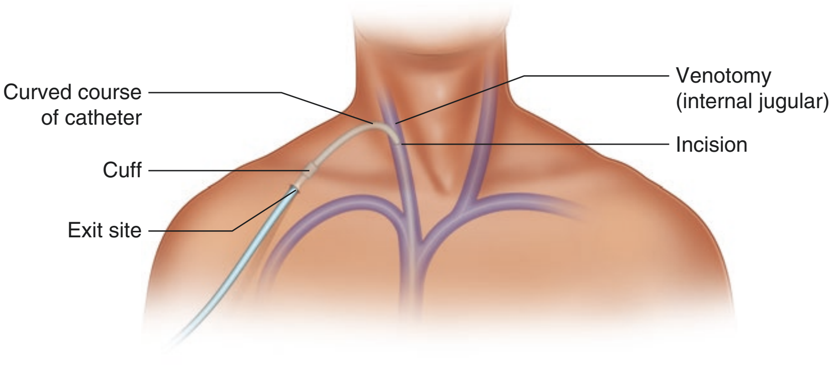

Fig. 21.1 Hemodialysis catheter placement

The commonest lines have two lumens. Because of the need for high flow to ensure efficient dialysis, the distal lumen frequently needs to sit within the right atrium, especially in smaller children. Bigger central vein calibre in older children and adults means that in these groups the tip of the line may be left in the conventional location just above the cavoatrial junction as long as flow is adequate. Sometimes two separate single lumen catheters may be placed for inflow and outflow respectively. If access is particularly difficult or tenuous then a single lumen line may suffice.

Sutures are placed in the skin incision and to secure the exit site of the line. The line is 'locked' at the end insertion with heparinised saline.

Expected Postoperative Course

The line should be usable immediately. This is another advantage over fistula formation which needs a period of weeks to mature into usable access. The dialysis prescription will vary from patient to patient and is independent of the line itself.

The actual procedure is day-case stay, but hospitalisation afterwards will depend on dialysis itself.

Follow Up

Efficiency and ease of dialysis are assessed 'live' during sessions. In the early postoperative phase, venotomy and tunnel problems can be seen easily.

Outcome

The desirable outcome for discontinuation of dialysis is recovery of function (in acute kidney injury where recovery is possible) or successful transplantation. Complications including those requiring CVC removal or revision are much more likely to occur in smaller children. In one study reviewing patients under 10 kg [3], the average lifespan of a CVC was 13 months. Another study of dialysis in under-two's [4] saw around 70 % requiring catheter removal (in groups on hemodialysis and peritoneal dialysis) for reasons other than end-of-use.

Risks and Complications

Bleeding

Normal clotting and platelet count should be ensured to minimise the risk. Bleeding from the venotomy, or tunnel, or exit site can usually be controlled with local measures. Life-threatening bleeding is very rare.

Hemopneumothorax

With ultrasound localisation of the vessel, or with open exposure of the jugular vein, this should be very rare. A chest radiograph is recommended after needle-puncture venotomy, in part to assess lung expansion, but also to assess line tip position.

Difficulty Positioning the Line Tip

This is most commonly due to central venous occlusion or stenosis (see below). For line revision, preoperative angiography (MR or direct) may be useful. Intraoperatively, use of a hydrophilic guide wire may be helpful. The services of interventional vascular radiologists may be required if angioplasty is required.

Infection

In one large registry study [5] bacteraemia occurs with a frequency of 1.9 per 1,000 catheter days and exit site infection 1.8 per 1,000 catheter days. Infection requiring line removal occurs 0.9 per 1,000 catheter days.

Line Malfunction

This can reflect line occlusion (by thrombus or fibrin sheath) or breakage. In the same registry study line malfunction requiring CVC change occurred 2.1 times per 1,000 catheter days.

Venous Stenosis

This occurs in about 10 % after jugular vein CVCs, and about 40 % after subclavian CVCs [6]. The latter are not recommended for dialysis because of this.

Peritoneal Dialysis

As with hemodialysis, access to the peritoneum can be created for short term (acute) or longer term use. PD tends to have fewer hemodynamic sequelae than hemodialysis, and has the advantage that machines for PD are relatively portable making it a dialysis modality suited for use in the patient's home. Dialysis can be done overnight, so as to interfere as little as possible with daytime activities.

Technique

In an acute setting, access to the peritoneum is made either by open cutdown or via needle puncture, aspirating to ensure no visceral breach, and placement of a guide wire. A catheter is then threaded into the general peritoneal cavity and can be used immediately.

For chronic PD access, placement either via open approach or via laparoscopic-assisted approach [7] is a matter of surgeon preference. They both involve small incisions in the peri-umbilical area. Many surgeons establishing PD access in children excise part or all of the greater omentum, as this lowers the risk of catheter entrapment and failure to dialyse [8].

The laparoscopic approach has the advantage of allowing an extraperitoneal tunnel for the catheter to be created under vision. This fixes the end of the catheter in the pelvis to lower the risk of catheter migration. It also allows the peritoneum and contents to be assessed (e.g. to assess colitis in cases of hemolytic uraemic syndrome, or to visualise the extent of intrabdominal adhesions that might affect dialysis). Through a small circumumbilical incision, a portion of omentum requiring excision can be delivered and removed. Either through the lateral end of the incision or through a separate site, a needle is inserted through the abdominal wall but remaining extraperitoneal under vision to create the extraperitoneal tunnel. A peel-away sheath large

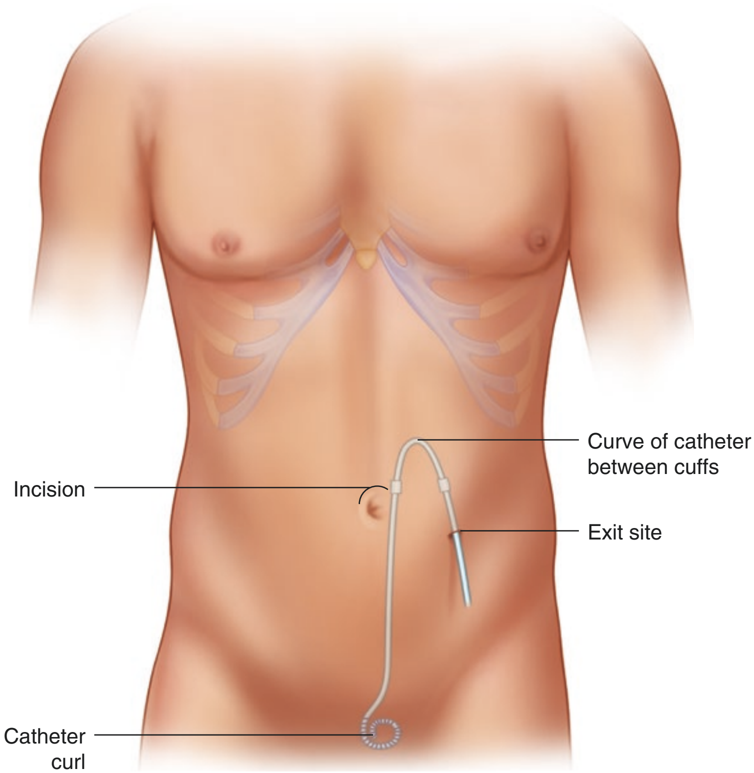

Fig. 21.2 PD catheter placement

enough to accommodate the catheter is inserted over a wire placed through the needle. The catheter is then placed under vision in the pelvis and the connector end tunnelled to the predetermined flank or iliac fossa. The fibrous cuffs of the catheter are left to implant in the abdominal wall. The abdominal wall is closed with sutures. Figure 21.2 illustrates the configuration of the PD tube on the abdominal wall.

An important point of planning is that the exit site should ideally be on the left side to avoid the planned incision for subsequent transplant. PD exit sites are safe on the same side as a gastrostomy [9] even when the two are established at the same time.

The abdominal wall is closed with sutures. The tube at the exit site should not be sutured, as it has a subcutaneous retaining cuff, to lower the risk of damage to the catheter or resistance to flow. Flow into and out of the catheter should be confirmed at the end of the operation.

Expected Postoperative Course

Pain relief is given as required. The incision will usually have been infiltrated with local anaesthetic. Dialysis can be started as soon as needed but if possible, a few days are allowed to lower the risk of leak of dialysis fluid through wounds.

Occasionally a degree of ileus can hold up restarting feeds. Patients on PD tend towards constipation that might require laxatives.

Follow Up

A period of supervised training by specialist staff is required to minimise technical and infective complications from handling the PD catheter. Monitoring biochemistry and weight to assess fluid balance is undertaken as routine in paediatric renal units. Parameters of dialysis efficiency are recorded by PD machines. Minimal routine surgical review is required – only to ascertain sound wound healing, or to troubleshoot problems such as leak or herniae.

Outcome

As for hemodialysis, the desired outcome is that the catheter is removed when no longer needed – either because of recovery or because of a successful transplant. Catheter survival is of the order of 80 % at 12 months, 60 % at 24 months and 35 % at 48 months [10, 11]. Smaller children (especially infants [4]) have an increased risk of catheter removal for problems.

Risks and Complications [12, 13]

Bleeding

Bleeding requiring reoperation is very rare, although exploration is mandatory if significant bleeding occurs, or if there is any concern about visceral injury.

Infection

Superficial wound infection is uncommon and amenable to treatment with systemic antibiotics. PD peritonitis is a common complication during the lifetime of PD in any patient and occurs at a rate of up to 20 %. It is treated by intraperitoneal antimicrobials, and intravenous if the patient is systemically unwell. Recurrent peritonitis occurs in 10–20 % of peritonitis cases and sometimes requires catheter revision or removal. This is more likely if peritonitis is fungal.

Dialysate Leak

This occurs in less than 10 % of patients, is usually early, and commoner in smaller children.

Catheter Malfunction (Blockage, Breakage, Migration)

Catheter occlusion and breakage both occur in around 10 % of patients. A fibrinous sheath or catheter entrapment within intestine or residual momentum can cause catheter occlusion. Occasionally with fibrinous occlusion, intracatheter fibrinolytic can help, but revision of the catheter is often required.

Development of Hydrocele and Hernia

These occur very commonly, in up to 50 % of patients on PD. They may not necessarily cause symptoms, but if they do, or dialysis efficiency becomes impaired, then surgery is required.

Sclerosing Peritonitis

This is a very rare complication in children (compared with adults) where dense encapsulation of the intestine can occur, causing gut failure. PD is discontinued, and if symptoms fail to settle, surgery in the form of peritonectomy might be required. This is a highly morbid condition.

Conclusion

Although CVCs are convenient for hemodialysis, and allow ‘no-needle’ dialysis, they can have significant morbidity, which may have an impact on the treatment of patients’ renal disease in adulthood where peripheral vascular access becomes more important. Peritoneal dialysis has less in the way of cardiovascular effects and can be performed more straightforwardly in the home environment at a time to minimise impact on daily life and education. Dialysis complications for both hemodialysis and PD are consistently more common in smaller children. A critical appraisal of dialysis options should be undertaken for each patient, with the goal ultimately being planning a successful transplant.

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Chapter 22
Kidney Transplantation

Alun Williams

> **Key Points**
> 1. A transplant is the best treatment option for kidney failure but should be planned in the knowledge that further transplants will be required.
> 2. There is a primary failure rate of 2–3 %, but more than 75 % of kidney transplants survive beyond 5 years.
> 3. Urological complications are commoner in children with an underlying urological problem.
> 4. Infection and rejection are common. Immunosuppression needs to be monitored to minimise the risk of both.

Lay Description

A successful kidney transplant is the gold standard renal replacement therapy. However, a transplant is only another treatment option for kidney failure, rather than a cure. Even though transplants now survive longer than in previous eras, it is almost inevitable that a child who has a transplant will need further transplants later in life

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Intended Benefit

When kidney function falls below about 10–15 %, a transplant is the best option to restore quality of life and freedom from dialysis and hospitalisation. A transplant confers most benefit if it can be performed before dialysis is required.

Work Up for Transplantation

This usually happens in parallel with treatment and investigation of kidney failure, and needs to include managing risk of infection by vaccination, nutrition and growth by dietary means (including supplementary feeds if needs be), and safeguarding biochemistry such as calcium, phosphate and potassium levels as these can cause symptoms that can become life-threatening. Sometimes dialysis becomes necessary.

The urinary tract needs to be investigated if it was associated with kidney failure in the first place or there are symptoms. Occasionally problems with the urinary tract need surgery prior to a transplant to provide a safe environment for the transplant. Heart function needs to be investigated as this can be affected by kidney disease and can affect the transplant if poor. Also, the transplant team needs to know that suitable blood vessels are present for implanting a new kidney. At the time an organ becomes available, it needs to be matched optimally (tissue type) to maximise its survival and to lower the risk of rejection, and 'cross-matched' to ensure that the recipient's immune system does not react immediately to cause loss of the kidney.

Organ Sources

The availability of an organ tends to be the rate-limiting step in transplantation. Kidneys can be from living or deceased donors. A living donor is frequently a relative (such as a parent) but not exclusively so and there is an increasing trend for altruistic donation. Deceased donors fall into two categories: donors after brain death and donors after cardiac death. The outcomes of transplants from both of these groups is similar. Living donors have a slightly better outcome in terms of life of the graft.

Timing of the Operation

A living donor transplant is a planned procedure, so allows the recipient to be as fit as possible. A deceased donor transplant frequently happens as an emergency as and when a suitable organ becomes available.

Technique

Before the transplant operation can begin, the kidney needs to be inspected and prepared for implantation. The kidney is stored and transported in cold preservation fluid surrounded by ice. Rarely, damage to the kidney or its blood vessels, or an abnormality such as a previously-unknown tumour, prevents its implantation. If there are multiple renal arteries, they sometimes need to be reconstructed to make the transplant easier. There is an increased risk of thrombosis with multiple vessels or if a vascular reconstruction is required.

The recipient is under general anaesthetic, and will have a central venous catheter and peripheral venous and sometimes arterial lines placed, and a bladder catheter either through the urethra or into a urinary reconstruction.

The incision varies from surgeon to surgeon, and according to the size of the recipient. It is usual for the incision in an older child or adult to be curved on one side of the abdomen. In a small child (for example under 15 kg), it is sometimes easier to open the abdomen through a midline incision (Fig. 22.1).

The recipient blood vessels for implantation are identified. In children above 15 kg, it is usually preferable to stay outside the peritoneal cavity – this preserves the peritoneum for dialysis and also lowers the risk of damaging intraperitoneal structures. In smaller children, or if the peritoneum cannot be reflected away, the vessels are prepared through the peritoneal cavity. A space is created for the kidney to lie.

Generally, the largest recipient vessels in relation to the donor vessels are selected. In small children, this usually means the aorta and inferior vena cava. In older children the iliac vessels are usually acceptable.

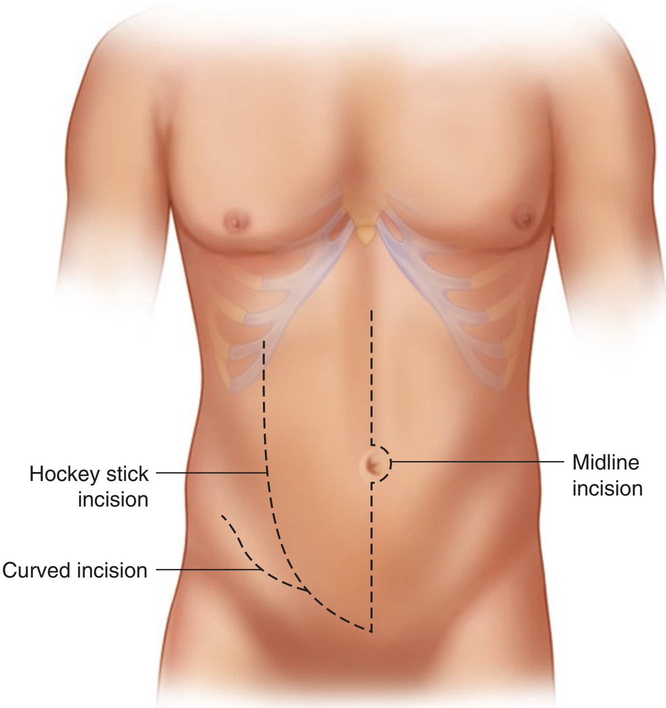

Fig. 22.1 Incisions for renal transplant

When the recipient vessels have been prepared, the kidney is removed from ice and the vessels are sutured into place on the recipient vessels. When the vascular anastomosis is complete, the kidney is reperfused with the recipient's blood. Any bleeding at this time can be controlled.

The bladder (or urinary reservoir) is then identified and the ureter of the transplant is sutured to this. Sometimes a ureteric stent is left in place, to be removed at a later date.

The abdominal muscles are closed if possible, usually leaving a drain. Occasionally, in smaller children, it is necessary either to use a patch of a prosthetic material, or leave the muscles open with a plan to return to the operating room to close the muscles when tissue swelling has resolved after a few days. It is important that the kidney is not under pressure if the abdomen is closed.

Postoperative Expected Course

Children are sometimes nursed in the intensive care unit after a transplant. This is more likely in smaller children who need more intensive monitoring of fluid balance and if they need medicines to regulate their blood pressure. Also, if they require ventilation after the surgery.

Between 2 and 3 % of kidneys fail to work – the cause of this is usually thrombosis in the vessels. Around 15 % of deceased donor kidneys have delayed graft function (DGF), where dialysis might be necessary while waiting for transplant function to begin. The incidence of DGF in living donor kidneys is lower at around 5 % [1].

Children can usually eat and drink within a day or 2 of their transplant, as long as they do not have an ileus. The drain in the wound will remain until its output is virtually nil, and the bladder catheter will stay in for 4–5 days normally. The central venous line is removed when no longer needed for fluid administration or monitoring.

Recipients will need to start oral immune suppressant medication immediately (this can sometimes be given intravenously at the start) and prophylaxis against infection. The exact medication will vary from patient to patient and on local protocols. Levels of immunosuppression will need to be monitored.

Follow Up

After discharge from hospital, transplant recipients require very intensive follow up on an outpatient basis. In the early weeks post-transplant, this can be several times per week, gradually decreasing. Medication needs frequent alterations and monitoring, but less so as time goes on and stable state is reached with a functioning transplant. If a stent was left in the ureter at the time of transplant, it will need to be removed via a cystoscopy at a later date. If a child was on dialysis before the transplant, the dialysis catheter may need to be removed under an anaesthetic in the operating room.

Some units have protocols for biopsies of the transplant. Sometimes a biopsy is require to exclude rejection or to follow up treatment of rejection. A biopsy can usually be done under local anaesthetic and sedation in older children, but may need general anaesthesia in younger children.

Outcome

Table 22.1 shows graft survival for all groups at 1, 3 and 5 years post transplant for living and deceased donor kidney transplants.

An important consideration in paediatric transplantation is that the majority of children who undergo transplantation will need transitional care into adulthood, with need for further renal replacement therapy (including retransplantation). The period of transition (aged 15–19 years) has the highest rate of graft failure and is therefore a critical time for ongoing monitoring and care.

Risks and Complications

Bleeding

Immediate bleeding from the vascular anastomoses is controllable directly by suturing. Vessels in the graft hilum may bleed but this will settle usually with pressure or suturing. Return to the operating room because of bleeding is rare (less than 5 %).

Vascular Thrombosis

This may be an immediate phenomenon, requiring oxne of the vessel anastomoses to be re-done because of poor flow, or may occur in the days after transplantation. Up to 7 % of recipients may suffer a venous or arterial thrombosis [2]. Venous

Table 22.1 Graft survival (%) at 1, 3, and 5 years post transplant according to year of transplant and organ source

		LD			DD	
	1 year	3 year	5 year	1 year	3 year	5 year
1987–1990	89	81	75	75	63	55
1991–1994	92	85	80	85	76	70
1995–1998	94	91	85	91	82	74
1999–2002	96	92	87	93	84	79
2003–2010	97	92	84	95	84	78

NAPRTCS Annual Report 2010 [1]
LD live donor, *DD* deceased donor

thrombosis is commonest in the first 5 days or so. It may present with pain over the transplant (as it becomes congested because of poor outflow), hematuria, or loss of function. Rarely, a graft rupture can occur needing emergency operation to control life-threatening bleeding. Arterial thrombosis is commonest in the first few days and presents usually with abrupt loss of urine output or rise in serum creatinine. If a thrombosis is suspected an urgent Doppler ultrasound is required to determine the need for surgical re-exploration of the transplant. Thrombosis can only very rarely be recovered. A graft nephrectomy is the usual outcome.

Ureteric Complications

These occur in up to 10 % [3] of transplants. A urine leak is an early phenomenon, and causes persistent or increased drainage via the wound drain (or through the wound), or pain or swelling under the wound with a rise in creatinine. A stent may delay the diagnosis. With a stent in place and catheter drainage, a urine leak may stop spontaneously. If it does not a diverting nephrostomy may need to be placed. A urine leak that cannot be managed by drainage alone will require exploration of the transplant to redo ureteric drainage.

A ureteric stenosis may present late with rising creatinine or hydronephrosis of the transplant. If a stent can be placed (usually from nephrostomy puncture of the kidney) that may be all treatment is required. If a stent fails to treat a stenosis, exploration of the transplant and revision of the ureter-bladder anastomosis will be required. This sometimes requires use of a native ureter or bladder flap if the length of stenosis is substantial.

Ureteric complications are slightly commoner in recipients whose underlying kidney disease was associated with a urinary tract malformation or urinary tract surgery.

Lymphocele

This is an uncommon condition where lymphatic fluid collects around the graft and can cause obstruction to ureteric drainage (and more rarely still to the vessels). Initially drainage is helpful, but it persistent drainage occurs then it can be treated by opening the lymphocele into the peritoneum to allow reabsorption of interstitial fluid.

Infection

Infection is very common as an early or late complication of transplantation. This is because of the need for immunosuppressant medication. Increasingly frequent or severe infections might be a marker of over-immune suppression. Systemic infection with bacteria or viruses need aggressive and early treatment, and in some cases monitoring (as in the case of cytomegalovirus [CMV] or Epstein-Barr virus [EBV]). If the transplant is from a know CMV positive donor into a CMV negative recipient, prophylaxis is usually recommended in many centres. It is also usual in the first few months after transplantation for recipients to receive antibiotic prophylaxis (with co-trimoxazole) against opportunistic infections such as Pneumocystis species.

Rejection

Up to 20 % paediatric recipients will suffer an episode of acute rejection, usually within the first 6 months post-transplant [1]. This usually presents with a rise in creatinine and requires a biopsy to diagnose and classify. Many episodes of rejection can be managed by an acute short course of steroids and modulating baseline immunosuppression. Sometimes extra treatment by lymphocyte depletion or plasma exchange is needed. A major cause of rejection in paediatric transplantation is non-adherence with immunosuppression. Monitoring drug levels sometimes helps, as does close supervision.

Long Term Risks

Transplant kidneys inevitably fail in the long term. This is usually because of a combination of factors. Some immune suppressants are relatively nephrotoxic (such as tacrolimus and cyclosporin) and cause damage and scarring within kidneys. Immunological 'chronic rejection' despite immunosuppression undoubtedly plays a part in long term graft loss. There are risks of transmission of occult tumours or infection from the donor (normally in the setting of deceased donor transplantation where donor workup is inevitably less complete than it can be in the living donor scenario). Immunosuppression per se increases the risks of malignancies in the recipient, especially skin malignancy, so regular clinical surveillance is important. Metabolic risks of chronic kidney disease and medications include hypercholesterolaemia and bone mineralisation problems are important to survey in recipient, as is the development of hypertension and proteinuria, both of which can accelerate graft loss as well as indicate the development of generalised vascular disease.

Conclusions

A successful kidney transplant is undoubtedly the best treatment for established renal failure, but it remains only a treatment rather than a cure. The majority of children who receive transplant will require a further transplant, so work up needs to be as comprehensive as possible, and the first kidney needs to be as well matched as possible to favour subsequent transplants. Lifelong follow up for long term as well as short term problems is required. Because urological problems are more common in the paediatric population, ongoing urology follow up for these recipients will also be needed.

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Part VII
Urogenital Tumours

Chapter 23
Wilms' Tumor and Other Renal Neoplasms

Armando J. Lorenzo

Name of Procedures

Open vs. laparoscopic radical nephrectomy, open partial nephrectomy, lymph node dissection.

Lay Description

A surgical procedure (operation) to remove a mass involving part or most of one or both kidneys. The goal of the intervention is to completely remove the tumor without breaking it or leaving part of it inside the body (so-called "positive margin"). At the same time, the surgery often includes removal of lymph nodes to assess for metastasis, help with proper staging and tailor treatment accordingly.

Surgical Technique

Renal tumors are commonly addressed surgically. Depending on patient characteristics (most notably age and presence of bilateral or metastatic disease), as well as the treatment protocol followed, neoadjuvant chemotherapy with or without

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confirmatory biopsy is sometimes recommended. Response to prior systemic therapy, presence of a contralateral normal kidney, tumor location and degree of compromise of the residual renal parenchyma dictates preference for radical nephrectomy (i.e. resection of the tumor and kidney) or nephron-sparing surgery (also called partial nephrectomy). In general nephron sparing surgery is favored in patients with underlying renal insufficiency, bilateral renal tumors, neoplasms in patients with a predisposition syndrome, and small renal masses in older children with renal cell carcinoma. Both procedures are performed in the operating room under general anesthesia, and can take 3–6 h, depending on the complexity.

For open surgery, the patient is positioned for supine. Depending on tumor location and patient size, the operating table may be gently flexed and the ipsilateral flank slightly elevated with a gel roll. Cystoscopy and stent placement is not routinely performed, with the exception of cases of complex nephron sparing surgery with a high likelihood of entry into the collecting system. Following traditional protocols, a generous trans-peritoneal incision is favored. In infants and young children, the abdominal cavity is often entered through a transverse incision. As the child grows, predilection shifts to either a Chevron or midline approach, heavily influence by surgeon preference. In some cases, including nephron sparing surgery, an extra-peritoneal approach through a flank (or modified flank) incision is employed. Exploration no longer includes routine surgical assessment of the contralateral kidney. Surveillance of the abdominal cavity and assessment for metastatic lesions or evidence of preoperative rupture is conducted. The kidney is then dissected from surrounding structures after reflecting the colon and ipsilateral pertinent structures (such as the duodenum and tail of pancreas). Careful attention is paid to controlling large parasitic vessels. The hilum is subsequently dissected, palpating the renal vein to verify absence of a tumor thrombus. Once defined, the renal artery (or arteries) are tied and transected, followed by the vein. The adrenal gland is not routinely removed, except for cases with a large upper pole mass, difficulty developing a dissection plane off the kidney, or concern for gross tumor involvement. Customarily, the ureter is transected as close to the bladder as possible, maneuver that is only likely to be of oncological benefit in cases with tumor botryoid extension. After removing the specimen, generous lymph node sampling is mandatory to complete proper surgical staging. Hiliar and peri-aortic or peri-caval nodes are harvested, along with any other suspicious ones. The wound is closed in layers using absorbable sutures. A closed suction drain is not routinely placed unless a partial nephrectomy was performed.

For nephron sparing surgery, lymph node sampling is often done first. Concurrently, the vascular hilum is secured by placing vessels loops around the artery (or arteries) and vein. The kidney is mobilized and Gerota's fascia opened paying close attention to preserve perinephric fat on the tumor area. Intraoperative ultrasound facilitates identification of the tumor and depth of resection, particularly in endophythic lesions. In complex cases, vascular clamping with or without regional hypothermia may be employed. However, there has been increasing interest towards "zero ischemia" resection with only selective arterial branch vascular control. The tumor is removed in its entirety along its pseudocapsule attempting to keep a small margin of normal appearing tissue with it. Bleeding areas and openings in the collecting system are controlled with absorbable sutures. Hemostasis is aided with local clotting agents or materials, packed in place by gently approximating the renal capsule over them.

Laparoscopic cases attempt reproduce the above-mentioned steps employing a minimally invasive approach. Most cases are well-selected children, often following neoadjuvant chemotherapy, who are candidates for a radical nephrectomy. Positioning is slightly different, with the child placed in a modified flank position with an axillary roll, well secured to the operating room table. An umbilical port is used for access and camera placement, while 5 or 10 mm additional ones are placed in the epigastric region and ipsilateral lower quadrant. On the right side, a laparoscopic liver retractor is often helpful. Depending on the anatomy, the hilum is controlled with Hem-o-lock clips or an endo-GIA (vascular loads). The specimen is removed through a Pfannesteil-type incision inside a laparoscopic bag (to avoid spill from rupture during delivery). It is critical to remember to perform a lymph node dissection.

Irrespective of the approach, at our institution patients undergo a frozen section evaluation of the specimen. If the results are fairly conclusive for a Wilms' tumor (or any other neoplasm that may benefit from systemic chemotherapy), a vascular access port is placed during the same anesthetic. Further technical details and complications from this part of the intervention are outside of the scope of this chapter.

What Happens Before the Operation?

Axial imaging (with CT scan or MRI) allows full characterization of the renal mass and presence of contralateral renal lesions, metastatic disease, intravascular involvement (tumor thrombus), and evidence of preoperative rupture. Often, axial imaging of the chest is obtained to rule out metastatic deposits in the lungs, and a contrast study with vascular phase is added if there is symptomatology suggestive of pulmonary embolism. When there is concern for tumor thrombus, the evaluation is often complemented with Doppler assessment of the renal veins and vena cava, with transthoracic or trans-esophageal echocardiograms obtained in cases with extension above the diaphragm or symptoms suggestive of embolization. Once surgery is considered to be the best next step forward, addressing the associated risks, benefits and alternatives for treatment, a standard consent form is signed. Due to the inherent risk for significant bleeding, a discussion about the possibility and risks of blood transfusion should be included. Similarly, anesthetic risks -including monitoring and analgesic strategies, such as an arterial line or epidural catheter placement- should take place.

Postoperative Expected Course

The patient is often kept with minimal oral intake until there is evidence of bowel function return, which often becomes evident in 48–72 h and appears to be expedited by minimizing opioid use and stimulating early mobilization. Analgesia is multi-modal, including epidural infusions, acetaminophen and judicious use of non-steroidal anti-inflammatory drugs. During open procedures, neuraxial adjuvant analgesia is commonly started after induction (in the form of an epidural infusion with local anesthetics and opioids). This will remain in place for 2–3 days. Despite the known side effects, patients will often require systemic opioids. A Foley catheter is routinely placed upon induction and removed in 48–72 h. Use of a nasogastric tube is optional, and current trends favor avoiding it altogether or removing it early after surgery (in the first post-operative day).

If a drain was placed after surgery (following partial nephrectomy), the output is monitored at least until removal of the indwelling Foley catheter, critical time at which leaks become clinically evident. When there is doubt about the presence of a urine leak, the fluid can be tested for creatinine level (and compared with a serum value). Depending on the result, the drain can be removed or closely managed during outpatient visits. Discharge occurs in approximately 5 days, when the child is pain-free on oral medications and tolerating a regular diet. It is wise to recommend the use of stool softeners as a prophylactic intervention against constipation or fecal impaction.

Follow-Up After Discharge

Patients are discharged from the hospital 3–7 days after surgery. Outpatient visits are often coordinated with evaluations with the Oncology team, as key information (i.e. the pathology result) will guide further treatment and dictate imaging protocols. Surgical assessment (wound healing, renal function, blood pressure check), can be done concurrently. Once all catheters and drains are out, postoperative urology visits rarely add to the already extensive evaluations performed by Oncology. In cases of nephron sparing surgery, a renal ultrasound is often performed 4–6 weeks after the procedure (or sooner depending on symptoms) to evaluate the appearance, Doppler flow and presence of associated fluid collections. If a stent was placed at the time of nephron sparing surgery, it is removed 4–6 weeks later, taking into account chemotherapy administration and blood counts. Before and after stent removal, the kidney is imaged with ultrasound.

Outcomes

Successful removal of the renal mass is the expected outcome. Rarely, in cases deemed to be too difficult to complete without sacrificing major adjacent structures or add significant morbidity to the procedure, an open or percutaneous biopsy is performed with the aim to provide specific adjuvant chemotherapy prior to re-attempt at resection. Survival and other critical outcome measures are rarely dependent on the surgical intervention, but other important parameters such as underlying diagnosis (type of tumor), stage, renal reserve, and need for other adjuvant treatment modalities (such as chemotherapy or radiation).

Informed Consent

Intended Benefits of Procedure

To remove the tumor along with part or the whole kidney, along with perinephric fat surrounding the mass and pertinent lymph nodes. Surrounding structures grossly involved by the tumor may have to be resected to complete the procedure. These include the ipsilateral adrenal gland, the psoas fascia, and a small portion of diaphragm. The main goal of this technical exercise is to completely remove the mass without rupture in order to provide pathological diagnosis and staging. Depending on the histology and staging, surgery may also provide stand alone therapeutic value. Often though, adjuvant chemotherapy or radiation may be necessary. The decision is driven by standardized protocols drafted by large cooperative oncology groups (such as the Children's Oncology Group and SIOP).

Risks of Procedure

General

Overall, complications after radical or partial nephrectomy are relatively low and often minor (Clavien-Dindo Grade I or II; i.e. conservative or pharmacologic management), however some can carry higher morbidity and demand additional procedures or interventions under anesthesia. As with any other surgery, families and caretakers are advised of issues from anesthesia (including neuroaxial analgesic interventions such as placement of an epidural catheter), and generic risks such as pain, bleeding, infection, and poor cosmetic/functional outcome. The most common complications and their incidence in a large up-front open nephrectomy series is presented in Table 23.1.

Risks, Open Radical Nephrectomy

1. Unresectable tumor: The current trend in North America for non-metastatic unilateral tumors without tumor thrombus up to or above the hepatic veins is to proceed with upfront nephrectomy. During such attempt, gross involvement of vital surrounding structures or need to sacrifice adjacent organs to complete the intervention is not recommended. Instead, the procedure is aborted after obtaining a biopsy to confirm the diagnosis and open the discussion for re-exploration after neoadjuvant therapy.
2. Tumor rupture and intra-peritoneal spill [1]: Despite meticulous dissection, some tumors are extremely friable and the capsule breaches during manipulation.

Table 23.1 Incidence of surgical complications after primary nephrectomy for Wilms' tumor

Complication	Percentage
Bowel obstruction	5.1
Significant bleeding	1.9
Hypotension	0.6
Wound infection	1.9
Vascular injury	1.5
Splenic injury	1.1
Liver injury	0.2
Pancreatitis	0.2
Diaphragmatic injury	0.4
Chylous ascitis	0.2
Incisional hernia	0.2
Pulmonary embolus	0.2
Respiratory failure	0.2
Pneumothorax	0.2
Pleural effusion	0.2
Urinary tract infection	0.2

Adapted from Ritchey et al. National Wilms Study Group data, 534 patients [4]

Similarly, preoperative rupture or bleeding may predispose to further disruption and spill. This complication increases the chance of local recurrence and, in cases of Wilms' tumor, upstages the patient to treatment arms that include flank or abdominal radiation.

3. Bowel obstruction: Bowel manipulation and dissection to remove the tumor can generate scar tissue and adhesions which can lead to bowel obstruction.
4. Damage to adjacent structures: Tail of pancreas, spleen, liver, duodenum, bowel/mesentery
5. Internal hernia: Dissection of the mesentery, which can be fairly attenuated by a large mass, can disrupt its integrity and allow for loops of bowel to become entrapped.
6. Wound infection: This is a rare complication, minimized by providing prophylactic antibiotics, practicing meticulous tissue handling, securing hemostasis and enforcing a sterile technique.
7. Incisional hernia, wound dehiscence
8. Postoperative ileus: Delayed resumption of oral intake due to Ileus (or dilated bowels), may be due to the use of opioids, electrolyte imbalance and slow return to ambulation.
9. Pneumothorax/hemothorax: Posterior dissection, particularly near the upper pole, can involve part of the diaphragm. Injury can lead to entry into the pleural cavity, accumulating air or blood, which is often evacuated at the time of surgery.
10. Atelectasis, respiratory compromise and infection, including pneumonia
11. Postoperative fever: Although a common occurrence, the development of fever often triggers an extensive workup, particularly in children who have received

chemotherapy prior to surgery. Broad spectrum antibiotics are given until blood and urine cultures are reported negative. To minimize the chances of a urinary tract infection, the indwelling Foley catheter is removed as soon as possible.

12. Bleeding requiring transfusion -and less commonly- need for exploration: Bleeding is most often not excessive, does not induce symptoms and can be managed conservatively. When significant bleeding does occur, a previously undiagnosed, preexisting bleeding condition might need to be excluded. In rare instances, patients with Wilms' tumor may have an acquired coagulopathy, thus screening with bleeding times is commonly performed.
13. Positioning injuries: These include compression from securing devices, injuries from stretching of joints or ligaments, and pressure from under-protected contact points. Depending on the location, pain, limited range of motion and peripheral neuropathy may occur.
14. Lymphocele formation or chylous ascites (if major disruption or lymphactics occurs during difficult dissection or lymph nodes, particularly if massively involved with tumor)

Additional Risks, Partial Nephrectomy

1. Urine leak with or without urinoma formation: If tumor resection involves entry into the collecting system (either inadvertent or by design), a urine leak may occur, usually in the early postoperative period. Large disruptions commonly trigger placement of a urinary stent, with continuous drainage of the bladder through an indwelling catheter, and a closed suction drain. In such circumstances, the leak will become evident or increase at the time of catheter removal. A urinoma may develop despite having a drain, thus symptoms (such as pain, fever and ileus) are evaluated with a renal ultrasound or abdominal CT scan. Most cases resolve with conservative management.
2. Development of an arterio-venous fistula or pseudo-aneurysm: These are the result of intra-renal vascular disruption during resection and/or repair of the parenchymal defect. Profuse of persistent bleeding requires selective embolization or, in rare cases, exploration and open control or nephrectomy.
3. Inability to perform nephron sparing surgery, requiring removal of the whole renal unit. This includes tumor involvement that precludes safe resection without breaching into the tumor and disruption of the kidney blood supply with resultant irreversible ischemia.
4. Incomplete resection (positive margins): Despite what appears to be an adequate rim of normal tissue removed with the tumor (outside of its pseudo-capsule), and negative intraoperative frozen sections, residual microscopic disease is an inherent risk of nephrons paring surgery. This has to be weighted against risk of removal functioning renal parenchyma with the tumor, and long term consequences such as renal insufficiency.

5. Acute tubular necrosis, ischemic nephropathy: Intermittent or prolonged occlusion of the kidneys blood supply can lead to transient or permanent loss of functioning parenchyma. In cases with a solitary kidney, pre-existing renal insufficiency or bilateral disease, the implications are clinically evident, with azotemia that may require renal replacement therapy. Permanent development of stage 5 chronic kidney disease is particularly worrisome, as these patients are not candidates for renal transplantation until demonstrating lack of recurrence (no evidence of disease) for at least 2 years.

Additional Risks, Laparoscopic Nephrectomy [2]

1. Injury to intra-abdominal or vascular structures during trocar placement
2. Gas embolism: Very rare complication, but can lead to catastrophic intra-operative hemodynamic collapse. It should be suspected in cases with temporary disruption of vein integrity and high CO_2 insufflating pressure.
3. Failure to progress of safely complete the case laparoscopically, with need for open conversion.
4. Tumor rupture during extraction.
5. Under-staging due to failure to perform or inadequate lymph node sampling.
6. Trocar site hernia.

Special Circumstances

1. Tumor thrombus: Renal neoplasms may extend into the venous system, with thrombus formation that can reach the atrium. The risk of resection in such cases involves difficulty with vascular control (which increases when the thrombus is adherent to the vena cava wall or extends to and above the hepatic veins), thrombus disruption with incomplete resection and pulmonary embolism, and compromise of the venous drainage for the contralateral kidney.
2. Pre-operative rupture: Patients may present with contained (retroperitoneal or within Gerota's fascia) or free rupture into the peritoneum. Resection may not be elective, but dictated by hemodynamic compromise, symptoms and risk for further tumor dissemination. The likelihood of severe bleeding due to difficulty in gaining and securing vascular control increases in this circumstances. Disruption and tumor spill can easily increase during manipulation of an already friable tumor with capsular breach.
3. Botryoid extension into collecting system: Tumor may extend down the ureter and into the bladder. If unrecognized, there is risk for incomplete resection if the bladder is not inspected and the ureter is not removed in its entirety.
4. Bilateral disease [3]: These patients are seldom candidates for upfront resection, and should be offered neoadjuvant chemotherapy with or without confirmatory

biopsy. Following this, bilateral nephron sparing surgery is the preferred treatment strategy. As indicated above, there are specific risks to partial nephrectomy, which are magnified based on the number and complexity of resections. Although recent evidence would suggest that it is possible to save enough normal parenchyma, additive effects of temporary ischemia, surgical resection and radiation lead to an increased risk of renal insufficiency over time.

Alternatives

Surgical resection is cornerstone in the management of most primary renal masses, and most cases are not amenable to treatment with chemotherapy or radiation alone. An open vs. laparoscopic approach and the decision to proceed with a total vs. partial nephrectomy is dependent on each case's particular characteristics, thus providing some alternatives in terms of risks and complications.

Conclusion

Surgical procedures offered for management of renal masses are fairly straight forward, based on a large published experience in adult populations and a growing body of evidence in children and adolescents. Because these operations are often necessary and indicated for staging and add therapeutic benefit for conditions that are life-threatening, the patient and family should be counselled extensively in the preoperative period about the rationale for the procedure and treatment plan. Discussion should clearly include relevant risk, benefits and alternatives as previously presented in this chapter.

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Chapter 24
Bladder and/or Prostate Rhabdomyosarcoma

Alonso Carrasco Jr. and Nicholas G. Cost

Introduction

A. Rhabdomyosarcoma (RMS) is a soft tissue malignant tumor of mesenchymal origin. It is the third most common solid tumor in children accounting for 10–15 % of all pediatric solid tumors [1]. The incidence of RMS is estimated at 4.5 per 1 million, with over 50 % of cases diagnosed in the first decade of life [1, 2]. RMS exhibits a bimodal distribution for age at presentation with a peak between 2 and 6 years, and then again between 10 and 18 years of age [3]. Approximately 15–20 % of RMS tumors arise in the genitourinary (GU) system primarily in the prostate, bladder, and para-testicular area [4]. Vaginal, cervical, uterine, primary retroperitoneal, and renal RMS are relatively unusual sites. The treatment of GU-RMS has drastically evolved over the years due to effective multidisciplinary, multimodal, and risk adapted therapies developed through clinical trials. Prognosis has significantly improved as a result of all these measures, currently with an expected overall 5-year survival >70 % [1]. Accordingly, the goals in treating GU-RMS have shifted towards minimizing morbidity and reducing therapy. Surgery has shifted from pelvic exenteration to organ preservation. Surgery is directed by stage, risk stratification, and response to chemotherapy/radiation therapy. Current relevant surgical procedures for the treatment of RMS include performing a biopsy to establish a diagnosis, and subsequent partial cystectomy, cystectomy ± prostatectomy (cystoprostatectomy), and pelvic lymphadenectomy. For para-testicular RMS the reader is recommended to review the chapter on testicular tumors and associated procedures such as orchiectomy and retroperitoneal lymph node dissection.

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Name of Procedures

A. Percutaneous, transurethral, or open biopsy.
B. Partial cystectomy, cystectomy ± prostatectomy (cystoprostatectomy), and pelvic lymphadenectomy

Lay Description and Intended Benefit

A. A biopsy is a procedure used to obtain a portion of tissue from a tumor for the intended purpose to have it analyzed by a pathologist. Based on microscopic characteristics of the sampled tissue, a pathologist can provide a diagnosis of the condition which can then help guide treatment strategies. Percutaneous biopsy in most cases can be performed under local anesthetic using a needle inserted through the skin into the tumor. Ultrasound or more advance imaging modalities are often times utilized to aid in identifying and sampling the tumor. This type of biopsy typically yields a small amount of tissue, and multiple biopsies are often obtained at the same operative setting.

   A transurethral biopsy accomplishes the goal of obtaining a tissue sample from within the bladder or prostate. This procedure is generally done under general anesthesia using a small endoscopic surgical instrument called a resectoscope. Under direct vision the resectoscope can be navigated into the urethra, prostate, and bladder. The resectoscope is equipped with an electrically heated wire loop which can be used to scrape or cut tissue away from the tumor. The resectoscope is able to obtain a large quantity of tissue in a minimally invasive way. A transurethral biopsy is not feasible when the patient's urethra is unable to accommodate the resectoscope, or the tumor obliterated the lumen of the prostate and bladder. In such cases, a percutaneous biopsy or an open biopsy is preferable.

   An open biopsy is performed under general anesthesia by making an incision and exposing the tumor. This is typically accomplished via a suprapubic incision. After obtaining the biopsy, a portion of the tumor is excised and sent for pathological analysis. When an open biopsy is done, and if feasible, a pelvic lymphadenectomy is often times simultaneously performed to better stage the patient.

B. Contemporary therapy for bladder/prostate (B/P) RMS consists of multimodal therapy with chemotherapy, radiation therapy, and surgical resection of residual viable tumors. B/P RMS is often times large and infiltrating, which makes complete resection with organ preservation challenging. Occasionally the tumor is isolated to a small portion of the bladder which renders it amenable to complete excision by performing a partial cystectomy. A partial cystectomy involves removing part of the tumor along with part of normal bladder. The remaining uninvolved bladder is left in place. A partial cystectomy is ideal for tumors that are located at the dome, in an area where a negative surgical margin can be easily obtained, and if there is sufficient uninvolved bladder to provide an adequate bladder capacity.

Since most tumors are unresectable, the majority of patients with B/P RMS undergo diagnostic biopsy followed by primary chemotherapy ± -radiation therapy. If after chemo-radiotherapy there is residual viable tumor, a partial cystectomy, cystectomy ± prostatectomy is performed to achieve local oncologic control. A cystectomy involves removing the entire bladder. In males, a cystoprostatectomy involves removing the bladder, prostate, and seminal vesicles. In females, a cystectomy may involve removal of the uterus, cervix, and vagina. Cystectomy ± prostatectomy is performed in conjunction with lower urinary tract reconstruction (e.g. neobladder, continent urinary diversion, or ileal/colon conduit).

A pelvic lymphadenectomy involves removing the lymphatic tissue surrounding the pelvic vessels (i.e. internal iliac, external iliac, and common iliac vessels) and the obturator fossa bilaterally. A pelvic lymphadenectomy is performed for staging and risk stratification which guide further therapy. At the time of an open biopsy, it is recommended to sample the pelvic lymph nodes when possible.

Technique

A. A percutaneous biopsy is typically performed by an Interventional Radiologist or Urologist and under general or local anesthesia. This can be done through the perineum, abdomen, or less commonly trans-rectally. A percutaneous biopsy is performed under sterile conditions and is generally done using ultrasound. A core needle biopsy device, which can range between 12 and 20 gauge, is use to obtain multiple tissue samples. This type of biopsy is typically well tolerated and results in minimal discomfort for the patient. The puncture site is cover with a small clean dressing for 24–48 h.

A transurethral biopsy is performed under general anesthesia. Patient is typically placed in the dorsolithotomy position. The perineum is prepped and draped in the standard fashion. Antibiotic prophylaxis to cover genitourinary flora should be administered prior to instrumentation. It is recommended to start the procedure by performing a diagnostic cystourethroscopy to evaluate the feasibility of a transurethral biopsy and delineate the anatomy. If there is ureteral obstruction, a stent placement can be performed prior to the transurethral biopsy. Depending on the size of the resectoscope and urethral anatomy, one may perform urethral dilation/calibration if needed. The resectoscope should be introduced into the bladder under direct vision as these tumors can obliterate the lumen and can increase the risk of iatrogenic injury. Once the resectoscope is in the bladder the ureteral orifices should be identified prior to performing any bladder biopsies. The resection can then be performed with either monopolar or bipolar electrocautery loop. For monopolar electrocautery 1.5 % glycine is recommended, and for bipolar electrocautery 0.9 % sodium chloride is utilized. The resection should be carried using low cutting current until enough tissue for analysis is obtained. Hemostasis is then ensured using coagulation current. The tissue is then extracted by using graspers or by allowing it to drain out through the resectoscope. It is important to remove all tissue fragments as these can potentially cause bladder outlet obstruction. When a resectoscope is not available, grasping forceps can be used to perform the biopsy, but this usually yields less tissue. It is recommended to decompress the bladder under direct visualization prior to removing the resectoscope to identify any additional bleeding sites which can be controlled with coagulation current. Depending on the extent of resection, it is recommended to leave a Foley catheter for 24–72 h.

An open biopsy should be performed under general anesthesia when a transurethral biopsy or a percutaneous biopsy cannot be performed. The patient is placed in a supine position, prepped, and draped in the standard fashion. Antibiotic prophylaxis to cover skin flora should be administered prior to incision. Typically an open biopsy is best approached via a suprapubic incision (midline or Pfannenstiel). A suprapubic incision has the added benefit of giving enough exposure to perform a pelvic lymphadenectomy for staging. The skin incision should be large enough to allow safe expose of the tumor. The subcutaneous tissue is dissected down to expose the fascia using electrocautery. The fascia is then incised transversely or longitudinally, and then the rectus muscles are separated longitudinally. Depending on the location of the tumor and size, the retropubic space (space of Reitzus) should be developed. For small tumors deep in the pelvis, the biopsy and pelvic lymphadenectomy can be performed in an extraperitoneal fashion by dissecting the peritoneum away from the bladder and/or tumor. Once the tumor is identified, a portion of the tumor is excised and sent for pathological analysis. Hemostasis is then ensured with the aid of electrocautery and/or hemostatic agents (fibrilar, gelfoam, or surgicel). If feasible the excision site edges should be approximated using absorbable suture. A pelvic lymphadenectomy is performed for staging when deemed feasible based on preoperative imaging (see section "Technique" B). The rectus muscle is then approximated in the midline with absorbable suture. The fascia is closed with absorbable suture in a running or interrupted fashion. The subcutaneous tissue is approximated and the skin is approximated with a sub-cuticular stitch using absorbable suture. The skin is then covered with a sterile dressing for 24–48 h.

B. A partial cystectomy is approached in the same fashion as an open biopsy (see section "Technique" A). Once the bladder and tumor have been completely mobilized, the tumor along with a small margin of uninvolved bladder is excised with the use of electrocautery. It is important to identify the ureteral orifices and ureter as one approaches the trigone to avoid iatrogenic injury of these. Of note, endoscopic placement of ureteral stents at the beginning of the procedure may aid in ureteral identification and avoidance of injury. A partial cystectomy is ideal for tumors that are located at the dome, in an area where a negative surgical margin can be obtained, and if enough uninvolved bladder is left to provide an adequate bladder capacity. The specimen should be oriented with clips or sutures for the pathologist. Utilizing intra-operative pathology including frozen section analysis is highly recommended to establish negative margins prior to reconstructing the bladder. A 0.5 cm margin is recommended, unless achieving such margin would result in significant structural/functional damage, or loss of organs [5]. While waiting for the frozen section results, a pelvic lymphadenectomy can be performed (see below). Once negative margins have been confirmed, the bladder should be closed with absorbable suture in one or two layers according to surgeon's preference. Depending on the extent of the partial cystectomy, an indwelling catheter is left in place for 1–10 days. A pelvic drain tunnel via a separate stab incision is optional. The abdominal/wound closure is performed in a similar fashion as to the open biopsy (see section "Technique" A).

Treatment of B/P RMS has been increasingly directed toward organ preservation. Complete primary resection by performing a cystectomy ± prostatectomy is currently uncommon, and this is typically reserved for patients who have persistent viable tumor after upfront chemotherapy ± radiation therapy. A cystectomy ± prostatectomy is approached in the same fashion as open biopsy and partial cystectomy (see section "Technique" A). Once the bladder and tumor are completely mobilized the ureters are traced as far distally towards the bladder, transected, and tagged with absorbable suture. It is recommended to have an indwelling catheter to aid in identification of the urethra. The endopelvic fascia is incised bilaterally. In males, the rectovesical and rectoprostatic space is developed in an effort to protect the rectum. In females, an attempt at preservation of the uterus, cervix, and vaginal canal should be done. This can be accomplished by developing the uterovesical (pouch of Meiring), vesicocervical, and vesicovaginal space. However, if the tumor is intimately involved with these structures, partial excision of these structures along with the bladder and tumor may be necessary. If complete excision of these structures is planned, the rectouterine space (pouch of Douglas) is developed instead. It is important to note that these planes may be difficult to develop due to post chemo-radiotherapy effect. The bladder, prostate, and uterine pedicles are ligated using a vessel sealing device (e.g. LigaSure or harmonic scalpel), a vascular stapler, or by tying off the pedicles with sutures or surgical clips. If an orthotopic neobladder is planned, a sizeable urethral stump must be preserved. In males, the urethra is transected distal to the prostate and in females just distal to the bladder neck. Once the specimen is completely freed it should be sent for pathological analysis. Frozen section is highly recommended to establish negative margins. While waiting for the frozen section results, hemostasis should be ensured and a pelvic lymphadenectomy performed (see below). Once negative margins are established and pelvic lymphadenectomy is complete, lower urinary tract reconstruction is undertaken. Viable options for urinary tract reconstruction include orthotopic neobladder creation, continent urinary diversion (e.g. Indiana pouch and Kock pouch), or a temporary or permanent ileal/colon conduit. Details on such urinary reconstruction can be found the corresponding chapter in this book. After reconstruction of the lower urinary tract, the abdominal/wound closure is performed similarly to open biopsy or partial cystectomy. Pelvic drains are highly recommended given the extensive lower urinary tract reconstruction, and the ureteral anastomosis are usually stented.

Lymph node involvement is noted in up to 12 % of patients with B/P RMS [6]. Management of lymph nodes in B/P RMS is not well defined, and there is no defined standard template. Pelvic lymphadenectomy is performed in conjunction to open biopsy, partial cystectomy, or cystectomy ± prostatectomy for staging and risk stratification. Pelvic lymphadenectomy typically involves removing any enlarged or suspicious lymph nodes, any lymphatic tissue surrounding the pelvic vessels (external, internal and common iliacs), and lymphatic tissue within the obturator fossa bilaterally. This requires meticulous dissection of the lymphatic tissue away from the vessels using a combination of sharp dissection and/or electrocautery. The obturator nerve should be identified early during dissection to avoid iatrogenic transection or clipping. Lymphatic tissue should be ligated distally using clips, sutures, or a vessel sealing device. Pelvic lymphadenectomy should be performed after the primary surgical intervention is accomplished and before lower urinary tract reconstruction is started.

Postoperative Expected Course

A. After a percutaneous, transurethral, or open biopsy patients are generally able to resume a regular diet shortly after recovering from anesthesia. Scheduled analgesia with non-narcotic medications (e.g. acetaminophen or non-steroidal anti-inflammatories) is sufficient, and narcotics can be used for breakthrough pain management. Anticholinergics can help alleviate symptoms due to bladder spasms from bladder biopsy or indwelling catheter. Antibiotics are typically not needed beyond surgery, but prophylactic antibiotics for urinary tract infection can be use given the indwelling catheter. For open excisional biopsy, patients may benefit from a caudal block or single shot spinal anesthetic. After a percutaneous and transurethral biopsy, patients can typically be dismissed home after recovering from the anesthetic. Open biopsy often times requires a 24–48 h observation in the hospital.
B. After a partial cystectomy and pelvic lymphadenectomy patients are generally able to resume a regular diet shortly after recovering from anesthesia. Scheduled analgesia with non-narcotic medications (e.g. acetaminophen or non-steroidal anti-inflammatories) is sufficient, and narcotics can be used for breakthrough pain management. Anticholinergics can help alleviate symptoms due to bladder spasms. Antibiotics are typically not needed beyond surgery, but prophylactic antibiotics for urinary tract infection can be use given the indwelling catheter. Patients typically can be dismissed home after 24–48 h of observation in the hospital.

After cystectomy ± prostatectomy, pelvic lymphadenectomy, and urinary tract reconstructions patients typically have a prolonged recovery. This is mainly due to the additional bowel work required for urinary tract reconstruction.

Following an enhanced recovery after surgery (ERAS) protocol, patients can typically resume clear liquids shortly after surgery, and regular diet within 24–72 h [7]. These use of scheduled non-narcotic medications, regional anesthesia (e.g. wound soakers or epidural), and the judicious use of narcotics can expedite the return of bowel function. Antibiotics are typically continued for 24 h. Patients can generally be dismissed home once they are on a regular diet, pain is adequately controlled on oral medications, and the parents are able to assume care of the patient at home (catheter/drain/stent care, irrigations, and stoma care) which is usually around 5–7 days after surgery. Drains and stents are removed at the discretion of the surgeon. Generally, stent are removed 7–14 days after surgery, and drain are removed once the output remains consistently low (less than 30 mL in 24 h).

Follow-Up

A. Follow-up after a biopsy is dependent on when the final pathology report is available and on the timing of starting chemotherapy/radiation therapy. Patients left with an indwelling catheter are seen back in clinic for catheter removal and voiding trial. Arrangements should be made for patients to meet with a medical oncologist and radiation oncologist to start therapy or to enroll in a clinical trial.

B. After a partial cystectomy patients are left with an indwelling catheter for 1–10 days. These patients are typically seen back in clinic for catheter removal and voiding trial if the catheter is not removed prior to dismissal from the hospital. Arrangements should be made for the patient to meet with a medical oncologist and radiation oncologist to start therapy or enroll in a clinical trial.

The follow-up after cystectomy ± prostatectomy is dependent on the type of urinary tract reconstruction performed. The readers is encourage to read the chapters dedicated to each type of reconstructions for further details regarding follow-up and additional studies needed. Arrangements should be made for the patient to meet with a medical oncologist and radiation oncologist once final pathology is available to discuss potential need for additional therapies.

Risks of Procedure

A. Generally patients undergoing any form of biopsy are at risk of infection, bleeding, and pain associated with the procedure. The complication rate for percutaneous, transurethral, or open biopsy of rhabdomyosarcoma of the bladder and/or prostate is less than 10 % [8]. While data for each potential complication are limited, it can be extrapolated from studies of other solid tumors or bladder tumors. Percutaneous biopsy of solid tumors has a diagnostic accuracy of over 90 % and major complications rate of less than 2.5 % [9–11].

Patients undergoing transurethral biopsy are at risk of bleeding (hematuria), urinary retention, infection, urethral injury, and bladder perforation. Although no specific data are available on B/P RMS, this can be extrapolated from data on transurethral resection of other bladder tumors in adults. The goal of transurethral biopsy of B/P RMS is to obtain tissue diagnosis, while the goal in adult patients with bladder tumors is to render them tumor free. Thus a more aggressive transurethral resection is performed in adults which results in an increased risk of complications. Despite this, the overall complication rate for transurethral resection of bladder tumor in adults is less than 10 %, with urinary retention accounting for 2.8 %, infection for 2.1 %, and bladder perforation for less than 0.5 % [12–14]. Most patients undergoing transurethral bladder biopsy should expect hematuria. These complications can be easily managed with antibiotics, indwelling catheter, and catheter irrigation. In addition to the risk of infection and bleeding associated with open biopsy, if a pelvic lymphadenectomy is performed the risk and potential complications of this additional procedure should also be taken into consideration (see section "Risks of procedure" B).

Partial cystectomy and cystectomy ± prostatectomy are each associated with early and long term complications. These procedures can be associated with risk of infection, bleeding, bowel obstruction/ileus, ureteral injury, bowel/rectal injury, and urine leak. The complication rates are highest among patients who were treated with radiotherapy preoperatively, but the complication rates for primary tumor resection can be as high as 25 % [8]. There are limited data available on B/P RMS postoperative complications as most studies focus on survival. Data on postoperative complications can be extrapolated from the adult experience with similar procedures. Partial cystectomy is associated with lower rate of complications than cystectomy ± prostatectomy, with a complication rate of less than 8 % [15–17]. Patients who undergo partial cystectomy or organ sparing surgery are at particularly increased risk of having long term complications related to chemo-radiotherapy. While bladder preservation is possible in up to 60 % of patients with RMS, over 40 % of patients experience bladder overactivity, diminish capacity, urgency, incontinence, decrease compliance, hemorrhagic/radiation cystitis, or chronic pain [18–21]. When these symptoms are severe, patients may require additional surgery such as urinary diversion or bladder augmentation. While complications for cystectomy ± prostatectomy are higher, these usually are related to more extensive resection and urinary tract reconstruction. Long term complications to urinary tract reconstruction include ureteral obstruction, stone formation, cutaneous fistulas, electrolyte disturbances, vitamin B12 deficiency, and issues with the catheterizable channel. For a more elaborate discussion on complications related to urinary diversions, the reader is encouraged to read the chapters dedicated to these. Pelvic lymphadenectomy for the treatment of other pelvic tumors (such as prostate cancer) can be associated with a complication rate between 4.1 and 10.6 % (see section "Risks of procedure" B) [22]. Most common complications of pelvic lymphadenectomy include lymphocele, vascular injury, postoperative bleeding, infection of lymphocele, and obturator nerve injury [22, 23]. With a focus now on improved quality of life, long term sexual function after treatment of B/P RMS has become a topic of interest. While there are few small studies available on this topic, it appears that over 40 % of the patients have adequate erections after cystoprostatectomy and the majority respond to erectogenic medications [24, 25].

Conclusion

A. B/P RMS remains a therapeutic challenge. While surgery can be challenging and variable, it continues to play a major role in the treatment outcomes of children with B/P RMS. It is important to manage these tumors in a multidisciplinary approach from the outset with the goals of preserving organ function and quality of life for patients.

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Chapter 25
Testicular and Paratesticular Tumors of Children and Adolescents

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Introduction

(a) Testicular and paratesticular tumors of children and adolescents are rare entities with similar surgical management. Testicular tumors represent 1–2 % of all pediatric tumors, with annual incidence of 0.5–2.0 per 100,000 children [1–3]. A bimodal age distribution is observed with first peak in the first 3 years of life, and then again after puberty [4]. Primary testicular tumors are divided into germ cell tumors (seminoma, yolk sac, embryonal carcinoma, choriocarcinoma, and teratoma) and stromal tumors (leydig cell, sertoli cell, and granulosa cell). Germ cell tumors usually present with painless testicular mass and have malignant behavior. Stromal tumors typically have a benign course, although some can become metastatic, and can be associated with hormonal production leading to precocious puberty. Treatment of testicular tumors is directed by the histologic type and age at presentation. The bimodal age distribution, histologic subtype, and stage encountered at different ages exemplifies the different molecular and behavior of these tumors. Of prepubertal tumors, 68–74 % carry a benign pathology, with mature teratoma making up the largest proportion (48 %) [5–7]. Furthermore, over 85 % of prepubertal patients present with stage I (localized) disease [8]. Regardless of the age at presentations, patients require a formal evaluation with scrotal ultrasound, tumor markers (AFP, βHCG, and LDH), and if indicated hormonal studies to evaluate for a stromal tumor. Scrotal ultrasound helps localize the tumor, determine if it has characteristics that will permit testicular sparing surgery, and it assess the status of the contralateral testicle. If metastatic disease is highly sus-

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pected based on tumor markers, physical exam, or symptoms, preoperative staging imaging of the chest, abdomen, and pelvis can be considered. Pre-operative staging imaging can also enable simultaneous central venous access placement at that time of radical inguinal orchiectomy if subsequent chemotherapy is warranted. The initial treatment of a testicular tumor is to establish a diagnosis. Prepubertal tumors are generally less aggressive, as is their treatment. For prepubertal stage I tumors, surveillance is recommended and for metastatic disease chemotherapy is the first line. Postpubertal tumors tend to have similar tumor characteristics and behaviors as adult tumors, as such they are treated more aggressively following established adult protocols.

Paratesticular tumors generally involve the spermatic cord or structures surrounding the testicle. Most common reported malignant tumors include liposarcomas, leiomyosarcoma, rhabdomyosarcoma, malignant fibrous histiocytomas, and fibrosarcomas. Of these, rhabdomyosarcoma is the most commonly encountered in children and adolescents. The genitourinary system is the primary site of Rhabdomyosarcoma (RMS) in 15–20 % of cases, and overall paratesticular tumors account for 5–6 % of RMS primary tumors [9, 10]. Paratesticular RMS represents over 40 % of all para-testicular tumors seen in children and adolescents [6]. These tumors are highly malignant, with up to 40 % of children presenting with metastatic disease [6]. Similar to testicular tumors, age at presentation impacts treatment. While multimodal treatment with chemotherapy, radiation, and surgery is warranted in all RMS, certain patients are treated with more aggressive therapy based on age. For instance, children under 10 years of age and stage I disease can be treated with orchiectomy and chemotherapy alone. However, children 10 years of age and older or those with evidence of retroperitoneal disease on staging imaging should undergo nerve-sparing ipsilateral retroperitoneal lymphadenectomy. Surgical procedures relevant to testicular and paratesticular tumors are similar. These include radical inguinal orchiectomy, testicular sparing surgical excision of testicular tumors, placement of testicular prosthesis, and retroperitoneal lymphadenectomy.

Name of Procedures

(a) Radical inguinal orchiectomy
(b) Placement of testicular prosthesis
(c) Partial inguinal orchiectomy (Testicular Sparing Surgery)
(d) Open retroperitoneal lymphadenectomy (RPLND)

Lay Description and Intended Benefits

(a) A radical inguinal orchiectomy is usually the first step in the multidisciplinary treatment of testicular and paratesticular tumors. It is a procedure used to remove the testicle along with the spermatic cord at the level of the internal inguinal ring. This procedure is both diagnostic, and in many patients curative.

(b) A testicular prosthesis is used to fill the void of a testicle in the scrotum. Testicular prostheses are made out of silicone shell and filled with saline, silicone gel, or soft silicone. Testicular prosthesis can be safely implanted at the time of radical inguinal orchiectomy or at a later time at the preference of the patient and/or surgeon. A testicular prosthesis is sized to the contralateral testicle, and as such it is advisable to delay placement until puberty is reached to avoid having to upsize prosthesis. In cases where further local surgery, systemic chemotherapy or radiation may be required, delaying placement of a testicular prosthesis may be ideal.

(c) Partial inguinal orchiectomy involves removing only the part of the testicle involved by a tumor, this is also known as testicular sparing surgery. This procedure is performed and approached in a similar fashion to a radical inguinal orchiectomy, but only the tumor is excised sparing the residual normal testicular tissue. This procedure is reserved for benign tumors, tumors with low risk of metastatic disease (stromal tumors), tumors under 2 cm in size, bilateral testicular tumors, and for patients with solitary testicle wishing to preserve fertility and avoid hormone supplementation.

(d) Malignant testicular and paratesticular tumors tend to metastasize in a well-known pattern. The first and most common site of metastatic disease is the retroperitoneal lymph nodes, which are located around the abdominal aorta and inferior vena cava (IVC). A RPLND is a staging/diagnostic and therapeutic procedure for testicular and paratesticular tumors. A RPLND involves mobilizing the bowel contents away from the aorta and IVC, and resecting the lymphatic tissue surrounding these vessels. A nerve-sparing RPLND is a term used when the nerves involved for ejaculation are dissected away and preserved during the lymphatic tissue. A primary RPLND is a term used when the RPLND is done prior to chemotherapy or radiation is administered, and it often is done in a nerve-sparing fashion. When patients have residual retroperitoneal tumor after chemotherapy, a RPLND is performed as a post-chemotherapy RPLND (PC-RPLND). A PC-RPLND can be a challenging procedure as the chemotherapy may cause the lymphatic tissue to become increasingly adherent to the surrounding structures. Additionally, a PC-RPLND may be associated with auxiliary procedures such as nephrectomy, bowel resection, and vascular reconstruction thus increasing the complexity of surgery and potential complications. The use of laparoscopic for surgery has expanded significantly in the last decades. Laparoscopic and robotic-assisted laparoscopic RPLND have been described as alternative approaches to RPLND in adults. By using small incisions a camera and small instruments are used to accomplish the same surgical task as an open RPLND. However, this procedure can be extremely challenging due to a steep learning curve. Laparoscopic RPLND will not be discussed as there are limited reports of these approach in the pediatric literature.

Technique

(a) Prior to performing a radical inguinal orchiectomy it is important to confirm laterality by repeating the physical exam and reviewing the preoperative imaging. After induction of anesthesia the patient should be placed in a supine position, prepped, and draped in the standard fashion. The patient should be prepped from umbilicus to mid-thigh (including the genitalia). Given the low risk of infection associated with inguinal procedures, antibiotic prophylaxis to cover skin flora are typically unnecessary unless placement of a testicular prosthesis is planned, or based on patient's risk factors [11–14]. A 4–7 cm oblique incision beginning above the external inguinal ring (2 cm cephalad and lateral to the pubic tubercle) is made. The incision should be big enough to allow the safe and atraumatic delivery of the testicle/tumor. The subcutaneous tissue (Camper's and Scarpa's facia) is then dissected down to the external oblique fascia using blunt dissection or electrocautery. The external oblique fascia is incised towards the external inguinal ring medially and internal inguinal ring laterally. It is important to identify and free the ilioinguinal nerve from the spermatic cord. Using gentle blunt dissection the spermatic cord is circumferentially dissected free. A 0.25 in. Penrose drain is passed posteriorly twice and clamped with a hemostat for early vascular control prior to manipulating the testicle/tumor. Alternatively, one can pass a heavy silk suture and ligate the spermatic cord. The testicle is delivered through the incision by applying gentle pressure from the scrotum towards the incision. If the tumor is too large, the incision should be extended towards the scrotum to facilitate delivery. Once the testicle is delivered, the hemiscrotum will be invaginated due to the gubernaculum attachment. This should be incised with electrocautery. The testicle and spermatic cord should then be dissected and doubly ligated high in the inguinal canal, preferable at the level of the internal inguinal ring, using heavy permanent suture and leaving long tails. If necessary later, the high ligation of the cord will facilitate removal of the intrabadominal portion of the spermatic cord at the time of RPLND. Hemostasis is then ensured, and the external oblique fascia is approximated using absorbable suture while safeguarding the ilioinguinal nerve. If placement of a testicular prosthesis was planned, at this point the prosthesis is placed (see below), otherwise a local anesthetic can be injected at this time. The subcutaneous tissue is approximated and the skin is approximated with a sub-cuticular stitch using absorbable suture. The skin is then covered with a sterile dressing for 24–48 h.

(b) Placement of a testicular prosthesis can be safely performed at the time of radical inguinal orchiectomy or in a delayed fashion depending on the clinical scenario. In children it may be preferable to delayed until patients reach puberty as this will allow for an appropriately sized/symmetric prosthesis to be placed. It is important to confirm that all sizes and spares of testicular prosthesis are readily available for use. Testicular prostheses are ideally placed via an inguinal incision. If the prosthesis is placed in a delayed fashion, the incision used for the radical orchiectomy can be used. After induction of anesthesia the patient should be placed in a supine position, prepped, and draped in the standard fashion. The patient should be prepped from umbilicus to mid-thigh (including the genitalia). Given the risk of infection it is recommended to administer perioperative antibiotics to cover skin flora, and to use antibiotic irrigation [13, 14]. The incision should be big enough to allow the delivery of the prosthesis with minimal skin contact. The subcutaneous tissue (Camper's and Scarpa's fascia) is dissected down to the external oblique fascia using blunt dissection or electrocautery. The neck of the scrotum is then dilated down towards the dependent portion of the scrotum with the goal of stretching the scrotum and identifying the most dependent point. This can be accomplished with Hegar dilators or large Pean clamp. Hemostasis is then ensured. Irrigation of the wound with warm antibiotic irrigant is recommended to uncover any bleeders. The most dependent portion of the scrotum is then invaginated out through the inguinal incision. A permanent suture is placed on the most dependent portion of the scrotum. An appropriately sized antibiotic soaked prosthesis is then anchored to the dependent portion of the scrotum. The prosthesis is then delivered though the inguinal incision down to the scrotum. Symmetry and dependent position of the scrotum is confirmed. The connection between the inguinal area and scrotum is then closed with a purse-string absorbable suture to prevent upward migration of the prosthesis. The subcutaneous tissue is approximated and the skin is approximated with a sub-cuticular stitch using absorbable suture. The skin is then covered with a sterile dressing for 24–48 h.

(c) Prior to performing a partial orchiectomy it is important to confirm laterality by repeating the physical exam and reviewing the preoperative imaging. A partial orchiectomy is approached in the same manner as a radical orchiectomy. However, the dissection of the spermatic cord can be performed distal to the external inguinal ring without the need to incise the external oblique fascia. The subcutaneous tissue should be dissected enough to allow for a radical orchiectomy if deemed necessary. Once the spermatic cord free, the testicle is delivered through the inguinal incision without excising the gubernaculum. The tunica vaginalis is then opened using electrocautery. Intraoperative ultrasound to identify small non-palpable lesions is highly recommended. A small gauge needle can be used on the operative field to localize the tumor under ultrasound guidance. The spermatic cord is then occluded using the 0.25 in. Penrose as a tourniquet. The tumor is excised along with a modest margin of uninvolved testicle. The tumor should then be sent for frozen pathological analysis. Any bleeding can be controlled with pinpoint cautery or suture ligation. The tunica albuguinea is then approximate using a running absorbable suture. The tourniquet is then released. Once the frozen pathology confirms benign or tumor with low malignant potential, hemostasis is ensured and the testicle is delivered back into the scrotum and pexed back into place with absorbable suture. If the pathology confirms a malignant tumor, then the external oblique fascia is opened to allow for dissection high into the inguinal canal and a "completion" radical orchiectomy is performed (see above). The subcutaneous tissue is approximated and the skin is approximated with a sub-cuticular stitch using absorbable suture. The skin is then covered with a sterile dressing for 24–48 h.

(d) Prior to performing a RPLND it is important to review the preoperative imaging to evaluate the retroperitoneum for suspicious lymph nodes or residual masses after chemotherapy, even those which may be outside the planned resection "template." An evaluation of vascular or other anatomic variations cannot be over emphasized. One should consider auxiliary procedures that may be required during the RPLND (e.g. nephrectomy, aorta/IVC reconstruction, and bowel resection) and discuss these with other surgical subspecialties. An RPLND, particularly a PC-RPLND, can often times require a multidisciplinary surgical team. For patients who have received chemotherapy it is also important to discuss with the anesthesiology team the fluid, oxygen, nasogastric tube, and central venous access considerations during the case, as well as the potential for the need of blood products.

After induction of anesthesia, the patient should be placed in the supine position, prepped (nipple to below pubic bone), and draped in the standard fashion. This procedure is commonly approached via a midline incision from xiphoid process to a few centimeters below the umbilicus, or if needed down to the pubic bone. This incision provides adequate exposure to the expected landing zone of testicular and para-testicular tumors. The subcutaneous tissue (Camper's and Scarpa's fascia) is dissected down to fascia using electrocautery. The preperitoneal space is then entered. An extraperitoneal approach can be done, but intraperitoneal approach is most commonly used. After entering the peritoneal cavity, the falciform is ligament is ligated and divided. A self-retraining retractor is then used to retract the abdominal wall. Depending on the side the RPLND will be performed, the ascending and descending colon are mobilized medially by incising the lateral attachments (white line of Toldt). The colon and small bowel mesentery is reflected medially to its root or as far as to the ligament of Treitz. The duodenum is reflected superomedially (Kocher maneuver). Bowel can be packed in the upper abdomen, over the chest, or contralateral abdominal cavity. Periodic inspection of bowel throughout the procedure is prudent.

There are several RPLND templates that can be used to determine the extent of dissection based on clinical scenario (Table 25.1). It is important to remove any suspicious lymph nodes or masses that one may encounter during the procedure. Margins of resection should not be compromised to maintain a template. Once the retroperitoneum is fully exposed, vital structures should be identified and retracted away from the area of dissection. If a nerve-sparing RPLND is planned, the sympathetic chain, post-ganglionic nerves, and hypogastric plexus should be identify, tagged with vessel loops, and preserved during the lymph node dissection. However, margin of resection should not be compromised to preserve these structures. Lymphatic tissue is dissected away from the abdominal vessels using the split and roll technique on the anterior surface of the vessels and dissecting the lymphatic tissue away from the aorta or IVC. Lymphatic channels should be ligated with small clips or silk suture particularly superior and posterior to the renal vessels. It is recommended to have small non-absorbable suture readily available to repair any vascular injuries encountered during the case. Dissection should be done meticulously to avoid damage to sympathetic nerves and lumbar vessels which penetrate posteriorly to the aorta and IVC. Lumbar vessels should be divided between silk ties to allow anterior reflection of the aorta and IVC and access to retroaortic and retrocaval lymph nodes. The residual abdominal spermatic cord should be identi-

Table 25.1 Retroperitoneal lymph node dissection templates

Template	Clinical indication	Unilateral border	Contralateral border	Superior border	Inferior border
Right modified template	Stage I disease	Unilateral ureter	Medial to aorta	Unilateral renal hilum	Diagonal line between the IMA to the crossing of the right ureter over right common iliac artery
Left modified template	Stage I disease	Unilateral ureter	Medial to IVC	Unilateral renal hilum	Diagonal line between the IMA to the crossing of the left ureter over right common iliac artery
Right bilateral modified template	Stage II disease	Unilateral ureter	Contralateral ureter down to IMA	Bilateral renal hilum	Diagonal line between the IMA to the crossing of the right ureter over right common iliac artery
Left bilateral modified template	Stage II disease	Unilateral ureter	Contralateral ureter down to IMA	Bilateral renal hilum	Diagonal line between the IMA to the crossing of the left ureter over right common iliac artery
Full bilateral template	Post-chemotherapy	Unilateral ureter	Contralateral ureter	Bilateral hilum	Crossing of the ureters over the common iliac artery bilaterally

IVC Inferior vena cava, *IMA* Inferior mesenteric artery fied at the internal inguinal ring and excised along with the ipsilateral gonadal vessels.

Once all suspicious lymphatic tissue is removed and no other suspicious lymph nodes are identified, the bowel is repositioned back into its normal anatomic configuration. It is recommended to run the bowel. The root of the small and large bowel mesentery can be approximated with absorbable suture to prevent volvulus. The ascending and descending colon can be retroperitonealized as well. The abdominal fascia is then closed with heavy interrupted or running absorbable suture. The subcutaneous tissue is approximated and the skin is approximated with a sub-cuticular stitch using absorbable suture. The skin is then covered with a sterile dressing for 24–48 h.

A PC-RPLND is performed similarly to a primary/nerve-sparing RPLND, but dissection of the lymphatic tissue can be more challenging due to the extensive desmoplastic reaction tumors have to chemotherapy. As a result, lymphatic tissue or residual masses can be extremely adherent to surrounding structures requiring en-bloc excision and extensive reconstruction.

Expected Postoperative Course

(a) After a radical inguinal orchiectomy patients are generally able to resume a regular diet shortly after recovering from anesthesia. Scheduled analgesia with non-narcotic medications (e.g. acetaminophen or non-steroidal anti-inflammatories) is recommended, and narcotics can be used for breakthrough pain. Antibiotics are typically not needed beyond surgery. Patients can typically be dismissed home after recovering from the anesthetic. Patients are recommended to avoid any strenuous physical activity for at least 4 weeks.
(b) The postoperative course after placement of testicular prosthesis is the same as a radical inguinal orchiectomy.
(c) The postoperative course after a partial orchiectomy is the same as a radical inguinal orchiectomy.
(d) The postoperative course after a RPLND can be widely variable. The extent of bowel mobilization, length of surgery, and auxiliary procedures performed can impact the postoperative course. Post-operative antibiotic prophylaxis is usually continued for less than 24 h [15]. Generally, after a primary/nerve-sparing RPLND patients are monitored on the general floor. Patients are kept NPO overnight, started on sips of clears by postoperative day 1, unrestricted clear liquids by postoperative day 2, and regular diet by postoperative day 3. Most patients are able to transition to home by postoperative day 4–6. For the PC-RPLND, the postoperative recovery can be slightly prolonged, and these patients may require overnight monitoring in the Intensive Care Unit.

Follow-Up

(a) The follow-up and subsequent evaluation after radical inguinal orchiectomy is dependent on the final pathology. When the likelihood of malignancy is high, obtaining preoperative staging imaging (chest, abdomen, and pelvis) should be coordinated prior to surgery in young children to avoid the need of a second anesthetic if central venous access will be needed for chemotherapy. Alternatively, waiting for final pathology report to proceed with further staging imaging, if necessary, will not significantly delay further patient care. The European Association of Urology (EAU) and the National Comprehensive Cancer Network (NCCN), Children's Oncology Group, and National Cancer Institute have developed comprehensive guidelines and recommendations for the treatment and follow-up of malignant testicular tumors [16, 17]. Given that most recurrences occur within the first 2 years, follow-up and surveillance imaging is frequent (every 2–3 months) during the first 2 years. Patients requiring additional therapies (RPLND, chemotherapy, or radiation therapy) should be referred to a medical oncologist or radiation oncologist. For benign tumors, follow-up can be at the discretion of the surgeon, but typically once 2–3 months after surgery seems prudent.

(b) Following placement of a testicular prosthesis, patients can be seen as per follow-up for radical inguinal orchiectomy. It is important to see the patient within 2–3 months after placement of prosthesis to assess the location of the prosthesis and patient satisfaction with it. Patients can follow-up per testicular tumor guidelines when indicated or as needed for issues with prosthesis.

(c) Much like a radical inguinal orchiectomy, follow-up after a partial orchiectomy will depend on final pathology. Generally it is recommended to have the patient follow-up in 2–3 months with a scrotal ultrasound. This is primarily to assess the viability of the testicle and establish a baseline ultrasound. Subsequent follow-up is based on guidelines or at the discretion of the surgeon.

(d) After a RPLND, the follow-up and need for additional therapies is dictated by the final pathology report. The EAU and NCCN guidelines should be followed closely after RPLND [16, 17]. If patient requires chemotherapy he should be referred to a medical oncologist. If surveillance is indicated, the guidelines dictate the frequency and studies needed for follow-up. Post RPLND surveillance is typically every 2–3 months for the first 2 years with imaging of the abdomen and pelvis annually.

Risk of Procedure

(a) Similar to open inguinal hernia repair in children, radical inguinal orchiectomy is associated with low complication rate. Similar to other inguinal procedures (hernia/hydrocele repair), most common complications include hematoma

(0.1–6.5 %) and wound infection (0.6–2 %) [12, 18, 19]. The ilioinguinal nerve is located within the inguinal canal, and damage or entrapment of it can lead to neuropathic pain. It is important to identify and protect this structure during radical inguinal orchiectomy. Despite this, up to 3 % of patients can experience chronic pain [20, 21]. In 238 adult patients who underwent unilateral radical orchiectomy without prosthesis, 25 % (60/238 patients) reported developing postoperative phantom pain at an average of 76 days [22].

(b) Issues relevant to placement of testicular prosthesis are infrequent but similar to those seen in radical inguinal orchiectomy. Other complications associated with testicular prosthesis include prosthesis malposition/migration, erosion, discomfort, and poor satisfaction. In 149 patients (76 children) device extrusion occurred in 2.6 %, with all extrusions occurring in prosthesis placed via a scrotal incision in pediatric patients [23]. Other minor complications reported include pain in 3 % and scrotal edema in 1.3 % [23]. The implications of wound infection are much more severe in the setting of a prosthesis, as this could lead to ex-plantation of the prosthesis. In a study of 86 patients who received a testicular prosthesis, no patient experience a complication, but 39 % felt that prosthesis was too high. A total of 15 % patients found the prosthesis bothersome during sexual activity, 5 % complained of inconvenience during physical exercise, and 10 % regretted the decision to have a prosthesis placed [24].

(c) Partial orchiectomy shares similar risk of complications to radical inguinal orchiectomy. Several studies have evaluated the use of testicular sparing surgery with complication rate of less than 6 % [25, 26]. Most common complications include testicular atrophy (2.5 %), hematoma (2 %), and infection (1 %) [26]. Additionally, there is the inherent risk that intraoperative assessment proves incorrect and either margins are inadequate or a malignant pathology is returned on final assessment, thus a later return to the operating room is needed to “complete” a radical orchiectomy.

(d) RPLND can be associated with significant intraoperative, perioperative, and long term complications. Much like postoperative care, the risk of complications is dependent on the extent of dissection, clinical scenario, addition of auxiliary procedures, and preoperative performance status. Primary RPLND is associated with intraoperative complication rate of 5 % with majority related to vascular injury followed by ureteral injury [27]. There is a less than 1 % risk of an unplanned auxiliary procedure during a primary RPLND, and less than 6 % risk of blood transfusion [27]. Early postoperative complication rate ranges between 10 and 24 % [27–29], with minor complications making up the majority of them. Ileus accounts for 0–18 %. Wound infection and deep vein thrombosis account for less than 1 %. Major complication can occur in up to 3 %. These include surgical exploration for small bowel obstruction (less than 1 %), chylous ascites (less than 2 %), and pulmonary embolism (1 %) [27, 29]. Late complications can occur in up to 7 % of patients, and these include incisional hernia (3.5 %), small bowel obstruction (1 %), and retroperitoneal fibrosis causing ureteral obstruction (1 %) [27]. Postop antegrade ejaculation with nerve-sparing RPLND can be preserved in over 90 % of patients [27, 29, 30]. Mortality after primary RPLND is much less than 0.1 %.

The reaction that tumors have to chemotherapy makes PC-RPLND a challenging procedure as the normal tissue planes are disturbed. This can be associated with an increased rate of complications and the need for additional (auxiliary) intraoperative procedures. The intraoperative complication rate during a PC-RPLND can be as high as 11 % along with a transfusion rate of 42 % [27]. Postoperative complications range between 3.7 and 32 % [31, 32]. Minor complications include ileus in 2.2–21 %, and wound infection in 0.3–3 % [31]. Major complications account primarily for chylous ascites in 3 %, pulmonary edema (bleomycin related) in 1 %, deep vein thrombosis in 1 %, and perioperative mortality of less than 1 % [31, 32]. Late complications can occur in up to 7 % of patients, and these include incisional hernia (4 %) [27]. Postop antegrade ejaculation after nerve-sparing PC-RPLND can be as high as 71–95 % [27, 33]. Risk of auxiliary procedures range between 24 and 45 % [31, 34–36]. Nephrectomy is the most common procedure, with rates between 7.3 and 14 % [31, 37]. Incidence of IVC resection reported in the literature ranges from 5 to 10 %, and aortic resection is less than 1.5 % [35, 38]. Mortality after PC-RPLND is extremely rare, and generally under 1 %.

Conclusion

Testicular and paratesticular tumors are rare and district entities with similar surgical management. It is important to continue to manage these tumors in centers with multidisciplinary expertise needed to adequately care for children and adolescents with these highly survivable tumors. Adherence to established guidelines for the management of these tumors is important.

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Part VIII
Trauma

Chapter 26
Genital Trauma

Vijaya Vemulakonda, David Chalmers, and Emily Serrell

Accidents and unintentional injuries are the leading cause of death in all children under the age of 14 [1], however, only 0.4–0.8 % involves injury to the external genitalia [2, 3]. This is largely due to the relatively protected anatomy of these structures [3]. Motor vehicle accidents, falls, straddle injury, sports injuries, and zippers are the most commonly associated mechanisms of injury [4, 5]. Recent literature regarding pediatric genital trauma includes identifying and treating sexual abuse injuries. In all cases of genital trauma, providers should be sensitive to signs or symptoms indicating sexual abuse. These may include but are not limited to laceration, bruising, abrasion, or scarring of labia, hymen, penis, scrotum, perianal tissue, or perineum or evidence of any sexually transmitted disease [6].

Female External Genital Trauma

Lay Description

Female external genital trauma includes any accidental or intentional injury to labia majora, labia minora, clitoris, hymen, vagina, and/ or urethra.

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Intended Benefit

Following trauma to external female genitalia, examination under anesthesia (EUA) or operative assessment/management occurs in between 5 and 20 % of cases [2, 3, 7]. The benefit of EUA is to allow a thorough assessment and recognize subtle injuries that may be missed with an unsedated physical exam, particularly if the clinician suspects sexual abuse. Proper identification aids in the avoidance of rectovaginal fistula, vesicovaginal fistula, urethral stricture, infection, adhesions, labial fusion, vaginal stenosis, anal laxity, incontinence/constipation, or other long-term complications [8]. Operative repair may improve future cosmesis, sensation, sexual function, and continence of urine or feces.

Management

Accidental genital trauma in young occurs along a spectrum of simple contusion to laceration and penetrating injury. Straddle-type injuries, commonly from monkey bars or bicycles, represent 70–80 % of these traumas and are unlikely to require operative intervention. Injuries that are penetrating or are above the labia are more likely to require more invasive assessment under anesthesia [3, 7]. Between 10 and 20 % of genital trauma assessed in an emergency departments involve the genitourinary or lower urinary tract, and these most commonly occur in cases of penetrating injury or pelvic fracture [9, 10].

Bedside physical exam may reveal labial bruising or bleeding at the vaginal introitus. Vulvar edema or hematuria may also be appreciated. Due to the proximity of the urethra and vagina, traumatic urethral injuries in girls should prompt an evaluation for associated vaginal injuries, and vice versa. Evaluation of external genital trauma should always include assessment of the urethra, vagina, rectum and pelvic stability for associated injuries [7]. Patients with pelvic fractures should always be evaluated for urethral injury. Examination in the emergency room can misidentify or underestimate the extent of injuries. Some institutions utilize urogenital trauma grading systems [9], while other studies recommend examination under anesthesia in all cases of blunt urogenital trauma [8]. There should be a low threshold for performing EUA. Relative indications include a young or uncooperative patient, inability to see the full extent of the injury, significant pain or fright, vaginal hemorrhage, expanding hematoma, or other concomitant severe injuries [8].

The majority of female genital trauma can be managed non-operatively with NSAIDs, sitzbath, and activity limitation during the first 1–2 days. Those that require further evaluation may require EUA with cystoscopy, vaginoscopy, anoscopy, sigmoidoscopy, or a combination of these modalities. Operative management is required in about 10–20 % of cases [7, 11, 12]. Any wound requiring debridement, hemostasis, or suturing should be performed under general anesthesia in the dorsal lithotomy position. Superficial laceration may be addressed with primary closure with a single layer of absorbable sutures. For extensive vaginal laceration, absorbable sutures should be placed deep to repair the perineal body and then more superficially in layers. External rectal sphincter or laceration through the rectal mucosa may require reconstruction. For trauma that extends into the rectum, a primary repair with temporary diverting colostomy may be required [13]. Foley catheters may be necessary for laceration repairs close to the urethra.

In cases of suspected urethral injury, retrograde urethrography should be the initial study for diagnosis. In the acute setting, there is controversy about whether preferred management is to immediately realign urethra endoscopically or to delay repair and manage conservatively. In adults, primary realignment has a high failure and complication rate [14]. In children, this is not as clear, particularly in girls, who may respond well to primary repair and experience higher complications of urethral stricture of vaginal stenosis if surgery is delayed [15–17].

Post-operative Expected Course

The majority of patients recover quickly and without complications following repair of genital injuries. Length of stay depends on the severity of injury. Some patients may be released on the same day as surgery, while others may require trial of void, pain control, or progression of diet back to solids. Given that the extent of genitourinary and anorectal injuries is frequently unknown when the decision is made to assess a patient under anesthesia or operatively manage her, perioperative antibiotics should be used to reduce the risk of secondary infection and wound dehiscence.

Pain management is initiated before surgery is complete with injection of local anesthetic at the surgical incisions. Most children's pain will be managed well by alternating Acetaminophen (Tylenol) and Ibuprofen (Motrin) for the first 24–48 h. Diet following surgery should be normal as tolerated. Sutures are absorbable and gauze bandage may be used over operative site. Wounds should be cleaned twice daily for the first 2 days, though children should avoid full water submersion during this time. Thereafter there are no restrictions on bathing or showering, and wounds may be cleaned and then covered with topical antibiotic ointment at these times.

Full recovery to normal activity is expected within a few days to weeks, and children will self-regulate their activity according to pain and energy. Typical recommendations include limiting straddle activities for 6 weeks. In general, some discomfort, redness, crusting, bruising, and swelling is normal, but worsening of these features or fever >101.3 °F requires follow-up with the surgeon or other healthcare provider. If a colostomy is necessary due to extent of anogenital injury, reversal surgery may be performed 2–3 months after the first operation.

Complications

Because pediatric genital trauma necessitating operative repair is uncommon with significant variability, it is difficult to predict complications rates. Furthermore, it may not be possible to assess what complications originate from the trauma as opposed to the surgery itself. Fortunately, the majority of studies assessing identification and management of pediatric genital trauma report few complications [10, 11, 18, 19]. Finally, it is important to recognize that many complications may present years after the primary repair, so long term follow-up is essential to rule out post-pubertal development or identification of some of these complications.

Infection: <5 %

General surgical wound classification would estimate a 5 % infection risk for external, lacerating trauma of female genitalia. If the wound extends into the gastrointestinal or genitourinary system, infection risk increases to 10 % [20]. This rate is similar to the 0–10 % rates published by several studies, as a single 116-patient evaluation reported a 10 % infection rate while another 91 girl study evaluating straddle-injuries reported one abscess [9, 10]. Only one study reported sepsis as a complication, with an incidence of 3 out of 116 children [9]. All cases of genital or genitourinary trauma also necessitate assessment of anorectal injury, as this can increase likelihood of infection or wound dehiscence.

Hematoma: <1 %

Following genital trauma, extravasation of blood may occur in the labia, along the vagina mons, or in the clitoral area creating hematoma that may persist for several weeks. Hematoma may be painful and cause functional urine retention or outright urethral obstruction requiring a urinary catheter. McCann et al. assessed 239 girls and described the types of injuries and times of resolution, outlining this general timeline of the healing: abrasions 3 days; edema 5 days; ecchymoses 2–18 days; labial hematoma 2–4 weeks; petechiae 1 day; blood blisters 24–30 days; superficial lacerations 2 days; deep lacerations 20 days [19]. The incidence of hematoma is less than 1 % in two retrospective studies assessing 271 girl [10, 19]. A single Austrian study reported hematomas in 16 % of their 91 patients [10]. Despite this disparity in rate of occurrence, all hematomas in all three studies resolved spontaneously by about 2 weeks. Related to hematoma, petechiae occurred in 65 % of prepubertal girls and 25 % of pubertal girls at their initial examination and had resolved within 24 h [19].

Urethral Stricture: <0.1 %

Trauma to the urethra is uncommon in girls due to its short course and protected positioning behind the bony arch of the pubis. The majority of injuries do not involve the urethra, so stricture is not reported as a complication in many studies [8–11, 18]. This complication is primarily a concern for children with pelvic fractures or direct transection with lacerating injury. In a review of 17 series of pediatric female patients with fractured pelvis and urethral rupture, nine cases of urethral stricture were reported with all females ultimately continent following either observation or operative management [15]. In girls, partial disruption or clean transection of the urethra is best repaired acutely with primary repair [15–17]. Conversely, in one small study, delayed repair with suprapubic cystostomy in acute presentation was associated with urethral stricture in four of five [15].

Fistula: <0.1 %

Acquired rectourethral, rectovaginal, urethrocutaneous, and vesicovaginal fistulae in pediatric patients may occur following severe trauma or the operative repair of that trauma. The mechanism appears to be a combination of pelvic fibrosis and adhesions creating weak areas that are susceptible to high pressure and form tracts between the rectum, urethra, and/ or vagina. The actual rate of fistula development is difficult to estimate, as most studies examine only individual or very few cases [9, 16, 21, 22]. The mechanism of injury is also important, as acquired fistulae have been reported to form after pelvic osteomyelitis [23] and pelvic fracture that may or may not involve genital trauma [22]. In seven girls suffering from "vaginal rupture" following pelvic fracture, delayed urethral reconstruction required simultaneous operative management of five urethra-vaginal fistula and four cases of vaginal stenosis [16]. In a review of 17 series of pediatric female patients with fractured pelvis and urethral rupture, 11 cases of fistula were reported with all females ultimately continent following either observation or operative management [15].

Vaginal Stenosis: <0.1 %

As a traumatic injury to the vagina heals, fibrotic stenosis and/or heterotopic ossification may occur about 3–5 cm deep to the hymen in the vagina. This may result in vaginal stenosis or stricture or in a transverse scarring that creates an outlet obstruction from the vagina or uterus that may be identified in adolescent or young adult women as hematocolpos or hematometra, respectively. Most cases reviewing vaginal stenosis occur after severe pelvic fractures through the pubic rami or symphysis, ileum, or ischium with associated genital trauma [16, 24, 25].

Sexual Dysfunction: <0.1 %

There is little data regarding adult sexual function and fertility rates in women who experienced genital or pelvic trauma as children due to the length of time between trauma and recognition of sexual dysfunction. There is, however, a wealth of information about intentional female genital mutilation performed on children that results in high incidence of infection, dyspareunia (RR 1.5), dysmenorrhea (RR 1.44), and urinary dysfunction (RR 2.6) [26]. Though this is not directly related to accidental trauma, it does reflect that a type of injury—damage to the vaginal or clitoral vasculature, nerves, or structures—will result in longterm sexual dysfunction. Further, up to 40 % of adult women complain of sexual dysfunction following a fractured pelvis, tough this depends on the severity, type, and pattern of fracture [27].

Testicular and Scrotal Trauma

Lay Description

Injury to the testicle, epididymis, and/or scrotum.

Procedures

Scrotal exploration, vasoepididymostomy (connection of vas deferens and epididymis), vasovasostomy (connection of vas deferens).

Intended Benefit

Testicular repair will minimize pain, swelling, bleeding or hematoma. In cases of severe trauma, operative treatment may help to preserve injured, torsed, or ruptured testis and its function of sperm and androgen production.

Management

The mechanism and severity of damage will determine management of the injured testis. All cases of testicular trauma must first exclude injury to adjacent structures or vasculature. Any patient who cannot be fully and properly examined should be examined under general anesthesia. Severe cases, as defined by Table 26.1, will in most cases require surgical intervention. As a rule of thumb, any injury that involves interruption of the tunica albuginea or penetration through dartos fascia will require operative intervention.

If an injury warrants surgery, this is done under general anesthesia. The patient is placed in supine position. A small scrotal skin-incision is used to access and

Table 26.1 Categories of testicle/scrotum damage and management

Injury type	Signs and symptoms	Management
Mild blunt	Minimal pain or scrotal swelling or ecchymosis; intact scrotum	Expectant management: bed rest, ice packs, supportive underwear, NSAID medication for pain management. Follow up in 48 h with primary care provider
Moderate blunt	Moderate pain or scrotal swelling or ecchymosis; superficial skin laceration	Testicular ultrasound. If normal, follow up in 48 h. Thorough washing and closure of skin wound with absorbable suture repair if warranted
Severe blunt	Severe testicular pain or scrotal swelling; intractable pain associated with nausea or vomiting; scrotal avulsion; presentation concerning for testicular rupture, fracture, torsion, dislocation, or avulsion; hematocele or hematoma with testicular compression	Testicular ultrasound Surgical exploration and repair under general anesthesia per surgeon discretion
Mild penetrating	Superficial penetration with preservation of dartos fascia	Cleansing and debridement of wound. Absorbable suture if warranted. Expectant management
Severe penetrating	Penetration through dartos fascia	Surgical exploration and repair under general anesthesia. Intra-operative wound culture depending on mechanism and severity. Antibiotics. Verification of up-to-date tetanus immunization

Table adapted from recommendation from Husmann in Campbell-Walsh Urology [28]

inspect the testicle and distal spermatic cord. Interruption of the tunica albuginea will be repaired with absorbable suture, followed by reapproximation of the dartos fascia and skin. In rare cases if the testicle cannot be salvaged or bleeding adequately controlled, orchiectomy may be performed.

Bite wounds are best treated by debridement and copious irrigation, which lowers the risk of infection from 59 to 12 % [29]. Prophylactic antibiotic use does not decrease the rate of wound-infections in cat or dog bites but does decrease the infectivity of human bites [30]. However, given the environment of a genital wound in a young child's diaper, prophylactic treatment is warranted with augmentin, clindamycin, or moxifloxacin with a particular goal of covering for *Pasteurella* species in dogs and cats, as well as *Bartonella henselae* in cats [30–32].

Postoperative Expected Course

Repair of severe penile trauma is frequently performed on an inpatient basis with an expected discharge within 1–2 days, although this is dependent on any additional injuries. Perioperative antibiotics are administered to prevent infection with *S. aureus* and enteric gram-negative bacilli. For cases of penetrating trauma, environmental bacilli may be introduced. Broad spectrum antibiotics and verification of up-to-date tetanus immunization is particularly important.

Full recovery to normal activity is expected within a few days to weeks, and children will self-regulate their activity according to pain and energy. Avoiding rigorous activity and straddle activities is surgeon dependent according to the severity of injury. Diet following surgery should be normal as tolerated. Pain management is initiated with local anesthetic following surgical intervention. Most children's pain will be subsequently well-managed by alternating Acetaminophen (Tylenol) and Ibuprofen (Motrin) for the first 24–48 h. Narcotic medications may be appropriate for older children.

Sutures are absorbable and gauze bandage may be used over the incision sites. Children should avoid full water submersion for at least 48 h but then have no restriction on bathing or showering. Antibiotic ointment should be administered for 1 week following superficial skin lacerations. Bruising and swelling of the scrotum may be severe and last several week.

Follow Up

Follow-up timing and frequency should be per surgeon's discretion. If there are signs of infection within 2 weeks of surgery, the patient should be seen in the office as soon as possible. Given the sensitive location and patient/family anxiety associated with genital trauma, families may prefer close follow up. In addition, when a traumatic event impacts the testicle, an ultrasound may be performed the subsequent months to confirm revasculatization and viability. Further follow-up depends on the specific clinical picture with an attempt to provide maximal reassurance to the family.

Complications

Complication rates associated with operative management of genital trauma largely depend on the mechanism of injury. More severe injuries are associated with greater risk of complications due to tissue contamination, destruction, and associated injuries [9]. Most studies are reported as small cohorts or case series. It should be noted that this lends to selection and publication bias, as failed cases are unlikely to be submitted or published.

Complications that should be reviewed with patients, although no specific incidence can be determined include scrotal edema (high); hematoma or hematocele (high if testicular fracture); and abscess (low, even in cases of lacerating trauma).

Infection: <5 %

Genital wound repairs tends to be superficial and so are classified as "clean" with low risk for contamination. However, penetrating or severely traumatic genital injuries have a high incidence of exposure to environmental, genitourinary, or gastrointestinal flora. As such, surgical repair should be considered "clean contaminated" with an associated <10 % risk of infection [20]. A retrospective review of 116 children with genital trauma and anorectal injury cited 10 % infection, 5 % wound dehiscence, and 3 % sepsis incidence, with complications correlating with severity of injury [9]. Alternatively, two studies evaluating 74 boys and 116 patients of mixed age with genital injuries that did not involve the anus/ rectum cited <1.5 % infection rates [33, 34]. This discrepancy likely reflects that wounds with lower morbidity are associated with fewer complications.

Cases of scrotal wounds caused by animal (dog, horse, donkey, pig) bites are rare but require particular attention to the patient's tetanus status as well as dog's rabies immunization status, if applicable [31, 35]. As there are only one to three cases of rabies annually in the United States, and 90 % of rabies is carried by wild animals, infection is exceedingly rare [36].

Shock: <0.1 %

There has been a single reported case of non-infectious SIRS and shock following a kick to the groin in an adolescent boy. The mechanism is thought to be that compression of the testicle against the pelvis caused compression and damage to induce a cytokine storm, and the cytokines (ex. Tumor necrosis factor alpha, interleukin 1) brought on a systemic response with pressor-requiring hypotension and a left-shifted leukocystosis [37].

Testicular Loss or Atrophy: Dependent on Intervention

Immediate (within 4–12 h) surgical intervention for patients presenting with severe testicular or scrotal trauma is associated with better outcomes, likely due to decreasing duration of ischemia. One study demonstrates that in cases of acute scrotum, testicular salvage could be attained in 100 % of cases if surgery occurred <6 h, 50 %

if 6–12 h, and 4 % after 12 h [38]. Another demonstrated that testicular rupture has a salvage rate of about 80–90 % if repaired <72 h or only 32–45 % if >72 h [39]. It should be noted that conservative management in late-presenting cases of testicular rupture may have equivalent outcomes to surgical intervention, though children were forced to decrease activity for longer with conservative approach [40].

This association between improved outcome and shorter duration of symptoms is also associated with fewer incidents of testicular atrophy. There is a rate of about 5 % if surgery is performed <12 h in testicular torsion [41] and up to 20 % if testicular atrophy is managed conservatively [39].

Decreased Fertility: Unknown, <0.1 %

Fertility in children who experience genital trauma is difficult to assess given the time between the injury and identification of infertility. Further, most fertility studies are completed with adults and animals, so the clinical implication of those studies is difficult to correlate with pediatric issues.

As written above, testicular salvage is maximized by repairing damage quickly. This rule seems to hold for fertility. A seven-boy study demonstrated normospermia after immediate repair of testicle rupture, though they were not necessarily followed into adulthood to assess for procreative capacity [42]. About 50 % of men with unilateral testis (predominantly from childhood etiology) will have a sperm count of less than 20 million/ml, though other studies indicate that semen volume and motility may remain normal [43–45]. Finally, a ten-adult review demonstrated that surgical salvage instead of orchiectomy of testis is associated with fewer endocrine and seminal abnormalities [45].

The longer-term effects that may impact fertility include endocrine changes, with testicular trauma associated with higher estradiol (inhibits GnRH) and decreased inhibin B (a correlator of spermatogenesis, testicular volume, and sperm counts) [46–50]. Another potential mechanism is an autoimmune response, with disruption of the spermatic pathway or blood-testis barrier exposing spermatic antigens to a naïve immune system. There may be a subsequent immunologic response with creation of antisperm antibodies that affect bilateral testes and immobilize sperm [45, 51–55]. Finally, testicular torsion is associated with vasoconstriction of the contralateral testis, which may cause subsequent long-term damage; this theory certainly could be applied to general testicular trauma [56].

Penile and Urethral Trauma

Lay Description

- Injury to the penis and/or penile urethra.

Intended Benefit

Trauma to the penis, scrotum, or testicles may be benign or serious depending on the extent of injury. The benefits of treating injury to the penis or urethra is to minimize pain, swelling, and hematoma; maintain cosmesis; preserve urethral integrity and continence; preserve erectile function and normal sensation.

Management

The mechanism and severity of penile trauma will determine management and operative treatment. Injuries in the neonate are commonly iatrogenic from circumcision. Toddler aged children are likely to experience trauma due to toilet seat injuries and zipper entrapment while school aged children injure genitalia with bicycling, kicks, falls, or sports related injuries [57]. Penile contusions and lacerations are usually mild and self-limiting, although amputations have been reported from assault or animal bites [58, 59]. Finally, blunt perineal trauma is the most common etiology for high-flow priapism secondary to a traumatic arteriovenous fistula.

Penile skin loss or dehiscence following neonatal circumcision is typically managed conservatively with antibiotic ointment and healing by secondary intention. Skin grafting may be considered in the immediate post-injury period.

Superficial skin injuries can be managed with wound irrigation with saline and application of topical antibiotic ointments. More severe lacerations that are not infected may require debridement of devitalized tissue and skin re-approximation with absorbable sutures utilizing local or general anesthesia. Zipper injuries to the penis more commonly occur in uncircumcised boys and may be treated with mineral oil to slip the trapped skin from the zipper or by cutting of the median bar of the zipper with bone cutters. Bites require verification of immunization status, including tetanus in the patient and rabies in a dog. Amputation injuries may be managed with primary re-anastamosis, with best results occurring within 8 h after injury.

Patients with penile trauma may have concurrent injury to urethra in 15–22 % of cases [34]. Injuries to the male urethra are divided into anterior (penile or bulbar) and posterior (prostatic and membranous). If there is concern for urethral injury, a retrograde urethrogram should be performed. Urethral lacerations may be immediately and primarily realigned, while blunt injuries are better treated with suprapubic drainage. There is some controversy regarding immediate vs. late repair, but current evidence indicates that it is the severity of injury and not treatment modality that determines later erectile dysfunction and urinary incontinence [28, 60].

Injuries to major vasculature can result in high-flow, non-ischemic priapism. This may be managed expectantly and with perineal compression for 6 weeks. If it does not spontaneously resolve, super-selective embolization and then surgical fistula-ligation may be performed, though the latter only in refractory cases due to increased risk of erectile dysfunction.

Postoperative Expected Course

Repair of severe penile trauma is frequently performed on an inpatient basis with an expected discharge within 1–2 days, although this is dependent on any additional injuries. Perioperative antibiotics are administered to prevent infection with *S. aureus* and enteric gram-negative bacilli. For cases of penetrating trauma, environmental bacilli may be introduced. Broad spectrum antibiotics and verification of up-to-date tetanus immunization is particularly important.

Full recovery to normal activity is expected within a few days to weeks, and children will self-regulate their activity according to pain and energy. Avoiding rigorous activity and straddle activities is surgeon dependent according to the severity of injury. Diet following surgery should be normal as tolerated. Pain management is initiated with local anesthetic following surgical intervention. Most children's pain will be subsequently well-managed by alternating Acetaminophen (Tylenol) and Ibuprofen (Motrin) for the first 24–48 h. Narcotic medications may be appropriate for older children.

Sutures are absorbable and gauze bandage may be used over the incision sites. Children should avoid full water submersion for at least 48 h but then have no restriction on bathing or showering. Antibiotic ointment should be administered for 1 week following superficial skin lacerations. More significant injuries like penile amputation will have a more prolonged recovery period. Erectile dysfunction and atrophy of the glans may occur but improve over months to years. Priapism may resolve with conservative measures or operative management as described above. Bruising, swelling should resolve within a few weeks if a procedure is required.

Follow Up

Follow-up timing and frequency are per surgeon's discretion. If there are signs of infection within 2 weeks of surgery, the patient should be seen in the office as soon as possible. Given the sensitive location and patient/family anxiety associated with genital trauma, earlier follow up can be conseridered. Further follow-up depends on the specific clinical picture with an attempt to provide maximal reassurance to the family.

Complications

Infection: <5 %

Genital wound repairs tend to be superficial and so are classified as "clean" with low risk for contamination. However, penetrating or severely traumatic genital injuries have a high incidence of exposure to environmental, genitourinary, or gastrointestinal flora. As such, surgical repair should be considered "clean contaminated" with an associated <10 % risk of infection [20]. A 116-child genital trauma and anorectal injury retrospective review cited 10 % infection, 5 % wound dehiscence, and 3 % sepsis incidence, with complications correlating with severity of injury [9]. See the "Scrotal and Testicular Trauma" section for a description of infection rates due to bite wounds.

Penile Amputation or Need for Reconstructive Surgery: <0.1 %

Studies of direct trauma to the penis are largely case reports of boys with varying degrees of severity, including skin or meatal injury, partial glanular amputation, and total amputation of the penis as a consequence of assault or circumcision complication. A 4 year old male with total penile amputation and replantation with microanastamosis within 6 h of an assault reported early complications of necrotized skin and progressive penile shortening, while longterm results were straight penis without deviation, intact penile sensation, full continence, no urethral stricture [61]. Other case reports and retrospective analyses of patients note similar excellent results [62–64] even with ischemia time up to 18 h [65].

Microanastamosis, the rejoining of corporal and dorsal arteries and veins, is superior to macroscopic repair with reanastamosis of the spongy tissue and fascia only [58, 64, 66]. Complications of penile amputation include skin necrosis, venous congestion, urethral fistula, urethral stricture, poor sensation, or absent/incomplete erection [58].

Historically, the families of male infants with inadequate genitalia secondary to trauma or congenital development were faced with a decision of gender reassignment. Fortunately, in more contemporary cases of severe penile trauma that could not be immediately addressed, phallic reconstruction with the suspensory ligament or corpora cavernosa have been reported with good cosmetic results and ability to achieve erection [59, 67].

Priapism: <0.1 %

Priapism is a prolonged penile erection lasting >4 h and is divided into three types: low-flow (ischemic), high-flow (non-ischemic), and stuttering (recurrent, self-limited). An estimated 1.5 per 100,000 cases of priapism occur annually in the general population, and there is an even lower, unknown incidence in children [68]. In the pediatric population, trauma accounts for 10 % of cases of priapism [69].

Low-flow priapism is a urologic emergency that leads to acute ischemia of the corpora cavernosa, while high-flow is non-ischemic but can lead to long-term fibrosis of the corpora cavernosa and subsequent disfigurement and erectile dysfunction [70–72]. High-flow priapism is most frequently caused by trauma that results in an arteriosinusoidal fistula, permitting arterial blood to flow into the sinusoids and causing a well-oxygenated and thus non-ischemic high-flow, prolonged, and painless erection. Typically, a boy presents with a few hours or days of firm but not fully engorged erection that descends when the perineum is compressed (Piesis sign). A trauma to the perineum or penis may have occurred minutes or days prior to the onset of priapism [73–75].

Initial conservative, 6-week "wait and watch" approach may be appropriate if the priapism is asymptomatic [71, 73]. However, there should be a low threshold for radiological embolization of damaged vessels with autologous clot or microcoils. In a review of 52 case studies, none reported on long-term sexual function or fertility, though many families reported successful erectile response judged by nocturnal erections [70]. In adult studies, men maintained sexual function in 80–100 % of high-flow priapism cases treated with embolization [71]. In the remainder of unresolved high-flow priapism, surgical ligation of the internal pudendal cavernosal arteries may be performed, though has a higher incidence of cavernosal scarring and erectile dysfunction [71, 76].

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Chapter 27
Urinary Tract Trauma

W. Robert DeFoor Jr. and Eugene Minevich

Major Renal Injury

Procedure(s)

Trauma nephrectomy, renorrhaphy, and partial nephrectomy

Lay Description

An operation to remove all or part of a severely damaged kidney due to a traumatic accident

Benefits

- Remove a damaged kidney or part of a damaged kidney
- Prevent life-threatening and/or ongoing blood loss
- Prevent leakage of urine in the abdomen
- Reduce risk of chronic high blood pressure
- Reduce risk of recurrent urinary tract infections

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Risks

- Death
- Anesthesia
- Bleeding (acute or delayed)
- Hematoma
- Intra-abdominal infection
- Wound infection
- Sepsis
- Urinary tract infection
- Urosepsis
- Urinary extravasation with partial nephrectomy
- Acute kidney injury
- End stage renal disease if solitary kidney removed
- Hypertension
- Damage to adjacent structures
- Subsequent procedure to remove ureteral stent if necessary
- Delayed diagnosis of concomitant unrecognized injuries
- Incisional hernia

Technical Points

- Thorough staging is mandatory prior to surgery in a stable patient. In an emergency situation with an unstable patient, a "one-shot IVP" can be helpful to assess contralateral renal anatomy and function.
- General anesthesia
- A midline abdominal incision (if concern for other injuries) or flank incision (if isolated injury or delayed procedure) can be performed.
- When performing a trauma exploratory laparotomy, an initial medial dissection is recommended to expose and control the hilar vessels.
- The decision is made intra-operatively to perform a total or partial nephrectomy depending on the location and severity of the injuries as well as the stability of the patient.

An externalized abdominal drain (Jackson Pratt) may be used with a partial nephrectomy to reduce the risk of post-operative urinoma.

Post-operative Expected Course

- The decision to recover the patient in the intensive care unit or the inpatient surgical ward is dependent on the patient's overall clinical status and hemodynamic stability.
- The time of convalescence in the hospital is typically dependent on the severity of any associated injuries and the overall clinical status.
- The eating and drinking status is dependent on the clinical stability as well as the severity of any associated bowel injuries.

- Analgesia can be provided by oral or intravenous narcotics or intravenous patient controlled analgesia (PCA). An epidural is very unlikely to be performed in an urgent trauma situation (particularly with hemodynamic instability) but may be considered in a delayed elective procedure.
- Peri-operative antibiotics can be discontinued 24 h after the procedure unless there are signs and symptoms of an active infection. With concomitant bowel injuries or significant urinary extravasation, antibiotic use may be deferred to the discretion of the attending surgeon.
- Discharge is dependent on the severity of concomitant injuries and the overall course of convalescence. In general, discharge is considered when the patient is tolerating a regular diet, having regular bowel and bladder emptying, and is comfortable on oral analgesics.

Major Ureteral Injury

Procedure(s)

Ureteral repair, ureteral reimplantation

Lay Description

An operation to repair a damaged ureter due to a traumatic accident including reimplantation of the distal ureter into the bladder

Benefits

- Repair an injured ureter
- Prevent leakage of urine in the abdomen
- Reduce risk of damage to the kidneys

Risks

- Death
- Anesthesia
- Bleeding (acute or delayed)
- Hematoma
- Intra-abdominal infection
- Wound infection
- Sepsis

- Urinary tract infection
- Urosepsis
- Urinary extravasation
- Acute kidney injury
- End stage renal disease if solitary kidney becomes obstructed by stricture/stenosis
- Hypertension
- Damage to adjacent structures
- Subsequent procedure to remove ureteral stent if necessary
- Delayed diagnosis of concomitant unrecognized injuries
- Incisional hernia

Technical Points

- General anesthesia
- A midline abdominal incision will likely be performed for an emergency exploratory laparotomy.
- A spatulated end to end ureteral anastomosis over a double J ureteral stent is preferable if the length of the ureteral injury is short. Injuries to the distal ureter require ureteral reimplantation.
- For more complex injuries, ureteral substitution options in an emergency setting are limited. Placing a nephrostomy tube to temporarily divert the urine with a delayed elective repair is an option. A ureterostomy may also be an option in rare cases.
- An externalized abdominal drain (Jackson Pratt) may be used to reduce the risk of post-operative urinoma.

Post-operative Expected Course

- The decision to recover the patient in the intensive care unit or the inpatient surgical ward is dependent on the patient's overall clinical status and hemodynamic stability.
- The time of convalescence in the hospital is typically dependent on the severity of any associated injuries and the overall clinical status.
- The eating and drinking status is dependent on the clinical stability as well as the severity of any associated bowel injuries.
- Analgesia can be provided by oral or intravenous narcotics or intravenous patient controlled analgesia (PCA). An epidural is contraindicated in an emergency trauma situation (particularly with hemodynamic instability) but may be considered in a delayed elective procedure.
- Peri-operative antibiotics can be discontinued 24 h after the procedure unless there are signs and symptoms of an active infection. With concomitant bowel injuries or significant urinary extravasation, antibiotic use may be deferred to the discretion of the attending surgeon.

- Discharge is dependent on the severity of concomitant injuries and the overall course of convalescence. In general, discharge is considered when the patient is tolerating a regular diet, having regular bowel and bladder emptying, and is comfortable on oral analgesics.

Major Bladder Injury

Procedure(s)

Bladder repair

Lay Description

An operation to repair a damaged bladder due to a traumatic accident

Benefits

- Repair an injured bladder
- Prevent leakage of urine in the abdomen
- Reduce risk of peritonitis

Risks

- Death
- Anesthesia
- Bleeding (acute or delayed)
- Hematoma
- Intra-abdominal infection
- Wound infection
- Sepsis
- Urinary tract infection
- Urosepsis
- Urinary extravasation
- Acute kidney injury
- Hypertension
- Damage to adjacent structures
- Subsequent procedure to remove ureteral stent or suprapubic tube if necessary

- Delayed diagnosis of concomitant unrecognized injuries
- Incisional hernia

Technical Points

- General anesthesia
- A midline abdominal incision will likely be performed for an emergency exploratory laparotomy.
- Consider a suprapubic tube as well as a urethral catheter to maximally divert the urine post-operatively.
- An externalized abdominal drain (Jackson Pratt) may be used to reduce the risk of post-operative urinoma.

Post-operative Expected Course

- The decision to recover the patient in the intensive care unit or the inpatient surgical ward is dependent on the patient's overall clinical status and hemodynamic stability.
- The time of convalescence in the hospital is typically dependent on the severity of any associated injuries and the overall clinical status.
- The eating and drinking status is dependent on the clinical stability as well as the severity of any associated bowel injuries.
- Analgesia can be provided by oral or intravenous narcotics or intravenous patient controlled analgesia (PCA). An epidural is contraindicated in an emergency trauma situation (particularly with hemodynamic instability) but may be considered in a delayed elective procedure.
- Peri-operative antibiotics can be discontinued 24 h after the procedure unless there are signs and symptoms of an active infection. With concomitant bowel injuries or significant urinary extravasation, antibiotic use may be deferred to the discretion of the attending surgeon.
- Discharge is dependent on the severity of concomitant injuries and the overall course of convalescence. In general, discharge is considered when the patient is tolerating a regular diet, having regular bowel and bladder emptying, and is comfortable on oral analgesics.

Renal Injury with Urinary Leak

Procedures

- Cystoscopy and ureteral stent placement

Lay Description

- An operation to place a stent in an injured kidney that has a urine leak due to a traumatic accident

Benefits

- Divert the urine to allow a renal leak to heal spontaneously
- Prevent leakage of urine in the abdomen
- Reduce risk of peritonitis and urosepsis
- Help stabilize renal function

Risks

- Death
- Anesthesia
- Bleeding (acute or delayed)
- Hematoma
- Intra-abdominal infection
- Inadvertent placement of stent outside collecting system
- Injury to urinary tract (bladder, ureter, kidney)
- Sepsis
- Urinary tract infection
- Urosepsis
- Urinary extravasation
- Acute kidney injury
- Hypertension
- Damage to adjacent structures
- Subsequent procedure to remove ureteral stent
- Delayed diagnosis of concomitant unrecognized injuries

Technical Points

- General anesthesia
- Perform retrograde pyelogram to delineate collecting system and find appropriate target for upper curl of stent (see Figs. 27.1 and 27.2)
- Leave urethral catheter to maximally drain urine
- Follow urinoma with serial ultrasound imaging
- Consider percutaneous drainage of urinoma outside kidney if signs of sepsis
- If stent unable to be placed then consider percutaneous nephrostomy

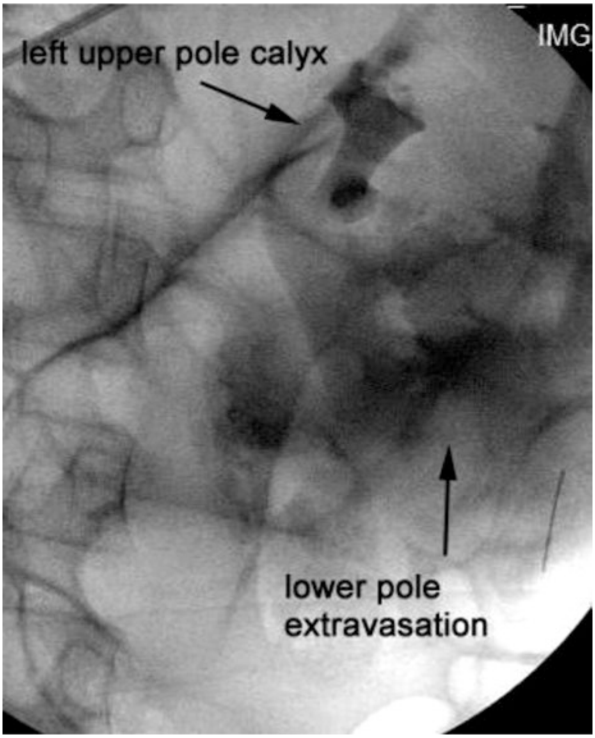

Fig. 27.1 Retrograde pyelogram of 14 year old boy with left grade IV renal injury from a fall showing a preserved upper pole calyx and lower pole urinary extravasation

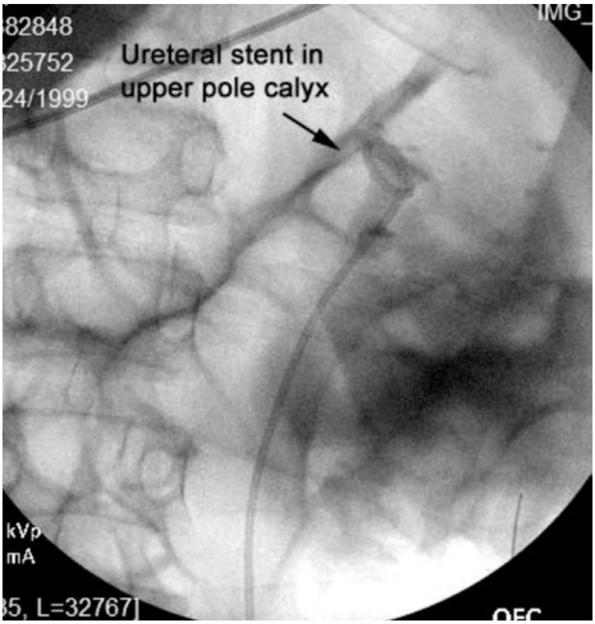

Fig. 27.2 Successful stent placement in an uninjured upper pole calyx in a grade IV renal injury with significant lower pole urinary extravasation

Post-operative Expected Course

- The decision to recover the patient in the intensive care unit or the inpatient surgical ward is dependent on the patient's overall clinical status and hemodynamic stability.
- The time of convalescence in the hospital is typically dependent on the severity of any associated injuries and the overall clinical status.
- The eating and drinking status is dependent on the clinical stability as well as the severity of any associated bowel injuries.
- Analgesia can be provided by oral or intravenous narcotics or intravenous patient controlled analgesia (PCA). An epidural is contraindicated in an emergency trauma situation (particularly with hemodynamic instability) but may be considered in a delayed elective procedure.
- Peri-operative antibiotics can be discontinued 24 h after the procedure unless there are signs and symptoms of an active infection. With concomitant bowel injuries or significant urinary extravasation, antibiotic use may be deferred to the discretion of the attending surgeon.
- Discharge is dependent on the severity of concomitant injuries and the overall course of convalescence. In general, discharge is considered when the patient is tolerating a regular diet, having regular bowel and bladder emptying, and is comfortable on oral analgesics.
- Long term imaging of the damaged kidney as well as clinical blood pressure monitoring is necessary.

Part IX
Surgery for Urinary Incontinence

Chapter 28
Surgery for Urinary Incontinence

Ahmet Ali Sancaktutar, Blake W. Palmer, and Bradley P. Kropp

Name of Procedure

Augmentation Cystoplasty and Diversion.

Lay Description

Augmentation Cystoplasty (AC) (also known as a bladder augmentation) is an operation to enlarge the bladder using a piece of the body's own tissue. This is usually the large or small intestine, but the ureters or even the stomach can be used [1].

Intended Benefıt

The aims of an AC are: (1) to provide low pressure storage for urine, (2) to protect kidneys from high pressures, (3) improve urinary continence. Ultimately the main goal is to improve a person's long term health and quality of life.

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Which Segments Should Be Preferred?

The selection of bowel segment to use for the AC is important. The risks and benefits of each option should be considered and this decision should be individualized for each patient based upon preexisting conditions and goals of surgery. Ileum is the most commonly used segment in AC for its ease of use, familiarity to the surgeon and it has been shown to be the most compliant segment [2].

What Happens Before the Operation?

All patients are admitted the day prior to the procedure for a mechanical bowel preparation. This involves drinking at least 4 l Golytely in 4 h. If the child is not able to drink this much fluid than a nasogastric tube will be placed to administer the Golytely.

Preoperative labs are drawn at the time of initial IV placement. The patient is allowed a clear-liquid diet until midnight [1].

Technique

A bladder augment surgery is performed in the operating room under general anesthesia. The procedure can take 3–5 h, depending on the complexity.

Where Is the Incision?

The incision will be on the abdomen. In most, but not all cases, it will be down the center or middle. The length of the incision will depend on the complexity of your child's surgery (Fig. 28.1).

How Is AC Performed?

AC is a type of abdominal surgery, which means that the surgeon makes an incision (cut) in the patient's abdomen to get to the bladder. After making the incision in the abdomen, the surgeon cuts open the bladder at the top to prepare it for enlarging.

The surgeon then removes a section of the intestine; and attaches it to the top of the bladder. The bladder and bowel are then sewn together to make the bladder larger [3]. The intestine is reconnected so that normal bowel function can be resumed.

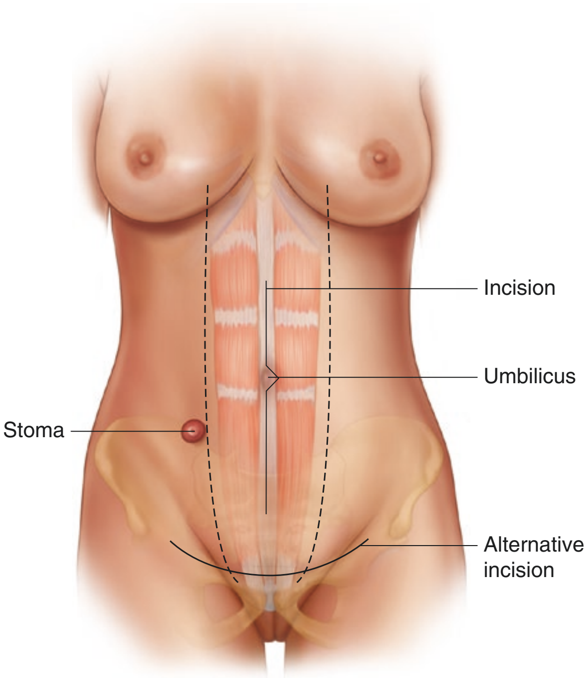

Fig. 28.1 The most common incision is in the midline sparing the umbilicus. Alternatively the incision can be made transversly in the pelvis or pfannenstiel location. The common sites for a stoma are in the Right lower quadrant for a right handed patient or at the umbilicus

Postoperative Expected Course

Many children will be given a caudal or epidural nerve block at the end of the procedure to minimize pain after surgery. The catheter for the epidural usually remains in place for about 2 days.

A suprapubic tube (SPT) is always placed out the native portion of the bladder to assist in post-operative drainage.

A Foley catheter is inserted through the urethra (where urine comes out) into the opening at the bottom of the bladder.

An NGT tube is placed during the procedure and removed on the morning of post-operative day 1.

Oral intake is initiated slowly once return of bowel function returns and the patient has bowel movement. Drainage of the augmented bladder is done via SPT and foley catheter via the newly created abdominal stoma or native urethra. Irrigation of drainage tubes is initiated immediately after surgery to ensure mucus does not obstruct.

Drainage is maintained for 3 weeks, at which time the foley catheter is removed and the augmented bladder is allowed to cycle with the SPT capped and performing CIC every 2–3 h. After this has proven successful, the SPT is removed at 4 weeks [4].

Follow Up

Patients will commonly be discharged from the hospital in 5–7 days after surgery. The catheters will be left in place when you leave the hospital, and the follow-up appointment to have the catheters removed will be almost 4 weeks after surgery. During your child's follow-up visit, many patients will sometimes have a cystogram (x-ray test of the bladder) to be sure healing is complete. The cystogram also shows us the shape and size of your child's new bladder. Three months after surgery patients should have another ultrasound of the kidneys and bladder, and sometimes urodynamic testing [1].

Outcomes

Success of AC is upwards of 95 % for enlarging the bladder. Less than 5 % of patients will require redo AC. It has many potential complication, both short and long term that are summarized below. Overall complications after major lower urinary tract reconstruction are relatively high (~40 %) commensurate with the complexity of surgery and consistent with similar procedures. Majorities of complication are considered minor Clavien Grade I or II. Patients with an obese BMI are at increased risk for complications [5].

Risk of Procedure

All surgery carries some risk and every effort to fully define these risks for each patient prior to surgery should be attempted. The entire medical team should work to optimize the patient's heath and minimize their risk prior to surgery.

Complications

Early

Early complications that happen during the first 1 month are similar to those of major bowel and pelvic surgeries. Wound infections and dehiscence occur in 1–6.4 %, Postoperative bleeding (0–3 %), small bowel obstruction (3–5.7 %) requiring reoperation, and mortality are very rare.

Long-Term

Because of this operation is irreversible, the patient and family should be counseled extensively in the preoperative period about the management of mucus, stones, and metabolic disorders that are possible after AC.

Long-term complications or side effects of the use of a GI segment in the urinary tract are commonplace and should be expected and are listed below [1].

Mucus Production

Inherent to the use of the lower GI segments is the continued production of mucus. Mucus mixed in the urine predisposes to UTIs, stones, and outlet obstruction, and is hypothesized to contribute to the risk of perforation. Regular irrigation also has been shown to reduce complications associated with mucus production in the augmented bladder, such as UTIs and stones. It's important to irrigate child's bladder frequently.

Urinary Tract Infections (UTI's)

Positive urine cultures are a very common finding after AC. This mainly comes in the form of bacteria colonization. Typically, this colonization is handled with saline irrigation of the bladder daily. Symptomatic UTI's are less common but do occur. Symptomatic UTI's should be addressed with oral antibiotics and aggressive bladder irrigations. There has been no significant benefit to patients to be on daily antibiotics and is not recommended. Recurrent symptomatic infections should prompt your physician to evaluate the bladder for stones or excessive mucus production.

Stone Formation

Stone formation, both of the kidney and of the bladder, occurs in 18–50 % of patients after augmentation. Struvite is the most common stone composition; thus, treatment should be initiated immediately for bacteriuria with urea-splitting organisms. Recurrent stones are common. A regular irrigation protocol can decrease the incidence of stones from 43 to 7 %.

Metabolic

Except for stomach segments, which cause hypokalemic hypochloremic metabolic alkalosis, most intestinal segments used for augmentation cystoplasty can cause metabolic acidosis. Hyperchloremic metabolic acidosis is seen in 16 % of patients. Chronic diarrhea is infrequent (10–23 %). Vitamin B_{12} deficiency can also be associated with AC. Therefore yearly blood test should be performed.

Perforation

Perforation is one of the worst complications. The incidence is 4.5–12.8 %, without a specific segment shown to be consistently safer or more at-risk than another. Often, they will have a history of reduced output with CIC, abdominal pain, abdominal distension, and fever. Patients are generally quite ill, and sepsis and death are possible.

Malignancy

Augmented bladders appear to be at greater risk for malignancy. Adenocarcinoma is the most commonly observed tumor, and all segments seem to be associated with the same level of risk. Therefore, it is recommended that episodes of blood in the urine be evaluated by a urology.

Conclusions

In conclusion, AC procedure has real risk and complications associated with it. Most of these early risks are the same as any other major abdominal procedure. Unlike other abdominal procedures, AC has certain long term risks listed above that must be followed life long.

Complications	Ileum/colon	Gastric	Ureter	Autoaugmentation
Mucus	Common	Decreased mucus production	None	None
UTI	13–23 %	8 %	Occurs	Occurs
Stones	18–30 %	Decreased incidence		
Metabolic	Hyperchloremic metabolic acidosis, not significant with normal renal function	Hypokalemic, Hypochloremic metabolic alkalosis	No	No
Perforation	5–19 % (highest with sigmoid colon, ileum 8 %)	5 %	Can occur	Can occur
Malignancy	1.5 % per decade	2.8 % per decade		

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Chapter 29
Appendicovesicostomy and Ileovesicostomy

Martin Kaefer

Name of Procedure:

Continent Catheterizable Channel (Appendicovesicostomy and Monti Procedure)

Lay Description

A Continent Catheterizable Channel (CCC) (also known as a Appendicovesicostomy or Monti Procedure) is an operation to create a tube through which one can pass a catheter into the bladder. This is achieved using a piece of the body's own tissue. This is usually created using the appendix or piece of small intestine, but the ureter or bladder wall itself can also be used for this purpose [1].

Intended Benefit

The aim in creating a CCC is to provide an alternate route (other than the urethra) through which the bladder can be emptied with a catheter. This may be needed for any condition in which the patient cannot empty the bladder spontaneously and includes conditions of impaired bladder function (e.g. spina bifida, spinal cord injury, bladder exstrophy) and impaired urethral anatomy (e.g. a surgically altered urethra or a strictured urethra from trauma). Ultimately the main goal is to improve a person's long-term health and quality of life.

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Which Segments Should Be Preferred?

The selection of tissue type to use for the CCC is important. The relatively uniform diameter of the appendix makes it the ideal structure for this purpose [2]. The appendix is easy to transfer from its position on the colon and has a good blood supply. When the appendix is not available, either because of previous removal or inadequate length, the technique of reconfiguring a piece of small intestine into a small tube with the diameter similar to the appendix can be used (i.e. the Monti Procedure) [3]. The additional time needed to create such a tube and the need for a small bowel anastomosis are the main drawbacks of this option. Finally, in cases where the surgeon wishes to stay out of the peritoneal cavity (e.g. patients with renal failure who require an intact peritoneal cavity for peritoneal dialysis), the ureter or a tube created out of the bladder wall itself can be utilized. The risks and benefits of each option should be considered and this decision should be individualized for each patient based upon pre-existing conditions and goals of surgery.

What Happens Before the Operation?

Patients have historically been admitted the day prior to the procedure for a mechanical bowel preparation. This involves drinking between 2 and 4 l of polyethylene glycol in 4 h. If the child is not able to drink this much fluid then a nasogastric tube may be placed to administer the laxative. Recently we have been performing bowel preparation at home with polyethylene glycol and having the patient present to the hospital the day of surgery. This option is generally chosen for patients with no prior abdominal operations whose families feel they can achieve the preparation in the outpatient setting. The patient is allowed a clear-liquid diet up until midnight on the night preceding the surgery.

Technique

Creation of a continent catheterizable channel is performed in the operating room under general anesthesia. The procedure can take 3–5 h, depending on the complexity.

Where Is the Incision?

The incision will be on the abdomen. In most cases, it will be down the center or middle. The length of the incision will depend on the complexity of your child's surgery and whether other procedures need to be performed simultaneously (e.g. bladder augmentation, ureteral reimplantation or bladder neck repair (Fig. 29.1).

Fig. 29.1 Midline scar from surgical procedure. Stomal opening (*arrow*)

How Is CCC Performed?

CCC is a type of abdominal surgery, which means that the surgeon makes an incision (cut) in the patient's abdomen to get to the intestine and bladder. After making the incision in the abdomen, the surgeon identifies the appendix and detaches it from the colon. If the appendix is not available then a piece of intestine is identified and taken away from the rest of the intestine to be made into an appendix like tube (The intestine is then reconnected so that normal bowel function can be resumed).

The surgeon then connects one end of the appendix or Monti tube to the bladder and the other end is attached to the skin. The location it is attached to the skin can be either in the belly button or halfway between the belly button and the right hip (Fig. 29.1).

Postoperative Expected Course

Many children will be given an abdominal, caudal or epidural injection at the end of the procedure to minimize pain after surgery.

A catheter is left through the channel in order to facilitate healing. If a bladder augmentation to increase bladder volume is performed simultaneously then a suprapubic tube (SPT) is placed in the native portion of the bladder to assist in post-operative drainage. A Foley catheter may be placed through the urethra.

An NGT (stomach drainage tube placed through the nose) tube is placed during the procedure and removed on the morning of post-operative day 1.

Eating and drinking resume slowly once return of bowel function returns and the patient has a bowel movement.

Drainage is maintained for 3 weeks, at which time the catheter in the channel is removed and the family is taught how to catheterize the bladder through the site. The

SPT is clamped and the bladder is allowed to cycle while performing clean intermittent catheterization every 2–3 h. After the patient and family have shown proficiency in catheterizing through the new channel, the SPT is removed (generally after 4 weeks).

Follow Up

Patients will commonly be discharged from the hospital in 5–7 days after surgery. The catheters will be left in place when you leave the hospital, and the follow-up appointment to have the catheters removed will be almost 4 weeks after surgery. Three months after surgery the patient will undergo an ultrasound of the kidneys and bladder to assure that there is adequate emptying of the bladder through the channel.

Outcomes

Complications of CCC can generally be minimized if proper surgical principles are observed and proper catheterization techniques are utilized after the procedure. The cornerstone of successful outcome includes adherence to a punctual catheterization schedule, generous lubrication of catheters and prompt reporting of any difficulty with catheterization to the physician. Failure to report difficulties with catheterization can result in scarring of the channel. A number of possible potential complications are summarized below. Overall complications after major lower urinary tract reconstruction are relatively high (~40 %) commensurate with the complexity of surgery and consistent with similar procedures. Patients with an obese BMI appear to be at increased risk of certain complications.

Risk of Procedure

All surgery carries some risk and every effort to fully define these risks for each patient prior to surgery should be attempted. The entire medical team should work to optimize the patient's health and minimize their risk prior to surgery.

Complications

Early Early complications that occur during the first month are similar to those of other major bowel and pelvic surgeries. Wound infections and dehiscence occur in 1–6.4 %, Postoperative bleeding (0–3 %) and small bowel obstruction (3–5.7 %) are among the most common.

Long-Term Because the operation creates a structure that is repetitively manipulated for emptying the bladder, complications can occur for the life of the channel [4].

Stomal Stenosis

Stomal Stenosis, which is narrowing of the channel opening at the skin, is the most common complication of CCC. This usually is seen in the first 2 years after surgery and occurs in between 8 % and 40 % of patients. Treatment initially consists of simple dilation and topical application of steroid cream to inhibit scar formation. Keeping a short stent in the channel between catheterizations can also help maintain adequate stomal diameter at the skin. If these simple measures are not successful then surgical revision by attaching a fresh skin flap to the channel is undertaken.

Stomal Prolapse

Stomal Prolapse occurs far less frequently than stenosis (2–5 %). Most cases occur in procedures where the channel was not brought through the center of the main muscles of the abdomen (e.g. rectus muscles) – hence leaving it poorly supported. Other causes include creation of an opening in the belly wall fascia that is too large or redundancy of the channel. Relocation of the stoma so that it has better abdominal muscle support is needed for definitive resolution of this complication.

Subfascial Conduit Complications

If one is able to pass a catheter at the level of the skin, but cannot advance it in to the bladder, one of several channel-based complications may exist (e.g. kinking, perforation). These types of complications occur in up to 25 % of patients. Prompt endoscopic evaluation to establish the nature of the problem and placement of a catheter to allow the channel to heal are necessary to minimize long-term injury. Often the knowledge obtained from endoscopic evaluation will provide clues as to how catheterization should be modified to avoid future injury. The catheter is generally removed one week later and catheterization is resumed. If problems with catheterization persist then open surgical revision may be required.

Stomal Incontinence

Incontinence through the channel may result from one or more technical or physiological problems. Inadequate length of implantation of the channel into the bladder and high bladder pressures (if the bladder is too small or stiff) are the most common causes. Bladder pressures should be measured, and if to high, should be ameliorated by using bladder relaxing medications or augmenting the bladder with a piece of intestine. If the channel length is too short then it can at times be buttressed by injecting biomaterials under the channel to improve its ability to close like a valve.

Conclusions

In conclusion, the CCC procedure has real risks and complications associated with it. Most of these early risks are the same as any other major abdominal procedure. CCC has certain long-term risks listed above that generally occur within the first few years after construction, but remain throughout the life of the patient. As such, close continued follow up and immediate access of patients to competent caregivers is critical for providing a reliable result.

Complication	Incidence	Etiology
Stomal stenosis	8–40 %	Tension at skin-conduit anastomosis
		Inadequate stomal diameter
Stomal prolapse	2–5 %	Failure to bring conduit through rectus muscle (inadequate conduit support)
		Fascial opening to large
Stomal incontinence	5–10 %	Elevated bladder pressure
		Insufficient conduit tunnel length
Subfascial complications (kinking, false passage)	25 %	Inadequate fixation of bladder to rectus fascia
		1. Conduit angle of entry into bladder may shifting as bladder fills
		2. Free unsupported segment of conduit may be more prone to false passage

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Chapter 30
Informed Consent Prior to Malone Antegrade Continence Enema: Surgery for Fecal Incontinence

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Key Points
- The ACE procedure is an effective and reliable way of managing the neurogenic bowel with associated fecal incontinence and constipation recalcitrant to conservative management.
- Stomal stenosis is the most common postoperative problem but has several minimally invasive management options.
- Compliance issues should be addressed in a nonjudgmental and supportive manner to help solve the underlying problem.
- A thorough discussion of potential complications should be undertaken prior to embarking on surgery for fecal incontinence.

Clinical Case

A 6-year-old female with a history of myelomeningocele with constipation and fecal soiling refractory to stool softeners and laxatives presents with her family to discuss surgical options for bowel management.

1. What are common complications and how can they be managed?
2. What is the role informed consent in preoperative decision making?

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Introduction

Bladder management in the pediatric patient with neurogenic bladder has been well standardized and described. The quality of life of these children has been greatly enhanced through the provision of independent bladder management and social dryness. The management of the associated neurogenic bowel tends to fall within the auspices of the Pediatric Urologist and can be very challenging for these children. The creation of the antegrade continence enema using the Mitrofanoff principle, popularized by Malone, has been used since 1990 for the management of the neuropathic bowel [1]. The antegrade continence enema (ACE) procedure involves taking either appendix or sometimes small bowel if appendix is not available and bringing to the skin as a catheterizable channel for flushing the colon. The catheterizable channel can be imbricated over the cecum to improve continence. The open technique has been enhanced with the minimally invasive mobilization of the cecum using laparoscopy [2–4]. The goals are for fecal continence, predictable bowel movements without soiling and complete colon evacuation [5]. Patients are able to perform their own stoma flushes for colon evacuation with high satisfaction rates [6]. Complications with the ACE procedure are rare with stoma related problems being the most common complication. One of the more frustrating complications is stool leakage caused by an incompetent valve. While newer minimally invasive techniques, such as Deflux injection can resolve the problem, open revision may also be required [7]. As in all reconstructive cases, patient selection with thorough preoperative patient education and good surgical technique are instrumental in optimizing surgical outcome.

Surgical Indications and Contraindications

Patients with chronic constipation as a result of neurogenic bowel malfunction are the ideal surgical candidates for the ACE procedure. Curry et al. confirmed better continence rates in patients with spina bifida and found success to be age dependent with children older than 5 years tolerating the ACE procedure better [8]. The surgery can be complicated by preexisting conditions such as previous abdominal surgeries with extensive adhesions, presence of a non-usable appendix that requires the creation of a catheterizable conduit from ileum [9] and intra-abdominal space limitation due to co-existing malformations of the spine that will not allow adequate insufflation of the abdomen. Even in the absence of previous abdominal surgeries, the presence of a ventriculoperitoneal (VP) shunt with its associated intra-abdominal adhesions can make the dissection more difficult, sometimes adding hours to the case.

Preoperative Investigation and Patient Preparation

After conservative measures for obtaining complete bowel cleanout have failed, the ACE procedure is an excellent option for children with neurogenic bowel and associated constipation and fecal incontinence. Thorough counseling of the patients and caretakers regarding compliance with the flushing regimen and correct use of the newly created stoma is imperative to ensure long-lasting proper function.

The procedure and the complication possibilities are explained in detail to the family and the patient. Preoperative imaging includes a plain abdominal film for constipation. A renal and bladder ultrasound is obtained to evaluate potential co-existing abnormalities of the urinary tract. The patient has to be evaluated towards the potential need for reconstruction of the urinary tract to combine procedures if necessary. In the authors opinion a complete bowel preparation is preferred to avoid intraoperative stool spillage and ensure fast postoperative return of bowel function. Other groups perform only a rudimentary bowel preparation and report no increased incidence of complications due to bowel spillage [19]. In our institution the patients are started on a clear liquid diet, enemas and oral laxatives at home the night before admission. The patients are admitted 1 day prior to surgery for a complete bowel preparation with oral Golytely and antibiotics. Laboratory studies are obtained at the time of the initial IV placement and values corrected if necessary. Intravenous fluids are run at maintenance and a half rate to compensate for fluid losses during the bowel preparation. Cefoxitin is started as a preoperative antibiotic as well as three doses of neomycin. Nafcillin and Gentamicin are used in patients with ventriculoperitoneal (VP) shunts with Vancomycin replacing Nafcillin in the Penicillin allergic patients. A nasogastric tube (NG) is placed for the administration of the Golytley unless the child is old enough to take it by mouth. An x-ray is obtained to confirm the position of the NG tube prior to starting the bowel preparation. The bowel preparation is continued until the stool output has no particulate matter then the NG tube is removed. Soap suds enemas are also administered as needed.

Operative Technique

Open

For the open technique, there is the need for a midline incision long enough to reach the hepatic flexure of the colon and this incision can be avoided if done laparoscopically. The initial description of the ACE by Malone involved dismemberment of the appendix from the cecum and reversing it prior to implantation into the submucosa of the cecum to create a flap valve mechanism [1, 5]. The appendix can be easily used in situ by mobilizing it along with its mesentery without detachment from the cecum, and this is the currently used technique. Windows are made in the mesentery between vessels. The continence mechanism is then constructed by applying a cecal wrap to the appendix or by either placing the appendix in a submucosal tunnel or imbricating the cecum over the appendix through the mesenteric windows. Some authors prefer not to imbricate the appendix but rather count on the ileocecal valve for continence and report continence rate between 90 and 95 % [10]. Depending on the site chosen for the stoma, either the umbilicus or right lower quadrant, the distal end of the appendix is delivered to the skin for later catheterization. Cecostomy buttons, cecostomy tubes, and tapered intestinal segments tunneled into the cecum can be used in the absence of the appendix.

Laparoscopy

The preoperative preparation is the same as for the open technique. Following the administration of a general anesthetic, the patient is placed in the supine position, padded and secured to the table. The patient is placed in Trendelenburg, a naso- or orogastric tube is placed and a foley catheter is inserted. The patient is then prepped and draped in the usual fashion. There are three minimally invasive techniques described for the laparoscopic ACE procedure. Laparoscopic ACE without imbrication of the conduit, laparoscopic ACE with intracorporeal imbrication and laparoscopic ACE with extracorporeal imbrication of the appendix [3]. The basic tenet of all three techniques is the laparoscopic mobilization of the cecum, appendix and ascending colon. We prefer the extracorporeal technique as described by Casale et al. as intracorporeal imbrication can prolong the procedure significantly.

In the beginning of the procedure, a triangular skin flap is mobilized from the right lateral sidewall towards the umbilicus for later stoma creation. The umbilicus is incised and partially cored out all the way down to the fascia. The fascia is freed up, and a small incision is made. A 5 mm trocar is placed inside the abdominal cavity and the abdomen insufflated to 12 mmHg. The camera is placed and the abdomen inspected for injuries. For mobilization of the cecum, two additional 5 mm trocars are placed. One in the right lower quadrant at McBurney's point and the second in the midline between the umbilicus and the xiphoid process to get better access to the hepatic flexure. The table is placed at 40° angle to the patient's left. The small bowel is swept medially. The appendix is mobilized on its mesentery, avoiding injury to the appendiceal artery. The cecum is mobilized superiorly all the way to the hepatic flexure to ensure enough mobility. The laparoscopic colon mobilization is sufficient if the cecum reaches the umbilicus with ease.

The fascial opening at the umbilicus is then increased and the cecum with appendix is brought to the skin level. The tip of the appendix is excised and a 10 French catheter used to intubate the appendix. Holding sutures are placed into the cecum, creating a plate with the teniae of the cecum in the center. Several windows in the mesentery to the appendix are created and the appendix is placed onto the teniae for support. Now, the cecum is taken left and right of the teniae and pulled through the mesenteric windows imbricating the appendix using 3-0 Ethibond sutures. The tunnel length should be at least five times the appendix circumference to ensure a continent valve. Other groups split the teniae in the middle, mobilize flaps, lay the appendix in the trough and close the flaps over the channel for imbrication. A 10 fr catheter is used to intubate the appendix and it should catheterize easily. The bowel is then placed back into the abdominal cavity. The cecum and or appendix are fixated to the fascia to prevent rotation and a free-floating appendix inside the abdomen. This step is preferred by the authors but not done by other groups. Sutures are placed into the fascia for later closure until only enough space is left for the appendix and its mesentery to pass through with ease. The fascial gap has to be wide enough to allow passage of the channel without causing obstruction but leaving it tight enough to prevent a hernia.

The appendix is spatulated and the previously created skin flap is placed into the spatulation with 4-0 Vicryls to create a wide, open anastomosis. The skin is attached circumferentially to the spatulated appendix allowing for a tension free, wide-open, concealed appendiceal ACE stoma. The stoma is catheterized again at the end of the procedure to ensure easy passage and a 10 or 12 French Foley is placed in the channel, secured with two silk sutures and left to drainage.

Now the abdomen is reinsufflated and inspected for any intraabdominal injuries. The trocars are then removed under direct vision and the fascia closed with 3-0 Vicryl. The naso- or orogastric tube is removed at the end of the procedure. More recently, there are a few case reports of using the laparoscopic single-site (LESS) procedure for the ACE with the added benefit of superior cosmetic outcome [4].

Postoperative Course and Follow Up

On postoperative day one the patient is encouraged to mobilize and the diet advanced after flatus is passed. As soon as the patient is started on oral diet, the ACE is flushed with 60 ml of normal saline daily and slowly advanced to the maximum volume. Pain is initially managed with intravenous Ketorolac and Acetominophen. Narcotics are avoided to allow rapid return of bowel function. The patient is discharged home after tolerating regular diet and the pain being well controlled on oral pain medication. Thorough discussion regarding instructions and expectations with the use of the ACE is mandatory. A well-established team that provides excellent instructions in the management of the stoma catheter along with the washout procedure is key. On our team, we have a dedicated nurse practitioner who instructs the family and patient intensively before discharge and provides thorough instructions, arranges supply of catheters and follow up appointments as well as contact information in case of complications, concerns or emergencies. While preoperative discussion is important, it is the postoperative close follow up and care that prevents early misunderstandings and complications. The patient is evaluated 3 weeks postoperatively and the catheter is removed. The family is instructed on the catheterization process and further follow up planned as needed. During the initial postoperative phase we

Table 30.1 Surgical outcomes based on primary diagnosis

Diagnosis	Full	Partial	Failure (%)
Myelomeningocele	63	21	16
Anorectal malformation	72	17	11
Hirschsprung's disease	82	9	9
Constipation	52	10	38
Miscellaneous	44	25	31

Source: Adapted from Curry et al. [8]

prefer that the patient flush their bowels with sterile normal saline. The desired flush volume is different for all patients depending on their age and body size with a range of 500–3000 ml. It can take anywhere from 30 min to 1 h for complete evacuation of stool. The flushing can be done with syringe or a 1000–2000 ml gravity enema bag.

Fecal continence rates in the neuropathic bowel and anorectal malformations have been reported to be close to 80 % (Table 30.1) [5] and in some series over 90 % [11].

The patients have to be alerted that although final continence rates are high, the road to achieve full continence can take several months as the family gets accustomed to the management of the ACE. It may take at least 1 month to obtain the volume necessary to attain fecal continence and complete bowel evacuation. Thus, discussion of expectations cannot be overemphasized.

Complications

Our patients usually go home on postoperative day two or three depending on how well they are tolerating oral intake. Problems relating to the stoma and difficult catheterization comprise the majority of the chronic and long-term complications. However, additional complications include constipation despite compliance with irrigation, and more rarely, metabolic disturbances. All of these potential pitfalls as well as solutions for each should be discussed preoperatively with the patient's family as well has his/her primary care physician.

Stomal Stenosis

Stomal stenosis at the level of the skin is the most commonly reported complication of the ACE [12], and it appears to be independent of technique, site or tissue used. Risk factors that have been identified to predict an increased rate of stomal stenosis are greater age at time of surgery in patients with neuropathic bowel [13]. The reasoning behind the older age children is the issue with increased parental supervision, which then translates into improved compliance with postoperative care. Obesity is a real problem as this can cause physical difficulties in reaching the stoma site due to the pannus. Frustration with the stoma then eventually leads to noncompliance and sometimes creation of false passages.

Two basic principles to help prevent stoma stenosis include the creation of a well-vascularized conduit with no tension and widely spatulated, broad based skin flaps. The former will help prevent ischemic necrosis and scar formation. The latter will ensure patent anastomosis and is the commonly reported site for stomal stenosis.

The basic management principle involves increasing the frequency of catheterizations on the part of the patient or caretaker to minor surgical corrections ranging from serial dilations in the office setting or in the operating room. The authors also favor placing a 3–5 cm catheter plug in the ACE tube when not in use to act as a passive dilator [14]. Another conservative measure we use is the application of Diprolene cream to the stoma site. Typically, when an open revision is warranted, a single procedure has been reported to suffice with a low re-stenosis rate.

Stomal Incontinence

Typically, this happens at the level of the imbricated tunnel and not at the skin level and tends to present in the early postoperative period. Minor leaks can easily be treated with endoscopic submucosal injection of bulking agents. Deflux has been used with success [7].

On a much smaller scale, smearing of stool at the stoma site can sometimes occur. Mucous plugs within the ACE tract can retain stool material that will occasionally smear at the stoma site. Patients pushing the catheter in too far during flushing of the ACE will typically bypass fecal material at the base of the conduit, which then can smear at the stoma site if left un-flushed for long periods of time. Larger leaks may require open revision of the failed flap valve mechanism.

Other Complications

False passages can occur secondary to kinking of a long and redundant channel or poor angles of catheterization. This can be best prevented by bringing the cecum directly to the abdominal wall and keeping the channel outside the imbricated tunnel as short as possible. During surgery the appendix and or the cecum is fixated to the fascia to allow for tunnel as straight as possible without kinking.

If false passage occurs, the authors recommend a properly placed balloon catheter, either under direct vision or fluoroscopically, left in place for about 10–14 day to help with healing of the injury. Other complications include stomal breakdown, prolapse and stricture. Barqawi reports false passage in 3 % of cases [13]. Strictures can be dilated but occasionally may require open or endoscopic revision in severe cases. Stomal prolapse is rare and corrected by placing a taking suture from the conduit to the abdominal fascia. Excessive tension on the vascular pedicle or constriction from a narrow fascial opening might lead to stomal breakdown, which has been reported to occur in 2% of cases [13]. The management of this requires a major revision with a new segment of bowel.

Electrolyte Abnormalities

A number of irrigation regimens have been described for Malone antegrade continence enemas including saline, hypertonic sodium phosphate, and plain tap water. With the exception of tap water, all solutions are hypertonic and have a stimulatory effect on transit [17]. Life threatening hypernatremia has been described in case reports following use of hypertonic solutions for antegrade washouts [18]. Fewer electrolyte abnormalities have been described with the use of normal saline or tap water. In a retrospective series of 71 patients, no patients experienced hyponatremia or water intoxication secondary to use of tap water [17]. Although the incidence of electrolyte abnormalities is low with the use of traditional irrigants is uncommon, periodic monitoring of serum electrolytes is recommended to ensure to reduce any potential morbidity. Furthermore, patients with preexisting metabolic abnormalities or renal dysfunction should utilize isotonic solutions to prevent life threatening electrolyte derangements.

Conclusion

The ACE procedure for the management of recalcitrant constipation and fecal incontinence in children with neurogenic bowel has been highly successful with reported fecal continence rate over 90% [11]. Complications occur but in experienced hands can usually be solved with conservative measures or minor surgical procedures. In private conversations, many reconstructive surgeons report very high satisfaction rate in the ACE patient population. This was echoed as patients are willing to accept the easily manageable complications for a better quality of life [15, 16]. Minimally invasive techniques for the construction of the ACE and management of the complications are very desirable for this complex patient population that might undergo multiple surgical procedures in their lifetime. When complications arise, clinicians need to re-evaluate the failure, to ensure the patient and caretakers have the necessary tools to overcome the issues. Obesity can be problematic as it relates to the stoma site, especially as these children can have poor mobility and sedentary lives. Even after years of noncompliance with the use of the ACE, clinical consequences are typically inconsequential and the ACE can be salvaged to help improve the patient's quality of life. However, adequate preoperative counseling will help reduce the likelihood of postoperative noncompliance. Thorough discussion of the procedure, expectations and the care for the ACE stoma cannot be overemphasized and a dedicated team member such as a nurse practitioner for such patients is a necessary asset.

Do's and Dont's

- **Do** create a tunnel length to be at least five times the appendix circumference to ensure continent valve.
- **Do** create adequate fascial opening to allow passage of the channel without causing obstruction but tight enough to prevent a hernia.
- **Do** engage in a thorough discussion of all potential complications of surgery for fecal incontinence as well as their management.
- **Do not** underestimate the need for a well-established team that delivers excellent instructions in the management of the stoma.

Clinical Case: Discussion

1. Common complications include stomal stenosis and incontinence and false passage. More rare complications include electrolyte abnormalities related to the irrigant used for flushing.
2. Preoperative counseling of the patient and caregivers is paramount to ensure that patients and families are familiar with the management of various complications should they arise.

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