

Edited by
Prasad P. Godbole
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SECOND EDITION

Pediatric Urology

Surgical Complications and Management



WILEY Blackwell

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Pediatric Urology

Surgical Complications and Management

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Preface

The editors are delighted to present the second edition of *Pediatric Urology: Surgical Complications and Management*. Since the first edition was published in 2008, pediatric urology has advanced in terms of our understanding of disease processes and technology. However, it is incumbent on any clinician undertaking pediatric urology practice to be able to maintain and sustain a safe, high quality and outcome oriented practice. While recognizing and managing complications of surgical interventions is important, it is imperative that prevention of these complications is also recognized. The second edition aims to address these issues.

The general format of the textbook has been changed to include some case-based discussions and a summary “dos and don’ts” take-home message for nearly every chapter. Where applicable, certain “tips and tricks” to maximize efficiency and minimize the risk of complications has been included. With the IT age, online videos demonstrating the techniques where appropriate also form an added feature of the online second edition.

There have been substantial updates to chapters and new chapters have been added based on feedback received from the first edition.

The editors believe that the second edition will be useful for practicing pediatric urologists, urologists in training, pediatric surgeons or indeed any surgeon undertaking office or specialist pediatric urology.

We are indebted to our contributors who have been very supportive in submitting such high quality chapters and within the tight deadlines required. The efforts of Jane Andrew and Rachel Wilkie at Wiley cannot go unrecognized and without whose assistance this book would have been a mere pipe dream.

Finally, as always, we are very grateful to our families who have stood by us and supported us by giving us the time to be able to undertake this worthwhile project.

PG, MK, DW
March 2015

PART I

Principles of Surgical Audit

CHAPTER 1

How to set up prospective surgical audit

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KEY POINTS

- Clinical audit is one of the keystones of clinical governance
- Audit can be conducted prospectively or retrospectively and robust data collected for patient benefit
- A well-performed audit can inform patients about surgical results and drive continuous quality improvement
- Data can be derived from local hospital statistics to nationally reported outcomes
- Paper based audit is time consuming and is being replaced by IT-based support to clinical care pathways

Introduction

Clinical audit is one of the “keystones” of clinical governance. A surgical department that subjects itself to regular and comprehensive audit should be able to provide data to current and prospective patients about the quality of the services it provides, as well as reassurance to those who pay for and regulate health care. Well-organized audit should also enable the clinicians providing services to continually improve the quality of care they deliver.

There are many similarities between audit and research but, historically, audit has often been seen as the poor relation. For audit to be meaningful and useful it must, like research, be methodologically robust and have sufficient “power” to make useful observations; it would be easy to gain false reassurance about the quality of care by looking at outcomes in a small or “cherry-picked” group of straightforward cases. Audit can be conducted retrospectively or prospectively and, again like research, prospective audit has the potential to provide the most useful data, and routine prospective audit provides excellent opportunities for patient benefit [1–4].

Much of the experience we draw on comes from cardiac surgery, where there is a long history of structured data collection, both in the USA and the UK. This was

initially driven by clinicians [1–7], but more recently has been influenced by politicians and the media [7,8]. Cardiac surgery is regarded as an easy specialty to audit in view of the high volume and proportion of a single operation (coronary artery bypass graft) in most surgeons’ practice set against a small but significant hard measurement endpoint of mortality (which is typically around 2%).

In the UK recently, increasing focus has been placed on national clinical audit. A Public Inquiry into the events at Mid Staffordshire NHS Trust found unsatisfactory care that had gone on for some time, despite the existence of data in the “system” that identified potential problems [9]. The UK Government’s response to these events has been to drive public reporting of outcomes down to the level of individual surgeons for 10 specialties, including gastrointestinal surgery, interventional cardiology and urology. These data were published in 2013, and the process has led to marked improvements in engagement with national clinical audit in the UK and has dramatically increased data quality and the utility of the audits [10,11].

Why conduct prospective audit?

There are a number of reasons why clinicians might decide to conduct a clinical audit (Box 1.1).

Box 1.1 Possible reasons for conducting clinical audit.

As a result of local clinical interests
 As a result of clinical incident reporting
 To comply with regional or national initiatives
 To inform patients about surgical results and support choice
 To drive continuous quality improvement
 To comply with health care regulation
 To engage patients in decisions about their health care
 To provide public reassurance

As a result of local clinical interests

Historically, many audit projects have been undertaken as a result of local clinical interests. This may reflect interest in a particular procedure by an individual or a group, or may reflect concern about specific outcomes for a particular operation.

As a result of clinical incident reporting

The major disciplines that ensure high quality care and patient safety are clinical risk management and audit. Most health care organizations should have sophisticated systems in place to report and learn from adverse incidents and near misses [8]. Reporting is usually voluntary and investigated according to a “fair and just culture” but it is unlikely that all incidents that occur are reported. If an adverse incident is recorded, the record identifies that it has occurred but gives no indication of how often it has happened previously, and only limited indication of the likelihood of recurrence. A mature organization should have clear links between risk reporting and audit, and choose topics for the latter based on data from the former.

To comply with regional or national initiatives

Increasingly audits are been driven by organizations that exist outside a hospital. These may include audit led by professional societies, regulatory bodies or regional/national quality improvement and transparency initiatives.

To inform patients

Across the world health care is becoming more patient-focused. The modern health care consumer

will sometimes want to choose their health care provider on the basis of that hospital or surgeon’s outcomes. Even if patients are not choosing between different hospitals, recent data from the UK suggest that patients are interested in outcomes of surgery by their doctors [13]. Patients’ views should inform decisions about what to audit, and they may be interested in many areas which will be dependent on the planned operation but may include data on mortality, success rates, length of stay, the incidence of postoperative infection and other complications, and patients’ experience data.

To drive continuous quality improvement

It has been shown quite clearly from cardiac surgery that structured data collection, analysis and feedback to clinicians improves the quality of outcomes. This has been detected both when data are anonymous and where named surgeon and hospital outcomes have been published [1–4]. The magnitude of this effect is large; in the UK, a system of national reporting for surgical outcomes was introduced in 2001 and has led to a 40% reduction in risk adjusted mortality [4]. The introduction of any drug showing a similar benefit would be heralded as a major breakthrough, but routine national audit has not been embraced by most surgical specialties. Simply collecting and reviewing data seems to drive improvement, but it is likely that the magnitude of the benefits derived and the speed at which improvements are seen can be maximized by developing a clear understanding of what data to collect and using optimal managerial structures and techniques to deliver better care. There is some debate about whether publicly disclosing health care outcomes encourages clinicians to avoid taking on high-risk cases [1,4,7,14,15], but recent experience from the UK certainly confirms that public reporting does drive compliance with national audit with all its inherent benefits.

To comply with health care regulation

Healthcare regulators have a responsibility to ensure that hospitals, and the clinicians working in them, are performing to a satisfactory standard. Whilst some assurance can be gained from examining the systems and processes in place within an organization, the “proof of the pudding” is in demonstrating satisfactory clinical results. This proof is important and can only come from

analyzing benchmarked outcomes data. Regulators of individual clinicians, such as the American Boards in the USA and the General Medical Council in the UK, are changing their emphasis so that it is becoming more important for clinicians to prove they are doing a good job rather than this being assumed. Routine use of structured outcomes data is now supposed to be in place and is included in the current proposals for professional revalidation in the UK (the process by which doctors now have to prove they are “fit to continue to practice” [16,17]).

To engage patients in decisions about their health care

As society becomes supported by better mobile devices and connectivity, people are looking to the internet to support many choices that they make, including choices about health care. It is vital that the medical profession and health care organizations accept this and provide patients and their carers with appropriate information to empower and engage them in the concept of “shared decision making” with their health care advisers.

To provide public reassurance

It is certainly true in the UK, but is possibly true more widely, that the trust that has traditionally been placed in the medical profession – and, indeed, medical professionals – is being eroded by repeated failure of clinical governance and increasing societal expectations. Maintaining a trusting relationship between an informed public and a trustworthy profession is in everyone’s best interests, and this can be supported by transparent clinical audit data.

What data can be used for audit?

Routine hospital data

Most health care systems are rich in data and poor in information. Medicare data in the USA and Hospital Episodes Statistics in the UK contain information about patient demographics, diagnoses, procedure, mortality, length of stay, day cases rates and readmissions. These information systems are developed for administrative or financial purposes rather than clinical ones, but may potentially contain much useful clinical data and will often have the capacity to

provide some degree of adjustment for case mix. In the UK this data has historically not been trusted by clinicians, but recently there has been increasing engagement between doctors and the data which is improving clinical data quality and increasing confidence. Many UK hospitals now have systems to benchmark their outcomes against national or other peer groups, to flag up areas of good practice, detect outlying performance and engage in quality improvement [18].

Ideally, hospitals should have clearly-defined systems in place to use the data: for example, they should regularly compare their outcomes for chosen procedures against an appropriately selected group of other hospitals. Significant “good” practice should be celebrated and shared with others inside and outside the organization, and bad outcomes should be investigated. It is not uncommon that high mortality or other clinical indicator rates may have a clear explanation other than that of “bad” clinical practice. The data may be incorrect, or there may be issues about classification or attribution that explain away an apparent alert, but structured investigation should improve the knowledge of both the organization and the clinician knowledge about their data systems and may lead to better knowledge that necessitates improvements in patient care.

Specialty-specific multi-center data

A number of surgical disciplines in the USA and the UK have embarked upon national programs to collect prospective disease- or operation-specific datasets. These are usually clinically driven and have benefits above routine hospital data in that a more useful dataset can be designed for specific purposes and, in particular, can look in more detail at subtleties of case mix and specific clinical outcomes in a way that is more robust and sensitive than that derived from routine hospital administration systems. Contemporary cardiac surgical datasets collect variables on preoperative patient characteristics, precise operative data and postoperative mortality, ICU stay, hospital stay, re-explorations, infection, renal failure, tracheostomy, blood usage, stroke rate and intra-aortic balloon pump use. The preoperative and operative data allow outcomes to be adjusted for case complexity to prevent comparison of “apples and oranges” by various algorithms such as the EuroSCORE [20]. Data for 10 such audits are now

published in the UK down to the level of individual surgeon [10,11].

Setting up specialty-specific multi-center audit raises a number of challenges including defining clarity of purpose, gaining consensus, agreeing a dataset, securing resource, overcoming information technology and methodology issues, and clarifying ownership of data, information policies and governance arrangements [21]. In cardiac surgery there is now increasing international dialogue between professional organizations, moving towards the collection of standardized data to allow widespread comparisons.

Locally-derived data

Individual hospital departments will often decide to audit a specific theme that may be chosen because of clinical risk management issues, subspecialist interest or other concerns. In the UK National Health Service, dedicated resources for audit were historically “top sliced” from the purchasers of health care to generate a culture of clinical quality improvement, but commentators are divided about whether significant benefits have been realized from this approach [13]. In the early stages, large amounts of audit activity were undertaken, but there were significant failures in subsequently delivering appropriate change. To maximize the chances of improving care as a result of audit the following should be considered. Will the sample size be big enough to be useful? What dataset is needed? Will that data be accessible from existing hospital case notes or will prospective data collection be necessary? Is there an existing robust benchmark to which the results of the audit can be compared? How will the “significance” of the results be analyzed? Does conducting the audit have financial implications? Will the potential results of the audit have financial implications? Are all stakeholders who may need to change their behavior as a result of the audit involved in the process?

Techniques of data collection

Historically, the majority of audit activity was conducted from retrospective examination of case notes, which was labor intensive and relied on the accuracy and completeness of previously recorded data. There has subsequently been increasing use of prospective

data collection, much of which has been based on paper forms. This obviously improves the quality of data, but again requires time and effort from clinical or administrative staff for completion. The development of care pathways whereby multidisciplinary teams manage clinical conditions in predefined ways is thought to improve patient outcomes and will generate structured data that are readily amenable to audit. The use of modern information technology to support care pathways is the “holy grail” of effective audit – all data are generated for clinical use and the relevant subset of that data can then be examined for any relevant purpose. The care pathway can be adapted to include new or alternative variables as required. All data collection can be networked and wireless, assuming issues about data access, confidentiality and security are resolved. Maximizing benefits from this approach raises a number of challenges, including implementing major changes in clinical practice and medical culture.

Good practice in audit

A clinical department should benefit from a clear forward plan about its audit activity that should be developed by the multidisciplinary team in conjunction with patients and their carers. The audit activity should include an appropriate mix of national, local and risk management-driven issues and the specifics should depend on the configuration of services and local preferences. The plan should include thoughts about dissemination of results to users and potential users of the services. The multidisciplinary team should include doctors, professions allied to medicine, and administration staff. Adherence to the audit plan should be monitored through the departmental operational management structures. For the department to be successful in improving care as a result of audit there should be clear understanding of effective techniques of change management.

Arguments against audit

In the UK, audit has been an essential part of all doctors’ job plans for a number of years, but audit activity remains sporadic. In some specialties, such as those included in the NHS England transparency agenda,

comprehensive audit is being led by clinicians and driven by politicians and the media [10,11]. In other areas there remains little or no coordinated national audit activity. This may be due to a perceived lack of benefits from audit by clinicians along with failure to meet challenges in gaining consensus or difficulties in securing adequate resources. The experience from cardiac surgery and many other national audits in the UK is that structured national audit improves the quality of mortality outcomes [1–4]. It is likely that other issues, such as complication rates, are also reduced with associated costs savings, and as such effective audit may well pay for itself.

Conclusion

In modern health care, patients are increasingly looking to be reassured about the quality of care they receive, and doctors are being driven towards demonstrating their competence rather than this being assumed. Hospital departments should have a robust clinical governance strategy that should include “joined-up” clinical risk management and audit activity. There are strong arguments that structured audit activity improves the quality of outcomes and for these benefits to be maximized there should be involvement of multidisciplinary teams supported by high-quality operational management.

DOS AND DON'TS

Do

- Continually work to evaluate the quality of care you deliver for patients
- Develop a strategy for clinical audit which incorporates the relevant area of your practice and is methodologically robust
- Benchmark your practice against accepted best practice
- Develop a link between learning from risk management and your clinical audit program
- Develop links between clinical audit and departmental/individual reflective practice
- Evaluate your personal surgical audit and know what to do if your results are not ‘as expected’
- Be transparent about your audit program and your results of care

Don't

- Undertake an audit that is not methodologically robust
- Fail to implement changes resulting from an audit which demonstrates unsatisfactory processes or outcomes
- Derive false reassurance from benchmarking against time-expired clinical standards
- Assume that patients and the public have no interest in the outcome of care derived from your audit program.

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CHAPTER 2

Evaluating personal surgical audit and what to do if your results are not “as expected”

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KEY POINTS

- Audit is the comparison of surgical results against a previously defined and accepted standard
- Published results may be better than the normal surgeon's
- Complexity specific audit is important
- Dealing with outlying performance can be 'directive' or 'collaborative' depending on the surgeon
- Surgeons are responsible for ensuring satisfactory quality of care

Introduction

Any well-conducted audit should give information about systems and outcomes related to patient care. Data collection that generates new information about patient outcomes should be classified as research; to be regarded as audit, results need to be compared against a previously-defined and accepted standard. Often an audit will demonstrate satisfactory outcomes and this in itself may be a useful finding which should be of interest to patients, clinicians, managers, commissioners and regulators of health care. It is hoped that structured and regular audit data collection will lead to ongoing improvements in quality as described in Chapter 1. On occasions, audit results will be unacceptable and it is essential that this is recognized and acted upon.

Presentation and analysis of data

Effective audit requires clarity of purpose. When an audit is conceived the clinical question should be clearly stated and the data required to generate an answer

should be defined. It is also important to be sure about the outcomes with which you will compare yourself, and there may be a number of options. Data on mortality or complication rates may be available from pooled national or regional registries [1–4]. Results of specific series of cases may be published through peer review journals for individual hospitals or individuals, but these outcomes may often be better than the “norm” because of submission and publication bias. False reassurance might be gained from comparing outcomes with outdated historical results; in cardiac surgery in the UK, a widely-accepted risk adjustment algorithm, the EuroSCORE [5], has been used to benchmark hospitals and surgeons in recent years. This was developed in a multi-center study in Europe in 1997 and improvements in overall quality of care in the UK are such that it no longer reflects current practice [6]. This concept of “calibration drift” for cardiac surgery has been seen in both the UK and the USA and is important to take into account when using benchmarking for audit [7].

It is possible to compare outcomes between units or surgeons simply by using “crude” or non-risk-adjusted data. Cardiac surgeons have focused on mortality as it is

a robust primary end point. In pediatric urology, mortality is not frequent enough to provide a meaningful measure; more appropriate endpoints need to be developed and this is a challenge for the profession.

Using non-risk-adjusted data has simplicity and transparency on its side but it is not embraced with enthusiasm by the majority of surgeons. It is clear that there are quite marked differences in patient characteristics between different units in cardiac surgery, and this variability is probably greater between surgeons who have different subspecialist interests [8]. These issues apply to other areas of surgery. Many surgeons are concerned that any attempt to produce comparative performance using non-risk-adjusted data will stimulate a culture whereby higher-risk patients are denied surgery to help maintain good results – so-called risk-averse behavior. In order to make data comparable between individual surgeons and units there have been a number of attempts to adjust for operative risk in cardiac surgery [9–12]. Other specialties will need to develop appropriate methodology and ideal tools should be accurate numerical predictors of observed risk (i.e. be calibrated correctly) and the ability to discriminate appropriately across the spectrum of risk (i.e. accurately differentiate between lower- and higher-risk patients).

In addition to the appropriate use of risk adjustment, some units have found graphical techniques of presenting outcomes data useful to monitor performance. Various techniques, such as cumulative summation or variable life-adjusted display plots, have been used to help analyze results and detect trends or outlying performance at an early stage. These curves may be adapted to include predicted mortality to enable observed and expected mortality to be compared. These techniques are well described by Keogh and Kinsman [2]. More recently, interest is developing for measuring outcomes using statistical process control charts, which are widely used in the manufacturing industry. These charts use units of time, typically months when institutions are under scrutiny and the outcome of interest is mortality, and display actual mortality against expected mortality using control limits to define acceptable and unacceptable performance [12].

The use of funnel plots is becoming popular as a way of displaying hospital or individual mortality [13]. These are simply a plot of event rates against volume of surgery, and include exact binomial control limits to allow excessive mortality to be easily detected. They

give a “strong visual display of divergent performance” [14]. They have been used to analyze routine data to define clinical case-mix and compare hospital outcomes in urology [15]. These methods have been used in the UK to display mortality rates to patients and the public [16].

Classical statistical techniques may be used to compare individual outcomes with a benchmark. When analyzing data from an individual hospital or surgeon it is probably appropriate to select 95% confidence intervals such that if significant differences are observed, there is a 1 in 20 probability that these are due to chance alone. Things become more difficult when many hospitals or surgeons are compared to a national benchmark. In the UK, there are over 200 cardiac surgeons and any comparison of the group against the pooled mortality. Using 95% confidence intervals with this group would raise a high probability of detecting outlying performance due to chance alone because of multiple comparisons, and it is appropriate to adjust for this. The choice of confidence intervals will always end up as a balance between ensuring that true outlying performance is detected without inappropriately creating stigma for surgeons with satisfactory outcomes [17]. It may be useful to select different confidence limits for different purposes. Tight limits may be appropriate for local supportive clinical governance monitoring; one hospital in northwest England launches an internal investigation into practice if a cardiac surgeon’s results fall outside 80% confidence limits but wider limits of 99% have been used to report those surgeon’s outcomes to the public [18].

Dealing with outlying performance

Detecting clinical outcomes that fall outside accepted limits does not necessarily indicate substandard patient care. However, any analysis which causes concern should trigger further validation of the data if appropriate. Then, if indicated, there should be an in-depth evaluation of clinical practice which may include analysis of subspecialty, case mix and an exploration of the exact mechanisms of death or complications. This process may lead to reassurance that practice is satisfactory. Ideally, this should be initiated by the clinician concerned who should be keen to learn from the experience to improve their practice. An excellent example comes from

pediatric cardiac surgery: a surgeon had concerns about his mortality outcomes following the arterial switch operation (which is complex, technically challenging, congenital surgery) [19]. He studied his outcomes in detail using CUSUM methodology and determined that things were worse than he would have expected from chance alone. He then underwent retraining with a colleague from another hospital with excellent outcomes, adapted his practice, and subsequently went on to demonstrate good outcomes in a further series of consecutive cases.

On occasions, the process of investigating outlying outcomes may be difficult for the individual hospital or surgeon involved. The investigation may raise significant methodological questions about the techniques of analysis and subsequent examinations. The cause of substandard results may be difficult to detect but may relate to failures in the systems of care in the hospital or department, or failures in the individual [17–23].

Clinical governance is an individual, departmental and hospital responsibility. Whilst the onus should be on the individual with unsatisfactory outcomes to investigate and change their practice, they may need support, advice and direction from their clinical and managerial colleagues. Over recent years, the roles of different organizations in clinical governance are becoming clearer. Most hospitals should now have increasingly effective management structures for promoting quality improvement and detecting suboptimal performance.

The investigation of unsatisfactory outcomes can be facilitated by appropriate clinical leadership, and different techniques may be necessary for different circumstances with the concept of “situational leadership” being useful to match the managerial intervention to the willingness and the readiness of the individual whose practice is being investigated [24]. Two examples make this point. A newly-appointed cardiac surgeon had three adverse outcomes following the same type of operation that, to colleagues, seemed to be due to a similar mechanism. Despite discussions, the surgeon involved had little or no insight into the problem. No confidence intervals for performance were crossed because of the small volume of cases involved but, due to the clinical concerns, the surgeon was subjected to forced but supportive retraining of his intraoperative techniques, which led to the reintroduction of full independent practice within a few months and excellent publicly-reported results for that operation several

years later. This would be described in a situational leadership model as a “directive” approach. A second example is that of a senior surgeon with a low-volume mixed cardiothoracic practice who had a “bad run” of cardiac results, which again led to outcomes that failed to generate statistically-significant mortality outcomes. On his own initiative, he involved his clinical managers and launched an in-depth analysis of his practice. He detected that he was conducting very high-predicted-risk surgery despite lower volumes of surgery than some single specialty colleagues. He was also suspicious of a potential common mechanism of adverse outcomes in several cases of mortality and some cases of morbidity. Along with colleagues, he changed his referred practice to make it more compatible with low volume mixed cardiothoracic surgery and adapted his technique of surgery to avoid further problems. This again resulted in excellent subsequent outcomes. This would be described in a situational leadership model as a “collaborative” approach. From a managerial perspective, both examples led to satisfactory ends, but adopting the appropriate leadership style was important in reaching the desired conclusions. In addition to having some understanding of leadership intervention models, we would also recommend that clinical managers have expertise in having “difficult conversations”, understand some change management theory, and have the ability to use an understanding of their personality characteristics and those of their colleagues to maximize the benefits, and the downsides, of managerial interventions.

In addition to the roles of the individual and the hospital in ensuring satisfactory outcomes, other agencies should be acting to support the process. In the UK, the Chief Medical Officer produced a report, *Good Doctors, Safer Patients*, about regulation of health care, and now the General Medical Council has responsibility for professional regulation, but passes significant responsibilities down to employers [25–28]. Professional revalidation for all 230,000 doctors in the UK has now started, and should include data from clinical audit to give positive affirmation that good care is being delivered. It is suggested that professional societies should set clear, unambiguous standards for care, and recertification of doctors should be dependent on achieving those standards. Patient consultation as part of this report has suggested that patients are keen to see that satisfactory outcomes of treatment by their doctors form part

of this process and this is now made available to the public from doctors working in 10 specialties [29,39]. UK cardiac surgeons have responded to this agenda by articulating clearly their responsibilities to patients and the public in their publication *Maintaining Patients' Trust* [33]. These themes have been reiterated clearly by the recent UK public inquiry into the events at Mid Staffordshire NHS Trust [20].

This direction of travel in the UK is a long way from the culture in which most doctors were trained. It will be a challenge for professional societies and the profession to deliver on this agenda.

Conclusion

Most audit projects will deliver results that demonstrate clinical practice is satisfactory. There is some evidence that scrutiny of results alone can contribute to improvements in quality. On occasions, audit will flag up concern about clinical processes or outcome, but it is important that the data and the methods are “fit for purpose”. Ensuring that satisfactory quality of care is given and demonstrated is the responsibility of all involved in health care delivery, including individual practitioners, employers, commissioners, professional societies and regulators.

DOS AND DON'TS

Do

- Act rapidly if audit data suggest results are not as expected
 - Ensure that data quality issues are resolved without unnecessary delay
 - Configure an improvement plan involving colleagues, the wider multidisciplinary team and organizational management
- Ensure that patient safety and outcomes remain the primary consideration in all actions

Don't

- Assume that poor outcomes are due to data quality issues
- Derive false reassurance on quality by benchmarking against a time-expired clinical standard
- Try to act on poor outcomes without engaging colleagues and the organization
- Underestimate the importance of excellent clinical leadership in optimizing the quality of clinical outcomes for patients

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CHAPTER 3

A critical assessment of surgical outcomes

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KEY POINTS

- Outcomes between units or surgeons can be compared using non-risk-adjusted data; for example cardiac surgeons focus on mortality; while simple it may lead to risk averse behavior amongst surgeons
- In pediatric urology, mortality is not frequent enough to use as a meaningful end point for comparison
- Other techniques to analyze data include cumulative summation or life-adjusted display plots, statistical process control charts, funnel plots and classical statistical techniques with a benchmark

Introduction

In 1979, Lewis Thomas said: "There is within medicine, somewhere beneath the pessimism and discouragement resulting from the disarray of the health care system and its stupendous cost, an undercurrent of almost outrageous optimism about what may lie ahead for the treatment of human disease if only we can keep learning" [1]. If he was concerned about the disarray and cost of the health care system in 1979, he would be disappointed today. Since 1979, health care costs have been rising at a much higher rate than the consumer price index, yet biomedical and technological research has been extremely productive [2]. In an attempt to control health care costs, an unfortunate consequence would be loss of new knowledge and innovation. Evidence suggests that around 40% of health care cost is due to waste or is non-value added to the patient, and reducing this waste should be the primary goal of health care providers [3].

As health care providers we can apply evidence-based medicine to provide the highest quality care to our patient population at the lowest possible cost.

Evidence-based medicine (EBM) was initiated by many in the medical field and is the critical evaluation of the current literature to obtain the best evidence and apply it to clinical practice. It is the

conscientious, explicit and judicious use of current best evidence in making decisions about the care of individual patients [4]. We, as health care providers, can apply this evidence to provide value (quality/cost) to the patients we treat.

It is now recognized that surgical outcomes vary by provider [5–7]. As such, surgeons and hospitals are being asked to provide evidence of the quality of care they deliver [8]. Social media is becoming more and more utilized by hospitals and medical professionals as a means of conveying general health information, sometimes even personalized help. Moreover, patients and their families are turning to the Internet and social media to make critical decisions about choice of surgeon and facility [9].

Surgeons have traditionally made therapeutic decisions based on personal experience, recommendations of surgical authorities and thoughtful application of surgical basic sciences. Evidence-based surgery emphasizes the need to evaluate properly the efficacy of diagnostic and therapeutic interventions before accepting them as standard surgical practice [10]. Evidence in clinical surgery, and especially in pediatric urology, varies in its quality.

Published research findings are sometimes refuted by subsequent evidence with ensuing confusion and

disappointment. Refutation and controversy is seen across a range of research designs [11]. There is also concern that false findings may be present in the majority of published articles. In a recent article in the *Journal of Urology*, Turpen et al. analyzed randomized controlled trials (RCTs) presented as abstracts at the 2002 and 2003 American Urological Association annual meeting and found that the current quality of reporting RCTs at urological meetings is suboptimal, and raising concerns about their use to guide clinical decision making [12]. Recently, De Sio et al. assessed the reporting quality of randomized and nonrandomized, controlled trials presented in abstract form at the European Association of Urology annual meeting over a 10-year period and determined the impact on subsequent publication. Unfortunately, they found that the reporting quality of European Association of Urology meeting abstracts did not improve in a decade. They stress the importance of improving the quality of abstracts following currently-available guidelines [13].

Pediatric urologists often make substantially different management decisions for similar clinical situations [14–16]. This variation in practice occurs in geographically close communities and is not always explained by differences in patient characteristics or preferences. More importantly, variation in management can be costly and often includes practices that are inconsistent with good evidence about optimal care [17,18].

Pediatric urologists should critically examine published evidence and then adjust their practices

accordingly. The purpose of this chapter is to assist them in doing so.

The main elements that will be discussed are: (1) asking focused questions; (2) finding the evidence; (3) critical appraisal of the evidence; and (4) making a decision.

Asking focused questions

One of the fundamental skills related to finding the evidence is asking a well-built clinical question. To benefit patients and clinicians, such questions need to be both directly relevant to patients' problems and phrased in ways that direct your search to relevant and precise answers. In practice, well-built clinical questions usually contain four elements (PICO: Population, Problem, Intervention, Comparison, Outcomes), summarized in Table 3.1. Included are some examples of asking these questions in pediatric urology [19].

By asking a concise and well-formed question it becomes straightforward to combine the terms needed to then query searching services such as PubMed.

Finding the evidence

You can now convert your PICO question into search words in a search engine such as PubMed as shown in Table 3.2.

Table 3.1 PICO elements in asking clinical questions.

	1: Patient or problem	2: Intervention (a cause, prognostic factor, treatment, etc.)	3: Comparison intervention (if necessary)	4: Outcomes
PICO	"How would I describe a group of patients similar to mine?"	"Which main intervention am I considering?"	"What is the main alternative to compare with the intervention?"	"What can I hope to accomplish?" or "What could this exposure really affect?"
Example 1	In adolescents diagnosed with varicocele and testicular size discrepancy	Would surgical varicocelectomy	When compared to nonsurgical treatment	Affect future fertility or sperm count
Example 2	In uncircumcised male infants diagnosed with SFU grade 4 hydronephrosis without vesicoureteral reflux	Would antibiotic prophylaxis	When compared to no antibiotics	Reduce the risk of febrile urinary tract infections

Table 3.2 Converting PICO question to search words.

Population	Adolescent and varicocele
Intervention	Varicocelelectomy
Comparison	Conservative
Outcome	Fertility

Critical appraisal of the evidence

Study designs

All study designs [20,21] have similar components (PICO):

- 1 A defined population (P) from which groups of subjects are studied.
- 2 Outcomes (O) that are measured.
- 3 For experimental and analytical observational studies an intervention or exposure (I) that is applied and
- 4 Compared to different groups (C) of subjects.

The first distinction is whether the study is *analytic* or *descriptive* (Figure 3.1).

A descriptive study uses the PO in PICO and therefore does not quantify the relationship. Instead it gives a picture of what is happening in the population, such as the incidence of urinary tract infections in males who are not circumcised in the first year of life. Descriptive studies include case reports, case series, qualitative studies and surveys (cross sectional) studies.

An analytic study attempts to quantify the relationship between the effect of an intervention or exposure (I) on the outcome (O).

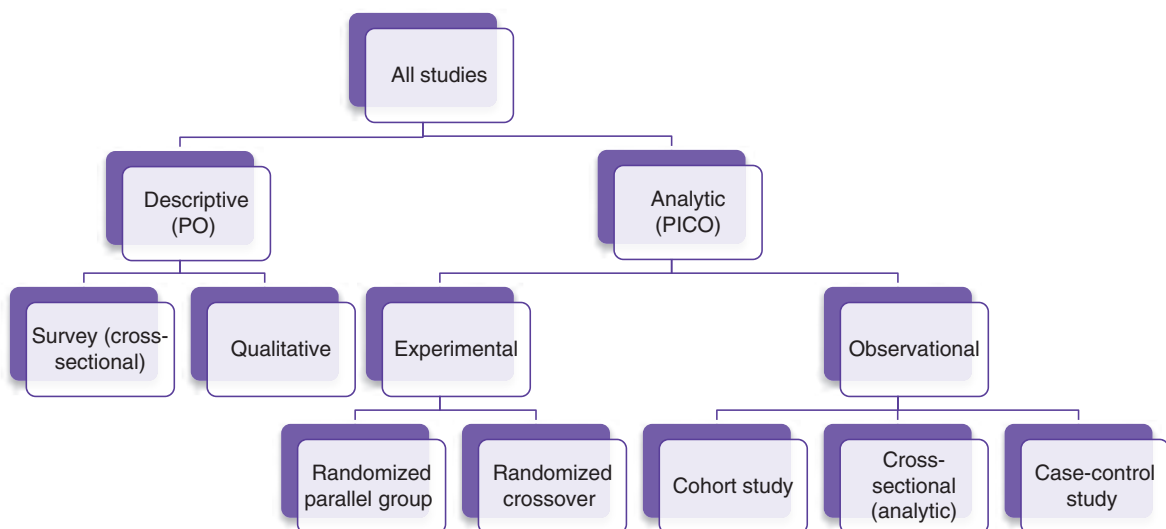
Whether a researcher actively changes the intervention determines if the study is *observational* or *experimental* (Figure 3.1).

In the experimental study the researcher manipulates the exposure, such as in randomized controlled trials. If adequately randomized and blinded, these studies have the ability to control for most biases. This, though, depends on the quality of the study, design and implementation. Therefore, not all randomized trials are high quality [12].

In the analytical observational study the researcher simply measures the exposure or treatments of the groups. These include case-control studies, cohort studies, and some population cross-sectional studies. This is the bulk of studies published in the pediatric urological literature.

The type of study can generally be appreciated by asking three questions [21]:

- 1 What was the aim of the study?
 - a To describe a population (PO question) – descriptive.
 - b To quantify the relationship between factors – analytic.
- 2 If analytic, was the intervention randomly allocated:
 - a Yes – RCT.
 - b No – observational.

**Figure 3.1** Summary of study designs.

- 3 If observational, when were the outcomes determined?
- a After the exposure or intervention – cohort study.
 - b At the same time as the exposure – cross-sectional study or survey.
 - c Before the exposure was determined – case-control study.

There are advantages and disadvantages of the designs as shown in Table 3.3 [21].

Systematic reviews and meta-analysis

Other study designs that are increasingly being reported in the pediatric urological literature are systematic reviews and meta-analyses. These are specifically designed to use with any one study that is underpowered to answer a research question. Meta-analysis is a method that pools the available data in the literature in an effort to increase statistical power.

An important consideration in appraising a meta-analysis is the homogeneity of the pooled studies. The quality of a systematic review and meta-analysis is dependent on the quality of the articles used in the review.

Significant heterogeneity as seen in most meta-analyses published in the pediatric urology literature creates more than chance variation in study outcomes and is a sign that the results of the included studies may not be compatible and should not have been pooled in the first place. This is particularly true in pediatric urology when observational and retrospective data is utilized. These studies tend to introduce confounding and bias and also have less control of variation [22].

Moreover, diagnostic accuracy may be overestimated when using certain study designs; thus, the inclusion of studies using different designs in meta-analyses may have important effects on their results, and influence clinical decision making [23].

Bias in pediatric studies

The *Oxford Dictionary* defines bias as: “Prejudice in favor of or against one thing, person, or group compared with another, usually in a way considered to be unfair” [24].

Bias is defined as any process that leads to the systematic deviation of study results from the truth. This

Table 3.3 Advantages and disadvantages of study designs.

Study design	Advantages	Disadvantages
Randomized controlled trial	Unbiased distribution of confounders Blinding more likely Randomization facilitates statistical analysis	Expensive Volunteer bias Ethically problematic at times
Crossover design (randomized to treatment A first then crossover to treatment B)	All subjects serve as own control, reducing sample size needed Statistical tests assuming randomization can be used	All subjects receive placebo or alternative treatment Washout period lengthy or unknown Cannot be used for treatments with permanent effects
Cohort study	Ethically safe Subjects can be matched Can establish timing and directionality of events Eligibility criteria and outcomes can be standardized Easier and cheaper to conduct	Controls difficult to identify Exposure may be linked to a confounder Blinding is difficult Randomization not present For rare disease large sample size or long follow-up required
Case-control study	Quick and cheap Only feasible method for rare disorders or those with long lag between exposure and outcomes Fewer subjects needed than with cross-sectional studies	Reliance on recall or records to determine exposure Confounders Selection of control group difficult
Cross-sectional survey (best for quantifying the accuracy of diagnostic test)	Cheap and simple Ethically safe	Potential bias – recall, selection, Neyman Establishes association at most, <i>not</i> causality Recall bias susceptibility Confounders Group sizes may be unequal Neyman bias (incidence-prevalence bias)

generally results from limitation in study design or reporting rather than from personal prejudices. Some forms of bias are inevitable but they need to be recognized and accounted for by the investigator.

Sackett [25] noted that bias occurs at different stages of the research and includes:

- 1 Pre-study literature review.
- 2 Selection of study sample.
- 3 Administration of intervention.
- 4 Measurement of outcome.
- 5 Statistical analysis.
- 6 Reporting and publishing results.

The Cochrane review database summarizes sources of bias well and can be used as a guide to assessing articles for bias [26]. It describes a useful classification of biases into the following categories: selection bias, performance bias, attrition bias, detection bias and reporting bias [27].

Selection bias

Selection bias refers to systematic differences between baseline characteristics of the groups that are compared. The unique strength of randomization is that, if successfully accomplished, it prevents selection bias in allocating interventions to participants.

For all potential sources of bias, it is important to consider the likely magnitude and the likely direction of the bias and to see it described in the discussion section of the article.

Performance bias

Performance bias refers to systematic differences between groups in the care that is provided, or in exposure to factors other than the interventions of interest. Blinding (or masking) of study participants and personnel may reduce the risk that knowledge of which intervention was received, rather than the intervention itself, affects outcomes. Blinding is not always possible, however. For example, it is usually impossible to blind people to whether or not major surgery has been undertaken.

Detection bias

Detection bias refers to systematic differences between groups in how outcomes are determined. Blinding (or masking) of outcome assessors may reduce the risk that knowledge of which intervention was received, rather than the intervention itself, affects outcome measurement. Blinding of outcome assessors can be especially

important for assessment of subjective outcomes, such as degree of postoperative pain.

Attrition bias

Attrition bias refers to systematic differences between groups in withdrawals from a study or exclusion from a study. These have the potential to lead to incomplete outcome data. There are two reasons for withdrawals or incomplete outcome data in clinical trials. Exclusions refer to situations in which some participants are omitted from reports of analyses, despite outcome data being available. Attrition refers to situations in which outcome data are not available.

Reporting bias

Reporting bias refers to systematic differences between reported and unreported findings. Within a published report, those analyses with statistically significant differences between intervention groups are more likely to be reported than non-significant differences. This sort of “within-study publication bias” is usually known as outcome reporting bias or selective reporting bias, and may be one of the most substantial biases affecting results from individual studies [28].

Other biases

In addition, there are other sources of bias that are relevant only in certain circumstances. Some of these include [29]:

- 1 **Prevalence–incidence (Neyman) bias:** When measurement of risk factors is selectively done on existing patients (prevalent) rather than newly-diagnosed (incident) patients. This may distort the results.
- 2 **Recall bias:** Cases and controls may differ in the accuracy of recalling past events.
- 3 **Confounding bias:** When a non-causal association observed between a given exposure and outcome is a result of the influence of a third variable, it is termed confounding, with the third variable termed a confounding variable. A confounding variable is causally associated with the outcome of interest, and non-causally or causally associated with the exposure, but is not an intermediate variable in the causal pathway between exposure and outcome. Stratification and multiple regression techniques are two methods used to address confounding, and produce “adjusted” odds ratios.

Investigators should be aware of potential sources of bias when designing studies and utilize methods to

reduce bias. Methods such as randomization, blinding in clinical trials, stratification, multivariate regression, propensity score, instrumental variable analysis and careful selection of matched controls are used to reduce bias and confounding effects.

Authors of observational studies should ideally address confounding and bias in two ways: analytically as discussed above and also in their discussion of the study's limitations. For variables that were not measured or that cannot be measured, the authors should itemize those variables and discuss the potential direction and magnitude of bias that could result from them.

Common description of outcome results

Odds ratio

When comparing outcomes between two groups, the most common method is to simply compare the ratio of the risk in one group versus the risk in another group and this is called the relative risk (RR). For example, if the risk of an outcome in group 1 is 50% and in group 2 it is 25% the relative risk would be $50/25 = 2$ meaning that the risk in one group is twice that in the other. Risk cannot often be calculated and the alternative is to compare the odds of an outcome between the two groups and this is called the odds ratio (OR) [30].

The OR can estimate the RR when the outcome of interest is rare (less than 10% per group). On the other hand, if the outcome is more common, the OR will exaggerate the RR.

Table 3.4 shows the math used in both calculations.

Sensitivity and specificity, positive predictive value and negative predictive value

One report [31] showed that the sensitivity and specificity of urine dipstick positive for leukocytes or nitrites

Table 3.4 Calculating relative risk and odds ratio.

	Have outcome	Do not have outcome	Total
Exposed to risk factor	A	B	A+B
Not exposed to risk factor	C	D	C+D

Relative risk (RR) = ratio of risks of outcome: $RR = A/A+B \div C/C+D$.

Odds ratio (OR) = ratio of odds of outcome: $OR = A/B \div C/D$.

Table 3.5 Calculating sensitivity, specificity, positive predictive value (PPV), negative predictive value (NPV) and likelihood ratio (LR).

	Disease present	Disease absent	Total
Test is positive	True positive (A)	False positive (B)	A+B
Test is negative	False negative (C)	True negative (D)	C+D
Total	A+C	B+D	A+B+C+D
	Sensitivity = $A/(A+C)$	Specificity = $D/(B+D)$	

PPV = $A/A+B$. Positive predictive value is the proportion of people with a positive test who have the condition.

NPV = $D/C+D$. Negative predictive value is the proportion of people with a negative test who do not have the condition.

LR+ = $[A/(A+C)]/[B/(B+D)]$ or sensitivity/(1–specificity).

LR– = $[C/(A+C)]/[D/(B+D)]$ or (1–sensitivity)/specificity.

in diagnosing a UTI in a woman was 51% and 99%, respectively [32]. What does that mean?

A test with high sensitivity (most patients with disease have positive test results) is most useful to rule out disease, and one with high specificity like the example above (most of patients without the disease will have a negative result) is most useful to rule disease (Table 3.5).

The test with high sensitivity will have very few false negatives – therefore, if negative it can be believed. A test with high specificity will have very few false positives – so a positive test can be believed.

A helpful mnemonic is SpPIIn and SnNOut: a test with high specificity, if positive, rules disease in; a test with high sensitivity, if negative, rules disease out [33].

Likelihood ratios and pre-test and post-test probability

Likelihood ratio (LR) is a measure of the accuracy of a diagnostic test – the degree to which a test does what you want compared with a gold standard. It is the probability of a given test result being seen in patients with the disease compared with the probability of the same result being seen in patients without disease [34].

LR for a test with a positive result is calculated as sensitivity/(1 – specificity) (Table 3.5).

The corresponding calculation for a negative test result is: (1 – sensitivity)/specificity.

How do we apply LR in clinical practice? LR indicates by what factor the result of a given diagnostic test would increase or decrease the pre-test probability of a condition.

If the test has an LR of 1, the post-test probability of the target condition is exactly the same as the pre-test probability or the test is useless.

If the LR is greater than 1, this results in a post-test probability that is higher than the pre-test probability. If the LR is lower than 1, this results in a post-test probability that is lower than the pre-test probability.

The size of the effect is related to the size of the LR.

1 LRs greater than 10 or less than 0.1 create large and often conclusive changes from pre-test to post-test probability.

2 LRs of 5 to 10 and 0.1 to 0.2 create moderate changes in pre-test to post-test probability.

3 LRs of 2 to 5 and 0.2 to 0.5 generate small changes in probability.

4 LRs of 1 to 2 and 0.5 to 1 are unlikely to alter probability to a clinically significant degree.

Once an LR is determined we can use that to link the pre-test probability to the post-test probability by using a Fagan Normogram (Figure 3.2). This allows the post-test probability to be read off by aligning the pre-test probability and LR for a clinical scenario; it can be

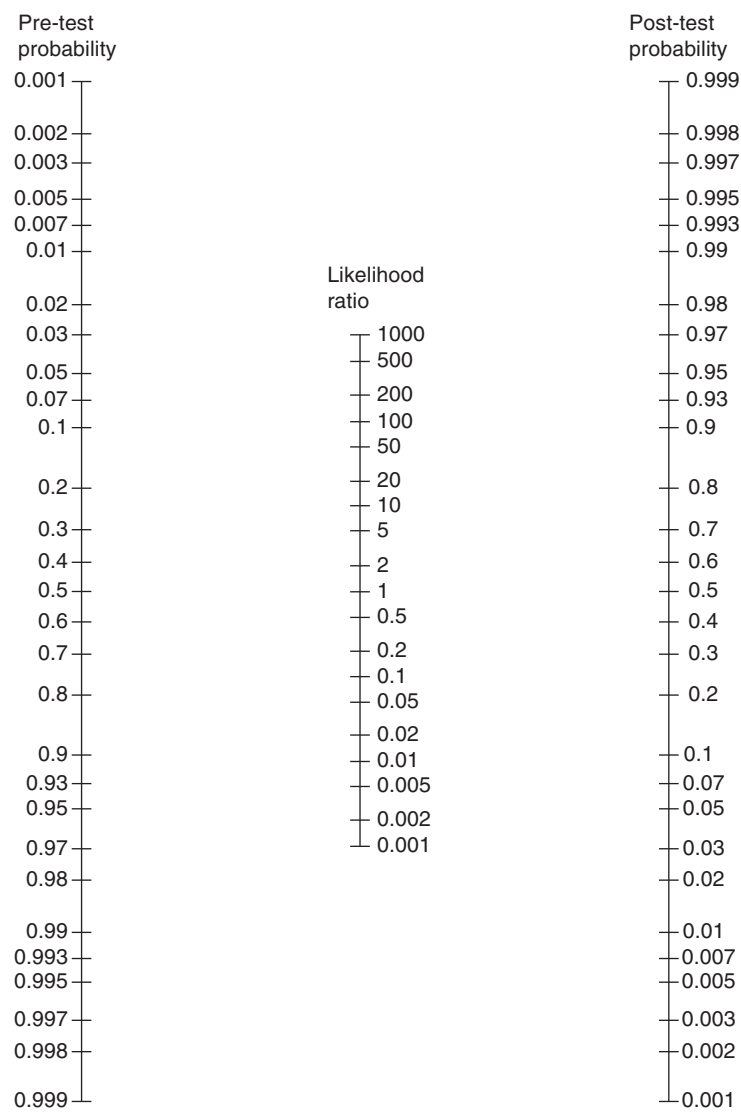


Figure 3.2 Fagan normogram. (Reproduced with permission from CEBM (www.cebm.net).)

Table 3.6 The usefulness of physical examination in the diagnosis of testicular torsion.

Physical finding	Sensitivity (95% CI)	Specificity (95% CI)	LR+ (95% CI)	LR- (95% CI)
Absent cremasteric reflex	96% (73%–100%)	88% (79%–93%)	7.9 (4.3–14.5)	0.04 (0.003–0.62)
Tender testicle	96% (73%–100%)	38% (28%–49%)	1.6 (1.3–1.9)	0.09 (0.006–1.46)
Abnormal testicular lie	46% (24%–70%)	99% (94%–100%)	72 (4–1215)	0.54 (0.33–0.88)
Tender epididymitis	23% (1%–50%)	20% (12%–30%)	0.29 (0.11–0.78)	3.95 (2.29–6.8)
Isolated tenderness (superior pole of testis)	4% (0%–27%)	83% (73%–90%)	0.21 (0.01–3.28)	1.17 (1.01–1.35)

CI, confidence interval; LR+, likelihood ratio of testicular torsion if the physical finding was present; LR-, likelihood ratio of testicular torsion if the physical finding was absent.

Adapted from Kadish and Bolte 1998 [36].

obtained from the CEBM (Centre for Evidence-Based Medicine) website (www.cebm.net).

In 2005, Schmitz and Safranek [34] from the University of Washington, evaluated the usefulness of physical examination in the diagnosis of testicular torsion and provided in Table 3.6. The patients all had testicular torsion, but how helpful were the exam findings?

If we apply the principles presented above only two findings are consistent with a high post-test probability and these are absent cremasteric reflex (LR- 0.04) and abnormal testicular lie (LR+ 72).

So, based on the high sensitivity of an absent cremasteric reflex, a positive test rules torsion out (SnNOut) but because of its lower specificity (88%) it is not as reliable in making the diagnosis of torsion (SpPIn).

Relatives and the number needed to treat [37]

Let us assume that the risk of UTI with the use of prophylactic antibiotics in patients with reflux is 2% and without antibiotics is 9%. The difference in rates is expressed as the absolute risk reduction (ARR) or risk difference ($ARR = 9\% - 2\% = 7\%$).

The same difference can be expressed as a relative risk reduction (RRR) ($(9\% - 2\%)/9\% = 0.78$ or 78%).

The relative measures including RRR, relative risk and odds ratio are helpful in understanding the efficacy of interventions.

The ARR can be converted into a useful quantity called the number needed to treat (NNT), which is the

number of patients one needs to treat to realize one improved outcome. This is the reciprocal of the ARR rounded to the next number ($NNT = 100\%/7\% = 15$). So, for the example above we would need to treat 15 patients in order to prevent one episode of urinary tract infection.

Confidence intervals

Of all numbers reported, the confidence interval is the most important in assessing clinical relevance.

P-values in scientific studies are used to determine whether a null hypothesis formulated before the performance of the study is to be accepted or rejected. In exploratory studies, P-values enable the recognition of any statistically noteworthy findings. Confidence intervals provide information about a range in which the true value lies with a certain degree of probability, as well as about the direction and strength of the demonstrated effect. This enables conclusions to be drawn about the statistical plausibility and clinical relevance of the study findings. It is often useful for both statistical measures to be reported in scientific articles because they provide complementary types of information.

Statistically, 95% CI describes an estimated range of values, calculated from the study data, that is likely to include a true population value. It is a way to indicate the precision of an estimate of a parameter from the study data.

A 95% CI that includes 0 (for studies evaluating absolute differences) or 1 (studies evaluating ratios) in its range indicates that the study has not rejected the null hypothesis.

The formula for calculating CI includes the number of patients in the study – the larger the number the narrower the CI and the more accurate the study.

Power calculations [39]

When comparing the efficacy of two treatments in a clinical trial, four outcomes are possible: (1) the study detected a true difference; (2) the study found a difference but there is no true difference (alpha error); (3) the study found no difference and there is none; and (4) the study demonstrated no difference but there is a “true” difference (beta error).

The *P*-value indicates the probability of an alpha error. The likelihood of beta error can be quantified before starting the study by providing a power calculation. The power is influenced by the number of study patients and the difference to be detected.

Power calculations are used when planning a study to determine whether a predetermined clinically meaningful difference, if present, will be detected. A power of 80% or 90% to detect this difference generally

is assumed to be sufficient to assure that a clinically meaningful difference will not be missed.

In a study that demonstrates no difference between two interventions, check to see whether the authors include a power calculation. Lack of a power calculation reflects a weakness of the study.

Multivariate analysis [40]

In most studies, the researcher is attempting to describe the relationship between an outcome variable and its several determinants. In order to examine the interactions and relations between a large number of determinants and the response variable simultaneously, simple univariate methods such as simple regression [41] would not be enough as it would provide a one-sided biased view.

Multivariate methods are based on multiple linear regression and they have the advantage of bringing in more information to bear on a specific outcome. They allow one to take into account the continuing relationship among several variables. This is very helpful in observational studies where total control is never possible [42,43].

The main methods of multivariate analysis are described in Table 3.7.

Table 3.7 Methods of multivariate analysis.

Method	Type of dependent variable	Type of independent variable	Purpose
Multiple regression analysis	Continuous (numerical)	Continuous but categorical may be used	To describe the extent, direction, and strength of the relationship between several independent variables and a continuous dependent variable
Logistic regression analysis	Categorical dichotomous	Continuous and categorical	To describe how many times more likely is the event in one group compared to the other
Analysis of variance	Continuous	All nominal	To describe the relationship between a continuous dependent variable and one or more nominal independent variables
Discriminant analysis	Nominal	Commonly all continuous	To determine how one or more independent variables can be used to discriminate among different categories of a nominal dependent variable
Factor analysis	Commonly continuous but may be any type	Commonly continuous but may be any type	To define one or more new composite variables or factors
Analysis of covariance	Continuous	Mixture of nominal and continuous variables	To describe the relationship between a continuous dependent and one or more nominal independent variables, controlling for the effect of one or more variable

The multiple regression analysis model assumes that the outcome variable is normally distributed. Unfortunately, in most research the outcome is not normally distributed and adjustments need to be made. To deal with this, a transformation is used to create a new variable that more closely approximates a normal distribution (taking the logarithm of length of stay) [42].

Propensity score analysis

This is an alternate method of risk adjustment [44]. In observational studies, investigators have no control over the treatment assignment. The treated and nontreated (that is, control) groups may have large differences on their observed covariates, which can lead to biased estimates of treatment effects. Even traditional covariance analysis adjustments may be inadequate to eliminate this bias. When the two groups are compared, logistic regression is used to calculate a given subject's risk or probability (propensity) of having the exposure of interest (robotic pyeloplasty versus open). The patient's calculated probability of receiving that treatment is the propensity score. It is defined as the conditional probability of being treated given the covariates, and this can be used to balance the covariates in the two groups, and therefore reduce this bias. In order to estimate the propensity score, one must model the distribution of the treatment indicator variable given the observed covariates. Once estimated, the propensity score can be used to reduce bias through matching, stratification, regression adjustment, or some combination of all three.

Generalizability

This refers to the ability to take information from a research study and apply it in the community. RCTs, for example, provide the highest level of evidence about the efficacy of competing interventions but where they take place (strict inclusion and exclusion criteria, conducted at academic centers) may limit the ability to reproduce the same quality and outcome in a non-research setting [45].

When reading articles, critical readers should consider why the outcomes or care patterns might not be generalizable in the broader community.

Determining causality using observational studies

One of the limitations of an observational study is that statistical analyses can only identify associations

between independent variables and the outcome of interest. Therefore be very cautious of observational studies in which the conclusion is that A causes B. Such studies can be hypothesis-generating and may show an association but causality can only be proven by a properly performed prospective trial [46].

Observational studies may reveal an association between exposure and outcomes. In order to infer causality the following must occur:

- 1 The exposure must happen before the outcome otherwise the exposure cannot plausibly lead to the outcome.
- 2 The association and hypothesized causal relationship must be clinically plausible.
- 3 The association between the exposure and the outcome must be large, and if there varying degrees of exposure there should be a varying magnitude of associations between exposure and outcome.

Critical appraisal of studies

When critiquing articles there are three basic questions that should be asked of all the articles:

- 1 Are the results valid?
- 2 What are the results?
- 3 Are the results applicable to my patients? [47]

Critical appraisal of prognostic studies

As many studies published in pediatric urology deal with prognosis (rate of complications after hypospadias surgery, enterocystoplasty, bladder exstrophy), a critical assessment of the study design, patient population and results is very important in determining whether the results of the study are valid.

Therefore in assessing these studies the following questions should be asked:

- 1 Was the representative sample of patients assembled at an early course of their disease or condition?

It is usually preferable to enroll patients at an early time in the disease (newborn with hydronephrosis). The patient population studied should also be representative of the underlying population. Therefore, studies published by pediatric urologists on the management or outcome of children with vesicoureteral reflux may not be representative of the population. A recent report from Stanford University showed that 39% of pediatricians tend to manage patients with vesicoureteral reflux without referring to a specialist [48]. So, for example, assessing the rate of urinary

tract infections in patients with vesicoureteral reflux using “urological” databases may be skewed or biased.

2 Was patient follow-up sufficiently long and complete?

Length of the study should be enough to detect the outcome of interest. This will therefore vary by the outcome. So if the outcome of interest in repair of hypospadias is fistula rate, follow-up until toilet training may suffice. On the other hand, if the outcome of interest is satisfaction with the appearance, function or satisfaction the follow-up will require several years.

3 Were outcome criteria clearly defined and objective?

A clear definition of outcomes should be provided and it is important that the individual determining the outcome does not know if the patient has a potential prognostic factor.

4 Did adjustments for important prognostic factors take place?

A prognostic factor is patient characteristic (e.g. age, grade of hydronephrosis, grade of reflux, ethnicity, circumcision status) that predict the patient’s eventual outcome. The study should adjust for known prognostic factors in the analysis so the results are not distorted.

5 How likely are the outcomes over time?

Often studies are reported as a rate (the number of patients experiencing the event) which is simple, and easily understood and communicated. Unfortunately, rates convey little information about important differences between studies within similar summary rates. Therefore, survival curves are used to estimate survival of a cohort over time. It is a useful method for describing any dichotomous outcome (not just survival) that occurs once during the follow-up period.

6 How precise are the prognostic estimates?

To determine the precision of the estimates we need to look at the 95% CI around the estimates. The narrower the CI, the more useful the information.

Clinical appraisal of diagnostic studies [49]

Questions to be asked while critiquing such articles are as follows:

1 Was the diagnostic test evaluated in a representative spectrum of patients and similar to those that would be treated in practice?

Ideally, the diagnostic test should be applied to a spectrum of patients. It is also best if patients are

randomly selected or consecutive patients are evaluated in order to reduce selection bias.

2 Was the reference standard applied regardless of the index test result?

Ideally both the index test and the reference standard should be carried out on all patients. This may not be applicable if the test is invasive and an alternative is to follow these patients long enough to see if they are truly negative.

3 Was there an independent blind comparison between the index test and an appropriate reference (gold) standard of diagnosis?

The reference standard should be appropriate and as close to the truth as possible. Second, those who interpret the results of one test should not be aware of the results of the other test.

4 Are test characteristics presented?

Two types of results are commonly reported in diagnostic test studies. One concerns the accuracy of the test and is reflected by the sensitivity and specificity. The other concerns how the test performs in the population tested and is reflected in predictive values (post-test probabilities).

5 What are the measures?

Sensitivity, specificity, PPV, NPV.

6 Were the methods for performing the test described in detail to permit replication?

Clinical appraisal of systematic reviews [50]

1 What question (PICO) did the systematic review address?

The main question being addressed should be clearly stated. The exposure and the outcome of interest should be expressed in terms of a simple relationship.

2 Is it unlikely that important relevant studies were missed?

Starting points are the major bibliographic databases, but should also include reference lists from relevant studies including abstracts presented at scientific meetings such as the APP or the AUA. The search should not be limited to the English language alone. The search should include both MESH and text words.

3 Were the criteria used to select articles for inclusion appropriate?

The inclusion or exclusion of studies in a systematic review should be clearly defined *a priori*. The

eligibility criteria used should specify the patients, interventions or exposure and outcomes of interest. In many cases, the type of study design will also be a key component of the eligibility criteria.

- 4 Were the included studies sufficiently valid for the type of question asked?

The article should describe how the quality of each study was assessed using predetermined quality criteria appropriate to the type of clinical question (randomization, blinding and completeness of follow-up).

- 5 Were the results similar from study to study?

The results ideally should be similar or homogeneous. If heterogeneity exists, the authors may estimate whether the differences are significant (chi-square test). Possible reasons for the heterogeneity should be explored.

- 6 What were the results?

A systematic review provides a summary of the data from results of a number of individual studies. If the results of the individual studies are similar, a statistical method, meta-analysis, is used to combine the results from the individual studies and an overall summary estimate is calculated. The meta-analysis gives weighted values to each of the individual studies according to their size. The individual results of the studies need to be expressed in a standard way, such as relative risk, odds ratio, or mean difference between the groups. Results are traditionally displayed in a figure called a forest plot as shown in the Figure 3.3).

The forest plot shown in Figure 3.3 depicts a meta-analysis of seven studies. The individual studies are represented in the squares and a horizontal line which corresponds to the 95% CI of the odds ratio. The size of the square reflects the weight of the study in the meta-analysis. The solid vertical line corresponds to “no effect” of treatment – an odds ratio of 1.0. When the

confidence interval includes 1 it indicates that the result is not significant at conventional levels ($P > 0.05$).

The diamond at the bottom represents the combined or pooled odds ratio of all seven trials with its 95% CI. In this case, the diamond overlaps the “no effect” line and therefore the pooled OR is not statistically significant.

Evaluating for heterogeneity

This can be statistically performed using statistical testing such as the Cochran Q test. It can also be eyeballed if one looks for overlap of the confidence intervals of the trials with the summary estimate. In the plot shown in Figure 3.3 there is significant heterogeneity as there is little overlap between the summary estimate CI and the studies.

Heterogeneity can also be assessed using the Cochran Q test. The level of significance for Cochran Q is often set at 0.1 due to the low power of the test to detect heterogeneity. If Cochran Q is statistically significant there is definite heterogeneity. If Cochran Q is not statistically significant but the ratio of Cochran Q and the degrees of freedom (Q/df) is >1 there is possible heterogeneity. If Cochran Q is not statistically significant and the $Q/df < 1$ then heterogeneity is very unlikely.

Braga et al. recently conducted a systematic review and meta-analysis on the use of antibiotic prophylaxis. The goal of this study was to conduct a systematic evaluation to determine the value of continuous antibiotic prophylaxis in reducing the rate of UTIs in this patient population. They found significant heterogeneity in the studies reported. Most of the studies were retrospective in nature and over 30% of the articles that were included in the study were of low quality. As a result, they concluded that there is suggestion that antibiotic prophylaxis may be beneficial in infants with high grade reflux but the level of evidence was moderate to low.

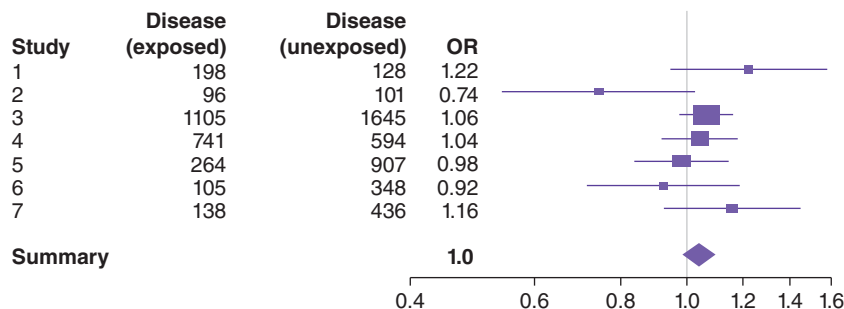


Figure 3.3 Example of a forest plot.

Are the results of the studies applicable to my patients?

This question should be asked by all pediatric urologists before incorporating trial recommendations into their practice.

If a randomized trial had very rigid inclusion criteria for enrolment, the results may not be applicable to the larger population of patients. Caution should be exercised in generalizing the results to other populations. Patient age in pediatric urology poses a problem when generalizing results. Before accepting any treatment as superior to another the relevance of the major outcome measures should be assessed. Most studies comparing laparoscopic pyeloplasty to robotic pyeloplasty to open pyeloplasty often use length of stay and narcotic use as major outcome measures. Narcotic use is not an objective measure as there are many factors that affect this measure, including variability in pain scores, experience of nursing staff in assessing pain and administering narcotics. Length of stay, even though an important measure to evaluate cost, is not the most clinically relevant outcome. The most important outcomes are resolution of obstruction and postoperative morbidity.

Finally, before applying any new practice we as surgeons should consider our skills in performing these procedures. Surgeons should ask themselves if they are proficient in performing a given operation. Proficiency with a particular new operation is an important component of its effectiveness. As such, surgeons are faced with three choices: (1) proceed with another operation; (2) refer the patient to a colleague who is proficient; or (3) seek additional training to master the operation.

A recent article by Lendvay et al. evaluated the importance of practice before performing tasks. They observed significant performance improvement and error reduction rates among surgeons of varying experience after virtual reality warm-up for basic robotic surgery tasks. In addition, the virtual reality warm-up reduced errors on a more complex task (robotic suturing), suggesting the generalizability of the warm-up [52].

More importantly, surgical proficiency highlights a fundamental difference between medical and surgical trials. A lack of surgical proficiency will bias a randomized control trial in favor of the simpler of two operations. On the other hand, a randomized trial of a very complex operation performed by a skilled surgeon may not be applicable to the larger surgical community.

Conclusion

The practice of pediatric urology is complex and it is the responsibility of all pediatric urologists to practice using the best available evidence. Hopefully, this chapter will serve as a guide for critically appraising the pediatric urological literature.

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CHAPTER 4

Implications of a poor surgical outcome

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KEY POINTS

- From the patient perspective a poor outcome could be anything from a minor delay in surgery, incorrect side surgery to loss of life
- An apology and explanation if a poor outcome or suboptimal result is achieved is strongly recommended. This also holds true where an error is made that is not immediately obvious to the patient
- Patient complaints can be managed by local investigation and resolution or through to litigation; the outcome for the surgeon may be decided locally by the employing organization or by the medical regulatory authority in their country

Introduction

One of the few stimuli prompting a unanimous response from any group of surgeons is the failure to produce the desired result, either from an operation or from a program of management. The response will be a mixture of empathy, regret, disappointment, frustration ... and a lingering fear that litigation, or worse, may ensue. In 21st-century practice, a “poor outcome” can encompass anything from the irritation of a minor delay due to a misplaced ultrasound report, to the loss of the wrong kidney, or possibly a life. This chapter deals with how to approach a patient with a poor outcome whilst describing the legal pathways that can be taken. It reviews the situation from the viewpoint of the English legal system. Every country has slightly different laws but this chapter remains relevant for those surgeons practicing within and outside of the devolved nations in the United Kingdom, as parallels can be drawn with most legal systems.

Disclosure of poor outcome

An apology

Doctors often debate the advisability of giving an apology in these circumstances, fearing that this could be

construed as an admission of guilt, analogous to the advice given to motorists by their insurance companies if involved in a collision. The analogy is flawed. A poor outcome from treatment *may* be the result of mismanagement, but the final determination of fault, if present at all, will result from a complex investigation and logical assessment. Whilst it could be argued that a fulsome apology may be seen as an indication of “guilt,” this will have a minimal effect on the process that will establish whether a doctor’s behavior has fallen below the reasonable standard (and, if so, whether this lapse has caused the harm that is alleged). This effect has to be balanced against the undoubted good that an apology will do, i.e. benefiting the patient and reflecting well on the doctor’s propriety and openness. An apology is therefore very strongly recommended. At the very least, failing this, an expression of regret for the plight in which the patient finds herself is mandatory.

An explanation

An explanation as to how the suboptimal result has occurred is also necessary. Our society has chosen to regard a patient’s autonomy, their right of self-determination, as the paramount consideration when dealing with their health. This manifests as a primacy for confidentiality and for the need to provide consent,

both of which are sometimes put ahead of what may be in the (medical) best interests of the patient.

It therefore follows that any information that a doctor possesses concerning a patient must be shared with that patient, or the parents. There is a theoretical tension when considering children, since a child who is judged to be competent to provide consent is also entitled to decide whether their information should be shared with their parents. In practice, this entitlement should be honored by simply checking with the competent child that they have no objection to their parents being told; in the vast majority of cases, there will be none. So a full discussion should follow, to enlighten the family as to how the poor outcome has occurred.

An obligation to disclose errors that are not immediately obvious to the patient

One particular difficulty is where an error has been made that is not immediately obvious to the patient, but has caused some tangible harm. There may be a temptation not to disclose. However, truth-telling is a cornerstone of a trusting relationship, and trust between individuals is central to civilized life. Since “morality” pertains to character and conduct and has regard to the distinction between right and wrong, truth-telling seems to be, inescapably, a moral activity. In families’ moral education of their children, the universal duty adults impose on children to “own up” to misdemeanors reflects this necessity to ensure that ordinary citizens are honest with each other. It also implies that “honesty” concerns the disclosure of hidden information, not simply the avoidance of the lie.

The relationship between doctors and their patients is not ordinary. It is described as a fiduciary relationship, emphasizing the necessity for mutual trust, confidence, and certainty (*L. fiderer*: to trust; *fides*: faith). In conclusion, considering the fiduciary relationship between doctors and their patients, and the lack of distinction between a lie and failing to disclose hidden information, there is a moral obligation to disclose.

Within the doctrine of behavioral ethics, the central “good” elements of human behavior rest upon honesty, probity, and truthfulness. From this perspective, disclosure of error would be considered as an ethical obligation.

How does the general public approach disclosure? In reality, generally by ignoring the ethical and moral obligations outlined above. The man who owns up to

scratching his neighbor’s boat while it was unattended would be perceived to have done the “right thing,” but such behavior might generate both mild surprise and congratulations on being “decent.” Failure to report the damage would lead to a disconsolate but unsurprised owner, resigned to the fact that “no one ever owns up these days.”

On a larger scale, viewing “acknowledging error” on Google reveals a robust avoidance of the obligation: “White House strategists conclude that acknowledging error is not an effective political tactic” [1]. Such comments recognize the moral obligation, but honor it in its avoidance. The general tenet of civil law is that the citizen should look after himself. There is no evidence of a civil obligation to report an error.

Thus the starting position is that there is a general moral presumption that citizens and organizations should be truthful. The National Health Service (NHS) is an arm of the state, and would normally be expected to be truthful. However, truth and candor are different qualities, and it is when harm occurs during clinical therapy that candor must be engaged.

The launch of the NHS Constitution for England in January 2009 included principles of “Being Open”, indicating that the “NHS also commits (to an arrangement that) when mistakes happen ... if anything goes wrong ... (the NHS organization will) acknowledge, apologise and explain the wrongful events to patients, relatives and carers”. The most recent changes to the NHS Constitution formalizes this promise into a contractual duty of candor [2].

However, the “Being Open” policy is explicitly predicated on patient safety incidents: “The Being Open process [3] *begins with the recognition* that a patient has suffered harm or has died as a result of a patient safety incident” (my italics).

The duty of candor has been canvassed at common law, and was adopted by the General Medical Council (GMC); thus [4] if a patient suffers “harm or distress,” doctors are admonished to put matters right.

Although this advice is not binding in the formal legal sense, it does represent the view of a recognizable body of medical opinion. Although civil judges do not invariably follow GMC guidance, they may be influenced by it and, if they choose to, may reflect the guidance in any future judgments. What damage could flow from failure to know that something has gone wrong? The patient’s eventual discovery may cause distress, but it

could be difficult to claim that this distress equates to personal damage of a degree that a court would view as worthy of financial recompense. *Chester v. Afshar* (2004) made lawyers (briefly) believe that there was an appetite among senior judges for expanding the law [5], creating a new tort of “failure to report a medical error,” a novel category of clinical negligence. Within a matter of weeks, a further House of Lords case (*Gregg v. Scott* 2005) indicated that radical expansion of this basis of liability was, for the time being, most unlikely.

Many NHS hospitals regard failure to report a serious untoward incident *to the hospital* as a disciplinary offence. Pending the introduction of the contractual duty of candor, there is no defined obligation to disclose the information to the patient, and one can see a potential conflict of interest on behalf of the hospital when deciding to disclose or not. However, should your hospital take a similar line, reporting a clinical error that could be construed as serious would seem prudent. But from a moral and ethical point of view, patients should also be told, before the hospital, because of the GMC’s guidance and the fiduciary relationship a doctor has with the patient. Failing to disclose to the patient has not yet created a liability in negligence, and even if the failure were admitted as falling below the reasonable standard of care, the claimant would have an uphill struggle in proving causation (*vide infra*). The National Patient Safety Authority’s (NPSA) safer practice notice [6] advises health care staff to “apologize to patients, their families or carers if a mistake or error is made that leads to moderate or severe harm or death, explain clearly what went wrong and what will be done to stop the problem happening again.” This, combined with the advice from the GMC, increases the chances of being liable to civil actions; but will only succeed if judges choose to follow the line of expanding the scope of clinical negligence.

However, from April 2013 in England, the constitutional change within the NHS ensures that hospital trusts (and thus their employees) will have a contractual duty to disclose harm that may not otherwise be obvious to the patient or to their relatives. It seems inevitable that the attitude of the employing hospitals, and in parallel, the courts, will harden to evidence of intentional nondisclosure.

Following a review and consultation by the Department of Health in 2014, the definition of “serious harm” when considering the threshold for candour was

aligned with the definition used in the management of adverse clinical events. Accordingly, the threshold for being candid with patients was set so that when as a result of clinical treatment the patient died; or suffered severe or moderate harm, the duty to be candid was engaged. Moderate harm was in turn defined as “requiring a moderate increase in treatment; significant but not permanent harm; or prolonged pain or psychological harm.” No advice was given as to whether an error or mistake of treatment was required. However, the eight professional regulators of healthcare in the UK then provided a joint statement, noting that “when something goes wrong with treatment or care which causes or has the potential to cause harm or distress,” the duty of candour is engaged.

From the professional point of view, given the GMC guidance, full disclosure is necessary. In the rare case where disclosure would cause clinical harm, perhaps psychiatric injury, the doctrine of therapeutic privilege will protect the doctor who correctly applies it and withholds disclosure. As a clinical decision, disclosure of medical error puts the doctor in an unsailable position. The hospital may wish disclosure had not occurred, but will hardly make their displeasure visible. Paradoxically, there is evidence that disclosure of error reinforces, rather than diminishes, the relationship between doctor and patient. Even if the admission leads to litigation, the court is likely to view the voluntary disclosure much more favorably than apparent concealment.

From the perspective of the patient

Local resolution

If the patient complains of the outcome, the complaint will initially be dealt with locally in the hope that resolution can be achieved. At the time of writing, this means that the claim is investigated by the hospital, which may involve experts from within or outside the organization to take an initial view on whether the hospital should accept liability for the poor outcome. If this initial investigation and suggested remedy satisfies the patient, the matter is brought to a close. If not, then the complainant may request a convenor to appoint a panel to hear the case. The panel consists of a lay chairman and two members independent of the hospital. Clinical assessors are appointed to advise

the panel when the complaint involves the exercise of clinical judgment. There is no provision for an appeal of the panel's decision.

The Ombudsman

If the complainant is refused panel review, he may refer that decision to the Health Service Commissioner [7] (Ombudsman), who may recommend that the decision to refuse a panel review be reconsidered. The Ombudsman is able to investigate complaints about clinical judgment, including those arising from independent providers of health services [8] and retired practitioners [9]. The Commissioner provides comments and recommendations, but has no power to award compensation other than *ex gratia* payments for out-of-pocket expenses. It is not allowed to refer the case to the panel or the Ombudsman if either civil or criminal proceedings have started.

NHS redress

Following a report [10] by the Chief Medical Officer (CMO) in 2003, there are arrangements in England to create a scheme for redress [11] without civil proceedings. This scheme will cover injuries caused to patients by an act or omission concerned with diagnosis of illness, care, or treatment. Despite what some Parliamentarians may have believed during debates, this is not a no-fault compensation scheme. It is anticipated that it will cover treatment by the NHS, even if provided in private hospitals. However, the scheme cannot be engaged if civil proceedings have begun, and will terminate immediately if they commence.

The scheme must comprise: an explanation, an apology, a report on the action proposed to avoid future similar cases, and an offer of compensation. The latter may be monetary, or could take the form of a contract to provide restorative care and treatment. If monetary compensation is awarded for pain, suffering, and loss of amenity, there must be an upper limit on the amount to be offered. The implementation of such a scheme will be dependent on political will and the funding that is diverted to pay for it. If it is implemented in the described form, there may be a perverse incentive for the hospitals to settle low-value claims irrespective of liability. If the complainant agrees to the offer, civil action will no longer be available to them. But even a relatively low guaranteed offer will seem attractive to both the hospital and the complainant, since it provides financial closure to both parties, a far cry from

the uncertainty and expense of civil actions. What it fails to provide is the guarantee that the doctor who is not at fault will have their blamelessness and, hence, good reputation publicly acknowledged.

Litigation

If the local or extended resolution processes fail, or the patient wishes for redress in the civil courts, litigation will commence. The action will usually be directed against the NHS hospital, only naming the defending surgeon if the complaint arose from private practice. The purpose will be ostensibly to provide the claimant with the full facts relating to the case, and financial compensation. The claimant is likely to be suing on the grounds of the tort (civil wrong) of negligence. To establish this, several separate elements will need to be established. First, that the surgeon was responsible for the claimant's care at the time of the incident. In hospital practice this is usually straightforward. A doctor has a single and comprehensive duty to exercise reasonable care and skill in diagnosing, advising, and treating the patient [12]. The test is whether the surgeon's conduct was reasonable, and this will be determined in comparison to the objective standard, which is the standard of his or her peers: "it is sufficient if he exercises the ordinary skill of an ordinary competent man exercising that particular art" [13]. Therefore, the claimant has to show that the surgeon's practice fell below the standard that would be set by his or her professional peer group [14]. It is this "standard of care" that is established by the expert witnesses who will be consulted concerning the case. However, the expert witnesses will also have to satisfy the judge that the standard they have identified can stand up to scrutiny, and be found to be coherent and logical [15]. Finally, the claimant has to show that the injury sustained was caused by the lapse in the standard of care, and that the damage must be such that the law regards it proper to hold the defendant responsible for it. The first of these two elements of causation is established on the basis of expert evidence, the latter by the court.

In private practice, the patient has the additional option of bringing an action in the law of contract, on the basis that they have purchased a service that has been imperfectly delivered. If a consent form has identified a particular surgeon, and the operation is performed by another, or if a surgeon fails to perform a promised procedure, then breach of contract will occur. An explicit and unequivocal guarantee – "I assure you that your

vasectomy will be successful and you will never father children again” – creates a contractual warranty on the basis of which action may be taken.

From the perspective of the surgeon

Local

The consequences for the surgeon are largely dependent on the magnitude of the damage caused. There is no doubt that close attention to and compliance with local procedures of incident reporting is vital to limit the negative consequences for the surgeon. The medical defence organizations emphasize the importance of alerting them to any potential claims as soon as a complaint has been made. If frequently-repeated irritating inconveniences are being caused to patients, it is likely that the surgeon will be asked to play his or her part in eliminating any procedural errors responsible. Where serious injury has been caused, many hospitals will investigate the matter either through a system of root cause analysis [16], or using some form of review group, usually composed of senior clinicians, risk managers, and executives. Should a serious breach of professional conduct be suspected, the Medical Director of the hospital will become involved and may refer the matter for local adjudication (the “Three Wise Men” approach) or may refer it on to the GMC. In the rare circumstance when a patient’s death may have been caused by gross incompetence, the possibility of a criminal charge may be considered. The police would interview all concerned, and the Crown Prosecution Service (in England) would then consider whether a conviction was likely on the basis of the written evidence, and whether it was in the interests of justice that the doctor should be charged with gross negligence manslaughter. It is noteworthy that the indemnity insurance that covers hospitals in England [17] is not available for criminal matters, reinforcing the importance of maintaining close contact with the medical defence organization.

National Clinical Assessment Service (NCAS)

This organization [18], soon to be part of the NHS Litigation Authority (NHS LA) [19], “work(s) with all parties to clarify the concerns, understand what is leading to them and make recommendations to help practitioners

return to safe practice.” It is therefore sometimes consulted about surgeons who have attracted the attention of their organization, through (amongst other things), unwanted surgical outcomes.

General Medical Council (GMC)

Having been established as a result of lobbying by the BMA in 1858 [20], the GMC [21] sets standards for doctors and investigates the facts relating to their performance and behavior when compared with those standards. This important role acts as a safety net, allowing cases to be considered that are not actionable at law but fall below the standard that should be expected of an ethical practitioner.

From the surgeon’s point of view, it is the hearing at the Fitness to Practise Panel, conducted not by the GMC but by its sister quasi-judicial Medical Practitioners’ Tribunal Service (MPTS) [22] that will determine whether his or her fitness to practice is impaired. If it is, a reprimand may be issued; conditions may be imposed on the doctor’s registration, or suspension, or erasure ordered. An appeal mechanism to the courts is available to doctors throughout the United Kingdom. But an appeal is also available to the GMC, if it is dissatisfied with MPTS’ decision-making.

The media response

The feeding frenzy created when a poor surgical outcome is reported in the media needs no further description, but has to be acknowledged as a key element that needs to be considered and “managed” in the broadest sense. For reasons possibly derived from self-interest, hospitals are becoming more accustomed to dealing with the media, and it would make sense to shelter under any cover that may so be provided. It seems unlikely that many surgeons will “win” in a direct encounter with journalists, and there is ample evidence that neither adequate recompense nor retractions will follow unjust or misleading reporting. Bold public assertions are therefore inadvisable.

From the perspective of the hospital

Adverse event reporting and clinical governance

The response to failures at either end of this spectrum should be proportionate, but follow a surprisingly

similar pattern. In each case, an apology and explanation is appropriate. In many institutions, all “adverse events” are collected into a central database, better to understand the systemic weaknesses that may have caused or contributed to the failure. These institutions in their turn share the database with those governing health care. In the context of the NHS, the data are collated with the Clinical Quality Commission, providing the overarching control of “quality” within the national service.

NHS Act 1977 Practice Direction 2006

This legislation gives a Strategic Health Authority (SHA) the power to issue an “alert notice,” naming an individual whom it considers poses “a significant risk of harm to patients, staff, or public, and who may seek work in the NHS” [23]. A draconian measure, the issue of such an alert may be requested by the chief executive or executive board member of an NHS body. The notice is sent to the National Clinical Assessment Service, the chief executive of each SHA in England, and the CMO for the other areas in Great Britain. It may then be sent to any NHS body that may be approached by the subject of the notice in search of work in the NHS. Such a notice must be reviewed at intervals of no more than six months. Alert letters have been available since 2002 [24], but the new guidance is a timely reminder of the efforts being made to protect the public from harm.

What is alarming is that the qualifying criteria for the issue of an alert could be interpreted by an executive in an ill-considered way, at a time when they were feeling vulnerable, perhaps having attracted unwelcome media interest. Although there is provision for revocation of a notice, the damage to a clinician’s reputation caused by a false allegation will be impossible to fully retract. Furthermore, the SHA is required to maintain a record of revoked notices for five years following revocation. It seems at least possible that this may disadvantage an “innocent” revokee who applies for work in the SHA area during the time period.

There is a requirement for the SHA to satisfy itself of the evidence supplied by the hospital, supporting the contention that there is a significant risk of harm. But it should be noted that it is the *risk of harm* that has to be significant, not the *degree of harm* itself. Should the climate develop in which unscrupulous health service managers behave aggressively

toward clinical staff, it could be seen how a poor surgical result, from the practice of a “troublesome” clinician, could lead to a disproportionate and unjust outcome. It would be incorrect to leave the impression that such devastating consequences are likely to flow from a poor surgical outcome. Such an outcome would be disproportionate and, in the present climate, exceedingly unlikely. However, the NHS is going through an unprecedented transformation of such magnitude that the “old rules” can no longer be relied upon. Government thinking appears to be challenging all “core values” relating to where patients are treated, and what level of training needs to be achieved as a prerequisite for treating them. In this climate, it will be prudent to acknowledge the potential, as well as the probable, consequences of a poor surgical outcome.

Conclusion

A poor surgical outcome is a miserable business for both patient and surgeon. Mercifully, few poor outcomes lead to litigation, and fewer still to the GMC or criminal courts. A prompt apology and full explanation will do a great deal more good than harm in the long term, and will make it more likely that the relationship between the surgeon and the family will recover and prosper. In many parts of the world, including England, there is a search for alternative modes of recompensing patients who claim to have suffered harm. This could lead to schemes that pay out small sums of money on the basis of scant evidence. Such schemes will flourish if they result in reducing the national financial burden of clinical negligence litigation, which is the reason for their existence. Hospitals may settle such claims irrespective of the effect on the surgeon’s reputation, and we all need to consider how we will handle that. As governments become more aware of the voters’ focus on the provision of health care, they are identifying measurable “quality” as a surrogate for success. In the United Kingdom, this scramble for the high ground of quality assurance has led government to provide itself with additional statutory tools of scrutiny and control. In this environment, careful compliance with local procedure in the event of an unwanted outcome becomes increasingly important.

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PART II

General Principles

CHAPTER 5

Endocrine and metabolic response to surgery and its management

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KEY POINTS

- Surgery evokes neuroendocrine, metabolic and inflammatory changes that characterize the stress response
- There is increased secretion of pituitary hormones and activation of the sympathetic nervous system
- There is an initial catabolic phase, with increased secretion of cortisol, catecholamines and glucagon to metabolize substrates and provide energy
- The catabolic phase is followed by the anabolic phase with increased secretion of insulin and growth hormone
- Fluid and electrolytes retention occur to maintain hemostasis
- Neonates have reduced glomerular filtration rate and impaired ability to concentrate urine and maintain electrolyte balance
- Postoperative hyponatremia is the most common electrolyte imbalance caused by administration of hypotonic fluids and increased production of antidiuretic hormone

Introduction

The “stress response” following surgery is complex and includes neuroendocrine, metabolic and inflammatory aspects resulting in substrates mobilization, muscle protein loss, sodium and water retention, immunological and hematological changes. Psychological and behavioral changes occur in association with the physiological events. All these events have been characterized in relation to surgery although similar features occur following trauma, burns and severe infections. The magnitude of the response is generally proportional to the surgical injury. The overall effect is the catabolism of stored body fuels.

The endocrine response to surgery

Initiation of the stress response to surgical trauma is characterized by activation of the sympathetic nervous system and increased secretion of pituitary hormones.

The pituitary hormones have effects on target organs with production of different secondary hormones resulting in increased catabolism, mobilization of substrates to provide energy and retention of salt and water to maintain fluid volume and homeostasis. In general, catabolic hormones such as catecholamines are released whereas anabolic hormones, such as insulin, are suppressed (Table 5.1). The overall result is an increase of the basal metabolic rate.

Sympathetic nervous system

Activation of the sympathetic nervous system results in release of catecholamines from chromaffin cells of the adrenal medulla and norepinephrine from pre-synaptic nerve terminals in response to hypothalamic stimulation. Impulses generated in afferent nerve endings (both unmyelinated C fibers and myelinated A fibers) at the site of tissue injury have a role in mediating the response to injury. Nerve impulses reach the thalamus via the dorsal horn of the spinal cord and the

Table 5.1 Hormonal response during surgery.

Gland	Change in secretion
Pituitary gland	Increased secretion of ACTH, GH, β -endorphin, prolactin, AVP Variable secretion of TSH, FSH and LH
Adrenal gland	Increased secretion of catecholamines, cortisol, aldosterone
Pancreas	Increased secretion of glucagon Decreased secretion of insulin
Thyroid	Decreased secretion of thyroxine and tri-iodothyroxine
Kidney	Increased secretion of renin
Other	Decreased secretion of testosterone and estrogen

ACTH, adrenocorticotrophic hormone; AVP, arginine vasopressin; FSH, follicle stimulating hormone; GH, growth hormone; LH, luteinizing hormone; TSH, thyroid stimulating hormone.

lateral spinothalamic tract. Afferent impulses reaching the thalamus mediate the response by activating the sympathetic nervous system. This leads to tachycardia and hypertension and release of renin from the juxtaglomerular cells of the kidneys, causing conversion of angiotensinogen to angiotensin I and subsequently to angiotensin II. In response to the production of angiotensin II there is secretion of aldosterone from the adrenal cortex leading to sodium and water in the distal tubule of the kidney. Aldosterone is responsible for 95% of the mineralocorticoid activity. Epinephrine promotes gluconeogenesis and inhibits insulin release, reducing peripheral glucose uptake, contributing to postoperative hyperglycemia. Finally, both epinephrine and norepinephrine increase free fatty acid mobilization and oxygen consumption.

Hypothalamic–pituitary–adrenal axis

Hypothalamic releasing factors are secreted into the hypothalamic–hypophyseal portal system during the stress response to surgery. They stimulate the secretion of anterior pituitary hormones. The anterior pituitary gland mainly releases adrenocorticotrophic hormone (ACTH) as part of the precursor proopiomelanocortin (from which β -endorphin and N-terminal precursor also originate), growth hormone (GH) and prolactin. The release of thyroid stimulating hormone (TSH), follicle stimulating hormone (FSH) and luteinizing hormone (LH) is generally unchanged during the stress

response to surgery. The hypothalamus also has direct neuronal control on the posterior pituitary gland controlling the release of antidiuretic hormone (ADH).

Adrenocorticotrophic hormone and cortisol

Adrenocorticotrophic hormone is a 39 amino-acids peptide that stimulates the adrenal cortical secretion of cortisol. The response is rapid, within a few minutes of the start of surgery. Cortisol accounts for 95% of the glucocorticoid activity and levels can increase 4–5 times within 4–6 hours of surgical trauma. The magnitude and duration of the increase is correlated to the severity of the insult and ACTH and cortisol levels remain persistently high up to 48–72 hours after major surgery as the usual negative feedback mechanism (high level of cortisol inhibiting ACTH production) is ineffective. This response is not abolished by the administration of corticosteroids but it can be modified by anesthetics intervention (see sections later in chapter). Cortisol has complex metabolic effects, including enhanced protein breakdown from muscles with increasing availability of plasma amino acids; it promotes gluconeogenesis in the liver and inhibits glucose use by cells resulting in increasing blood glucose concentration. Cortisol promotes lipolysis with production of gluconeogenic precursors from the breakdown of triglycerides into glycerol and free fatty acids. Cortisol has also mineralocorticoid effects, encouraging potassium loss and sodium and water retention. Finally, cortisol has anti-inflammatory effects mediated by the inhibition of macrophage and neutrophil infiltration in areas of inflammation and reduction of synthesis of inflammatory mediators such as leukotrienes, prostaglandins and cytokines.

Growth hormone

Growth hormone is a 191 amino-acids peptide with a major role in growth regulation, particularly in the neonatal period and childhood. GH has a minor role in the perioperative period and it has mixed catabolic and anabolic effects. Its effects are mediated by insulin-like growth factors (IGFs), mainly somatomedin C (IGF-1), produced by liver, muscle and other tissues. GH and IGF-1 levels increase significantly following trauma, returning to normal levels within few days [1]. The effects on metabolism include increased protein synthesis and inhibition of protein breakdown, thus promoting tissue repair. GH promotes lipolysis and has anti-insulin activity. Anti-insulin activity inhibits uptake of glucose from

peripheral cells leading to hyperglycemia. Finally, GH stimulates glycogenolysis in the liver.

Prolactin and β -endorphin

The release of these two hormones is increased during surgery although their role in the stress response is yet to be clarified. Prolactin has little metabolic activity and is mainly produced during pregnancy to stimulate milk secretion. Prolactin levels are higher in children following surgery, particularly in female subjects [2]. β -endorphin, an opioid peptide of 31 amino acids, is produced from the precursor proopiomelanocortin and has no major metabolic activity. It is an agonist of the opioid receptors and is important for its role as a natural pain-reliever in the body. In addition, it participates in the regulation of hunger, and it is linked to the production of sex hormones. β -endorphin is also released by leucocytes present in inflamed tissue. Both hormones might be implicated in the mediation of immune responses.

Thyroid hormones

The thyroid hormones tri-iodothyroxine (T3) and thyroxine (T4) are stored in the thyroid gland and secreted in response to TSH. T3, mainly formed in the tissues by the mono-deiodination of T4, is about five times more active than T4. They are mostly bound to albumin, thyroxine-binding pre-albumin and thyroid-binding globulin with only a low hormone concentration free in the circulation and active. They stimulate oxygen consumption in most tissues except brain, spleen and anterior pituitary gland. Their activity increases the metabolic rate and promotes heat production. T3 and T4 increase carbohydrates absorption from the gut, stimulate central and peripheral nervous system and influence growth and development. T3 and T4 also promote the activity of epinephrine and norepinephrine by increasing the number and affinity of the β -adrenoceptors in the heart. During surgery there is reduction in TSH concentrations that returns to normal limits within 2 hours after surgery. T3 and T4 production is also reduced and returns to normal limits in a few days. The mechanism of T3 and T4 reduction during surgery is unclear, but it has been shown that high cortisol levels after surgery induce T3 levels reduction in serum [3].

Gonadotropins

The significance of changes in FSH, LH, testosterone and estrogen after surgery are not clarified. Testosterone

and estrogen levels are reduced for several days post-surgery while LH levels are variable [4,5].

Antidiuretic hormone

Antidiuretic hormone is released by the posterior pituitary gland under direct hypothalamic neuronal control. ADH has a direct antidiuretic effect on the renal tubules. It is an important vasopressor causing arteriole constriction and decreasing output from sweat glands. It promotes the release of ACTH from the anterior pituitary gland.

Insulin and glucagon

Insulin is a polypeptide with two chains (21 and 30 amino acids) while glucagon is a 29-amino acids peptide, respectively produced by the β - and α -cells of the pancreas. The reduced secretion of insulin, mediated by the α_2 -adrenergic inhibitory effects of catecholamines, leads to breakdown of glycogen in the liver and muscles with consequent availability of glucose and lactate and mobilization of free fatty acids. Decreased insulin levels in the perioperative period are associated with reduced cellular response to insulin (so-called “insulin resistance”) with a state of “functional insulin deficiency” that contributes to the catabolic, hyperglycemic response. Although there is an increased release of glucagon from the pancreas this does not apparently play a major role in the hyperglycemic response.

The immune response to surgery

Following tissue injury there is an acute phase response activated by the production of different cytokines from macrophages, fibroblasts, endothelial and glial cells. Cytokine production reflects the degree of tissue trauma. Cytokines have an important role in mediating the immune and inflammatory response and they exert their effects locally (paracrine action) and systemically (endocrine action). Cytokine production is limited by negative feedback action of cortisol. The complex acute phase response is summarized in Box 5.1. The effects of interleukin-1 (IL-1), interleukin-6 (IL-6) and tumor necrosis factor- α (TNF- α) have been better characterized. IL-1 stimulates hepatic lipogenesis and gluconeogenesis and promotes muscle proteolysis. IL-6 increases within 30–60 minutes after commencement of surgery, reaching significant levels 12–24 hours following injury.

Box 5.1 The acute phase response.

Tissue injury and cytokines production
 Changes in vascular permeability
 Pyrexia
 Leukocytosis, neutrophilia and lymphocyte differentiation
 Acute phase proteins production: CRP, fibrinogen, α_2 -macroglobulin, serum amyloid A and P, metal binding proteins, complement, coagulation proteins, proteinase inhibitors.
 Reduction in serum concentration of: albumin, transferrin, α_2 -macroglobulin, iron and zinc
 Increase in serum concentration of: ceruloplasmin, copper
 Muscle proteolysis

It stimulates the production of ACTH *in vitro* and is a marker of the stress response, particularly in neonates. TNF- α promotes hepatic lipogenesis and muscle proteolysis and increases glucose transport.

Thermoregulation

Maintenance of normothermia is important in reducing the extent of the body's metabolic response. Patients are frequently mildly pyrexial for 24–48 hours following surgery and the metabolic rate increases by 6–10% for each 1°C change in body temperature. This occurs mainly because cytokines reset the temperature-regulating centers in the hypothalamus but also as a complication of infection occurring after injury. Children, and especially neonates, have more difficulty in regulating their body temperature than adults mainly because of the increased surface area/body mass ratio. The thermoregulating mechanisms are also immature until the end of infancy, and body temperature in children tends to be higher than in adults. It decreases toward adult levels during childhood and slowly declines to normal body temperature through puberty, stabilizing at 13–14 years in girls and 17–18 years in boys.

Neonates and infants produce heat by “non-shivering thermogenesis” using adipose tissue (brown fat). Brown fat makes up about 5% of the neonate body mass and is located on the back, along the upper half of the spine and toward the shoulders. Brown fat has a large number of mitochondria as the metabolism occurs locally,

in the brown fat cell. The mitochondria have a special protein (uncoupling protein 1) which allows the proton gradient of the external mitochondrial membrane to be dissipated without making ATP. The result is heat production. This is accompanied by an increase of the metabolic rate and an increase in the plasma concentration of noradrenaline. Surgery typically involves exposure to a cold environment, administration of unwarmed intravenous fluids, and evaporation from within surgical incisions potentially leading to hypothermia. Body temperature should be monitored during general anesthesia exceeding 30 minutes in duration and measuring body temperature (and maintaining normothermia) is now the standard-of-care during prolonged general anesthesia.

The metabolic response to surgery

The overall effects of the acute response consist of mobilization of carbohydrates, lipids and proteins together with retention of salt and water. The classic description of the metabolic response identifies three phases: the “ebb” (or “shock phase”); the “flow” and the “convalescence” phase. The ebb phase occurs soon after surgery or injury; it lasts a short period and is associated with a decline in body temperature and oxygen consumption. The flow phase is characterized by high metabolic rate, breakdown of proteins and fats, net loss of body nitrogen (negative nitrogen balance) and weight loss. This phase can last a significant period of time in relation to the level of injury and prompts significantly increased oxygen consumption and energy expenditure. During the convalescence phase proteins and fat reserves are restored and weight regained [6].

Carbohydrate metabolism

After surgery begins there is a rise in blood glucose concentrations. Hyperglycemia results from an increase in glucose production and mobilization, facilitated by catecholamines and cortisol. It is a major feature of the metabolic response to surgery. Blood glucose levels are directly related to the intensity of the surgical injury and remain high for >24 hours after surgery. Care is therefore necessary when utilizing glucose infusions and blood products. Hyperglycemia is also promoted by the initial reduction in insulin secretion, increased insulin resistance and decreased peripheral utilization

of glucose. Levels of blood glucose $>10\text{--}12\text{ mmol litre}^{-1}$ can cause impaired wound healing, increased wound infection and ischemic damage to myocardium and nervous system [7].

Protein metabolism

Initial response is characterized by inhibition of protein metabolism, followed by enhanced catabolism, stimulated by cortisol and various cytokines. Visceral muscle proteins and predominantly skeletal muscle proteins are degraded. Catabolism can lead to significant muscle wasting and weight loss. It is estimated that after major surgery in adults up to 0.5kg of protein per day might be lost. Protein loss can be estimated by measuring nitrogen excretion in urine. The amino acids released are utilized to maintain blood glucose levels as substrate for energy production and to produce the acute phase proteins in the liver. However, the production of albumin is reduced with possible imbalances in extracellular volume. It is important to highlight that standard nutritional support, enteral or parenteral, has proven to be ineffective in preventing protein catabolism. The use of “immune-nutrition” (glutamine, arginine, omega-3 fatty acid) has been shown to improve recovery in adults [8] although results in children are controversial [9]. Insulin infusion, with or without glucose, may reduce protein breakdown.

Lipid metabolism

Cortisol, catecholamines and GH enhance the conversion of stored triglycerides into glycerol and fatty acids. Glycerol is utilized in the liver for gluconeogenesis and free fatty acids are oxidized in the liver and muscles and converted into ketone bodies or re-esterified. The use of heparin during surgery can activate lipoprotein lipase with consequent lipolysis.

Water and electrolytes metabolism

Renin converts the plasma protein angiotensinogen to angiotensin I in the liver. Angiotensin I is then converted to angiotensin II by the angiotensin-converting enzyme in the lung capillary bed. The result is consequent release of aldosterone from the adrenal cortex promoting sodium and water retention as well as potassium and hydrogen ions excretion in the renal tubules. The concomitant reabsorption of chloride and bicarbonate ions helps to correct the acidosis secondary to tissue injury and maintain the normal pH. Aldosterone levels

increase promptly after surgery and its effects last up to a week [10]. Angiotensin II also acts on smooth muscles of the arteriolar wall, promoting vasoconstriction and increase in blood pressure. Water retention and potassium loss also occur as a result of the secretion of ADH. These effects may continue for 4–5 days after surgery. The atrial natriuretic peptide secreted by the cardiac atria in response to atrial stretching promotes the excretion of both water and sodium and suppresses the excretion of ADH, renin and aldosterone.

Perioperative fluid management in children

Perioperative fluid management in children has been the focus of considerable interest and debate in recent years with no consensus guidelines. Clinical practice varies considerably. The neonatal kidney has lower concentrating ability and reduced ability for tubular reabsorption of sodium and secretion of potassium and hydrogen ions compared to the older child and adult kidney. Glomerular filtration rate (GFR) increases in the first 2 years of life until it reaches adults levels. The normal infant will also lose up to 10–15% of its body weight in water during the first few days of life. Fluid management should take into account the administration of maintenance fluid, the replacement of any fluid deficit and the replacement of any losses, as well as the age of the child.

Maintenance of fluid requirements

Water requirement in children is normally 100 ml per 100 kcal/kg of energy expended. Maintenance of fluid requirements is routinely calculated according to the recommendations of Holliday and Segar [11] for children and infants older than 4 weeks of age, using body weight:

- 0–10 kg → 4 ml/kg/h
- 0–20 kg → 40 ml/h + 2 ml/kg/h above 10 kg
- >20 kg → 60 ml/h + 1 ml/kg/h above 20 kg

A maximum 24-hour volume of 2000 ml in females and 2500 ml in males is normally considered.

In neonates, with a gestational age >36 week, maintenance fluid volume requirements are reduced in the first few days after birth. In the neonatal period, maintenance fluids should be 0.18% saline in 10%

dextrose given at a rate of 4 ml/kg/h (100–120 ml/kg/day). Maintenance fluid requirements may need to be increased in children with pyrexia, excess sweating, and hypermetabolic states such as burns or when radiant heaters or phototherapy is used.

Oral fluid management prior to surgery

Children having minor surgery, such as circumcision and inguinal herniotomy, should be managed with oral fluids alone. There is general consensus that the administration of oral clear fluids up to 2 hours before surgery does not increase the risk of aspiration during anesthesia and helps to prevent dehydration. Most hospital guidelines will recommend fasting infants for 3 hours after breast milk and 4 hours after formula milk. Older children should be fasted of solids and milk for 6 hours prior to surgery.

Fluid deficit prior to surgery

Precise calculation of water deficit due to dehydration using clinical signs is usually difficult. In mild dehydration the useful clinical signs/symptoms are increased thirst and dry mucous membranes. In moderate dehydration the child can present with tachypnoea, cool/pale peripheries, prolonged capillary refill time, decreased skin turgor and sunken eyes. In severe dehydration the child can present with irritability, lethargy, deep (acidotic) breathing, sunken fontanelle and may also be hypotensive, which is a late premonitory sign. A more reliable method takes into account the difference between the current body weight and the immediate premonitory body weight if available. A child's water deficit in milliliters can be calculated following an estimation of the degree of dehydration expressed as a percentage of body weight (e.g. a 10 kg child who is 5% dehydrated has a water deficit of 500 ml).

The child having minor elective surgery usually has only a minor fluid deficit, which it is not necessary to correct.

In dehydrated children about to undergo major surgery it is accepted to give initial boluses of an isotonic fluid (0.9% sodium chloride or Ringer lactate/Hartmann's) of 10–20 ml/kg in the first hour. In dehydrated children who require urgent surgery, hypovolaemia should be corrected with an isotonic fluid or colloid, followed by a slower correction of residual dehydration with an isotonic fluid, taking into account ongoing losses, serum electrolytes and urine output.

Dextrose management during surgery

One of the issues is whether or not smaller children undergoing surgery need glucose administration. Small babies are capable of mounting a stress response secondary to surgery and it has been shown that it is possible to manage even small children by using glucose-free solutions without causing hypoglycemia during short surgical procedure. The rationale for this is that pronounced degrees of hyperglycemia may cause problems including osmotic diuresis, increased rate of wound infection, and less favorable outcome in situations where the patients may be at risk for periods of cerebral ischemia (i.e. cardiac and neurosurgical procedures). Neonates in the first 48 hours of life and small children having prolonged surgical procedures should be given dextrose during surgery (1–2.5% dextrose). Children having extensive regional anesthesia that will reduce the stress response should also receive dextrose.

Management of fluid losses during surgery

Fluid losses during surgery are normally replaced with an isotonic fluid such as 0.9% sodium chloride, Ringer lactate/Hartmann's solution, a colloid or a blood product, depending on the child's hematocrit. Children over 3 months of age may well tolerate a fall up to 25% of the hematocrit before requiring a blood transfusion. Stable, critically ill children might tolerate a hemoglobin level of 7 g/dl without having increased risk of adverse outcomes. Artificial colloids should be used instead of human albumin as there are no advantages demonstrated with the use of albumin. Third space losses due to sequestration of fluid into tissues at the site of surgery should be replaced with an isotonic fluid. Estimation of third space losses is made with 1–2 ml/kg/h for superficial surgery, 4–7 ml/kg/h for thoracic surgery and 5–10 ml/kg/h for abdominal surgery but could be less if procedures are performed laparoscopically.

Postoperative fluid management

During surgery the significant release of ADH and the use of hypotonic fluids in the postoperative period may cause hyponatremia. Ringer lactate/Hartmann's solution or 0.9% sodium chloride with 5% dextrose should be used in the postoperative period as these have been proven to reduce the incidence of hyponatremia compared to 0.45% sodium chloride [12]. In some patients, fluid restriction to 60–70% of full maintenance and

additional boluses of isotonic fluid given as required might be indicated. Losses from drains or nasogastric tubes should be replaced every 4–6 hours with 0.9% sodium chloride with added KCl (10 mmol in 500 ml). Stoma losses are replaced if >20ml/kg/day with 0.9% sodium chloride with added KCl.

Monitoring of fluid therapy

Healthy children prior to elective surgery do not normally require serum electrolytes measurement in the preoperative period. Serum electrolytes in the preoperative period should be measured in all children undergoing elective or emergency surgery who require i.v. fluids prior to surgery and every 24 hours in all children on i.v. fluids in the postoperative period. A fluid input/output chart, along with daily weight measurement, should be used to monitor the daily fluid balance.

Management of hyponatremia

Sodium requirement in children is 2–3 mmol/kg in 24 hours. Hyponatremia (serum sodium <135 mmol/l) in the postoperative period is the most common electrolyte abnormality mainly related to the use of hypotonic maintenance fluids. In severe hyponatremia serum levels can be <120 mmol/l. The use of medications, such as opioids that enhance the action of ADH and non-steroidal anti-inflammatory drugs (NSAIDs) that cause inhibition of prostaglandin synthesis, can contribute to the hyponatremia. Losses from stoma and nasogastric tubes also contribute to hyponatremia. Early signs of hyponatremia include headache (difficult to assess in very young children), lethargy and irritability. Late signs include seizures, not responding to antiepileptic drugs, or respiratory arrest requiring PICU admission, and are associated with poor outcomes [13,14]. In symptomatic children, initial management must be rapid and serum sodium corrected within 1–2 hours with infusion of 3% sodium chloride (1 ml/kg of 3% sodium chloride will raise the serum sodium by 1 mmol/l). The amount of Na required can be calculated using the following formula:

$$\text{mmol of Na} = (130 - \text{present serum Na}) \times 0.6 \times \text{weight (kg)}$$

Children with asymptomatic hyponatremia and normal or increased volume status should be volume-restricted at 50% of maintenance rate.

Management of hypernatremia

Hypernatremia (serum sodium >150 mmol/l) can occur from excess sodium intake, free water deficit (diabetes insipidus, fever, radiant warmers, phototherapy), or combined sodium and water deficit (postobstructive diuresis, emesis, diarrhea). Signs of hypernatremia are more severe when it develops rapidly, possibly leading to brain hemorrhage when the serum sodium is >160 mmol/l. Chronic hypernatremia is often well tolerated. Management of hypernatremic dehydration consists of initial volume replacement with 0.9% sodium chloride given in boluses of 20 ml/kg to restore normovolemia. Water deficit is calculated with the formula:

$$5 \times 0.6 \times \text{body weight in kg} \times 3 [12 \times (145/\text{current sodium})]$$

Complete correction should be done very slowly over at least 48 hours to prevent cerebral edema, seizures and brain injury. The serum sodium should be corrected at a rate of no more than 12 mmol/kg/day with 0.45% sodium chloride or 0.9% sodium chloride in dextrose. Potassium should only be added to maintenance fluids once good urine output is established.

Management of potassium imbalance

Potassium requirement in children is normally 1–2 mmol/kg in 24 hours. The immature kidney is less efficient in the secretion of potassium, making neonates and infants more susceptible to hyperkalemia. Hypokalemia (serum potassium <3.5 mmol/l) in the postoperative period is commonly related to excessive vomiting and diarrhea or iatrogenic, related to large stoma and nasogastric losses or loop diuretic use. Signs and symptoms include cramps, arrhythmias, reduced cardiac contractility and paralytic ileus. In children able to tolerate enteral feeding, oral supplements of 3–5 mmol/kg/day can be used. In severe hypokalemia (serum potassium <3 mmol/l), intravenous correction should be no faster than 0.25 mmol/kg/h using a maximum peripheral concentration of 40 mmol/l KCl. Hyperkalemia (serum potassium >5.5 mmol/l) can develop in renal failure, bilateral high-grade obstruction, extreme acidosis, and insulin deficiency. Fictitious hyperkalemia can be secondary to capillary phlebotomy or usage of intravenous sites running potassium-containing fluids for blood sampling. Signs and symptoms include skeletal muscle weakness and

ECG changes (low amplitude and widened P waves, widened QRS complex, fusion of QRS-T, loss of the ST segment and tall tented T waves). Treatment of hyperkalemia is done with 100 micrograms/kg of 10% calcium gluconate which is equal to 0.5 ml/kg of 10% solution (1 ml 10% calcium gluconate contains 0.22 mmol calcium). It is also important to increase intracellular shift of potassium by giving 1–2 mmol/kg of sodium bicarbonate, an infusion of 0.3–0.5 g/kg/h of glucose with 1 unit of insulin for every 5 g of glucose or to give 2.5 mg to 5 mg nebulized salbutamol (5 micrograms/kg in neonates i.v.). Removal of potassium from the body is done by giving 125–250 mg/kg calcium resonium rectally or orally, by use of furosemide 1 mg/kg or by dialysis or hemofiltration.

Management of calcium imbalance

Hypocalcaemia is defined as corrected total calcium <2 mmol/l in children or <1.5 mmol/l in neonates. Neonates are at high risk of hypocalcaemia because of low albumin levels. There is also a physiological fall in calcium levels soon after birth and return to normal levels after the second day of life. Common causes of hypocalcaemia in neonates include encephalopathy, renal failure, Di George syndrome, maternal diabetes and iatrogenic following thyroid surgery. Symptoms of hypocalcaemia include twitching and jitteriness, perioral, finger and toe paraesthesia, masseter and carpopedal spasm, prolonged QT interval on ECG and reduced cardiac contractility. Immediate treatment is with 10% calcium gluconate 0.5 ml/kg to a maximum of 20 ml over 10 minutes or 10% calcium chloride 0.2 ml/kg to a maximum of 10 ml over 10 minutes under continuous ECG monitoring.

Postobstructive diuresis

Postobstructive diuresis refers to the marked polyuria that occurs after the relief of bilateral ureteral obstruction or obstruction of a solitary kidney. Release of obstruction can lead to marked natriuresis and diuresis with the wasting of potassium, phosphate, and divalent cations.

There are several factors that can lead to the development of this condition in children. Common causes include posterior urethral valves, stones, catheter occlusion, urinary retention after reimplantation, obstruction of a catheterizable stoma, or, rarely, obstructing malignancy such as rhabdomyosarcoma. The mechanisms in-

involved are both physiological and pathological. Physiologic factors include excess sodium and water retention and accumulation of urea and other non-reabsorbable solutes resulting in an osmotic diuresis. Pathologic factors include decreased tubular reabsorption of sodium secondary to altered proximal and distal sodium transporters. Also there is inability to maximally concentrate urine, secondary to a decreased medullary concentrating gradient, leading to decreased response to ADH, increased tubular transit flow time and reducing equilibration time for absorption of sodium and water. Finally, the increased production of prostaglandins immediately following relief of obstruction contributes to the post-obstructive diuresis.

Children with post-obstructive diuresis need to be monitored closely, especially if they are unable to eat and drink on their own. Once the accumulated excess of sodium and water has been excreted, dehydration and hypokalemia can occur and it is important to monitor urine output closely. Once the patient has diuresed to the point of euvoemia, fluid replacement should be administered by replacing 75% of the urine losses with 0.45% saline. Post-obstructive diuresis is usually self-limiting and resolves over 24–48 hours or several days in children with normal kidney function.

Metabolic consequences after incorporation of bowel into the urinary tract

Bladder augmentation is associated with possible metabolic consequences and other long-term complications including the formation of bladder stones, chronic bacteruria, mucusuria, bladder perforation, bowel complications and malignancy. The gastrointestinal tract is a relatively poor substitute for urothelium and its semipermeability can lead to fluid and electrolyte abnormalities. Bowel segments commonly used include ileum and sigmoid colon, with stomach being used infrequently. Exposure of colon or ileum to urine results in the absorption of ammonium and chloride ions which can cause hyperchloremic metabolic acidosis proportional to the patient's renal function and the amount of intestine used for bladder reconstruction. Chronic metabolic acidosis can lead to skeletal growth impairment and osteopenia. The stomach can be also used instead of small or large intestine for bladder augmentation. The normal stomach function is secretion of acid and enzymes for digestion rather than absorption

and gastric augmentation can therefore eliminate the absorptive problems with intestine including metabolic acidosis. However, secretion of acid can irritate the bladder and urethra, leading to hematuria and dysuria. Most cases can be controlled pharmacologically with H₂ histamine blockers or proton pump inhibitors. The secretion of acid and chloride can predispose patients to develop severe hypochloremic, hypokalemic metabolic alkalosis. Given the risks of metabolic complications, serum electrolytes should be monitored routinely during follow-up after bladder augmentation. Patient with ileal bladder augmentation can have reduction in serum B12 levels after surgery and it is suggested that they should have their B12 levels regularly checked and be appropriately supplemented, particularly in the first 5 years postoperatively [15].

Factors affecting the endocrine and metabolic response to surgery

The magnitude and duration of the response to injury are influenced by many factors. Some of these are directly related to the patients and recent evidence shows there is genetic predisposition for how an individual responds to injury and infection. The presence of coexisting disease, such as cancer and chronic inflammatory disease, and the use of anti-inflammatory or immunosuppressive therapy prior to surgery may also influence the response. Malnourished patients may have decreased immune function or deficiency in important substrates and malnutrition prior to surgery or trauma is associated with poor outcomes. Greater tissue damage is associated with a greater metabolic response and minimally invasive surgery could play a significant role in reducing the extent of the response. Anesthesia also plays a role in the endocrine and metabolic response to surgery.

Minimally invasive surgery

Minimally invasive surgery causes less tissue injury with reduction of the mediators of the acute phase response leading to shorter hospital stay, reduced analgesia requirement and a more rapid postoperative recovery [16–18]. The systemic immune response to carbon dioxide (CO₂) pneumoperitoneum has been investigated primarily in adults. The response after laparoscopic surgery seems to be characterized by a brief

period of immune suppression with a small decrease in the inflammatory response and little or no difference in the metabolic response [18,19]. Peritoneal macrophage functioning is affected, probably because of the CO₂ effect on pH in peritoneal fluid. There is also a diminished production of cytokines and a decline in the intrinsic functioning of monocytes. Serum CRP and IL-6 levels are appreciably lower after laparoscopy than after laparotomy but other cytokine parameters do not show a marked difference between the two operating procedures [20]. This inclination toward less immunosuppression is not associated with real clinical effects. A study looking at open versus laparoscopic Nissen's fundoplication in children did not show any difference in concentrations of TNF- α or IL-1 between the two groups although there was a slight increase in postoperative immune suppression in the open group [21]. In children, it has also been found that laparoscopy is associated with an intraoperative hypermetabolic response, increased oxygen consumption and rise in core temperature [22]. However, a study during emergency laparoscopy and laparotomy in children found no difference in the levels of cortisol, glucose, insulin, lactate, and adrenaline between the two groups [23]. Insufflation of the abdomen during laparoscopy alters cardiovascular performance because of both the effects of hypercarbia as well as the change in intra-abdominal pressure. Mild hypercarbia (PaCO₂ of 45–50 mmHg) has little effect on hemodynamics, whereas moderate to severe hypercarbia has both direct and indirect effects on cardiac function [24]. At a PaCO₂ of 55–70 mmHg, hypercarbia and acidosis cause significant hemodynamic changes. In addition, elevated CO₂ directly causes myocardial depression and vasodilation. These effects are counteracted by a centrally mediated sympathetic stimulation that causes tachycardia and systemic vasoconstriction with increase in heart rate, mean arterial pressure, central venous pressure, pulmonary artery pressure, cardiac output, and stroke volume. Hemodynamic changes are also mediated by the absorption of the CO₂ through the peritoneal surface, causing adverse effects on the cardiovascular system requiring increasing minute ventilation by 20–30% to prevent hypercarbia [25,26].

General anesthesia

Aspects of the patient's stress response to surgery are influenced by the anesthesia. Opioids suppress the

hypothalamic and pituitary secretion, especially at very high doses. However, the use of high-dose opioids will prolong postoperative recovery and the need for prolonged ventilation. Etomidate, used for anesthetic induction, suppresses the production of cortisol and aldosterone for up to 24 hours if infused for 1–2 hours. Midazolam and diazepam have an inhibitory effect on steroid production although the mechanism is not completely clear. Clonidine decreases the sympathetic and vascular response to surgery.

Local anesthesia

Epidural and spinal anesthesia can reduce the levels of patient serum epinephrine, ACTH, cortisol, GH and ultimately glucose since both the afferent input from the operative sites to the central nervous system and the efferent autonomic pathways are blocked. Although no real benefits have been demonstrated in terms of morbidity and mortality with the use of anesthetic regional techniques, the reduced thromboembolic complications, improved pulmonary function and reduction of paralytic ileus have proven benefits in the surgical patient.

DOS AND DON'TS

Do

- Correct dehydration without hypovolemia slowly
- Correct hypovolemia rapidly to maintain cardiac output and organ perfusion
- Replace blood losses during surgery initially with crystalloids and colloids and then with blood products when the hematocrit falls below 25%
- Use artificial colloids in preference to human albumin as there are no proven advantages demonstrated with the use of albumin
- Employ fluid restriction to 60–70% of full maintenance volume with additional boluses of isotonic fluid after major surgery to avoid fluid overload and hyponatremia
- Monitor serum electrolytes every 24 hours in children receiving parenteral fluids in the postoperative period.
- Monitor vitamin B12 levels in patients with ileal bladder augmentation as they may develop a deficit especially in the first 5 years after surgery

Don't

- Use hypotonic fluids in the postoperative period as they are a major cause of hyponatremia

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CHAPTER 6

Perioperative anesthetic and analgesic risks and complications

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KEY POINTS

- Mortality associated with anesthesia has reduced dramatically and is as low as 0.1 per 10,000, although higher in neonates and prematurity
- Peripheral regional anesthesia associated nerve damage is rare, especially with ultrasound guidance
- Central neuraxial blockade (CNB) provides excellent analgesia with a low risk of major complications
- Enhanced recovery after surgery (ERAS) protocols have been shown to reduce complication rates and speed recovery in children
- Large trials are under way to establish the effects on neurodevelopment of multiple anesthetic exposures in neonates
- Perioperative genomic testing is emerging as a means of avoiding adverse outcomes strongly associated with specific genetic predispositions

CLINICAL PROBLEM/CASE

You find yourself in discussion with the anesthetist regarding optimal analgesia in a neonate booked for a laparoscopic nephrectomy secondary to multicystic dysplastic kidney.

- 1 What multimodal analgesia would you consider appropriate?
- 2 Would you prescribe NSAIDs postoperatively?
- 3 In the event of the procedure converting to open, what alternate analgesic strategies might be more suitable?

Introduction

Modern anesthesia is extremely safe. Indeed, the risk of serious injury or death is so small that it becomes difficult to measure accurately. National Confidential Enquiry into Perioperative Deaths (NCEPOD) data in the UK, the Australian Incident Monitoring System (AIMS) project in Australia [1] and the closed claims process in the USA [2] are the best sources of recent data.

Perioperative and anesthetic-related mortality is still a major issue in the developing world [3]. Limiting our discussion to developed medical systems, however, shifts focus to morbidity and quality issues. Mortality

associated with anesthesia has decreased dramatically from 6.4 per 10,000 in the early 1950s to as low as 0.1 per 10,000 by 2000 [4,5]. There are several reasons for this: advances in anesthetic agents, techniques, monitoring, audit and subspecialist training in areas like pediatric anesthesia [6]. There is good evidence that a trained pediatric anesthetist decreases the incidence of perioperative events [7,8].

In 2009, the UK Royal College of Anaesthetists published useful patient information leaflets explaining anesthesia-related risks (rcoa.ac.uk/patients-and-relatives/risks). Quoted risk values like those above are clearly dependent on individual patient factors.

For example, children under 1 year have a greater incidence of anesthetic mortality. American Society of Anesthesiologists (ASA) status, prematurity, obstructive sleep apnea, congenital abnormalities, pulmonary, cardiac or hematological comorbidities, weight less than fifth percentile, surgery duration over two hours and intraoperative bradycardia all adversely affect risk [9,10]. Nevertheless, human error and equipment failure still account for approximately 90% of critical incidents. The ability to reduce risk therefore lies to a large part with the anesthetist, their team and with systems designed to minimize the scope for these errors.

This chapter adopts a systems approach to complications of general anesthesia (GA) before looking specifically at those arising from pharmacological, intravenous and regional anesthetic interventions. It will also attempt to point to emerging and future areas of concern, research and improvement.

Gastrointestinal

Postoperative nausea and vomiting (PONV) is common, the incidence ranging from 9% to 43% [11], increasing with age (peak 11–14 years). Incidence is highest following strabismus and ENT surgery, appendectomy and orchidopexy. High-risk patients include those with prior PONV and/or motion sickness [12]. Anesthetic techniques with less PONV and pre-emptive anti-emetics should be considered in high-risk cases.

The incidence of perioperative aspiration is low and generally has a good outcome. It is reported as between 1 and 10 per 10,000 with a very low incidence of pneumonia or need for admission to intensive care [13].

Respiratory

Control of a patient's airway is a vital aspect of any anesthetic and adverse respiratory events are common perioperative problems faced by anesthetists [14,15]. The incidence is higher in younger children due to the relatively narrow infant airway (Figure 6.1) coupled with the high incidence of respiratory tract infections in this population. The most common events are episodes of arterial desaturation associated with laryngospasm, and bronchospasm [15].

Laryngospasm is the reflex closure of the glottis by adduction of the vocal cords (Figure 6.2). It can persist after cessation of the stimulus. Common causes include local stimulation of the larynx, e.g. by saliva, blood, or foreign body including a laryngoscope or endotracheal tube. It can also occur in response to other stimulation, e.g. surgery, movement, or stimulation of the anus or cervix. The reflex is abolished in deeper planes of anesthesia. It presents with noisy breathing and stridor and can lead to hypoxemia, hypoventilation and negative pressure pulmonary edema. Treatment consists of removing the stimulus, 100% oxygen and positive end expiratory pressure (PEEP) via the breathing circuit. If the spasm does not resolve with these maneuvers, a small dose of intravenous induction agent or muscle relaxant can be used to break the spasm.

Airway difficulties arise due to inability to provide adequate ventilation by bag-mask ventilation – a “can't ventilate” situation, or when intubation is difficult “can't intubate” (estimated incidence between 1.5% and 13%). It is the extremely rare and much feared “can't intubate, can't ventilate” combination that can tragically lead to death or permanent brain damage [16].

More minor adverse events associated with the airway include postoperative sore throat and oral tissue or dental damage. These latter complications account for a significant proportion of medicolegal claims against anesthetists in adult practice [17]. Injuries tend to occur during laryngoscopy and intubation. A dental history should be obtained preoperatively and children with wobbly teeth (peak ages 6–8 years) should be warned of potential loss. Postoperative sore throat has a reported incidence of 12%, 24 hours after surgery in daycase adult patients [18]. The incidence is higher after intubation than following insertion of a laryngeal mask airway (Figure 6.3).

Certain childhood syndromes are associated with difficult airways and these should be highlighted preoperatively to enable appropriate anesthetic planning. These include the Pierre Robin sequence, Crouzon syndrome, Apert syndrome, Pfeiffer syndrome, Treacher–Collins syndrome, craniofacial microsomia, and Goldenhar syndrome [19].

Children with symptomatic viral upper respiratory tract infections (URIs), wheezing, purulent secretions and fever should be postponed for >2 weeks due

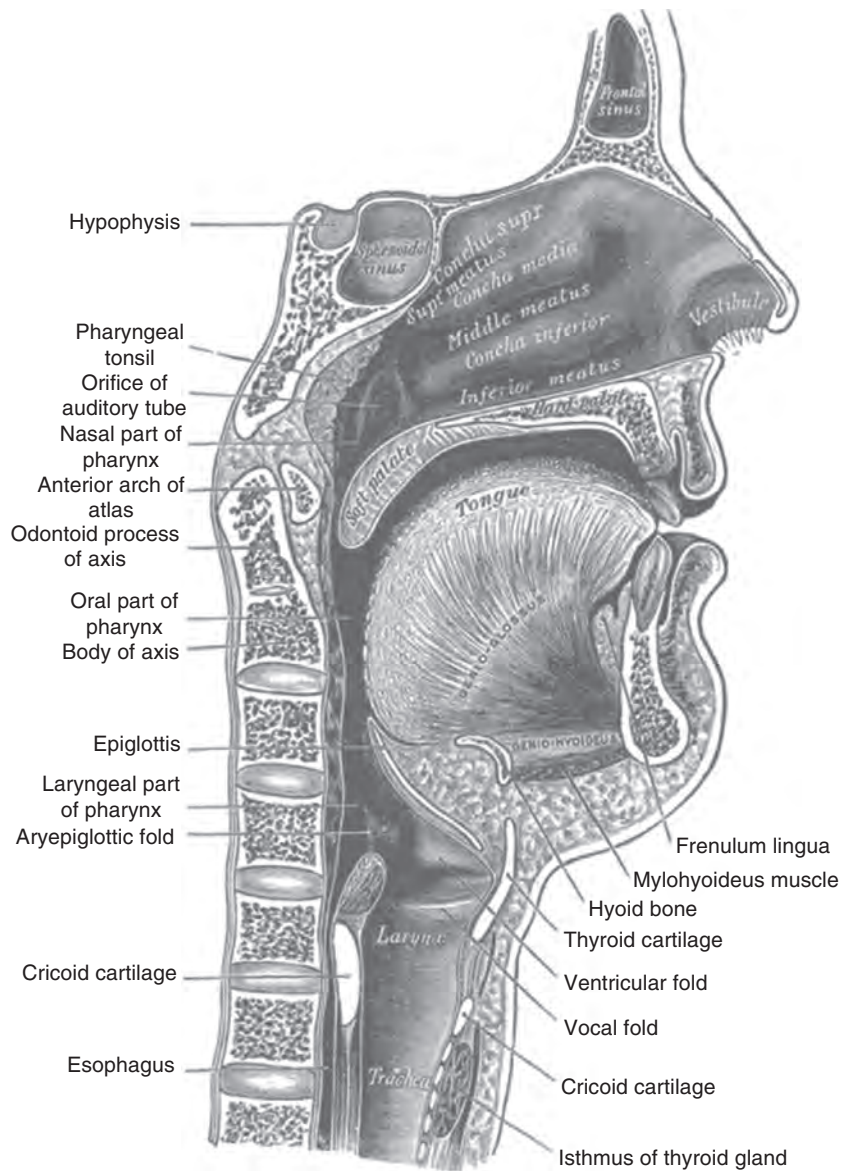


Figure 6.1 Sagittal view of human airway. (From Gray 1918 [73].)

to a sevenfold increase in respiratory complications [20,21]. Additional risk factors for developing adverse respiratory events in children with URIs include: the use of an endotracheal tube, age <5 years, prematurity <37 weeks, a history of reactive airway disease or nasal congestion, parental smoking and surgery on the airway [22].

Cardiovascular

Adverse cardiac events are only a quarter as common as respiratory complications during anesthesia. Common events include arrhythmias and bradycardias, and hypertension or hypotension [14,15]. The results of the Paediatric Perioperative Cardiac Arrest Registry (POCA)

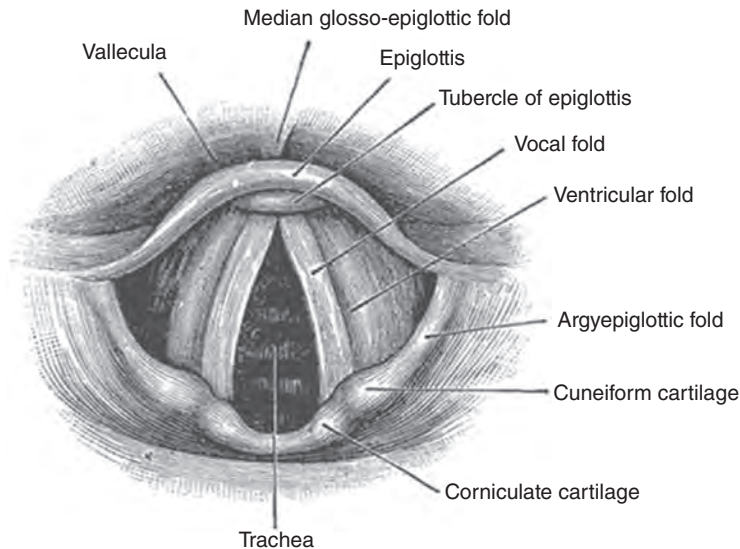


Figure 6.2 Superior view of vocal cords. (From Gray 1918 [73].)

found an estimated incidence of anesthesia-related cardiac arrest of 1.4 per 10,000 with a mortality rate of 26% in those affected [5]. Pharmacological (overdose) and underlying cardiac disease were the most common causes; 55% of events occurred in children under the age of 1 year.

Congenital heart defects (Figure 6.4) have an incidence of 6–8 per 1000 live births. The Center for Disease Control reports cyanotic heart defects occurred in 56.9 per 100,000 live births in the USA in 2005. The

incidence of congenital heart disease in premature infants is increased at 12.5 per 1000 [23].

Neurological

Peripheral nerve injury under GA has a reported incidence of 1 per 1000 in adults, although data in children are harder to come by. The most commonly affected are ulnar nerve 30%, brachial plexus 23% and lumbosacral nerves 16% [24]. The usual mechanism is ischemia due to compression by surgical retractors, leg stirrups, or other equipment and is more likely to occur during periods of poor peripheral perfusion due to hypotension. Brachial plexus injuries can be caused by traction through excess shoulder abduction. Damage is avoided through meticulous care with patient positioning, padding of protect pressure points, and avoidance of extreme joint positions. Most nerve injuries recover over a period of months; all should to be reviewed by a neurologist.

The commonest type of ocular complication is corneal abrasion, causing blurring of vision and usually resolving over 1–2 months [14,15]. Protective reflexes are lost during anesthesia and eyes need to be taped shut to protect the cornea.

Perioperative visual loss is extremely rare and associated more with cardiac, spinal and head and neck



Figure 6.3 Laryngeal mask airway. 2006. (From Ignis (own work). GFDL (<http://www.gnu.org/copyleft/fdl.html>), CC-BY-SA-3.0 (<http://creativecommons.org/licenses/by-sa/3.0/>) or CC-BY-SA-2.5-2.0-1.0 (<http://creativecommons.org/licenses/by-sa/2.5-2.0-1.0>), via Wikimedia Commons.)

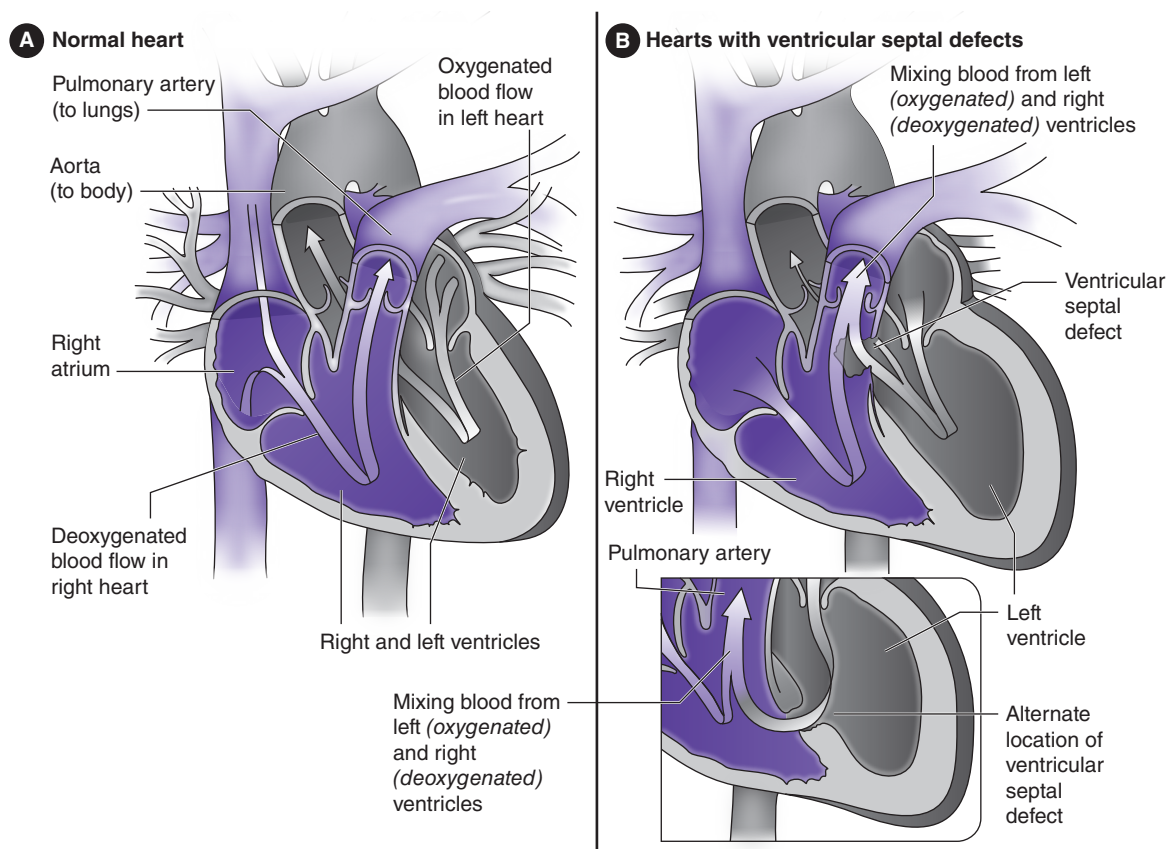


Figure 6.4 Congenital heart disease, i.e. ventricular septal defects. (From National Heart Lung and Blood Institute, <http://www.nhlbi.nih.gov/health/health-topics/topics/chd/types.html>.)

surgery. The most common mechanism of injury proposed is optic nerve ischemia. Risk factors include duration in the prone position, decreased ocular perfusion pressure, anemia, hypotension and hypoxia [25]. Care must be taken to avoid pressure to the eyes with extra padding and protection whilst prone.

Psychological

Maladaptive postoperative behaviors (anxiety, sleep disturbance, night terrors and a return to bed wetting) can occur in up to 65% of children postoperatively. Age, degree of patient and parent preoperative anxiety, type of surgery, and level of postoperative pain have been reported to predict the occurrence of behavioral changes [26]. Prolonged upset can occur even after daycase surgery. Up to 32% of affected children exhibit negative behavioral changes one month after surgery

[27]. Sevoflurane is the volatile anesthetic agent that is most widely used for inhalational induction and can be associated with temporary post-anesthetic delirium. However, there is no greater incidence of maladaptive behaviors with sevoflurane compared against either isoflurane or halothane [28,29].

Awareness

Awareness during anesthesia is a state of consciousness revealed by explicit or implicit memory of intraoperative events, associated with post-traumatic stress disorder. It was the focus of the large UK NAP5 study 2012. From adult data, the incidence of conscious awareness with explicit recall and severe pain is estimated to be 1 per 3000. Conscious awareness with explicit recall but without pain is more common at 0.1% and 0.7% [30]. The incidence is higher where neuromuscular blocking

agents are used. The commonest cause is drug error: either inadvertent paralysis of an awake patient or failure of delivery of volatile anesthetic agents.

Pharmacological

Clinically indistinguishable anaphylactic and anaphylactoid reactions during anesthesia are rare but potentially life-threatening events. Their most severe manifestations present with cardiovascular collapse, bronchospasm, and laryngeal edema. Serum histamine and mast cell tryptase levels confirm the diagnosis [31]. Patients require follow-up skin-prick testing and radio allergy sorbent tests (RASTs) for specific IgE antibodies to identify triggers. The incidence of anaphylaxis is approximately 1 per 6000 anesthetics [32]. The most commonly linked agents are neuromuscular blocking drugs 58%, latex 16.7% and antibiotics 15%. A history of atopy, asthma and food allergy is more frequent in cases of latex allergy [33].

Malignant hyperthermia (MH) is a rare autosomal dominant condition triggered by volatile anesthetic agents and suxamethonium. It has an incidence of 1 in 15,000 in children and 1 in 50,000–100,000 in adults [34], has a mortality of 10% and requires prompt and efficient recognition and management [35]. Diagnosis is made by muscle biopsy and relatives of the index case must be investigated. Anesthesia in susceptible individuals requires careful planning to avoid triggers.

Suxamethonium is a short acting depolarizing muscle relaxant, metabolized by plasma cholinesterase. Suxamethonium apnea occurs in patients with reduced cholinesterase activity, primarily of genetic etiology. It presents as prolonged nonreversible paralysis. Management involves continued sedation and ventilation until the drug has been metabolized.

When any drug is administered the potential for error exists (Figure 6.5). It is possible to give the wrong drug (in the case of an allergic history for instance), in the wrong dose, via the wrong route. Assuming we give appropriate drugs to appropriate patients, however, it is worth considering the recognized problems associated with common analgesic drugs.

Paracetamol

Paracetamol's pharmacokinetics are better known than any drug in the pediatric pharmacopoeia. The main



Figure 6.5 Drug administration with the potential for error. (From ICU IV1 by Calleamanecer (own work). Licensed under Creative Commons Attribution-Share Alike 3.0 via Wikimedia Commons (http://en.wikipedia.org/wiki/File:ICU_IV_1.jpg).)

risks in its use relate to the potential for overdose. Suggested maxima are 25 mg/kg per 24 hours at under 30 weeks postconceptional age, 45 mg/kg at less than 34 weeks, 60 mg/kg in term neonates and infants and 90 mg/kg thereafter [36]. Paracetamol is metabolized in the liver mainly by glucuronidation and sulfation but if these pathways become saturated the hepatotoxic oxidative metabolite *N*-acetyl-p-benzoquinoneimine may accumulate. Risk factors for this include dehydration and sepsis.

Non-steroidal anti-inflammatory drugs (NSAIDs)

In a large study, the risk of administering a short-term course of ibuprofen was low and similar to paracetamol [37]. Childhood asthma does not seem to be affected by NSAIDs in the same way as adult asthma [38]. Impaired renal function and bleeding tendency remain contraindications, however. The use of NSAIDs in children below 3 months of age is ill advised because of the risk of pulmonary hypertension and alterations in prostaglandin-dependent cardiac, cerebral and renal regional blood flow. NSAIDs can be used for the closure of patent ductus arteriosus in neonates but there is specific analgesic-related guidance in the pediatric British National Formulary for its use above one month [39].

Opioids

The main side effects of this group are respiratory depression, nausea and vomiting, constipation, urinary retention, itching, and sedation. From a safety

perspective, clearly the most important of these is respiratory depression. It is readily treatable with naloxone but due to its relatively short duration of action, repeated administration or infusion may be necessary. Urinary retention and itching have been successfully treated with lower doses of naloxone whilst itching often responds to chlorpheniramine.

Since moving away from intermittent intravenous bolus and intramuscular injections, the risk has diminished as infusions and patient-controlled analgesia PCA pumps decrease the potential for high-peak plasma concentrations. A 10,000 child study at Great Ormond St Hospital has shown nurse-controlled analgesia (NCA) with morphine to be a safe and effective analgesic strategy for major surgery [40]. The addition of high background infusion rates to PCA regimes increases the risk of sedation, hypoxia, nausea, and vomiting [41]; however, low background rates can offer improved analgesia without these problems [42]. Nausea and vomiting occurs in 30–45% of children on PCA/NCA and can be reduced with prophylactic anti-emetics [43,44].

Fluid management

Children are more susceptible to hypovolemia so volume correction with either crystalloids or colloids is crucial and should compensate for deficits and maintenance. Fluid deficits consist of preoperative deficits (fasting, GI, renal, or cutaneous losses), hemorrhage and third space losses. Goal-directed therapy aims to target volume correction to organ perfusion and in adults is often guided by intraoperative esophageal Doppler assessment of cardiac output.

There has been a shift from hypotonic to isotonic infusions in pediatric patients [45,46] due to their association with acute hyponatremia. Hyponatremic encephalopathy can lead to permanent neurological damage or death as sudden water shift into brain tissue causes cerebral edema. Over 50% of children with a serum sodium of <125 mmol/l will develop some degree of hyponatremic encephalopathy [47] and mortality can be as high as 50% [48].

Children are at particular risk due to a higher number of brain cells and a larger brain-to-intracranial volume ratio compared with adults [49]. Surgical patients are also at elevated risk, due to excess secretion of antidiuretic hormone (ADH) in the postoperative period

triggered by pain, stress, anxiety, nausea and vomiting, and morphine [49]. ADH reduces the ability of the kidneys to excrete free water, leading to hyponatremia and oliguria.

The UK National Patient Safety Agency has recommended the removal of hypotonic NaCl 0.18% from use and that hypovolemia should be corrected with isotonic solutions, i.e. NaCl 0.9% or Hartmann's solution [50,51]. NaCl 0.45% with glucose can be prescribed as maintenance fluids but close monitoring with regular weights and measurement of plasma sodium is advised.

Transfusion

In general, children tolerate hemodilution well and perioperative levels of 6 g/dl are acceptable in a hemodynamically stable child. There are many potential complications of transfusion: hypocalcemia, hyperkalemia, hypomagnesemia, metabolic acidosis, and hypothermia. However, these are generally correctable [52]. Of greater concern to parents and children is the infective risk.

Infectious risk of transfusion

These risks have decreased dramatically secondary to advances in blood screening (Figure 6.6) and pathogen inactivation. Incidence varies between countries and is dependent upon the prevalence of domestic infections and screening programs. In the USA, blood is screened



Figure 6.6 Transfusion screening: bags of blood. (From Antonia from Mijas, España (Siempre hace falta sangre/Always need blood). CC-BY-SA-2.0 (<http://creativecommons.org/licenses/by-sa/2.0>), via Wikimedia Commons.)

for hepatitis B virus, hepatitis C virus, HIV1 and 2, human T-lymphotropic viruses I and II, West Nile virus, syphilis and Chagas' disease (www.aabb.org).

Besides viral pathogens, bacterial contamination can occur and is most commonly seen with platelets [53]. Other infectious diseases potentially transmissible via transfusion include Chagas' disease, Lyme disease, malaria and Creutzfeldt-Jakob disease (CJD) although donor screening and the deferral of those with potential symptoms has greatly reduced transmission.

Incompatibility and immune considerations

Human error is the most common cause of mismatched transfusion. Severe acute hemolytic reactions most often result from immunologic destruction of red cells because of ABO incompatibility but less frequent serologic incompatibilities, not detected by standard antibody screens, also occur. Anaphylactoid reactions with bronchospasm, laryngeal edema, and urticaria are dangerous but rare and typically occur in IgA-deficient individuals. The mandatory use of leukocyte-depleted products in the UK has significantly reduced transfusion reactions and immune modulation [54].

Regional anesthesia

Pediatric regional anesthesia is gaining popularity and carries a very low risk in appropriately selected infants and children [55,56]. With all blocks, however, the potential for local anesthetic (LA) toxicity exists. The risk is greatest when LA is injected into vascular tissue or, inadvertently, intravascularly. An upper limit of 2 mg/kg for bupivacaine (0.8 ml/kg of 0.25%) and 3 mg/kg for lidocaine is advised. The introduction of levorotatory bupivacaine is welcome due to decreased cardiotoxicity, which can manifest as ventricular extrasystoles and shock-resistant VF. Intralipid® therapy and supportive management is the mainstay of LA toxicity protocols.

Caudals

Caudals are the commonest regional technique used in pediatric anesthesia, being relatively simple to perform and providing effective analgesia for 3–10 hours [57]. There are reports in large series of caudals showing very few side effects [58,59]. Transient leg weakness, urinary retention and risk of prolonged block should always be anticipated, however.



Figure 6.7 Central neuraxial blockade: spinal anesthesia. (From DocP (own work). CC-BY-SA-2.0-de (<http://creativecommons.org/licenses/by-sa/2.0/de/deed.en>), via Wikimedia Commons.)

Epidurals

National Audit Project (NAP4) was a UK-wide review of central neuraxial blockade (CNB) assessing all major complications of CNBs (Figure 6.7) performed over one year, with expert analysis of cases. The incidence of permanent injury was pessimistically 4.2 per 100,000 and optimistically 2.0. The incidence of paraplegia or death was pessimistically 1.8 per 100,000 and optimistically 0.7. The data concluded there was a low incidence of major complications, many of which resolved within six months [60].

A UK national audit of pediatric epidural complications from 21 centers over five years resulted in data from >10,000 epidurals. Ninety-six serious clinical incidents were reported giving an overall incidence of 0.9% [61]. Out of these, 40 were judged by an expert panel to be coincidental. Of the remaining 56 incidents, the commonest was local infection.

Continuous epidural anesthesia has been shown to significantly reduce the need for opioid administration, allowing early extubation and faster postoperative recovery. The technique is considered safe and effective for pain management following major reconstructive surgery in pediatric urology [62].

Despite a good safety profile, there is still debate as to the merits of epidural analgesia over systemic pain relief, itself not without complications. Complications arising from opioids are more likely to be attributed to patient and drug characteristics. Epidural complications, which can cause significant disability, may more readily be apportioned, fairly or not, to technique and operator error.

Peripheral blocks

Peripheral regional anesthesia-associated nerve damage has an incidence of $<0.1\%$ and the prognosis is good [63]. There are some specific complications depending on puncture site, i.e. pneumothorax. Permanent nerve injury, however, is rare with an estimated incidence of 0.03% [63]. The pathophysiology and etiology of nerve injury depends on needle-nerve trauma, cytotoxicity of LA, and patient premorbid factors. Ultrasound guidance (Figure 6.8) is now strongly recommended in both pediatric and adult populations [64]. Nerve stimulation techniques can also aid placement and reduce risk.

Adrenaline-containing solutions should be avoided near end arteries such as dorsal nerve block or ring block to help avoid the risk of necrosis. Ilioinguinal blocks and transversus abdominis plane (TAP) blocks are commonly performed for groin surgery and are considered safe and effective. The main risk for these is tracking of LA to the femoral nerve with possible leg weakness and delayed discharge. Penile blocks are also well established for circumcisions and hypospadias repairs.

TAP blocks and catheters offer an alternative to a CNB for patients undergoing major abdominal surgery. Administration of low dose, low concentration local anesthetic through TAP catheters has been shown to provide children with excellent postoperative analgesia post spinal dysraphism surgery [65].



Figure 6.8 Ultrasound-guided femoral nerve block. (From PhilippN (own work). GFDL (<http://www.gnu.org/copyleft/fdl.html>) or CC-BY-SA-3.0 (<http://creativecommons.org/licenses/by-sa/3.0/>), via Wikimedia Commons.)

The future

There is strong evidence that anesthetic techniques including supplemental oxygen and avoidance of hypothermia, blood transfusion, hyperglycemia or large swings in blood glucose reduce postoperative infection rates. There is also evidence that the use of regional anesthesia reduces chronic postsurgical pain and the complications thereof [66]. Appreciation of these perioperative influences has been developed by the Copenhagen abdominal surgeon Henrik Kehlet into the concept of “enhanced recovery after surgery” (ERAS). Benefits of these fast-track concepts have been confirmed in colon, pediatric and urological surgery [67]. ERAS protocols and regional anesthesia usage are becoming more common, improving recovery profile, mobilization and complication rates [68].

The incidence of childhood obesity ranges today from approximately 8% to 17%, and is becoming increasingly significant in pediatric anesthesia [69]. Obese children not only have more comorbid diseases like asthma, diabetes and hypertension, but also a higher incidence of anesthesia-related complications [69].

Recent animal and human studies have suggested a dose-dependent association between multiple exposures to anesthetic agents in early childhood, neuronal damage and adverse neurodevelopmental outcomes [70]. Exposure of animals to agents above threshold doses and durations during a critical neurodevelopmental window causes widespread neuronal apoptosis and subsequent abnormal behaviors. The relevance of such animal data to humans is unknown although it is worth considering that untreated neonatal pain and stress are themselves associated with neuronal death and maladaptive behavior [70]. Ongoing prospective investigations are under way: the Pediatric Anesthesia Neuro-Development Assessment (PANDA), the GAS and SmartTots studies. The results of these may clarify such concerns, with particular focus on the neonatal population undergoing multiple GAS. No change in current practice is yet indicated.

A high proportion of adverse outcomes is strongly associated with genetic predispositions, including thrombosis, bronchospasm, cardiac dysrhythmia, organ failure and sepsis [71]. Many of these associations are monogenic. Drug toxicity, inefficacy and prolonged neuromuscular block reflect pharmacokinetic and pharmacodynamic genomic susceptibilities that are unknown in advance of exposure to drugs first encountered in perioperative care [71].

At present, genetic screening is not performed prior to surgery, exposing genetic outliers to significant risk. This is likely to change as genotype technology becomes more available and the complex epigenetics of phenotype response to anesthesia and surgery become clearer. Standardized microarray gene tests for preoperative blood samples have already been developed, currently testing for 48 anesthesia-relevant polymorphisms [72].

Conclusion

Despite substantial progress, there are clear areas of concern and potential risk reduction. The future of pediatric anesthesia will most likely be influenced by much-needed large prospective studies, providing further insight into patient safety and personalized perioperative medicine.

DOS AND DON'TS

Do

- Know the common cautions and side effects of analgesics like paracetamol, NSAIDs and opioids – this is core knowledge
- Prescribe intravenous fluids and blood products with the same caution as drug therapy
- Consider introducing ERAS protocols into your practice (erassociety.org)
- Follow the WHO surgical checklist (who.int/patientsafety/safesurgery)

Don't

- Ignore the increased risk associated with symptomatic respiratory tract infections in children. Consider postponing elective cases
- Forget the benefit of multimodal analgesia. Would your patient benefit from the addition of regional anesthesia?

CLINICAL PROBLEM/CASE: DISCUSSION

- Local anesthetic infiltration combined with simple analgesics and oral opioids are usually adequate.
- NSAIDs are recognized to affect glomerular filtration rate, and although renal function in this patient group is likely to be normal most practitioners would be cautious about their use.
- Open procedures are likely to benefit from combined GA and regional techniques. An epidural, ultrasound-guided transversus abdominis plane TAP block, or morphine nurse-controlled analgesia NCA would all be considered appropriate.

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PART III

Open Surgery of the Upper Urinary Tract

CHAPTER 7

Nephrectomy

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KEY POINTS

- With the advent of laparoscopic surgery, open simple nephrectomy is rarely performed
- The retroperitoneal approach is preferable to the transabdominal approach as it does not lead to the formation of intraperitoneal adhesions
- Conversion of laparoscopic to open nephrectomy is usually achieved by joining or extending port site incisions
- Preoperative imaging provides valuable anatomical information useful in planning nephrectomy, particularly when the kidney is ectopic
- Careful preoperative planning and surgical technique can minimize the morbidity associated with hemorrhage or damage to perirenal structures

CLINICAL PROBLEM/CASE

Case 7.1

A 1-year-old boy is seen in clinic one month following an open nephrectomy. Mum complains that his side bulges intermittently.

- 1 What is the most likely diagnosis?
- 2 What is the appropriate management?

Case 7.2

A 2-year-old girl presents to the emergency department five days after discharge following a technically difficult open left nephrectomy. She has abdominal pain and her wound is discharging clear fluid.

- 1 What is the most important potential diagnosis?
- 2 What is the appropriate management?

Introduction

The advent of laparoscopic surgery has made open nephrectomy an infrequent undertaking in pediatric urology. The most common indications for open nephrectomy now are malignant disease and trauma.

Contraindications to elective, simple laparoscopic nephrectomy are increasingly rare, with several centers now reporting successful laparoscopic surgery for xanthogranulomatous pyelonephritis (XGP) [1–4]. These facts, together with the absence of contemporary publications on elective open simple nephrectomy, would

seem to confirm that laparoscopic surgery has now taken over as the gold standard approach.

Although infrequently applied, a good working knowledge of open approaches to the kidney remains important to the pediatric urological surgeon. In particular, the ability to convert rapidly from laparoscopic to open nephrectomy, whether it is to control hemorrhage or for non-progression, remains an essential surgical skill. In addition, familiarity with the choice of open approaches to both normally positioned and ectopic kidneys is vital on the rare occasion that laparoscopy is contraindicated.

This chapter covers open approaches to the kidney, concentrating on the anatomical basis, advantages, and potential complications of each approach.

Surgical approaches to the kidney

The first intentional nephrectomy was performed by Gustav Simon in 1869 in the treatment of an uretero-vaginal fistula. The first pediatric nephrectomy for a Wilms tumor was performed in Leeds by Richard Jessop in 1877. Retroperitoneal flank approaches had a lower incidence of postoperative peritonitis and were the approach of choice in the first half of the 20th century. Advancements in the surgical technique led to a revival of transabdominal approaches in the 1950s. The modern surgeon has numerous choices of approach, which can be tailored to the needs of each individual case.

Retroperitoneal approaches

The majority of surgeons prefer a retroperitoneal approach to simple nephrectomy, the principal advantage being avoidance of the peritoneal cavity and the associated risk of forming intraperitoneal adhesions. Open access to the retroperitoneum can be achieved through loin, lumbotomy, and anterior approaches. Readers are referred to excellent operative texts of the various approaches while the advantages and disadvantages of the various approaches are described in Box 7.1.

Transabdominal approaches

The increased postoperative recovery time and risk of intraperitoneal adhesion formation means that these approaches are generally reserved for malignant or traumatic cases. The advantage of these approaches is that they allow excellent access to the renal pedicle and great vessels. Transverse/subcostal or midline incisions are most frequently employed, although occasionally a thoracoabdominal incision may be used.

The advantages/disadvantages of the transverse/subcostal approach have been discussed; converting it to a transabdominal procedure simply involves opening rather than retracting the peritoneum. This approach allows excellent access to the lateral and superior portion of the kidney. If required, the incision can be extended across the midline, although this involves further muscle division.

Box 7.1 Advantages and disadvantages of different surgical approaches.

Loin (flank) approach

Advantages: Good access to renal parenchyma and collecting system [5]. Good access in obese patients

Disadvantages: Exposure of renal pedicle inferior to anterior approaches. Relatively large incision with higher incidence of wound pain and muscle bulge

Dorsal lumbotomy

Advantages: Rapid access without cutting muscle [6]. Useful for bilateral procedures without patient repositioning. Less postoperative pain and wound bulge. Fresh approach in those with previous loin or abdominal surgery

Disadvantages: Exposure may be limited and access to the kidney and pedicle inferior, so more suitable for low, small, or cystic kidneys

Anterior subcostal/transverse

Advantages: Good access to the renal pedicle

Disadvantages: Retraction of peritoneal cavity can limit access

A midline incision is generally preferred in traumatic cases or when the patient has a narrow subcostal angle. It allows for rapid access without muscle cutting via incision of the linea alba and provides good access to the entire peritoneal cavity. The incision is made from the xiphoid to the caudal aspect of the umbilicus but is easily extended inferiorly. Mass closure is performed.

A thoracoabdominal approach tends to be reserved for large upper pole tumors [7] and has the obvious disadvantage of entering the thoracic cavity and cutting costal cartilage.

Independent of incision choice, once the peritoneal cavity has been entered, the colon can be reflected medially by incising its lateral peritoneal attachment. On the left side, this is facilitated by dividing the splenocolic ligaments, which also prevents undue traction on the spleen. On the right side, the duodenum is reflected along with the colon. Exposure may be maintained with the use of a ring retractor. Vascular anatomy is variable, but on the right side the renal vein normally receives no tributaries, but it is short and care must be

taken not to damage the vena cava. If retraction of the right renal vein is difficult, the renal artery can be taken between the vena cava and aorta as opposed to lateral to the cava. On the left, the renal vein is long and typically receives gonadal, adrenal, and lumbar tributaries. These are ligated and divided to facilitate retraction of the renal vein and ligation of the renal artery. The kidney is mobilized by sharp dissection starting laterally to provide better access to posterior hilar area where friable veins may be present.

Surgical approaches in specific situations

Conversion from laparoscopic to open nephrectomy

The choice of conversion technique depends on the reason for conversion and the laparoscopic approach employed. Emergency conversion for severe hemorrhage should be via whichever route the surgeon feels will allow fastest vascular control. If possible, pressure should be applied to the bleeding point with a pledget or open swab pushed through a port site. If the reason for conversion is less precipitous, then the surgeon can be more circumspect about the incision. In retroperitoneoscopy, conversion is usually achieved by joining the port sites in the form of a subcostal loin incision. In transabdominal laparoscopy, the port site best placed to give access to the area of difficulty is extended.

Approaches to ectopic kidneys

Ectopic kidney position can be a result of abnormal migration (e.g. pelvic kidney) or abnormal fusion (e.g. horseshoe kidney and crossed fused renal ectopia). The choice of incision will depend on the position, size, and vascular supply of the renal unit to be removed, as determined by the preoperative imaging [8]. An extraperitoneal iliac fossa approach is often the best for a pelvic kidney. An oblique skin incision is deepened by dividing the external oblique aponeurosis in line with its fibers. The internal oblique and transversus abdominis muscles are divided by muscle splitting. The peritoneum is bluntly mobilized and retracted medially to give access to the retroperitoneal space and the kidney. The ectopic renal vessels can derive from the lower aorta or iliac vessels and the anatomy must be carefully defined before the vessels are taken and kidney removed.

An anterior subcostal extraperitoneal incision is usually the favored approach for a horseshoe kidney heminephrectomy, although a transverse supraumbilical transabdominal approach is better if access to both sides of the kidney is required. A midline incision is often preferred in older children and adolescents. The vascular supply varies considerably between individuals, and preoperative CT reconstruction can provide valuable information. The lower poles and isthmus frequently receive blood from the common iliac vessels, which can be intimately related to the gonadal vessels and ureters. Again, careful dissection is required to define the vascular anatomy. The isthmus can be thin and fibrous and, if so, can be divided between clamps and the edges oversewn. In a thicker isthmus, the renal capsule is incised and the arcuate vessels are ligated individually, with any calyceal breaches repaired.

Complications

Complications are uncommon in simple open nephrectomy in children. While there are numerous contemporary publications outlining complication rates following laparoscopic nephrectomy, there are no recent series with outcome data from open nephrectomy. In this section, the more frequently encountered and the potentially more serious complications are discussed, together with the methods by which they can be avoided and treated.

Intraoperative complications

Hemorrhage

Intraoperative hemorrhage is very unusual in open nephrectomy in children, and blood is usually only cross-matched if the surgeon anticipates difficulties, e.g. XGP nephrectomy. Preoperative contrast CT is rarely required.

If hemorrhage is encountered, it is vital that the anesthetist is immediately made aware of the problem. As a general rule, brisk hemorrhage is best initially dealt with by applying pressure whilst ensuring that suction and vascular equipment is available. Proper exposure of the area allows for precise vascular control and is more reliable than blind diathermy or clip application. Venous hemorrhage tends to be more troublesome and difficult to locate than arterial bleeding. The sites of

venous bleeding are to some extent predictable, with four areas most commonly encountered:

- 1 *Lumbar veins entering the posterolateral aspect of the vena cava at each vertebral level.* These may be damaged by traction on the cava and should be identified and ligated if the cava needs to be mobilized. If a lumbar vein is avulsed, compression should be applied to the cava above and below. The cava is then rolled medially and the ostium clamped with Allis clamps. The defect is then closed with a vascular suture. The proximal end of the lumbar vein can retract back into the psoas muscle leading to troublesome hemorrhage. If the vein cannot be grasped with a clip, the area of hemorrhage is oversewn.
- 2 *Right gonadal vein entering anterolateral surface of the vena cava.* Similarly, this thin-walled vein is at risk during mobilization or traction of the cava and avulsion can be repaired as described above.
- 3 *Lumbar veins draining into the left renal vein just lateral to the aorta and into the vena cava close to the entry of the right renal vein.*
- 4 *Right adrenal vein draining into the vena cava.*

Venous injury is less commonly encountered if dissection is undertaken in the relatively bloodless plane immediately adjacent to the cava's wall. If the cava itself is damaged, repair is best facilitated with pressure above and below the tear, using Allis or Satinsky clamps to allow the placement of a vascular suture.

Hemorrhage can also rarely be encountered as a result of splenic or hepatic injury. Traction is the most common mechanism of injury to the spleen. This can be prevented by taking down the lienorenal and splenocolic ligaments early in the procedure. Small, superficial tears in either organ can be controlled with pressure and application of hemostatic gauze (Surgicel). Deeper lacerations may require repair with mattress sutures, over Surgicel bolsters, if necessary. More extensive damage to the spleen can be managed by placing it in a bag or mesh to apply external pressure. Splenectomy is rarely necessary and only used as a last resort.

Bowel injury

The duodenum is particularly at risk in right nephrectomy and the colon can be damaged on either side. Careful mobilization (Kocherization) of the duodenum reduces the risk of unwitting injury from retraction or diathermy. If the duodenum is breached, it should be sutured directly, after debriding the area in the case of

diathermy injury. The same holds for colonic injury, with a defunctioning stoma reserved for a large or severely contaminated injury.

Pancreatic injury

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

Pneumothorax

The pleura is not infrequently breached, both deliberately and inadvertently, during nephrectomy. Small defects can be closed with running sutures, taking care to avoid the lung. Once the sutures are loosely in place, the lung is inflated to push out the fluid and air that have accumulated in the pleural space, before tightening the suture line. This process can be facilitated by placing a tube in the pleural space, which is removed when the lung is fully inflated. Larger defects are closed as fully as possible, leaving chest drains *in situ*. A postoperative chest radiograph should be taken to confirm resolution of the pneumothorax.

Early postoperative complications (≤30 days)

Pancreatic fistula

This presents in a similar fashion to acute pancreatitis and with fluid discharge from the wound. Ultrasound or CT scans will reveal a retroperitoneal collection and fluid analysis shows high pH and amylase. Treatment may involve percutaneous drainage of the collection to prevent pseudocyst formation. The majority of fistulae close spontaneously but extended periods of drainage and dietary support may be required.

Ileus

Ileus is more commonly encountered following trans-abdominal incisions. Most cases will resolve with nasogastric drainage and careful fluid and electrolyte replacement. Although the majority are due to bowel handling, care must be taken not to miss a more serious cause such as bowel injury or pancreatic fistula. Ileus lasting more than a few days or accompanied by systemic sepsis should be treated with suspicion.

Wound infection and dehiscence

Superficial wound infections are managed by opening superficial layers if necessary, and appropriate

dressings. Unless accompanied by systemic infection or spreading cellulitis, many do not require use of antibiotics. Wound dehiscence is rarely encountered with modern suture materials and wound closure techniques and generally reflects poor surgical technique. Deep wound dehiscence, particularly in transabdominal incisions, requires early surgical intervention but most can simply be resutured.

Chest infection

Atelectasis is common after nephrectomy, particularly with flank incisions. Both the operative and non-operative sides can be affected by the surgery itself and the flexed intraoperative position. The risk of chest infection is increased by inadequate postoperative analgesia, leading to poor chest expansion, expectoration, and mobilization.

Secondary hemorrhage and hematoma

The presentation depends on the briskness of the bleed. Severe hemorrhage is usually obvious, presenting with signs of shock and abdominal distension. Dry wound drains do not rule out the diagnosis and the hemoglobin does not necessarily fall in the acute phase of bleeding. Immediate surgical intervention is required in conjunction with resuscitation with intravenous fluid and blood. Slower hemorrhage may be less obvious and lead to hematoma formation, particularly in retroperitoneal incisions. Abdominal distension, abdominal wall bruising, and a fall in hemoglobin are common findings. Treatment can be conservative or by radiological or surgical drainage depending on the extent of the collection.

Renal insufficiency

Preoperative renograms are always obtained to give differential function, so unpredicted postoperative renal impairment is exceptionally rare following nephrectomy for unilateral renal disease. If nephrectomy is contemplated in bilateral renal disease, a pediatric nephrologist

is usually involved, and it may be necessary to assess glomerular filtration rate formally prior to surgery. Where unexpected renal insufficiency is encountered postoperatively, acute reversible causes need to be actively ruled out and, again, a nephrology opinion sought.

Late postoperative complications (>30 days)

Wound bulge

Loin incisions are frequently accompanied by postoperative wound bulge, especially in infants. This usually represents a localized muscle weakness secondary to muscle stretching or subcostal nerve neuropraxia, and tends to resolve spontaneously. Incisional hernia are rare but can complicate any of the approaches and, when encountered, surgical repair should be considered. Mesh can be used depending on the site and size of the defect.

Pain

Although rare in children, chronic wound pain can be encountered after any incision but is more common after loin approaches. In the majority of cases the wound appears well healed and there is no readily appreciable cause for the pain. In many individuals the pain is due to local nerve injury. If simple analgesia is not effective, input from the pain team should be sought.

Conclusion

Most pediatric urologists will rarely need to perform open simple nephrectomy. A good understanding of the surgical technique and the issues that surround the procedure remains important, particularly to laparoscopic surgeons who may have to convert to an open approach as a matter of urgency.

DOS AND DON'TS

Do

- Use the retroperitoneal approach if possible
- Maintain knowledge of open nephrectomy as rapid conversion from the laparoscopic approach may occasionally be necessary

Don't

- Forget that open nephrectomy remains a useful approach in selected situations

CLINICAL PROBLEM/CASE: DISCUSSION**Case 7.1 discussion**

The most likely diagnosis is a localized muscle weakness secondary to muscle stretching or a subcostal nerve neuropraxia. This is likely to resolve spontaneously over the next few months, and reassurance is all that is required. An incisional hernia would be very rare but needs to be excluded, as this would require surgical repair.

Case 7.2 discussion

This child may have sustained damage to the tail of her pancreas with subsequent pancreatic fistula formation. The fluid should be sent off for analysis and a high pH and high amylase content would confirm its pancreatic origin. The diagnosis should be confirmed with an ultrasound scan $\pm$ CT scan. If a collection is demonstrated, it should be drained percutaneously to prevent the formation of a pseudocyst. The majority of pancreatic fistulae close spontaneously, but they may require a prolonged period of drainage and dietary support.

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CHAPTER 8

Partial nephrectomy

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KEY POINTS

- Partial nephrectomy is indicated for ureteral duplication and associated polar renal dysplasia
- Up to 10% of patients require reoperation on the remnant ureteral stump
- Hemorrhage and urine leak are rare but problematic complications following partial nephrectomy

CLINICAL PROBLEM/CASE

Part A

A healthy, asymptomatic 2-month-old infant girl presents with antenatal hydronephrosis. Follow-up ultrasound (Figure 8.1) at 1 month of age shows upper-pole hydronephrosis and dysplasia of the left kidney, a cystic structure within the bladder, and a normal left lower pole and right kidney. Physical exam is unremarkable.

- 1 What further work-up and treatment should be considered for this child?
- 2 How should the parents be counseled regarding surgical options, including risks and outcomes?

Part B

A voiding cystourethrogram (VCUG) shows left-sided ureterocele and no evidence of associated reflux (Figure 8.2). MAG3 nuclear renography shows that there is

no appreciable function in the left upper pole (Figure 8.3). The child undergoes upper-pole partial nephrectomy at 6 months of age.

- 1 What are the keys for minimizing complications to the remaining lower-pole moiety?
- 2 What are options and outcomes for management of the distal ureteral stump?

Part C

The upper-pole moiety is removed via a flank incision with the distal ureteral stump left open. After an uneventful hospital course, she presents to the emergency department on postoperative day 15 with fevers of 103°F (39.5°C).

- 1 What is the appropriate work-up, including imaging, for this patient's fever in the setting of her recent surgery?
- 2 Ultrasound reveals a 5 cm fluid collection near the distal ureteral stump. What is the appropriate management of this complication?



(a)



(b)

Figure 8.1 (a) Renal ultrasound demonstrating a duplicated left collecting system with a dilated and tortuous upper-pole ureter with echogenic and thinned upper-pole parenchyma. (b) Longitudinal view of the bladder showing a debris-filled ureterocele and associated dilated ureter.

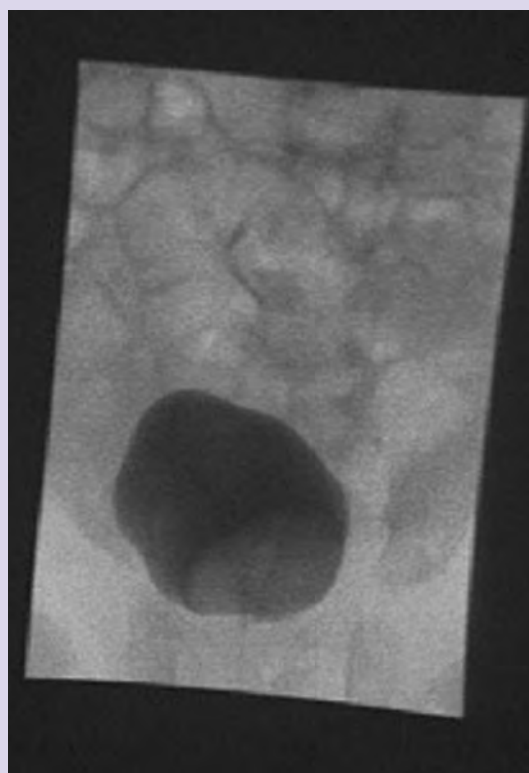


Figure 8.2 Cystogram showing a filling defect at the left base of the bladder, representing a large ureterocele.

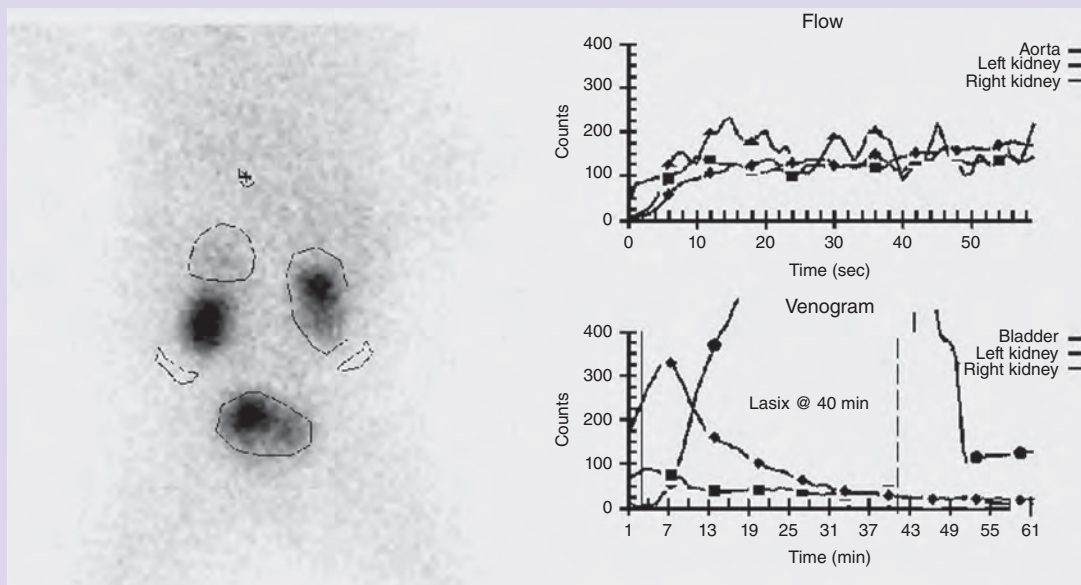


Figure 8.3 Nuclear renography with MAG3 showing absent function involving the left upper-pole segment.

Indications and evaluation

Partial nephrectomy in children is performed for ureteral duplication with renal dysplasia and rarely for malignancy and segmental renovascular hypertension in children.

Wilms tumor is the most common renal malignancy in children and may be managed with a partial nephrectomy in the instance of stage V bilateral tumors, a solitary kidney, or even for low-to-intermediate risk histology after good response to chemotherapy [1]. Renal cell carcinoma accounts for 2–6% of all renal tumors in children and may be managed by partial nephrectomy in select cases [2].

Ureteral duplication anomalies represent the most frequent surgical indication for partial nephrectomy and is the primary focus of this chapter. It has an estimated incidence of 0.3–2%, with a 2:1 female-to-male preponderance, and may be associated with ureterocele and ectopic ureter of the upper pole and/or vesicoureteral reflux (VUR) of the lower pole [3]. Ureteroceles are the primary finding in duplication requiring surgical correction [4,5]. Occasionally, segmental renovascular hypertension associated with a duplicated moiety may be managed by removal of the affected renal segment [6]. Renal dysplasia is noted in 43–70% of all ureteroceles with duplicated systems [7,8]. The principles of surgical repair in these conditions are to eliminate obstruction, reduce risk of infection, manage incontinence in girls, and preserve renal function of the healthy renal moiety.

Ureteral duplication commonly presents with an abnormal prenatal ultrasound, though febrile urinary tract infections (UTIs), flank mass, pain, bulging vaginal mass, renal disease, hematuria; continuous incontinence (in girls) may also be noted [9]. The history focuses on presenting symptoms, general health of the child, toilet habits, and associated urinary issues. Renal ultrasound is a mainstay of diagnosis, defining anatomy and degree of renal parenchymal loss. VUR is common and should be evaluated with voiding cystography. Nuclear medicine renography may elicit renal function and endoscopy can help to further define anatomy [5].

Procedure

In benign conditions, partial nephrectomy may be achieved via an open flank approach or, more recently, by using minimally-invasive laparoscopic or

robotic assisted approaches. Regardless of approach, the mainstays to a safe operation include clear identification of both the abnormal and main renal hila, segmental vessels and urinary collecting systems. Most authors prefer ligation of the abnormal renal blood supply first, to aid in demarcation of the dysplastic and normal renal parenchyma [10]). The dysplastic moiety may then be removed safely along this line, avoiding entry into the normal collecting system. Others prefer to resect the abnormal parenchyma first, to then aid in hilar identification [11]. The associated ureter is then easily identified and dissected as distally as reasonably possible. As the duplicated ureters reach the bladder, most enter the trigone in a common ureteral sheath with closely associated blood supply. Thus, care must be taken not to devascularize the healthy ureter. Dissection of the affected ureter while leaving the sidewall alongside the healthy ureter helps preserve the ureteral blood supply [5].

Outcomes

Partial nephrectomy for ureteral duplication with associated ureterocele

An upper-pole partial nephrectomy is indicated in a ureterocele with associated dysplastic upper-pole renal parenchyma. Endoscopic decompression for a single-system ureterocele results in an 80–90% success rate, without need for additional procedures [9,12]. However, following endoscopic management for ectopic or duplex ureteroceles, up to 50% of patients will have persistent obstruction or reflux, and up to 64% require reoperation [12,13]. Comprehensive management at the level of both the kidney and bladder calls for partial nephrectomy along with lower urinary tract reconstruction via separate incisions. This single-staged approach is associated with a high degree of surgical complexity and length of operative time [14]. Alternatively, upper-pole partial nephrectomy alone offers a high success rate whilst minimizing morbidity [15].

Several concerns regarding the success of partial nephrectomy for ureterocele include the fate of the remaining renal unit, the fate of the retained ureterocele and ureteral stump, and the rate of *de novo* VUR. The degree of preoperative VUR and presence of VUR into multiple moieties appear to predict the need for

future operations. Up to 40% of patients with low grade VUR into a single moiety preoperatively, and 98% of those patients with higher grade (III–V) VUR or VUR into multiple moieties preoperatively required re-operation [16]. Incontinence following upper-pole partial nephrectomy for an associated ureterocele is rare and is thought to arise from an intrinsic muscular defect at the bladder. Notably, these cases often require bladder neck reconstruction and/or artificial urinary sphincter placement for management of incontinence [16].

Partial nephrectomy for ectopic ureter

Up to 70% of ectopic ureters are associated with duplex collecting systems. Females often present with incontinence, with ureteral ectopia into the urethra, vestibule, vagina or, rarely, into the uterus or Gartner's duct cysts. In males, ectopic ureters may drain into the prostatic urethra or Wolffian structures such as the vas deferens, seminal vesicles, or ejaculatory ducts [17]. Males are continent but may present with obstruction or UTIs, especially if the ectopic ureter communicates directly to the vas deferens resulting in recurrent epididymitis [5].

Management options include ureteroureterostomy of the upper- to lower-pole ureter or upper-pole partial nephrectomy. Some authors champion ureteral reconstruction, citing the lack of significant dysplasia found in upper-pole partial nephrectomy specimens [7]. However, upper-pole partial nephrectomy for the ectopic ureter appears to be associated with as an equally low complication rate as ureteral reconstruction and adequately manages incontinence [7,18].

Partial nephrectomy for lower-pole vesicoureteral reflux

VUR can often be managed conservatively or with ureteral reimplantation, although lower-pole reflux nephropathy may necessitate partial nephrectomy. The approach for lower-pole partial nephrectomy is essentially the same as for an upper-pole defect. Complications of lower-pole partial nephrectomy appear similar to that of upper-pole partial nephrectomy, although one series reported a 9% transfusion rate. The presence of VUR does not appear to impact the outcome of the distal ureteral stump [19].

Management and outcomes of the retained ureteral stump

Excising the ureter completely reduces the theoretical risk of urinary stasis or reflux of urine into the ureteral stump but exposes the healthy ipsilateral ureter to risk. As such, leaving a remnant distal ureteral stump is generally recommended as an initial measure [15]. Typically, the ureteral stump is ligated only if VUR is present into the affected system and is otherwise left open to allow decompression and prevent stasis of urine [20,21]. Some authors routinely leave refluxing ureteral stumps open with minimal postoperative complications [19]. Surgeon preference varies with regards to postoperative drains. We prefer to leave a small Penrose drain near the open non-refluxing stump for postoperative drainage. The natural history of the retained ureteric stump has been described in several retrospective reviews. Up to 8–10% of patients managed with partial nephrectomy and partial ureterectomy required further excision of both refluxing and non-refluxing ureteral stumps, often within one year of surgery [15,22]. A more recent series reported a lower rate of stump excision, occurring in only 1 of 142 patients (0.07%). If VUR is present preoperatively, it is likely to persist postoperatively. However, reflux has been reported to resolve in up to 50% of patients undergoing partial nephrectomy for reflux nephropathy [23]. Thus, a small proportion of patients may require excision of the ureteral stump for management of persistent symptoms. Of those that require excision, many will have an obstructive component to the ureteral stump, while simple reflux alone into the ureteral stump rarely results in clinically significant symptomatology.

Specific complications of upper-pole heminephrectomy

Renal loss

Renal loss may occur due to misidentification and direct ligation of the normal renal vasculature or vasospasm resulting from aggressive dissection and handling of the main renal hilum. Complete renal loss may be slightly higher in minimally invasive techniques, with reports varying from 0–9%, although whether this is related to technique, pressure from pneumoperitoneum, or

more vigorous tracking and reporting of complications in the current era is unclear [20,24–26]. Patients may present with flank pain, fever, or hypertension during the postoperative period. Renal ultrasound may show renal blood flow into the proximal renal hilum with abrupt cessation of flow prior to reaching the remaining renal parenchyma, indicating a vascular etiology of renal loss [24]. However, complete loss of function can also present late with minimal or no symptoms. Completion nephrectomy of a nonfunctioning remnant is indicated for symptoms of pain or hypertension. Asymptomatic nonfunctioning renal units may be observed [20,24]. Some loss of renal function is expected, with mean loss of ipsilateral renal function reported at 2–6% based on nuclear medicine studies. Significant loss of overall renal function is less common, with only 5% of patients having a decrease in renal function of greater than 10%. In most cases, creatinine remains unchanged, owing to a normal contralateral renal unit. Neither proteinuria nor hypertension has been noted in patients with mild loss of ipsilateral renal function [27,28]. As renal loss can be a delayed complication of partial nephrectomy, periodic monitoring for hypertension and proteinuria seems reasonable. Renal torsion around the hilar vessels is a rare but devastating complication following complete mobilization of the kidney and has been primarily described in renal transplantation [29]. Although this complication has not been specifically described following partial nephrectomy, some authors advocate nephropexy to prevent renal torsion [19]).

Renal cysts

Renal cysts arising from residual dysplastic renal parenchyma are present in 18–34% of patients following partial nephrectomy [20,26,28]. Intermediate length follow-ups have not reported any adverse effects of retained dysplastic segment and have even suggested some spontaneous regression of up to half of the cysts [28]. Renal ultrasound is useful for following these cysts, although the necessity of long-term monitoring has not been defined.

Hemorrhage

Renal hemorrhage, often defined as need for blood transfusion, presence of a hematoma, or postoperative hematuria, is rare following partial nephrectomy.

Postoperative need for blood transfusion has been reported in up to 0.9% of patients, although most series report no need for intraoperative blood transfusion. Postoperative hematuria or incidentally discovered hematomas are rare and often managed conservatively [19,20,24,26]. These rare rates of hemorrhage are in contrast to higher rates of bleeding complications noted in the adult literature, where hemorrhage rates range from 1.6% to 4.2% in one large series [30]. The discrepancy likely arises from incising through otherwise healthy or even hypervascular renal parenchyma near tumor in adults as opposed to dysplastic and hypovascular renal parenchyma in children.

A theoretical late complication of partial nephrectomy, reported in adults but never to our knowledge in the pediatric population, is an arteriovenous fistula. This clinical entity often presents with persistent or intermittent gross hematuria several weeks after the initial procedure, and may result in hemodynamic instability or acute blood loss anemia. While computed tomography with excretory urography or Doppler ultrasound may identify a likely vascular source, renal angiography may be both diagnostic and therapeutic allowing for embolization of the lesion upon identification [31]. Such presentations are likely to be rare in the pediatric population owing to the dysplastic nature of the renal parenchyma being excised.

Urine leak

In the setting of partial nephrectomy for renal duplication, urine leak may arise from entry into the healthy ipsilateral collecting system, or persistent refluxing urine leakage from the retained ureteral stump. Such cases may present with fever, pain, leakage of urine through the flank incision, or may be asymptomatic. Rates of urinoma or urine leak have been reported in up to 5% following partial nephrectomy in children [20]. These cases may be managed with ureteral stenting and/or percutaneous drainage though others may resolve spontaneously [24,32]. While open repair for closure of a persistent urinary leak may be considered if conservative measures fail, in the absence of failed or inaccessible minimally invasive management with maximal urinary drainage, this is often not necessary.

DOS AND DON'TS**Do**

- Ensure adequate exposure and precise dissection to allow identification of the renal hilum, segmental vasculature and both ureters.
- Remember that *de novo* VUR of the retained ureteral segment may spontaneously resolve following heminephrectomy.
- Obtain a renal ultrasound to evaluate for renal infarction, hematoma, or urinoma, especially in case of unexplained postoperative fevers or pain.

Don't

- Ligate a non-refluxing ureteral stump that might cause trapped urine.
- Devascularize the healthy ipsilateral ureter during the ureteral resection.
- Induce vasospasm of the main renal hilum with aggressive tissue handling.

CLINICAL PROBLEM/CASE: DISCUSSION**Part A**

- 1 This patient is presenting with a duplicated system and upper-pole ureterocele with resulting obstruction of the upper-pole moiety. Voiding cystourethrogram should be obtained, as VUR is present in the majority of duplicated systems and may predict surgical outcomes [3,16]. Nuclear medicine renography can define renal function and cystoscopy may further define anatomy. Antibiotic prophylaxis for obstructing ureteroceles should be initiated to maintain sterile urine and prevent urosepsis; UTI may be as low as 3% in patients on antibiotic prophylaxis [16].
- 2 Surgical options include endoscopic ureterocele decompression, upper tract ureteral reconstruction, upper-pole partial nephrectomy alone or partial nephrectomy combined with excision of the remaining ureteral stump. Endoscopic decompression of a single system ureteroceles is generally successful, although the majority of patients with duplicated systems will require further intervention [9,12,13]. Upper tract ureteral reconstruction or partial nephrectomy alone have similar complication rates and outcomes with regards to the ureteral stump. Complete excision and reconstruction of the abnormal duplicated system has higher operative complexity but eliminates risks of the ureteral stump.

Part B

- 1 The key principles of the partial nephrectomy include proper identification and ligation of vasculature to the affected renal moiety with gentle handling and preservation of the main renal hilum. Ligation of the

blood supply results in demarcation between healthy and unhealthy parenchyma, easing excision. Dissection of the affected ureter is carried to the lowest point possible, whilst identifying the normal ipsilateral ureter [11].

- 2 A non-refluxing distal ureteral stump should be left open and is commonly ligated in the presence of VUR. Preoperative VUR is unlikely to resolve, though most *de novo* VUR (present in up to 20% of patients) will resolve spontaneously. In general, 10% of patients may require reoperation for symptoms of recurrent UTI [21,23,27].

Part C

- 1 Serum lab analysis and urine culture will help guide initial management and antibiotic therapy. Ultrasound of the kidney *with attention to the area of the ureteral stump and bladder* is key. Doppler ultrasound will evaluate renal blood flow, as renal infarction may present with fever [24]. A urinoma or hematoma may be present. The ureterocele should be evaluated to ensure adequate decompression. An ultrasound focusing only on the kidney may miss ureteral stump leak or inadequately decompressed ureterocele, especially in larger children.
- 2 The fluid collection may represent abscess, infected hematoma or infected urinoma, and in the presence of infection should be managed with percutaneous or open drainage (if percutaneous access is not feasible). Urinary stent placement in this healthy moiety may aid with healing of urine leak from damage to the normal ipsilateral collecting system [24].

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CHAPTER 9

Ureteropelvic junction obstruction

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KEY POINTS

- Gold-standard Anderson–Hynes success rate > 95%
- Age, presentation, and preoperative criteria do not affect outcome
- Most common complications are urinary leak and urinary infection
- Use of stents and/or nephrostomy tubes may decrease complications and improve outcomes
- Urine leaks and obstruction should be managed with immediate diversion with stent preferred
- Failures can be treated with endopyelotomy, but repeat pyeloplasty remains the gold standard

CLINICAL PROBLEM/CASE

A 1-year-old infant undergoes an open unilateral dismembered pyeloplasty for ureteropelvic junction obstruction. Preoperatively, the patient had grade 4 hydronephrosis on the affected side, which had been followed secondary to prenatal hydronephrosis. The preoperative renal scan demonstrated delayed drainage on the affected side. The patient was otherwise asymptomatic, with no prior history of urinary tract infection. An internalized stent was not placed at the initial surgery.

1 A postoperative ultrasound at 1 month demonstrates persistent hydronephrosis. What further studies would one obtain at this point?



2 A repeat ultrasound is obtained at 3 months postoperatively. Grade 4 hydronephrosis is still present. The patient remains asymptomatic. What is the next step?



3 The decision is made to proceed with a revision pyeloplasty. What approach could be used? Would drains be utilized with a revision pyeloplasty?

Surgical techniques

Kuster first described a “uretero-pyeloneostomy” as a direct anastomosis of the ureter to the renal pelvis in 1891. In 1892, Fenger adapted for urology the Heineke–Mikulicz, a general surgical technique for pyloric stenosis. The Fenger technique splits a stenosed ureteropelvic junction (UPJ) longitudinally to close transversely. In an attempt to achieve a smooth pelvic-ureteral transition with minimal excess tissue, the Foley Y-plasty evolved. This procedure advances a Y-shaped incision to close as a V [1]. A variety of flaps then ensued, such as the spiral flap by Culp–DeWeerd [2,3], the vertical flap by Prince–Scardino [4], the advancing V-flap by Devine [5], and the dismembered V-flap by Diamond–Nguyen [6].

The now common Anderson–Hynes dismembered pyeloplasty was first described in 1949 by British plastic and urologic surgeons J. C. Anderson and Wilfred Hynes [7]. As evidenced by the myriad other techniques described above, its elevation to gold-standard status was not immediate. In its original description, an L-shaped wedge of redundant renal pelvis was excised. The vertical arm of the L was closed primarily whilst the lower arm was anastomosed to a spatulated ureter. The original description did not include a stent, an issue under much current debate. In older studies, the Davis intubated ureterotomy, Anderson–Hynes, and Prince–Scardino reported success rates >80% [4,8] (Figure 9.1).

Outcomes

As the indication to perform a pyeloplasty is varied, so is the definition of outcome. These can include improved symptoms (e.g. flank pain or urinary tract infection), improved hydronephrosis on ultrasound, stabilization or improvement of renal function and T1/2 on radio-nuclide scan. Large historic series consisting mostly, but not exclusively, of dismembered repairs have reported successful repairs in as high as 210/214 (98%) [9] and 152/153 (99%) [10].

In a more recent and large series of dismembered repairs, Sheu et al. [11] retrospectively reviewed 102 patients with 109 kidneys. Their average age at operation was 21.7 months with a mean follow-up of 44 months (Table 9.1). All patients received a perinephric Penrose drain, but no collecting system drains were used. Four patients (3%) required reoperation. These consisted of repeat pyeloplasties and one nephrectomy. Renal function improved or remained stable in 99 (97%) patients. Preoperatively 92.5% of patients had T1/2 >20 min. This decreased to 7.5% postoperatively.

Another large series, by Tal et al. [12], retrospectively reviewed 103 patients. The mean age at operation was 12 months with mean follow-up of 32 months. The use of collecting system drains were left to the discretion of the surgeon. This series reported

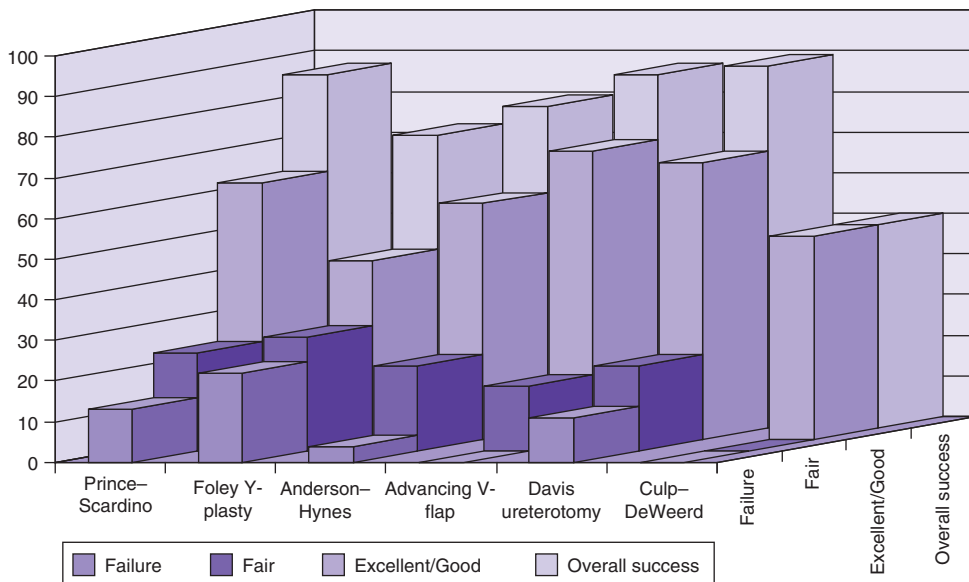


Figure 9.1 Historic success rates from 1961 and 1982. (Adapted from Prince and Scardino 1962 [4] and Tynes et al. 1981 [8].)

no incidences of reoperation, sepsis, or mortality. Decreased hydronephrosis was seen in 83 (80.6%) patients, improved drainage by renal scan seen in 90 (87.4%) patients, and improved or preserved renal function by renal scan seen in 92 (89.3%) patients (Table 9.1).

Outcomes by preoperative criteria

Whether one can predict functional improvement, i.e. who are the best operative candidates, remains a subject of active investigation. Zaccara et al. [13] retrospectively reviewed 69 patients who underwent stented Anderson–Hynes repairs. In attempting to correlate age, anterior–posterior diameter, parenchymal thickness, glomerular filtration rate, and differential renal function with postoperative improvement in differential renal function, no association could be found. The authors concluded that improvement based on renal scan criteria was a random event.

Outcomes by presentation

It has been shown that the manner of presentation, whether prenatal hydronephrosis, urinary infection, or pain, has no effect on initial differential renal function [14]. In assessing whether the mode of presentation af-

fects outcome, most series agree that initial presentation has little effect.

In the series by Sutherland et al. [15], 108 patients under 1 year of age presented with a prenatal ultrasound in 86 (80%) and urinary infection in 9 (8.3%). Conversely, the presentation of 101 patients greater than 1 year of age was dominated by pain in 47 (48%), urinary infection in 29 (24%), incidental in 13 (11%), and hematuria in 12 (10%). Outcomes did not differ between the two groups.

Tal et al. [12] noted that patients presenting with a urinary tract infection versus prenatal detection were more likely to be females and older. This was associated with an increased complication rate, but did not affect overall outcomes as measured by ultrasound or renal scan.

Salem et al. [16] in their series of 95 patients observed that symptomatic patients (e.g. urinary infection, abdominal pain, or palpable mass) showed a significantly greater improvement than asymptomatic patients presenting with ultrasound findings alone. This significance, however, did not persist on logistic regression analysis. The lack of association between outcomes and age or presentation supports that of a prior study by Macneily et al. [14].

Table 9.1 Modern presentations and outcomes of Anderson–Hynes dismembered pyeloplasty.

		Sheu et al. [11]	Tal et al. [12]
	No. patients	109	103
	Mean age (months)	21.7	12
	Mean follow-up (months)	44	32.4
Presentation (Tal) or surgical indication (Sheu)	Prenatal ultrasound	31%	77.5%
	Differential function, 40%	60%	N/A
	T1/2.20 min	22%	N/A
	Urinary infection	3%	7.8%
	Flank pain	14%	4.9%
Postoperative imaging	Decreased/stable hydronephrosis	98%	80.6%
	Improved/stable renal function	97%	89.3%
	T1/2 improved	76%	87.4%
Complications	Fever	N/A	31.1%
	Urinary infection	3.7%	12.6%
	Leakage	3.7%	7.8%
Outcome	Repeat pyeloplasty	3%	0
	Nephrectomy	1%	0

Adapted from Sheu et al. 2006 [11] and Tal et al. 2005 [12].

A significant difference that did persist on logistic regression analysis was renal function improvement as predicted by preoperative differential renal scan. Salem et al. found that patients with preoperative differential functions of <40% were significantly more likely to improve at least 5% than those with >40% function. This finding is likely influenced by the fact that well-functioning kidneys have little room to improve.

Outcomes by age

A consequence of increasing diagnoses by prenatal ultrasound is the evolution of patient age. Most series prior to the 1980s consisted exclusively of symptomatic older toddlers and children whereas most series from the turn of the century are now populated by asymptomatic infants. Whilst early studies reported higher infection and complication rates in those under 1 year old [17], more recent studies support successful outcomes in infants.

Sutherland et al. [15] reviewed their series of pyeloplasties from 1974 to 1994. This yielded 234 renal units who underwent Anderson–Hynes repairs via a flank incision. The use of stents or nephrostomy tubes depended on surgeon preference and did not correlate to age. Of these patients 108 were <1 year old and 119 were >1 year old. Decreased dilation on postoperative intravenous pyelogram or ultrasound was seen in 95% in those under 1 year old and 96% in those over 1 year old. They concluded that pyeloplasties were effective regardless of age.

Salem et al. [16] reported a series of 95 patients who received renal scans pre- and postoperatively. They found no difference in functional improvement in age groups ranging from <3 months to >5 years. As a group, about one-third of patients showed improved function and two-thirds showed stability.

If age does not affect outcome, two conclusions can be made. One is that surgical anatomy and technique is unhindered in the young infant, and the other is whether prompt surgery on young infants is indicated given equivalent outcomes at a later age.

Outcomes by delayed repair

Surgical versus nonsurgical management of ureteropelvic junction obstruction remains controversial. Immediate surgical repair was favored in the past; however, a study by Ransley et al. [18] revealed the possibility of conservative management. In this nonrandomized study, only 23% of children with an initial differential

renal function, .40% function, managed nonsurgically eventually deteriorated to require surgical correction. Not all patients regained or improved their renal function after pyeloplasty. This prompts the question whether waiting until the development of deterioration affects overall outcomes.

In a study by Apocalypse et al. [19], 77 children were managed surgically or with watchful waiting. Of 38 children initially conservatively managed, 12 (32%) eventually required surgical intervention due to deterioration on imaging or urinary tract infections. Though this group experienced a transient decrease in renal function, after repair there was no difference in renal function by DMSA scan between the conservative, early surgery, and delayed surgery groups.

Another study by Chertin et al. [20] retrospectively reviewed 44 patients who underwent delayed pyeloplasty for a >5% worsening of renal function by renal scan. All patients (100%) showed improvement in hydronephrosis by ultrasound. Forty-two patients (95%) showed improvement of their renal function with 36 (82%) regaining initial levels of renal function. They found no correlation between age, degree of hydronephrosis, or initial renal function and subsequent improvement of renal function.

Outcomes by surgical technique

Laparoscopic, robotic, and endoscopic techniques are discussed in other chapters of this book. In open repairs, the Anderson–Hynes is now used almost universally. One variable to the dismembered pyeloplasty is the choice of flank versus dorsal lumbar incision. Though the flank incision is more common in modern days, the dorsal lumbar incision has historic roots, being described as early as 1870 [21]. The dorsal lumbar approach offers exposure of the renal pelvis and hilum at the expense of the upper pole and distal ureter.

Wiener and Roth [22] described 33 consecutive children undergoing an Anderson–Hynes pyeloplasty. The first 17 patients received a flank incision with the next 16 patients all receiving a dorsal lumbar incision. Overall success rates were similar. One patient in the flank incision required reoperation while none required reoperation in the dorsal lumbar group. Overall complication rate in the flank incision group was 22% with two stent placements and two urinary tract infections. The complication rate for the dorsal lumbar incision group was 12% with stents required for a urinoma and

worsening hydronephrosis. One statistically significant finding was the decrease in operative time in those older than 12 months by dorsal lumbotomy.

A similar study by Kumar and Smith [21] retrospectively examined 91 infants. The choice of incision was surgeon dependent. The authors concluded that the dorsal lumbotomy was superior given the significantly decreased hospital stay (3 versus 7 days), faster time to oral intake (48 versus 83 hours), perceived superiority of exposure, and minimal learning curve.

Braga et al. [23] reported a similar study in 2010, retrospectively examining a total of 67 patients older than 3 years who underwent dorsal lumbotomy or flank incision pyeloplasties. Operative time was similar between the two techniques (113 minutes for dorsal lumbotomy and 117 for flank incision). Mean hospital time was also similar between the two incisions – 3.3 days for dorsal lumbotomy and 3.6 days for flank incision.

Both groups support the dorsal lumbar incision based on its excellent operative time, time to recovery and decreased hospital stay. Both groups also note that operative and recovery time are similar to those of endopyelotomy while providing an improved success rate. More recent studies show even shorter hospital stays between 1–2 days [24].

Complications

Prevention of complications during surgery starts with gentle tissue handling, preserving blood supply, and providing an adequate, tension-free anastomosis. Other factors such as identification of a crossing vessel, drainage tubes, and surgical approach can also affect postoperative recovery.

Most complications are present in the immediate postoperative period. Failures usually present within 2 years, with some being described as late as 8 years postoperatively in the pediatric population [25–27]. A review of literature by Smith et al. [28] compiled 833 patients from prior studies with an overall complication rate of 13%. Urinary leakage, urinary tract infection, and wound infection make up almost 90% of these complications (Figure 9.2).

Complications by use of drains

The use of indwelling stents, externalized stents, or nephrostomy tubes is heterogenous and largely

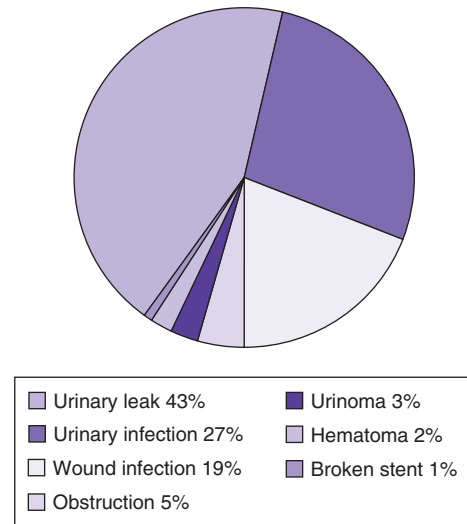


Figure 9.2 Distribution of complications by type. Total infection rate 108/833 (13%) patients. (From Smith et al. 2002 [28]. Reproduced with permission of Elsevier.)

dependent on surgeon preference. In most studies, a perinephric Penrose drain is utilized. The theoretical advantages to collecting system drains include decreased urinary extravasation, decreased urinoma formation, decreased obstruction secondary to postoperative edema, ability to assess radiographically via a nephrostomy, and optimization of alignment of the anastomosis. Those who do not use drains cite possible higher infection rates, dislodgement of tubes, continued possibility of obstruction or extravasation, need for further anesthesia in tube removal, and the questionable quality of a dry anastomosis.

Woo and Farnsworth [29] presented 54 patients, 13 of whom received no tubed drainage, six of whom received nephrostomy-only drainage, and 34 of whom received stent-only drainage. Results regarding need for repeat pyeloplasty and incidence of urinary infection and leakage favored the stent-only group. Repeat pyeloplasties were required in two (15%) of the tubeless drainage group, one (17%) of the nephrostomy group, and none of the stented group. Similarly, there were no incidents of urinary leakage among the stented group, but it occurred in 31–50% of the other two groups. The authors noted that while there was one stented patient (3%) with a urinary infection, this patient did not need further surgical intervention. Conversely, of the four (31%) patients in the tubeless group who developed a

urinary infection, 50% later went on to require a repeat pyeloplasty. This observation led to the conclusion that an infection in the face of leakage could have devastating consequences on anastomotic scarring. Therefore, these authors advocated a stent in all patients.

In assessing the utility of both a stent and a nephrostomy, Smith et al. [28] presented a retrospective, nonrandomized series of patients. Fifty-two patients had an externalized stent and nephrostomy tube while 65 patients were tubeless. This series found a similar overall complication rate between stent plus nephrostomy versus tubeless (13% and 17%, respectively). There was a trend toward a higher urinary tract infection rate with tubes (6%) than without (1.5%). While no patients with tubed drainage required repeat pyeloplasty compared to three patients in the tubeless group, this finding did not reach statistical significance. A review of the literature presented in this chapter demonstrated significantly more follow-up procedures needed in the tubeless group (9%) versus the tubed repairs (4%).

In support of the routine use of nephrostomy tubes, Austin et al. [30] presented findings on 132 patients. All patients underwent postoperative nephrostograms prior to nephrostomy removal. Notably, nephrostomy tubes were capped early in the postoperative course on postoperative day 0 or 1. Average length of follow-up was 2.1 years. Though 9% of nephrostograms initially showed extravasation, all nephrostograms later showed patent anastomoses with no subsequent obstruction. Their rate of urinary infection was 1.5%, comparable to other series. These authors concluded that their low rate of complications warranted the use of a nephrostomy tube as a means to achieve temporary diversion, perform radiographic assessment, and reduce extravasation.

In a randomized prospective trial, Arda et al. [31] divided patients into externalized stent versus no stent groups. Stents were removed on postoperative day 3 and Penrose drains on postoperative day 4, unless urine leakage was observed. In patients with Penrose drains only, drains were removed on postoperative day 3, unless urine leakage was observed. Arda et al. found no difference in hospital stay, urine leakage, or favorable results between those with an externalized stent and those without. A persistent urinary tract infection was present in one patient in the non-stented arm of the study. Based on this investigation, the authors

Table 9.2 Combined rates of complications in tubed drainage versus no tubed drainage.

	Tubed drainage	No tubed drainage
No. patients	244	94
Repeat pyeloplasties (%)	1 (0.4)	5 (5)
Overall complications (%)	17 (7)	19 (20)
Urinary tract infections (%)	6 (2)	6 (6)
Urine leak/obstruction/urinoma (%)	21 (9)	13 (14)

Adapted from Ransley et al. 1990 [18], Smith et al. 2002 [28], Woo and Farnsworth 1996 [29] and Austin et al. 2000 [30].

recommend the use of a stent only in selected patients such as those with poor renal function, severe hydronephrosis, a solitary kidney, or a revision pyeloplasty.

Table 9.2 shows combined data from the above studies. There exists a trend toward decreased complications of all types and a decreased need for repeat pyeloplasties in those with any type of tubed drainage. Though these data are not definitive, they do suggest that tubes do not increase complications and may even improve outcomes.

A retrospective review of 100 patients was performed in 2012 by Liss et al. [24], evaluating persistent urinary leakage after open dismembered pyeloplasty. All patients had a Penrose drain placed within Gerota's fascia at the end of the case. Seven patients also had a double-J stent placed, prior to the adaptation of a stentless technique. Of these, 86% were able to have their drain removed 1 week postoperatively, per protocol. Fourteen patients had persistent urinary leakage that required the Penrose drain to be left in place for longer: seven patients were dry at 14 days, six at 21 days and one at 27 days. Only two patients overall required redo pyeloplasty, one in the dry group and one in the persistent urinary leakage group. Their study demonstrated an internal ureteral stent was not necessary and did not lead to increased numbers of obstructions requiring redo pyeloplasties.

The cost-effectiveness of transanastomotic drainage has been evaluated in several studies. Braga et al. [32] examined 470 age-matched children who underwent primary open pyeloplasty over an 11-year period. They looked at a JJ internal ureteral stent and an externalized pyeloureteral stent (Salle stent). The externalized stent had a decreased cost of 565 Canadian

dollars per patient, secondary to avoiding a secondary anesthesia for stent removal. The overall complication rate was equivalent for the two stent types. Yiee and Baskin [33] developed a deterministic model evaluating no stent, external stent and internal stent during pyeloplasty. Their study found that an external stent was superior to no stent, and no stent was superior to an internal stent. This made internal stents the most inferior method, consistent with the prior study by Braga et al. [32].

Managing complications

As described above, the most common complications of pyeloplasties include urinary leakage, infection, and obstruction. The ultimate goals in managing complications are to preserve renal function and to prevent repeat surgery. Management of certain complications is a part of general medical knowledge. For example, most physicians would manage urinary tract infections with cultures and tailored antibiotic care. Management of other complications is not as straightforward. Specifically, how should leakage, obstruction, or malfunctioning drainage tubes be treated in order to minimize distress to the patient whilst maximizing function?

Initial management

Initial management of urinary leakage, obstruction, urinoma, or infection focuses on relieving symptoms and

trying to prevent scar formation. Urinary extravasation, especially in the setting of infection, is thought to promote scar and subsequent long-term failure. Therefore, all infections should be treated promptly, especially in the setting of extravasation.

It is also agreed that urine should be diverted to limit anastomotic scarring. Nephrostomies and stents are thought by some to be equally adequate modes of drainage; however, several authors [15,29] believe a stent is superior in maintaining a patent anastomosis. Sutherland et al. noted that of their patients who developed urinary leakage or obstruction, 3 of 4 (75%) patients treated with a nephrostomy eventually needed a repeat pyeloplasty whereas 0 of 5 patients treated with a stent eventually needed a repeat pyeloplasty.

After diversion and sterile urine are achieved, the patient may be reassessed in 6–8 weeks after an adequate period of recovery (Figure 9.3).

Definitive management

If obstruction persists after a trial of urinary diversion, definitive management should be attempted. Options include balloon dilation, endopyelotomy, repeat pyeloplasty, or ultimately nephrectomy.

Anatomic factors contributing to failure include scar tissue, a redundant pelvis, and crossing vessels that had been missed or are new since the

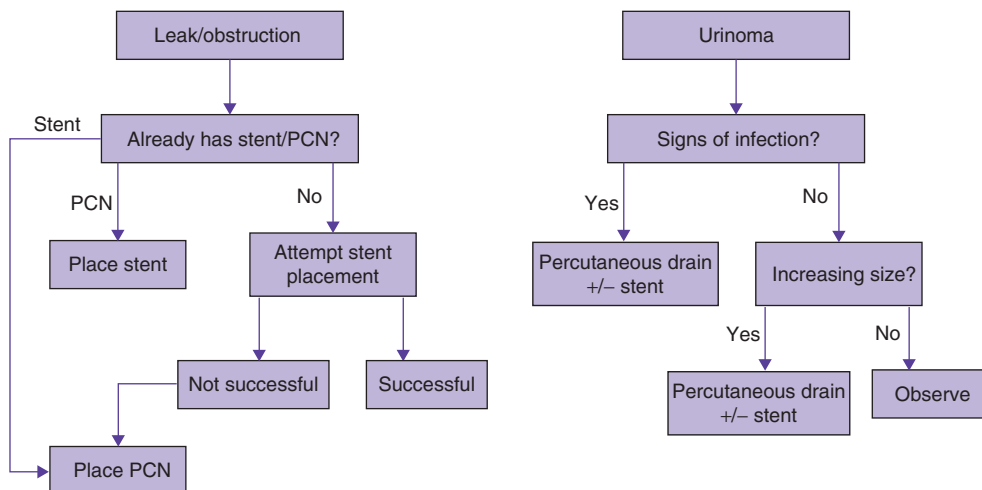


Figure 9.3 Initial treatment algorithm for urinary leak, obstruction, or urinoma PCN, percutaneous nephrostomy.

prior surgery. Methods used to maximize success in reoperation vary. Some advocate antegrade or retrograde pyelograms prior to any reoperation [34] to define the anatomy. Others believe CT scans before the primary or salvage surgery are warranted to evaluate for crossing vessels. Rohrmann et al. [34] utilize a transperitoneal approach to gain access to virgin tissue planes. In the cases of a ureterocalicostomy, employment of an omental wrap can decrease leakage and enhance blood supply. Ironically, though the use of stents and nephrostomy tubes is variable during primary repairs, their role in salvage repairs is more widely accepted.

The collective results of five case series show all 37 (100%) attempted repeat pyeloplasties were successful (Table 9.3). An additional six patients underwent successful ureterocalicostomy. Three patients underwent nephrectomy as the primary salvage technique due to renal function, intraoperative findings, and family preference. Therefore, in 42 attempted salvage repairs of recurrent ureteropelvic junction obstruction, all were successful.

Though open surgery is the most definitive approach, the use of endopyelotomy or balloon dilation is attractive given decreased morbidity. Jabbour et al.

[35] reported a series of patients undergoing endopyelotomy after failed pyeloplasty. Though children were a part of this group, the mean age was 35 years. The interval between pyeloplasty and endopyelotomy averaged 57 months. Mean follow-up at 88.5 months revealed that 63 of 72 patients (87.5%) had success as defined by resolution of symptoms and improved intravenous pyelogram findings. All patients who failed presented within one year. Failures were subsequently treated with nephrectomy (44%), repeat pyeloplasty (33%), endopyelotomy (11%), and ileal interposition (11%). The authors did not advocate preoperative CT scan to assess for existing vessels, but Clayman reports that doing so has decreased his postoperative bleed rate from 6.9% to 0% [36]. This technique provides an acceptable success rate while obviating the need to re-explore a scarred system. Unfortunately, this technique has yet to be well studied in the pediatric population.

A review of endopyelotomy in a pediatric population was published by Kim et al. in 2012 [37] of their 25-year experience. A 94% success rate (29/31 patients) was reported in their secondary endopyelotomy group, with a median follow-up of 61 months. Two patients developed postoperative hematoma/

Table 9.3 Presenting symptoms and outcomes of recurrent ureteropelvic junction obstruction.

		Thomas et al. [40]	Lim et al. [27]	Rohrmann et al. [34]	Persky et al. [26]	Sutherland et al. [15]
	No. patients	103	127	336	N/A	227
	No. failed (%)	7 (93)	3 (98) + 9 referrals	10 (97) + 6 referrals	8 (N/A)	9 (96)
Presenting symptom	Pain/obstruction	6 (86)	2 (17)	4 (25)	1 (13)	5 (56)
	Abnormal imaging	1 (14)	6 (50)	2 (13)	7 (88)	N/A
	Prolonged leak	0	3 (25)	5 (31)	1 (13)	4 (44)
Initial treatment	Stent	4	4	0	0	4
	Nephrostomy	1	2	10	0	5
	Balloon dilation (no. successful)	5 (1)	0	3	0	0
	Endopyelotomy (no. successful)	1 (0)	0	0	0	0
	Success with initial treatment (%)	1 (14)	0	0	0	6 (67)
Definitive treatment	Repeat pyeloplasty	3	10	13	8	3
	Ureterocalicostomy	3	0	3	0	0
	Nephrectomy	0	3	0	0	0
	Crossing vessel found (%)	2 (33)	2 (17)	0	0	0
Percentage of successful kidney salvage		100%	75%	100%	100%	100%

urinoma that extended their hospital stay to six and 11 days, both managed nonoperatively. One patient developed a postoperative ileus, leading to a 7-day hospital stay. No patients required a blood transfusion after their procedure. Success was defined in this study as absence of symptom recurrence and improved radiographic features on ultrasound, CT scan, IVP or diuretic renogram.

Balloon dilation in the treatment of primary pediatric ureteropelvic junction obstruction has reported success rates from 47–70% [38,39]. No studies dedicated to balloon dilation for recurrent ureteropelvic junction obstruction exist. Combined data from Thomas et al. [40] and Rohrmann et al. [34] (Figure 9.3) suggest that balloon dilation as a treatment for recurrent ureteropelvic junction obstruction has only a one-eighth (12.5%) success rate. Though morbidity is minimal with this intervention, its low success rate does not make it a definitive treatment.

Conclusion

The Anderson–Hynes dismembered pyeloplasty offers a >95% success rate for a common diagnosis in pediatric urology. The criteria for and timing of operation are often surgeon dependent; however, age preoperative factors, delayed surgery, and mode of presentation have not been shown to affect outcomes. Outcomes are affected by choice of incision, with a lumbar dorsal incision providing a shorter hospital stay and time to gastrointestinal recovery over the more common flank incision. The use of stents and nephrostomy tubes is also largely surgeon dependent, though data may suggest that complication rates decrease with tubed urinary drainage. When complications do occur, any urinary extravasation or obstruction should be managed with diversion. A stent is preferred when possible over a nephrostomy tube. Pyeloplasty failures are definitively treated with a repeat pyeloplasty, though adult literature suggests endopyelotomy may be an option.

DOS AND DON'TS

Do

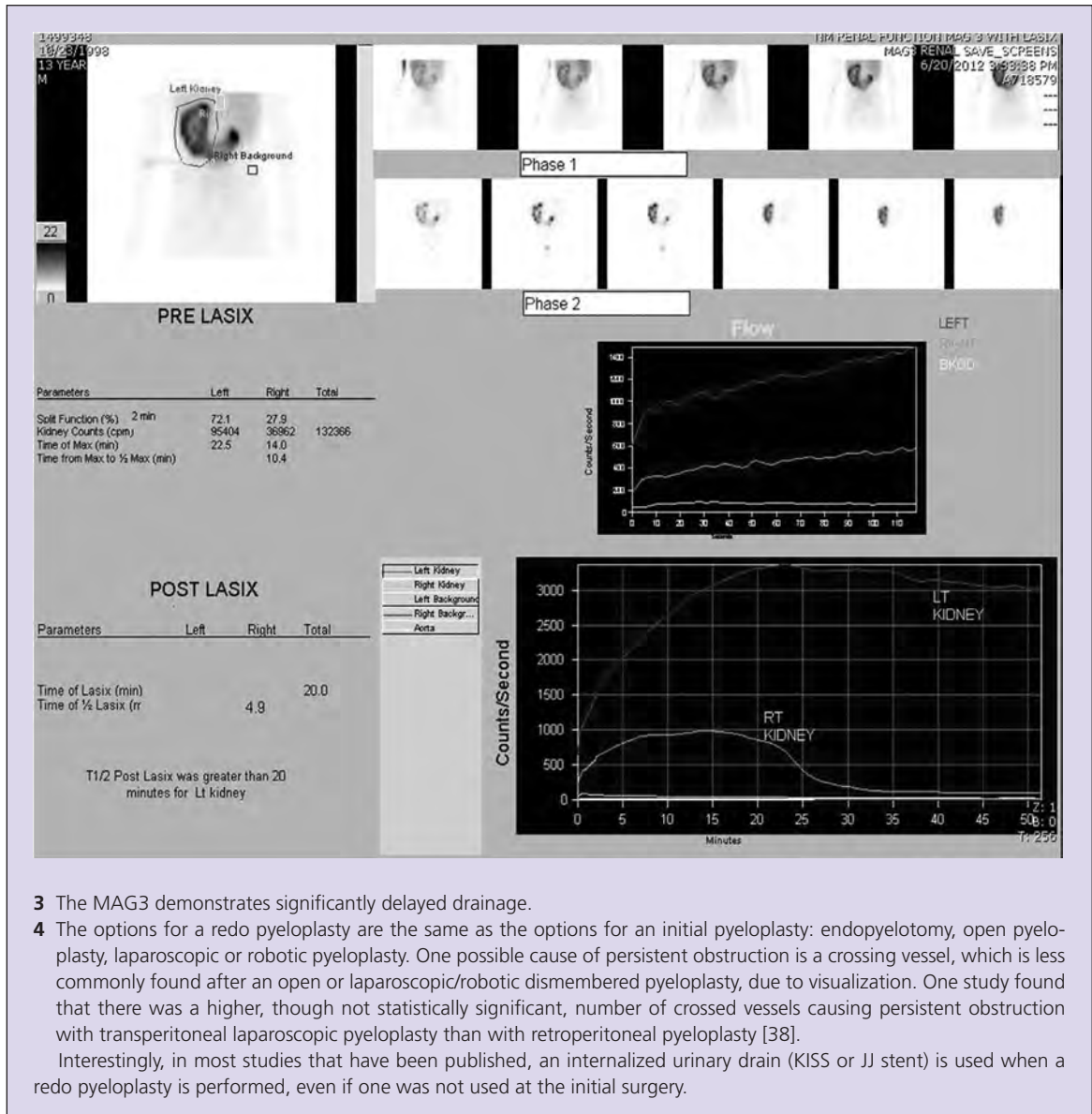
- Prevent complications by using the following surgical techniques:
 - gentle tissue handling
 - preserving blood supply
 - creating a patent, tension-free anastomosis
- Realize there is no proven method of drainage post-surgically that is ideal – the use of indwelling stents, externalized stents, or nephrostomy tubes is heterogenous and largely dependent on surgeon preference

Don't

- Delay in treatment of urinary infections, especially in the setting of urinary extravasation, after a pyeloplasty. Urinary extravasation, especially in the setting of infection, is thought to promote scar and subsequent long-term failure.

CLINICAL PROBLEM/CASE: DISCUSSION

- 1 Persistent hydronephrosis seen on ultrasound only one month after surgery can be attributed to postoperative changes: edema, continued decompression of a capacious urinary system. Published studies often specify that the first follow-up imaging was performed at least two months after the initial surgery. The best option would be to wait another month or two and allow the postoperative changes to subside before reassessing the urinary system.
- 2 Now a renal scan would be indicated. A MAG3 Lasix renogram is preferred, due to the lower radiation associated with the study and the information it gives regarding excretory function.



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CHAPTER 10

Complications of antireflux surgery

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KEY POINTS

- Open ureteral reimplantation has a success rate of 95–98%
- Laparoscopic/robotic ureteral reimplantation has published success rates comparable to open surgery but long-term data are lacking
- Failure to achieve an adequate tunnel length-to-ureteral diameter ratio in cases of high-grade reflux can lead to persistent vesicoureteral reflux (VUR) postoperatively
- Endoscopic management corrects reflux by injection of a bulking substance that allows elevation and coaptation of the ureteral orifice and wall
- Success rate for endoscopic therapy after a single injection ranges between 70% and 93%, depending on technique, material and experience
- Persistent/recurrent VUR is frequently caused by failure to recognize secondary reflux associated with neurogenic bladder or untreated bladder dysfunction
- Antireflux surgery has been shown to reduce the occurrence of febrile urinary tract infections
- The majority of postoperative complications are transient; however, progressive or symptomatic ureteral obstruction requires renal drainage
- The endoscopic method currently achieving the highest success rates is the double hydrodistention implantation technique (HIT)

CLINICAL PROBLEM/CASE

Case 10.1

A 2-year-old boy with bilateral grade III vesicoureteral reflux and recurrent urinary tract infections despite antibiotic prophylaxis undergoes an uncomplicated bilateral extravesical ureteral reimplantation. Urine output intraoperatively is adequate and the urethral catheter is removed at the conclusion of the case. Eight hours later the child has not voided despite IVF running at 1.5 times maintenance and receiving 10 cm³/kg fluid bolus.

- 1 What is the most likely etiology of the child's lack of urine output?
- 2 If an indwelling catheter is placed with no return of urine, what is the next step?

Case 10.2

A 7-year-old girl is seen in clinic 3 years after successful endoscopic injection of dextranomer-hyaluronic acid copolymer (Deflux®, Salix Pharmaceuticals, Raleigh, NC) for bilateral vesicoureteral reflux. She has had three culture-documented *Escherichia coli* urinary tract infections in the past 6 months and her primary care provider obtained a voiding cystourethrogram (VCUG), which demonstrated right grade II VUR. Upon questioning, the child states that she holds her urine and does not void while at school. Her mother also endorses urinary urgency, incontinence and constipation.

- 1 What is your initial management?
- 2 Why is this child not a good candidate for surgical intervention?

Introduction

Vesicoureteral reflux (VUR) is one of the most common urologic diagnoses affecting the pediatric population, with an estimated prevalence of 0.4–1.8% in the general pediatric population and 30% in those with a febrile urinary tract infection (UTI) [1,2]. Optimal management remains controversial, and options include observation with or without antibiotic prophylaxis and surgical repair. Surgical intervention may be necessary in children with persistent reflux, renal scarring or recurrent febrile urinary tract infections. An individualized risk-based approach that takes into consideration a multitude of demographic, radiographic, and clinical factors should guide management [3]. Ureteral reimplantation has a high success rate, and prevents VUR by increasing intravesical ureter length, which facilitates compression of the ureter against the backing of the detrusor muscle during bladder filling and emptying. Endoscopic repairs correct reflux by injection of a bulking substance that allows elevation and coaptation of the ureteral orifice and detrusor tunnel [4]. This chapter focuses on the surgical correction of VUR, patient selection, and potential complications of management.

Surgical techniques

Various open reimplantation techniques have been described, including both intravesical and extravesical approaches. More recently, robotic-assisted laparoscopic ureteral reimplantation has been reported.

The technique utilized for extravesical ureteral reimplantation is the Lich–Gregoir or one of its modifications. In this procedure, the juxtavesical ureter is dissected and a detrusor trough is created by incising the serosa and detrusor down to the mucosa, extending laterally from the ureteral hiatus. The bladder mucosa is not violated. The ureter is placed into the trough, and the detrusor is closed over the ureter, creating a flap valve mechanism without opening the bladder [5,6].

The Cohen cross-trigonal reimplant is the most widely utilized intravesical ureteroneocystostomy technique, due to reliable results and broad applicability. It maintains the same ureteral hiatus in the bladder wall. The ureter is advanced through a submucosal tunnel across the trigone to the contralateral bladder wall with the new orifice located superiorly [7–9]. It is well-suited

for small or thick-walled bladders, as ureteral advancement across the back wall of the bladder rarely results in kinking or obstruction. In the Politano–Leadbetter reimplant, the ureter is mobilized intravesically and then brought through a new muscular hiatus located superior and lateral to the original mucosal orifice [10,11]. The Glenn–Anderson technique is similar to the Cohen cross-trigonal but involves the ureter being advanced distally through a submucosal tunnel extending inferomedially towards the bladder neck [12].

Laparoscopic approaches to ureteral reimplantation were initially reported in the 1990s [13–15]. More recently, robotic-assisted laparoscopic approaches have been described, negating some of the longer operative times and greater technical difficulty associated with pure laparoscopy [16,17]. Long-term studies are required to demonstrate cost and quality-of-life benefits of robotic surgery over standard open repairs.

Endoscopic treatment of VUR using a bulking agent was pioneered by Matouschek in 1981 as an alternative to antibiotic prophylaxis and ureteral reimplantation [18]. The technique was further developed by O'Donnell and Puri, who performed subureteric injections (STING approach) in 13 patients [19]. Modification of the injection technique further evolved to include two intramural injections (proximal and distal), to achieve total ureteral tunnel coaptation via the hydrodistention implantation technique (double HIT) [20,21]. The ideal injectable material should be durable, effective, safe, and not extrude or migrate. Currently, the only FDA-approved bulking agent is dextranomer hyaluronic acid co-polymer (Deflux®). Due to its minimally invasive nature, endoscopic injection has become the most common means of correcting VUR worldwide.

Patient selection and timing of surgery

Spontaneous resolution of primary reflux is common due to remodeling of the ureterovesical junction (UVJ), elongation of the intravesical ureter and stabilization of bladder voiding dynamics. Resolution depends on initial grade of reflux, gender, age, voiding dysfunction, presence of renal scarring and timing of VUR [3,22]. Surgical intervention is therefore individualized and based on patient age, health, VUR grade, clinical course, renal scarring and parental preference. Indications for

surgical correction of VUR include moderate-to-high grade reflux (grades III–V), low probability of spontaneous resolution, renal scarring, recurrent pyelonephritis, breakthrough febrile UTI while on continuous antibiotic prophylaxis and parental preference [9,45]. While high grades of VUR are more likely to result in recurrent urinary tract infections and renal damage, low grade of reflux associated with other risk variables can also result in renal damage.

Assessment of outcomes

Clinical success following any antireflux surgery may be based on radiographic (absence or down-grading of VUR) and clinical findings (absence or decrease in febrile urinary tract infections). Current data do not always define success in these terms and thus true surgical outcomes are difficult to determine [23].

The Cohen cross-trigonal procedure is the most widely utilized transvesical approach, with a success rate of 98% in cases of primary reflux [24,25]. Results are similar for the Politano–Leadbetter repair and Lich–Gregoir extravesical reimplant, with published success rates of >95% [26,27]. Robotic ureteral reimplantation lacks long-term follow-up, but recent reports suggest an operative success rate similar to that of open surgery [28,29]. The average resolution of VUR following a single endoscopic injection is 83% based on aggregate data, though success rates have ranged from 70% to 95% [45].

Factors affecting surgical outcomes

Recurrent febrile UTI is the most common complication following both endoscopic injection and ureteral reimplantation surgery, having been reported in up to 28% of cases. Persistent VUR is the next most common complication of antireflux procedures. Endoscopic failures may be secondary to implant migration, disruption (mucosal breach) or dissolution with decrease in implant volume [4]. Although persistent reflux following reimplantation may be due to technical error, more often it is secondary to overt bladder pathology or failure to recognize underlying bladder dysfunction. Pathology can be in the form of neurogenic bladder, anatomic anomalies or voiding dysfunction. It also may be due to a large

ureteral diameter such as with a megaureter, where an adequate 5:1 tunnel length is not achieved. In rare cases, urodynamic studies may be needed to identify such problems when recurrent surgical failures arise.

The etiology of a neurogenic bladder will often be obvious, such as in the case of a myelomeningocele or spinal cord injury. A more subtle presentation would be an older child with an occult presentation of tethered spinal cord. In most cases, reflux will resolve or improve by directly decreasing bladder pressure via intermittent catheterization, anticholinergic therapy, or bladder augmentation. In the rare infant experiencing recurrent infection, especially with ongoing renal scarring or progressive hydronephrosis despite adequate medical management, vesicostomy is a temporary option that assures bladder drainage. Anatomical causes of bladder dysfunction that may negatively impact the results of reimplantation include the presence of a ureterocele, paraureteric diverticulum, or bladder outlet obstruction, such as posterior urethral valves.

Voiding dysfunction or dysfunctional elimination by far accounts for the majority of treatment failures. These patients can be identified based on a history that elicits symptoms such as urinary tract infection, incontinence, urgency, frequency, and constipation. Aggressive treatment, including strict adherence to voiding and bowel regimens will lead to resolution of reflux as well as associated lower urinary tract infections. The 2010 American Urological Association (AUA) Guidelines recommend management of any suspected bladder/bowel dysfunction, preferably prior to any surgical intervention [45]. If these children have failed reimplantation surgery, correction of the underlying voiding dysfunction will often eliminate the need for further intervention.

Preventative measures to avoid complications

In addition to proper patient selection, there are a number of technical concepts that help to ensure a successful outcome. Paramount among these surgical principles is adequate detrusor muscle backing. Regardless of the surgical approach utilized, the submucosal tunnel or trough length should be at least five times the ureteral diameter [30]. Adequate mobilization of the distal ureter should be accomplished without tension or damage

to the delicate ureteral blood supply. It is important that the chosen technique be done in such a fashion that excessive trauma to the tissue is avoided in order to prevent secondary obstruction.

Thus, minimal handling of the ureteral tissue cannot be overemphasized. The mucosa of the orifice should not be touched in order to prevent edema. The judicious use of retraction sutures facilitates minimal touch techniques. These sutures will also allow the surgeon to maintain proper orientation and avoid kinking of the ureter.

Importance is placed on a tension-free anastomosis. The direction of the new hiatus should be in a location so as to avoid stenosis, angulation and torsion of the ureter. The anastomosis should be fixed securely. Placement of an anchoring stitch through full-thickness ureter and both bladder mucosa and detrusor helps to ensure that the ureter does not retract into the submucosal tunnel – thus shortening tunnel length. Finally, the original detrusor hiatus is closed to avoid a bladder diverticulum, with care taken to not occlude the ureter.

The need for routine ureteral stenting remains controversial. Stenting should at least be considered when the distal ureter has been tapered and in those patients with a thickened or scarred bladder or reoperative cases in order to minimize temporary postoperative obstruction. Stenting is often performed in infants undergoing ureteroneocystostomy due to the small ureteral caliber. Preoperative stenting of the ureter may also be advantageous in a laparoscopic/robotic approach where the stiffness can assist with easy identification of the ureter. Routine placement of perivesical drains is no longer advocated. In the rare event of urinary extravasation from the suture line, a urethral catheter can be placed making re-exploration rarely necessary.

Likewise, there are technical errors that need to be avoided with endoscopic injections as well. Failures can result from improper injection technique, ineffective injection site, inadequate volume injected or ambiguous endpoints for injection.

In the traditional STING procedure, the needle is introduced under the bladder mucosa 2–3 mm below the refluxing orifice at the 6 o'clock position and the injection is continued until there is a prominent bulge with the orifice assuming a crescent-like shape. Alternatively, the hydrodistention implantation technique (HIT) and double HIT require that the tip of the cystoscope be placed at the ureteral orifice, and a pressured stream achieved by placing the irrigation bag approximately 1 m above the

bladder on full flow. Hydrodistention allows visualization of the intraureteral injection site, grading the severity of ureteral tunnel laxity, and also assessment of treatment progress (ureteral coaptation). With the double HIT methodology, the needle is inserted at the mid-ureteral tunnel at the 6 o'clock position, and bulking agent injected to produce a bulge which coapts the ureteral tunnel. A second implant is injected within the most distal intramural tunnel, which leads to coaptation of the ureteral orifice. When hydrodistention ceases to dilate the orifice, appropriate coaptation has been achieved. The HIT methodology requires that an adequate volume of injection be utilized and directly correlates with the hydrodistention grade of the ureteral orifice [4].

Risks and complications

Complications are categorized into those that occur in the immediate postoperative period and those that manifest later, up to several years from the time of surgical intervention (Table 10.1).

Early complications

Early complications occur within the first few days following surgical intervention and are typically transient. They include low urine output, hematuria, bladder spasm, retention, voiding dysfunction and infection. Obstruction can also occur following ureteral reimplantation, and in the acute postoperative period is often due to technical issues such as twisting or kinking of the ureter in its new tunnel, intramural blood clots or extramural compression at the anastomotic site [31]. Endoscopic management of VUR carries a 0.6% risk of ureteral obstruction, and is often associated with bladder dysfunction and markedly dilated ureters [4].

Table 10.1 Early and late complications of surgical intervention for vesicoureteral reflux (VUR).

Early complications	Late complications
Hematuria	Obstruction
Bladder spasms	Persistent VUR
Urinary retention	Urinary tract infection
Voiding dysfunction	
Urinary tract infection	
Obstruction	

Preoperative dehydration, obstruction at the level of the ureter (edema) or bladder outlet (clot or catheter balloon) can cause decreased urine output in children following ureteral reimplantation. Initially, the position of the catheter and absence of clots should be assured with gentle bladder irrigation. Often, inadequate hydration is uncovered as the culprit. Prevention of this problem is paramount by assuring vigorous hydration during the intraoperative and immediate postoperative periods. If anuria or oliguria persists beyond 24–48 hours despite these interventions, a renal bladder ultrasound and serum creatinine level should be obtained to look for obstruction or, more rarely, urinary extravasation. Extravasation may be treated initially with prolonged Foley catheter drainage, whereas complete obstruction requires placement of ureteral stents or nephrostomy tube drainage. Placement of ureteral stents should first be attempted endoscopically. However, if the cross-trigonal approach was used, placement of feeding tubes into the newly-positioned ureteral orifices may require reopening the incision and bladder for open ureteral catheterization. Since this is typically done within 48–72 hours of the original surgery, little wound healing has taken place and the skin, fascia, and bladder incision can be reopened with little effort. The new orifices are typically edematous but will accept a stent initiating a brisk postobstructive diuresis. Placement of nephrostomy tubes avoids reoperation but has its own disadvantages. Nephrostomy tubes may require more intensive postoperative care to prevent dislodging and can be more uncomfortable for the child. They do offer an option for antegrade contrast studies to determine if the obstruction has resolved or antegrade stent placement in the rare occasions where this may be necessary.

Hematuria and bladder spasms are frequent complications of transvesical ureteral reimplantation and usually resolve within two weeks of surgery. The intravesical techniques are associated with a greater incidence of postoperative hematuria and bladder spasm. These symptoms can be distressing; however, reassurance and selective use of anticholinergics are typically all that are necessary. Gross hematuria and bladder spasms are rarely seen following extravesical reimplantation or endoscopic treatment.

Transient voiding dysfunction has been reported after open surgery. Symptoms include urge incontinence and nocturnal enuresis due to inflammation, which resolves over several weeks. The extravesical approach has been

associated with postoperative urinary retention, possibly as a result of transient bladder denervation [40,41]. The incidence of this complication is higher in cases of bilateral extravesical reimplantation, and such patients are discharged with an indwelling catheter or on clean intermittent catheterization for a period of time. Prolonged use of bladder catheters and avoidance of bladder over-distention may decrease the risk of acute urinary retention [42]. Patients with a previous history of voiding dysfunction must be encouraged to continue their voiding regimen. Some surgeons place a temporary bladder drainage system at the time of the bilateral extravesical reimplantation, such as a suprapubic tube or Mic-Key button. This facilitates monitoring of post void residuals without the need for repeated urethral catheterizations. While the incidence of transient bladder dysfunction after bilateral ureteral reimplantation varies, nearly all cases resolve within two months of surgery. Permanent bladder dysfunction may occur following reimplantation surgery; however, the literature suggests that such an occurrence would be extremely rare.

It is not uncommon for patients to develop a febrile urinary tract infection after ureteral reimplantation or endoscopic injection [32]. This can often be avoided by obtaining a preoperative urine culture. Since a preoperative urine culture is not always practical, a urinalysis or urine dip the morning of surgery will alert the physician to the possibility of infection.

Late complications

Ureteral obstruction is the most serious late surgical complication of reimplantation. As mentioned previously, anuria or oliguria persisting beyond 24–48 hours mandates ultrasound imaging to rule out obstruction. Kinking, excessive angulation or devascularization of the distal ureter can lead to obstruction following ureteroneocystostomy. A recent review of 28 articles reported an overall post-reimplantation obstruction rate of 0.4% [45]. Complete obstruction necessitates nephrostomy tube and/or ureteral stent drainage. Although there are anecdotal reports of resolution, the majority of these patients will require reoperation, most often a redo ureteral reimplantation, via either an intravesical or dismembered extravesical approach. In cases where the affected ureter is not healthy for reimplantation, a transureteroureterostomy may be necessary. It is not unusual to see temporary resolution with continuous

Foley catheter drainage. Angulation may not occur in the decompressed bladder and is seen only with filling.

Less than 4% of children undergoing endoscopic VUR therapy complain of flank or nausea postoperatively, and nearly all resolve with analgesics. Ureteral obstruction following endoscopic injection is approximately 0.6%, and is frequently associated with voiding dysfunction and larger ureters where a high volume of bulking agent is injected [4,45].

Postoperative reflux may be due to persistent reflux in the reimplanted/injected ureter(s) or new onset contralateral reflux. For open bladder surgery, the reported incidence of persistent VUR is up to 2% [33]. In almost all cases, observation is the preferred treatment as spontaneous resolution occurs over time. Treatment failure following endoscopic therapy ranges from 7–50%, and is dependent on technique, VUR grade and surgeon experience [34]. Success rates for the double HIT technique have approached those of ureteral reimplantation [35].

While not a true surgical complication, the development of contralateral VUR after unilateral uretero-neocystostomy or endoscopic injection may require continued medical or surgical treatment. New onset contralateral VUR has been observed in up to 19% of open unilateral reimplants [36], with similar rates reported in endoscopic cases [4]. This complication has been attributed to various conditions, including interference with the normal ureteral attachment during intravesical ureteral dissection, occult reflux, and the possibility that high-grade VUR may be a pop-off mechanism for high bladder pressure, which when corrected may destabilize the contralateral ureter. Resolved contralateral reflux is a risk factor for new onset occurrence and is seen in 45% of patients where only a single side is surgically corrected [37]. The majority of these patients will resolve with time and observation, often on continuous antibiotic prophylaxis [36,38]. While many experts in the field consider reflux to be a bilateral process and will correct abnormally-appearing contralateral orifices to prevent new reflux from occurring, the true risk benefit has not been determined.

While open, robot-assisted laparoscopic and endoscopic approaches to reflux correction have all been shown to reduce the occurrence of febrile UTI, recurrent cystitis and febrile infections remain the most common complication following VUR surgery. In children with postoperative febrile UTI or recurrent infections following surgical treatment of VUR, evaluation for

bladder and bowel dysfunction or recurrent VUR is recommended [9].

Suggested follow-up after surgical correction of VUR

Patients should be kept on prophylactic antibiotics until postoperative studies have been obtained, especially if there was a history of recurrent urinary tract infections preoperatively. A renal ultrasound should be obtained approximately 3 months following ureteral reimplantation to assess for hydronephrosis and possible obstruction. Mild dilation is expected due to transient edema. Moderate or worsening hydronephrosis may be a sign of significant obstruction and requires further testing. Comparing the degree of postoperative hydronephrosis to the preoperative reflux grade is helpful in assessing asymptomatic postoperative hydronephrosis. In cases where obstruction must be excluded, a nuclear diuretic renogram can be obtained to confirm these findings. Based on the 2010 AUA Guidelines, a voiding cystourethrogram (VCUG) may be obtained following open reimplantation but can be limited to high-risk cases or those with postoperative pyelonephritis or new hydronephrosis, due to the high success rate of the procedure [45]. Long-term sonographic follow-up is not routinely necessary [31,39].

As with ureteral reimplantation, a renal ultrasound should be obtained 4–6 weeks following endoscopic treatment of VUR. A postoperative voiding cystography is also recommended by the most recent AUA Guidelines [45], but there remains wide variability in postoperative imaging dependent upon the individual patient and the surgeon's clinical experience and success rate [23].

The long-term impact of VUR and renal injury on individual patients is unknown. Although the incidence of serious side effects is low, they increase with length of follow-up. Screening for late-occurring complications of VUR is performed yearly and includes measurement of blood pressure, selective renal sonography, and a urinalysis, to monitor proteinuria, renal growth, hydronephrosis and infection. Normal values for spot urine protein are inconsistent. Measurement of urine protein is more likely to be of benefit in those individuals with bilateral scarring.

Patients with recurrent febrile urinary tract infection after successful surgical treatment of VUR should be evaluated for elimination dysfunction and recurrent reflux.

Conclusion

Ureteral reimplantation is a safe and effective intervention for management of VUR. The short- and long-term complications of ureteral reimplantation are well documented. The majority of ureteral reimplantation complications are transient, and managed conservatively during the immediate postoperative period. Endoscopic injection of bulking agents has emerged as a highly

successful, minimally invasive alternative to uretero-neocystostomy, with minimal morbidity. Progressive ureteral obstruction is a serious complication of either treatment modality, and although occurring in less than 1% of children, it requires intervention with ureteral stenting or nephrostomy tube placement to achieve renal drainage. Persistent reflux is often managed conservatively, and bowel/bladder dysfunction should be addressed prior to surgical revision in most cases.

DOS AND DON'TS

Do

- Thoroughly counsel families regarding various VUR management options
- Screen all children for and treatment of bowel/bladder dysfunction prior to ureteral reimplantation or endoscopic therapy, and continue the therapy postoperatively
- For ureteroneocystostomy, ensure the tunnel length to ureteral diameter ratio should be at least 5:1
- Obtain a renal ultrasound following ureteral reimplantation to assess for obstruction; renal ultrasound and postoperative voiding cystography is recommended following endoscopic injection of bulking agents, but depends on the surgeon's success with the procedure

Don't

- Handle ureteral tissues excessively during ureteral reimplantation, and respect the ureteral blood supply

CLINICAL PROBLEM/CASE: DISCUSSION

Case 10.1 discussion

Preoperative dehydration is a common cause of low urinary output in the initial postoperative setting; however, this child has received adequate IV hydration as well as an appropriate fluid bolus, making a pre-renal etiology unlikely. The extravascular approach to ureteral reimplantation offers the benefit of decreased hematuria, fewer bladder spasms, and overnight hospitalization in most cases. However, it has been associated with postoperative urinary retention in up to 30% of bilateral cases, possibly as a result of transient bladder denervation [40,41], or bladder overdistention [42]. The incidence of this complication is higher in cases of bilateral extravascular reimplantation, and is the most likely cause of lack of urine output in this clinical scenario.

If placement of a urethral catheter does not result in urinary drainage, the child has anuria rather than postoperative urinary retention. The catheter should be gently irrigated to ensure there are no clots, and also to confirm isovolemic return and therefore rule out significant urinary extravasation. If anuria or oliguria persists beyond 24–48 hours despite these interventions, BUN and creatinine should be evaluated and a renal bladder ultrasound performed to look for urinary extravasation or hydronephrosis. Extravasation may be treated initially with prolonged Foley catheter drainage, whereas complete obstruction requires placement of bilateral ureteral stents or percutaneous nephrostomy tubes, the former technique may be technically difficult early following ureteral reimplantation depending on the technique used.

Case 10.2 discussion

This child has numerous symptoms related to elimination dysfunction. There is a clear co-existence of lower urinary tract symptoms and VUR [43,44]. Aggressive treatment of both the patient's constipation and voiding dysfunction, including strict adherence to a bowel regimen, increased hydration and timed voiding schedule is the initial management strategy in this scenario.

The 2010 AUA Guidelines recommend management of any suspected bladder/bowel dysfunction, preferably prior to any surgical intervention [45]. If these children have failed reimplantation or endoscopic therapy, correction of the underlying voiding dysfunction will often eliminate the need for further intervention.

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CHAPTER 11

Ureteral surgery

Ureteroureterostomy and transureteroureterostomy

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KEY POINTS

- Ureteroureterostomy is a safe, effective procedure for managing ureteral duplication anomalies
- Urinary drainage with stents and/or drains is crucial for preventing postoperative complications
- Vesicoureteral reflux (VUR) can develop or persist after surgical intervention and usually can be managed conservatively
- Symptomatic VUR into remnant distal stumps is uncommon in distally performed operations
- Problems with anastomosis patency are uncommon
- Transureteroureterostomy can be a viable option for salvage procedures and diversion/undiversion

CLINICAL PROBLEM/CASE

A 6-month-old girl was referred to the pediatric urology clinic with a four-month history of two febrile urinary tract infections that both required hospitalization. Renal ultrasound is shown (Figure 11.1), demonstrating left upper-pole dilation and megaureter. Further investigations demonstrated no reflux and <10% of total function emanating from the upper pole on voiding cystourethrogram (VCUG) and nuclear scans respectively. She was taken to the operating theatre for unilateral left upper- to lower-pole end-side ureteroureterostomy with stent placement. Postoperatively, several hours after her Foley catheter had been removed on the first postoperative day, she had a fever of 38.9°C (102°F) and was lethargic. Ultrasound revealed a urinoma and cystography showed a urine leak at the anastomosis (Figure 11.2). A Foley was reinserted and she improved over the next 48 hours. The Foley was removed as an outpatient 10 days later after ultrasound revealed no increase in size of the urinoma and cystogram showed no leak. Four weeks later, the stent was removed cystoscopically.

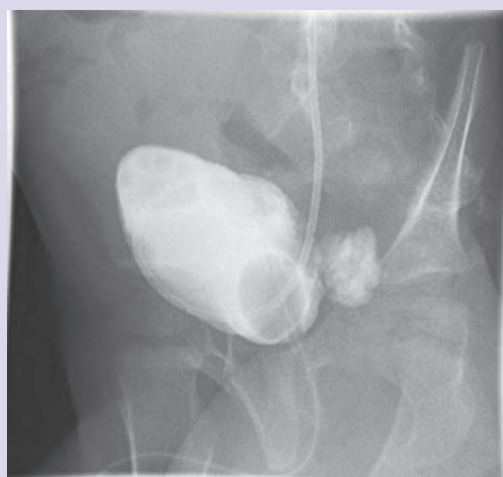


Figure 11.1 Renal ultrasound demonstrating left upper-pole dilation with a dilated proximal ureter.

Follow-up ultrasound showed resolution of the urinoma and dilated ureter, despite persistent upper-pole dilation (Figure 11.3). She remains free of urinary tract infections postoperatively.

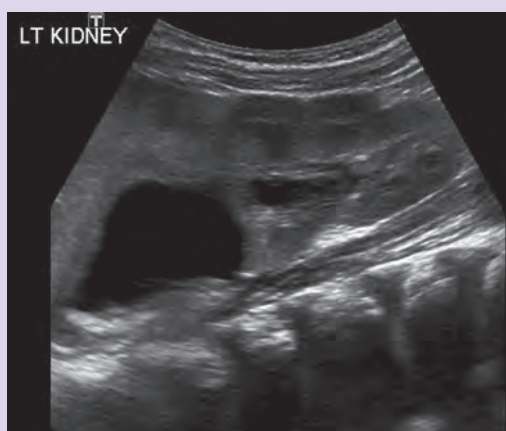


(a)

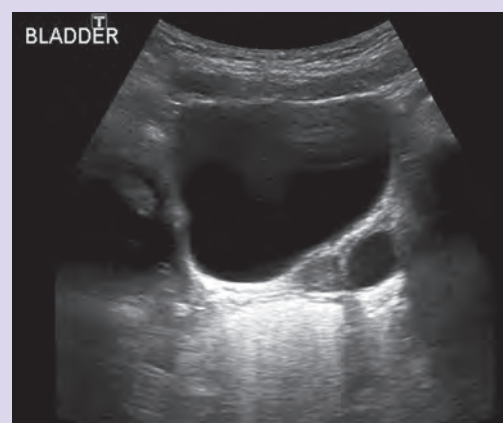


(b)

Figure 11.2 (a) Ultrasound showing a peri-vesical urinoma, necessitating ureteral stenting. (b) Cystogram demonstrating anastomotic leak after ureteral stent placement.



(a)



(b)

Figure 11.3 (a) Ultrasound demonstrating improvement in the proximal ureterectasis, with persistent upper-pole hydronephrosis. (b) Ultrasound showing distal ureterectasis, but resolution of the urinoma.

Introduction

Ureteral duplication anomalies with complete ureteral duplications can present in many different ways. Vesicoureteral reflux (VUR) often associated with the lower-pole can be seen in conjunction with an upper-pole ureterocele and/or ectopic upper-pole ureter. There are a number of ways to surgically approach this, including incision/excision of the ureterocele, upper-/lower-pole

heminephrectomy, pyelopyelostomy, or common sheath reimplantation. One potentially underutilized technique is the ipsilateral ureteroureterostomy (U-U). Evidence in the literature suggests that U-U can be used with high success with minimal morbidity and complication.

Another use of U-U is transureteroureterostomy (TUU) for salvage procedures as well as diversion/undiversion.

This chapter addresses these surgical techniques and attempts to troubleshoot complications and pro-

vide steps for management. Although focused on open techniques, these procedures have been reported both laparoscopically, and robotic assisted.

Surgical techniques

Ipsilateral ureteroureterostomy

The initial use of U-U was first described by Buchtel in 1965 [1]. The patient is placed supine on the operating table. Most cases can be performed through an inguinal, modified Gibson or Pfannenstiel incision. If cystoscopy is necessary for stent placement into the recipient ureter, this can be done in the dorsal lithotomy position with or without fluoroscopy. The ureteral complex is located as it passes below the obliterated umbilical artery and the ureters are separated above the common distal blood supply. The donor ureter is then transected and ligated distally if necessary. The recipient ureterotomy is then performed longitudinally to match the diameter of the donor ureter. The ureteral anastomosis is performed end-to-side with 7:0 polydioxane (PDS) absorbable suture in an interrupted or running fashion. An indwelling stent can be placed prior to the anastomosis. Ureterocelex excision and recipient ureter reimplantation can be performed through this same incision. After the surgery is finished, the incision is closed and a Penrose drain can be brought out through the incision if desired.

Transureteroureterostomy

The technique for TUU involves greater exposure because of the need to mobilize the donor ureter to the recipient ureter for the anastomosis. This usually involves a larger, midline transperitoneal incision. The posterior peritoneum is incised to expose the retroperitoneum and access to the ureters. The bowel is mobilized cephalad to provide maximum exposure. The donor ureter is then mobilized and ligated as distally as possible, taking care to preserve the adventitia with the blood supply. The gonadal vessels often have to be ligated for mobilization. The donor ureter is then brought across the midline to the recipient ureter above or below the inferior mesenteric artery – this decision is based intraoperatively, depending on which will bring the donor ureter to the recipient ureter without tension for a good anastomosis. An end-to-side anastomosis is performed similar to the ipsilateral U-U. An indwelling stent is placed prior to completion of the anastomosis. External drainage with Penrose or Jackson-Pratt drains can be placed to monitor for any urine leak, although we often elect to forgo placement of any drain.

Outcomes from operations

Outcomes from ipsilateral U-U have historically been very good. The largest series by Lashley et al. [2] of 100 ureteroureterostomies had an average patient age of 28 months with a hospital stay of 4.6 days. The anastomoses patency was 94% and they found that the most common complication was prolonged Penrose drain output. Of 13 patients with prolonged drain output, one required percutaneous drainage of the kidney and one required percutaneous drainage of a urinoma. Other complications included fever of unknown origin (2 patients) and blood transfusions (2). In addition, one patient each had ileus, retained drain, gastroenteritis, febrile urinary tract infection (UTI), and pneumonia. Six patients were considered failures. Three patients had U-Us requiring revision secondary to obstruction. Two patients had VUR that subsequently underwent ureteral reimplantations, and one patient had a non-draining ureteral stump that required excision. None of the complications were seen in these six patients with a failed procedure.

Another series from Chacko et al. [3] of 41 U-Us had an average age of 31 months and hospital stay average of 1 day. The patency rate was 100%. Complications seen were *de novo* VUR in two patients who had U-U alone. One patient underwent ureteral reimplantation, and the other underwent subureteric injection. Two patients with U-U and concomitant common sheath reimplantation had persistent VUR that was treated with observation in one patient and subureteric injection in the other. In children having concomitant U-U and reimplantation, two patients without indwelling stents developed transient postoperative urinomas that required subsequent drainage. Another patient presented with transient ipsilateral urinary obstruction that required percutaneous drainage which resolved spontaneously.

Bieri et al. [4] reported on 24 U-Us with an average age of 4 years and a hospital stay of 3 days. Complications noted were UTI in one patient and U-U revision for prolonged drain output. Long-term patency rates were 100%.

Jelloul and Valayer [5] performed 19 U-Us on patients with a mean age of 3.5 years that averaged 6 hospital day stays. They did not note any complications at an average follow-up of 38 months.

U-U can be used as a primary and a salvage procedure. Choi and Oh [6] reviewed their management of ureteral duplication pathology. Eighteen U-Us were performed of which 13 were primary U-Us. Five U-Us were performed as salvage procedures for prior failed

approaches in which all succeeded. They noted that U-U had the lowest failure rate of the different techniques they used. Overall, they observed an 89% success rate for U-U. Two patients with primary U-Us required intervention. One patient underwent remnant ureterocele excision, while another had a ureterocele excision with ureteral reimplantation for VUR with infection.

Bochrath et al. [7] reported on 13 U-Us on patients ranging from 2 to 14 years of age. The hospital stay ranged from 2 to 10 days with a median of 3 days. Complications noted were VUR into three ureteral stumps, and persistent VUR in one patient. None required further intervention.

A more recent series by Lee et al. [8] reported on 74 proximally performed U-Us through a flank incision in an effort to prevent yo-yo reflux. At an average follow-up of over 5 years, they did not report patency rates, but did note a re-operation rate of 14.9%. Nine (12.2%) were due to UTI and VUR of the distal stumps. This is likely due to the proximal anastomoses and flank incisions, making distal excision less complete. When compared to

those who did not require distal excision, the preoperative maximal upper-pole ureteral diameter was the only variable that carried statistical significance as a risk factor. The mean diameter of the ureters that had postoperative symptomatic VUR and UTI was 17mm, vs. 9mm in the group without complications, with a P -value <0.001 .

Prieto et al. [9] reported similar results on 23 patients operated through inguinal incisions. One patient had the recipient ureter inadvertently transected, requiring double barrel cutaneous ureterostomy, followed by re-implantation 6 months later. Three additional planned U-Us were aborted because of a small recipient ureter or purulent drainage from the upper pole. All were successfully anastomosed as U-Us 6 months later. Two patients had UTIs postoperatively, and one developed VUR requiring subureteric injection.

Outcomes for ipsilateral U-Us are displayed in Table 11.1.

Outcomes for TUUs in pediatric patients have been reported by a number of groups. An early study

Table 11.1 Outcomes of ipsilateral ureteroureterostomies (U-Us).

Study	Number	Age	Hospital stay	Follow-up	Success rate (U-U patency) (%)	Complications
Chacko et al. [3]	41	31 months	1 day	3–34 months (average 12 months)	100	VUR (4); urinoma (2); transient obstruction (1)
Lashley et al. [2]	100	28 months	4.6 days	2.5–24 months	94	Prolonged Penrose output (13) – one required PNT, one required percutaneous drain; U-U obstruction (3); non-draining stump with UTI (1); fever (2); ileus (2); VUR (2); retained drain (1); gastro-enteritis (1); pneumonia (1); no yo-yo reflux
Choi et al. [6]	18	0–12 years	–	7.6 years	89	VUR (1) required reimplant; excision ureterocele for UTI (1)
Bieri et al. [4]	24	4 years	3 days	41.4 months	100	UTI (1); U-U revision (1); no yo-yo reflux
Jelloul et al. [5]	19	3.5 years	6 days	38 months	100	UTI (1); VUR (2); no yo-yo reflux
Bochrath et al. [7]	13	2–14 years	3 days	55 months	100	VUR stump (3); VUR (1); no yo-yo reflux
Lee et al. [8]	74	75 days	–	60.5 months	100	Hemorrhage requiring laparotomy (1); prolonged Penrose output (2) requiring anastomosis revision in one; VUR stump with UTI requiring ureterectomy/ureterocele excision (9)
Prieto et al. [9]	23	10 months	0.6 days	26 months	100	UTI (2); unplanned 2-stage repair after cutaneous ureterostomy (2); VUR requiring subureteric injection (1); inadvertent recipient ureteral transection (1)

PNT, percutaneous nephrostomy tube; UTI, urinary tract infection; VUR, vesicoureteral reflux.

reviewing TUU in children was performed by Halpern et al. [10]. They performed TUU in 38 children – 14 TUUs alone and 24 TUUs with cutaneous ureterostomy. The majority of patients had underlying bladder pathology. Complications included avulsion of the TUU resulting in eventual death and two patients with ureteral necrosis resulting in ileal conduit diversions.

Hodges et al. [11] reviewed their 25-year experience with 100 patients. The age ranges were 1–83 years. Two cases resulted in postoperative death from other sources of pathology. Complications included prolonged urinary drainage (2), inferior mesenteric syndrome (2), anastomotic disruption (1), VUR (1) and acute pyelonephritis (5).

Hendren and Hensle [12] described experience with 75 cases of TUUs. The main indications were for failed ureteral reimplantations or urinary undiversion. They noted no deaths, anastomotic leaks, or nephrectomies. Three patients developed reoperative complications. One patient required a revision for an anastomosis that was too anterior on the recipient ureter, causing excessive angulation. Another patient had a kink in the recipient ureter distal to the anastomosis that required resection. The last patient needed revision of a bowel segment used to drain a donor renal pelvis into a recipient ureter, which was subsequently moved to drain into the opposite recipient renal pelvis.

Rushton et al. [13] used TUU for 31 patients for urinary diversions/undiversions and failed ureteral reimplantations. They noted neurogenic bladders in 26

patients and four others with bladder pathology. Complications included two stent placements for transient obstruction and ureterocutaneous drainage. Another patient developed obstruction after tapered reimplantation requiring cutaneous ureterostomy. Lastly, one patient developed a large urinoma due to ischemic necrosis of the upper ureter that resulted in nephrectomy. Two complications including partial small bowel obstruction and vesicocutaneous fistula, both resolved without intervention. There were no complications with any of the TUU anastomoses.

A review of 69 TUUs by Mure et al. [14] for multiple indications, for salvage and reconstruction, for diversion/undiversion noted a low complication rate. Three patients required reoperation – one for postoperative urinoma and another for common ureteral trunk ischemia that required separate ureteral reimplantation into the existing sigmoid conduit. The last patient required nephrectomy for deterioration and infection. No complications with the TUU anastomoses were noted.

Lastly, Pesce et al. [15] had 70 patients requiring TUUs with the majority (97%) for salvage procedures for failed ureteral reimplantations. Complications included one temporary obstruction requiring stent placement and one distal obstruction of the recipient ureteral reimplantation, treated with balloon dilation. Four patients with neurogenic bladders developed complications unrelated to the TUUs. There were no complications related to the TUU anastomoses. Results are shown in Table 11.2.

Table 11.2 Outcomes of transureteroureterostomies.

Study	Number	Age	Salvage/diversion-undiversion	Follow-up	Success rate (U-U patency) (%)	Complications
Pesce et al. [15]	70	2–13 years	68/2	4–21 years (average 10.8 years)	100	Temporary obstruction requiring PNT (1); distal obstruction requiring dilation (1); 4 patients with neurogenic bladder: progression of pre-existing renal disease (2); renal stones donor pelvis (1); distal ureteral stenosis requiring reimplant (1)
Mure et al. [12]	69	1 month to 21 years (mean 8.6 years)	22/47	Median 6 years	100	Urinoma (1); ureteral trunk ischemia (1); nephrectomy donor kidney (1)

(continued)

Table 11.2 (Continued)

Study	Number	Age	Salvage/diversion-undiversion	Follow-up	Success rate (U-U patency) (%)	Complications
Rushton et al. [13]	31 to 17 years	5 weeks	8/23	>1 year	100	Transient obstruction requiring stent (1); obstruction tapered reimplant (1); ureterocutaneous fistula requiring stent (1); nephrectomy (1); partial small bowel obstruction (1); vesicocutaneous fistula (1); 30 patients with neurogenic bladder or bladder pathology
Hendren and Hensle [12]	75	Newborn to 36 years	35/40	–	98.6	Angulation at U-U (1); common stem obstruction (1); dilated bowel to ureter problem (1)
Hodges et al. [11]	100	1–83 years	22/78	>1 year	99	Prolonged urinary drainage (2); inferior mesenteric syndrome (2); U-U disruption (1); VUR (1); pyelonephritis (5)
Halpern et al. [10]	38	4 months to 10 years	7/31	–	97	U-U avulsion resulting in death (1); ureteral necrosis requiring ileal conduit (2)

PNT, percutaneous nephrostomy tube; U-U, ureteroureterostomy; VUR, vesicoureteral reflux.

Complications

Reported complications with ipsilateral U-U are depicted in Figure 11.4. The most common problems were prolonged drain output and VUR, whether *de novo* or persistent vs. VUR into the distal remnant stump. Few of the patients that had prolonged drain output required intervention and most were managed conservatively until it resolved.

VUR after surgery invariably is lower grade and often can be managed with conservative measures including subureteric injection and observation. Kaplan et al. [16] reported on conservative management of patients with complete duplication and VUR and, of the observed group, 48% had resolution of VUR. In addition, the high success rates of subureteric injection with lower-grade VUR can and has been applied to VUR seen after U-U. Lastly, VUR into the remaining stump after U-U can be minimized by dissecting the stump to the level of the bladder, which can be easily accomplished with the use of a low incision.

Obstruction of the U-U anastomosis was uncommon and, in cases where revision was necessary, it was easily

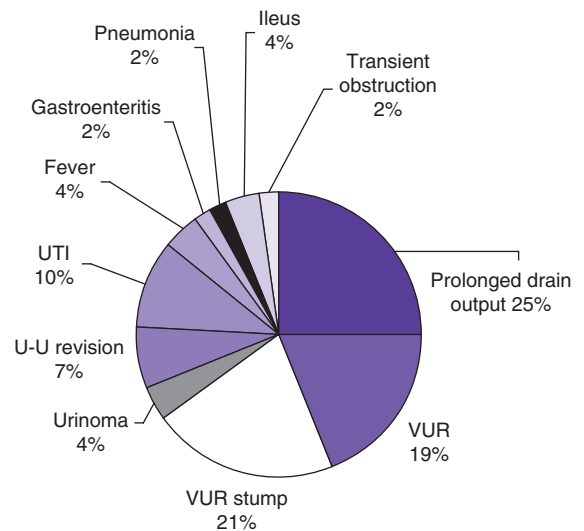


Figure 11.4 Complications of ipsilateral ureteroureterostomies.

accomplished with 100% success after the revision. Urinoma formation was relatively uncommon and the chance for leakage from the anastomosis can be minimized with drainage. All studies advocated using some form of urinary drainage, whether it was urinary stent, drain, or both.

One concern many have with U-U is potentially leaving behind a dysplastic upper pole. Smith et al. [17] compared U-U vs. upper-pole nephrectomy. They found that 9.5% of patients on pathology of the upper pole had evidence of marked dysplasia and half of the patients had no dysplasia or insignificant evidence of pyelonephritis. The decision for U-U was based on the appearance of the upper pole intraoperatively, and if it appeared healthy and U-U was technically feasible, it was performed. Husmann [18] reviewed consequences of leaving tissue behind in multicystic dysplastic kidneys and duplicated dysplastic segments and found them very rarely associated with urinary infection, hypertension, and renal tumors.

Another concern is ureteral disparity during U-U anastomosis and the potential for yo-yo reflux between the two ureters. This appears to be an academic concern and no investigators have reported any problems with ureteral disparity from donor to recipient ureter and no instances of yo-yo reflux have occurred.

Ureteral anastomosis complications with TUU are uncommon and all studies have shown excellent success. Since the TUU is mainly used for salvage and more complex reconstructive and creative surgeries, the chance for complications invariably increases. Many cases are reoperative procedures from the beginning and often the patients have other comorbidities that can increase the risk for complications. The most important point is to anticipate problems and have a higher suspicion for complications postoperatively.

Preventing complications

Preoperative

Preoperative evaluation is crucial because of the variety of anomalies seen in complete ureteral duplication. Information that is necessary consists of presence or absence of ureterocele, location of the insertion of the upper-pole moiety and whether it is ectopic, obstructed, or refluxing. In addition, lower-pole VUR and contralateral VUR need to be evaluated. Also, the character of the upper-pole parenchyma is important. Most of these answers can be elucidated by a renal/bladder ultrasound (RBUS) and voiding cystourethrogram (VCUG).

Anatomical definition not seen on VCUG can be evaluated by intravenous pyelogram (IVP). Further evaluation of upper-pole function can be performed using nuclear medicine imaging. An upper-pole function of >10% should be considered for U-U. Often, the less invasive RBUS can give an idea of the quality and amount of upper-pole parenchyma.

Intraoperative

Prior to making the incision in the lower abdomen, cystoscopy with retrograde ureteropyelography can be performed if preoperative imaging does not provide an accurate map of the duplicated anatomy. It may be helpful to identify ectopic ureteral orifices. A ureteral stent or open ended catheter may be placed at this time as well. If cystoscopy is not warranted, ureteral stenting can be performed at the time of the U-U anastomosis.

At the time of identifying the ureteral complex, care must be taken when exposing the ureters to preserve the blood supply, particularly to the recipient ureter. Minimal tissue handling is important to prevent postoperative urine leak and stricture, as is placement of a stent/drain. Ureteral luminal disparity from donor to recipient ureter has not been a problem, but it is crucial to make an appropriate length incision on the recipient ureter to match the dilated donor ureteral diameter. Using a lower abdominal incision allows for concurrent procedures to be performed including ureterocele excision or low resection of donor ureteral stump, in addition to U-U with reimplantation if VUR exists in the lower-pole ureter. If VUR exists on the contralateral side, reimplantations are best done intravesically to avoid potential bladder neuropraxia from bilateral extravesical manipulation.

In patients with TUUs, the same principles of tissue handling and urinary drainage apply. Other key points are to create a tension-free anastomosis, as well as making sure the donor ureter crosses the abdomen without interference from the inferior mesenteric artery.

Postoperative

Patients postoperatively generally do well. It is important to monitor drain output and remove it when it is dry to prevent urinoma formation. Although not usually necessary, if a patient presents with UTI, pain or fever, they warrant urinalysis and culture and possible imaging with CT scan or RBUS.

Normal routine follow-up should include RBUS at six weeks postoperatively. In addition, patients with other

renal and bladder comorbidities should have routine creatinine and blood pressure monitoring.

Managing complications

Initial

Management of initial complications consists of evaluation of symptoms. Early signs of problems can manifest as fever, malaise, or urinary symptoms. All patients with a documented, febrile UTI need evaluation for VUR with a VCUG. Evaluation for urinoma or obstruction is best carried out by CT scan with contrast to assess size and location of urine leak as well as hydronephrosis.

Definitive

Problems at the U-U can manifest in two forms: obstruction or leak. Obstruction at the U-U site will manifest as dilation in the donor renal segment. Management consists of percutaneous nephrostomy drainage and empiric antibiotics until infection is proven. After the acute period resolves, antegrade nephrostogram should be performed to assess patency of the anastomosis. If the U-U is not open and draining, surgical intervention and revision may be necessary.

Urinary leak at the U-U site can occur and the risk increases if a stent and/or drain are not left in place postoperatively. If urinoma does occur, percutaneous drainage or open drainage is necessary. After resolution of the urine leak, most studies show no problems with patency with the U-U anastomosis. Of patients that did

require anastomotic revision, all of them resulted in patency postoperatively.

VUR after surgery was a common complication. The most definitive treatment historically performed was ureteral reimplantation. However, other management strategies that have been successful, specifically observation or subureteric injection, offer less morbidity. Yo-yo reflux in all studies has not been an observed phenomenon and is more of an academic concern.

Conclusion

Ipsilateral U-U is a safe and effective technique for managing ureteral duplication anomalies. Postoperative complications can be minimized based on patient selection and recognizing pitfalls early and treating them appropriately as they arise. For the right patient, it offers good cosmesis, short recovery, and excellent success rates.

TUU is also a safe and effective procedure for both salvage procedures after failed ureteral reimplantations and urinary diversion/undiversion procedures in more complex patients. Although postoperative problems are uncommon, when they occur, they can be challenging to manage. However, this should not deter the pediatric urologist from using TUU as one of many methods for managing these difficult cases. Finally, laparoscopic and robotic approaches to these procedures have been reported with good initial success rates. However, larger series with longer follow-up are required before they can be universally recommended.

DOS AND DON'TS

Do

- Place a stent or ureteral catheter cystoscopically to identify recipient ureter
- Consider leaving a drain and/or stent, which may reduce the risk of urinoma
- Create a wide open anastomosis with watertight closure
- Manage urine leaks conservatively at first by assuring drainage if clinically stable
- Manage symptomatic urinomas, if they manifest with fevers/illness, with percutaneous drainage
- Consider ureteroureterostomy in cases of duplex renal units with pathologies of either or both poles
- Consider transureteroureterostomy in cases where the distal 1/3 of the donor ureter is diseased or deficient

Don't

- Dissect too closely to the ureter, disrupting the blood supply
- Create anastomoses under tension
- Worry about the concept of yo-yo reflux
- Worry about discrepancy between large donor anastomosis to normal caliber recipient ureter (no different than a pyeloplasty)

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PART IV

Surgery of the Bladder

CHAPTER 12

Complications Epispadias and bladder exstrophy

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KEY POINTS

- The most important prognostic factor is the success of the initial closure. An experienced surgeon, ideally with experience in the management of exstrophy, should perform initial closure
- Short-term complications include bladder dehiscence, fascial dehiscence urethrocuteaneous fistula formation, cast complications, and penile ischemia
- Intermediate complications include urinary obstruction and vesicoureteral reflux
- Late complications include renal failure, incontinence, malignancy, reproductive challenges, and psychological burdens
- These patients need life-long management and follow-up

CLINICAL PROBLEM/CASE

You are consulting on a 1-month-old infant status-post complete primary repair of bladder exstrophy at 1 day of life who presents with leakage of urine at the peno-pubic site.

1 When do you elect to repair the fistula?

2 What type of evaluation is warranted for this problem?

3 The fistula is successfully closed; however, 5 years later, the child is still in diapers. What are the management options?

4 A bladder neck reconstruction is indicated; however, the child is still incontinent 1 month later. What is the next step?

Introduction

The epispadias–exstrophy complex (EEC) consists of epispadias, classic bladder exstrophy, cloacal exstrophy, and exstrophy variants. The complex spans a spectrum of genitourinary malformations characterized by deformation of the lower urinary tract, with varying involvement of the lower abdominal wall, pelvis and hindgut. Interestingly, classic bladder exstrophy and epispadias often occur in isolation, while cloacal exstrophy or exstrophy variants are usually associated with anomalies of intestines, neurological system, upper urinary tract,

and skeletal system. The complications associated with this condition include those of the untreated state as well as those that occur as a consequence of surgical intervention [1]. In this chapter, we discuss complications associated with classic bladder exstrophy and epispadias.

No treatment

Although bladder exstrophy is not a lethal anomaly in infancy, these patients are often social pariahs because of associated odor and hygiene problems. In addition, 66–75% of affected patients die by age 20 due to

pyelonephritis and renal failure [2]. The untreated exstrophic patient has a 17.5% risk of bladder neoplasia after 20 years of age. These malignancies tend to carry a high mortality rate, even after cystectomy [3]. As such, continued observation is not an acceptable management option, and all children should be managed by closure or urinary diversion.

Surgical intervention

The choice of surgical management is a function of surgeon preference, patient anatomy, and access to tertiary care facilities. The goals of all surgical intervention include renal preservation, urinary continence (for primary closure and continent diversion), and optimization of phallus length.

Anatomical reconstruction of the epispadias–exstrophy complex

Prior to the 1970s, early attempts at bladder exstrophy closure were limited by high morbidity and poor long-term continence and renal function rates; many surgeons favored the use of primary urinary diversion [4,5]. However, the advent of the staged repair demonstrated that anatomic reconstruction of the exstrophic bladder was feasible and safe [6]. This approach led to significant improvements in continence, renal function, and cosmesis [7,8]. Contemporary repairs, including the modern staged repair bladder exstrophy (MSRE) [9] and complete primary repair of bladder exstrophy (CPRE) [10–12] are the repairs of choice at most centers at this time. While there is no consensus on the exact timing of closure, data suggest that neonatal closure may be associated with higher continence rates than repairs delayed greater than 6 months [13,14]. Continence rates vary in the literature, with some patients requiring clean intermittent catheterization (CIC) and bladder augmentation or diversion to achieve dryness [15,16].

Epispadias repair, whether performed for primary epispadias or as part of a staged exstrophy closure, is traditionally associated with a high rate of fistula and/or stricture disease [17]. Modern closure methods (Cantwell-Ransley and Mitchell penile disassembly techniques) use dissection of the corporal bodies and transposition of the tabularized plate to the ventral aspect of the penis, resulting in improved cosmesis and

a decreased rate of complications overall [18,19]. The optimal timing of closure is controversial; however, studies suggest improved bladder growth when repair is done at less than one year of age [20].

The different surgical techniques (CPRE, MSRE, etc.) are designed to optimize long-term functional and cosmetic outcomes. Regardless of approach, surgical outcomes are dependent on strict adherence to Halstedian surgical principles, quality of the tissue, surgeon experience, and proper postoperative management. The degree of exstrophy also dictates outcome: whilst isolated epispadias can be successfully closed by a single operation in 80–91% [17,18], classic bladder exstrophy requires a median of five operations per patient [21].

Early complications

Fascial dehiscence

Fascial dehiscence affects up to 3% of patients and typically presents within the first week of closure. Identification of a fascial dehiscence requires immediate surgical repair if it is not associated with a fascial infection. In the setting of infection, this must be treated before addressing the dehiscence. Fascial dehiscence may not affect the integrity of the bladder or the urethra [12].

Catheter malfunction

Catheter patency should be confirmed routinely during the initial closure. Loss of the suprapubic catheter within two weeks of closure increases the risk of bladder dehiscence. We recommend immediate replacement with a urethral catheter when possible or, if necessary, open suprapubic tube insertion. Similarly, early ureteral catheter malfunction may also precipitate leakage or azotemia necessitating immediate repositioning with fluoroscopy [22] or open surgery [11]. We recommend that all catheters be secured and checked diligently for patency and positioning with each encounter in the immediate postoperative period.

Bladder dehiscence

Bladder dehiscence is rare, with an incidence of only 3%. In addition to errors in surgical technique, associated risk factors include postoperative abdominal distension, bladder prolapse, lack of osteotomy, and loss of ureteral stents prior to postoperative day 7 [23]. Distinction should be made between bladder and

fascial dehiscence, as the latter necessitates immediate exploration and repair. Isolated bladder dehiscence requires a recovery period of at least 6 months prior to reclosure. Tension-free reclosure with osteotomy, preoperative testosterone (for boys), and combined epispadias-bladder repair are important factors in subsequent closures [12,24].

The importance of successful primary closure cannot be overemphasized. The risk of subsequent failure after reclosure is as high as 40%. Following two closures, the likelihood of eventual continence is as low as 30%, compared to 70–80% after successful initial repair [25]. This number drops to less than 20% after more than two closure attempts [26,27]. Furthermore, in patients with failed initial repair, the potential to achieve bladder capacity large enough for bladder neck reconstruction is as low as 68%; after two failed closures, only 40% obtain a capacity for bladder neck reconstruction [26,28].

Urethrocutaneous fistula formation

Penopubic fistula occurs in 14–35% of newborn exstrophy closures, making it the most common surgical complication of CPRE [12,21]. This rate increases to 26–52% in delayed or redo cases [13,14]. Overlapping suture lines at the penopubic junction are almost impossible to avoid during CPRE; however, small intestinal submucosa onlay has been reported to help decrease fistula formation [29]. After staged exstrophy repair, urethrocutaneous fistula develops in 21% and 25% of patients with intact plates and paraexstrophy flaps, respectively [30].

Spontaneous fistula closure rate has been reported in 25–100% of patients within 7.5 months. Persistent fistulae should be evaluated with urethroscopy to determine the extent of fistula and rule out the presence of distal urethral stricture. Surgical repair should be delayed at least 6 months following initial closure. The effect of bladder neck fistula on eventual continence is uncertain [14,29].

Orthopedic complications

Within the first 48–72 hours, the pubis remains elastic and can often be reapproximated without significant tension during an exstrophy repair. If there is any doubt regarding the pliability of the pubic bones, osteotomy is indicated. Evidence suggests that osteotomy decreases the risk of developing prolapse or wound dehiscence

[31]. Osteotomy can result in femoral nerve palsy, delayed union of osteotomy, persistent sacroiliac pain, leg length inequality, and/or osteomyelitis in up to 6% of patients [31–34]. Authors suggest that the majority of complications are due to osteotomies performed using a posterior approach; major complications are not as common with combined anterior innominate/transverse iliac or oblique approaches [34].

Additionally, both traction and spica casting can cause pressure sores and compartment syndrome if the lower extremities are wrapped too tightly [25]. Generally, though, spica casts are well-tolerated and are associated with a low risk of complications [35]. We recommend that urologists be present for all spica casting to ensure adequate room for growth, ample padding of all potential pressure points, and appropriate abduction of the hips to reduce tension on the closure.

Male genital complications

Atrophy of the corporal bodies, glans, and/or urethra has been described for both the staged reconstruction [36,37] as well as CPRE [38,39]. The glans, urethra, and corporal bodies are at risk for devascularization if the terminal arteries to the penis or bulbar artery are damaged during dissection of the corporal bodies from the pubic symphysis. The glans should be assessed after approximation of the rami since the rotational tension created by the reapproximated symphysis on the corporal bodies can cause venous congestion, compromising blood supply to the penis. Color change in the glans or corpora should be managed with papaverine, followed by replacement of the pubic sutures higher on the pubis [25]. In a review of 24 cases of penile ischemia, Cervellione and colleagues noted that 83% of patients did not undergo osteotomy during initial closure. They suggested that osteotomies may have a protective effect and should be done if there is any question of decreased penile perfusion [40,41].

If ischemia of the glans and penile skin is recognized within 24–48 hours after closure, more than half of the cases resolve with observation alone. Vasodilators may also be helpful. Otherwise, acute ischemia should be treated with reoperation with release of the pubic sutures and osteotomy (if not already performed); however, this is not helpful in setting of prolonged ischemia [39]. If intervention is not successful, urethral plate and penile skin loss can be replaced with regional and distant grafts and flaps at later dates [24]. In the

catastrophic setting of penile amputation secondary to ischemia, salvage phalloplasty has been described [42].

Intermediate outcomes

Bladder outlet obstruction

Patients are at risk for obstruction secondary to neourethral stricture or a narrow bladder neck after the initial repair. Patients with para-exstrophy skin flaps have a 10–67% risk of stricture development at the proximal anastomosis [43]. Whilst the use of para-exstrophy skin flaps has largely fallen out of favor over the past decade, skin flaps may occasionally still be necessary to position the bladder into the deep pelvis if there is deficiency of urethral tissue. The use of urethral substitution flap/graft is also a risk factor for stricture disease. Given that 50–77% of urethral plates in exstrophy patients are inherently short, we recommend converting these plates into hypospadias urethras rather than risk complication with a flap/graft interposition or elongating the urethra as described by Caione and colleagues [12]. Erosion of the intrapubic stitch into the posterior urethra can also result in outlet obstruction. In this situation, authors have suggested treating early obstruction (<4 months postoperative) with a suprapubic tube; after 4 months, the stitch should be removed through a small suprapubic incision. Patients undergoing delayed or redo closure are also at higher risk for stricture disease with rates as high as 10.5% reported [13,14].

Obstruction may have an insidious onset; patients should be carefully followed with surveillance imaging for at least 4–6 months postoperatively, as 7.5% of patients may develop renal deterioration after closure [12,16,25]. Any suggestion of obstruction should be immediately assessed with cystoscopy [44]. Dilation of the bladder neck may be helpful in the setting of outlet obstruction. Institution of clean intermittent catheterization should also be considered.

Vesicoureteral reflux

Almost all patients experience vesicoureteral reflux following bladder closure. Spontaneous resolution can be seen in up to 16% [12,21]. While asymptomatic bacteriuria can be seen in up to 70% of patients, the rate of febrile urinary tract infection may be as high as 22% [21]. Given the low rate of spontaneous resolution, patients with recurrent febrile episodes should be treated

early with either ureteral reimplantation or injection of a subureteral bulking agent. If open surgery is elected, a cephalo-trigonal technique is preferred over a cross-trigonal repair, as the latter may complicate future bladder neck reconstruction [45]. The clinical scenario of recurrent febrile urinary tract infections should also raise suspicion for outlet obstruction.

Late outcomes

Renal deterioration

Renal damage, defined as renal scarring and/or moderate-to-severe hydronephrosis, can be seen in 7.5–25% of children following bladder closure [10,12,16,46]. Serum creatinine remains normal in 97% of children, with 1.5% developing mild renal insufficiency, and 1.5% eventually requiring kidney transplantation [1,12]. Up to 23% of children may demonstrate transient hydroureteronephrosis following successful closure; this is less frequently due to obstruction than as a result of gradual accommodation as the bladder begins to cycle urine in the presence of vesicoureteral reflux. Half of children with hydroureteronephrosis may require CIC even without evidence of anatomic bladder outlet obstruction [12,46]. While spontaneous improvement is seen in some children, persistent or worsening hydroureteronephrosis may be indicative of a poorly compliant bladder necessitating treatment with anticholinergic, endoscopic botulinum toxin A injection, and/or augmentation cystoplasty.

Incontinence

Most articles define continence in bladder exstrophy patients as a 3-hour dry interval. While some patients undergoing primary bladder closure are continent by the age of toilet training, most patients experience some degree of daytime and/or nighttime incontinence [47]. Incontinent children typically demonstrate low leak point pressure, wide bladder neck, with or without impaired compliance and capacity. While medications like anticholinergics, desmopressin, or imipramine can be helpful [48,49], 60–87% of these patients achieve continence from procedures that increase bladder outlet resistance. Endoscopic injection of Deflux into the bladder neck has been shown to improve continence in up to 50% of patients but is considered to lack durability [50,51]. Formal bladder neck reconstruction and

Mitrofanoff urinary diversion, with or without augmentation cystoplasty, is indicated in cases of persistent incontinence [12]. Following bladder neck reconstruction, up to 20% of patients experience *de novo* outlet obstruction requiring temporary urethral catheterization [25]. Following resolution of edema, continence is often delayed: mean time to daytime continence is 14 months; nighttime continence can take a mean of 22 months [8,25,52,53]. These patients need long-term follow-up, as up to 60% will require future augmentation cystoplasty and/or diversion in the second decade of life due to gradually worsening compliance [54].

In patients with persistent incontinence, salvage endoscopic deflux injection can be employed with up to a 45% success rate in achieving continence [50,55]. Salvage revision surgery options for children with persistent or recurrent incontinence include placement of an artificial urinary sphincter, revision bladder neck reconstruction, bladder neck wrap [56] or bladder neck closure with diversion [57]. Augmentation cystoplasty is almost always necessary to achieve continence in this scenario, as initial failure is usually secondary to poor bladder compliance or small bladder capacity [58].

Malignancy

The risk of malignancy in an unrepaired exstrophic bladder is 17.5%. Over 90% of these malignancies are adenocarcinomas and 5% squamous cell carcinomas [59]. Early closure reduces this rate to 3.3%–7.5% at a median age of 42 years [3,59]. The risk of malignancy is increased in those with intestinal substitution or using clean intermittent catheterization [60]. These tumors tend to be aggressive and most are managed with radical cystectomy [61]. Lifelong surveillance with screening cystoscopy is an option, particularly if there is intestinal substitution; however, no accepted screening protocol exists. Current recommendations include annual cystoscopy beginning at 10 years following enterocystoplasty.

Male sexual and reproductive health

While libido, erection, and orgasm are typically intact, males with exstrophy are typically psychosexually delayed by at least 2–4 years compared with their peers [62]. Of men with bladder exstrophy, 50–70% describe engaging in serious and long-term intimate relationships [63–65]. Up to 66% of patients report concerns over penile appearance, sexual inadequacy,

and incontinence as impairing their ability to engage in meaningful sexual relationships [66].

Despite having a history of extensive reconstruction, 63–90% of males are able to ejaculate [67]. Elevated FSH is evident in 19%. Semen analysis demonstrates impaired sperm quality and counts in up to 84% [67,68]. This may be secondary to partial obstruction, retrograde ejaculation, slow seminal emission and/or recurrent infections. Half the men with bladder exstrophy require assisted reproductive techniques to father children [69]; however, it is unclear whether subfertility is due to poor semen parameters, impaired ejaculation, inadequate phallus length or a combination of factors [63–65].

Female sexual and reproductive health

Female genital reconstruction, if necessary, can be performed during adolescent years. Lower midline defects can be repaired with monsplasty, using adjacent and hair-bearing skin to reconstruct a more aesthetically pleasing mons [70]. Some women require vaginal dilation or a cut-back/Y-V vaginoplasty to allow for sexual intercourse [71]. Whilst initiation of sexual activity tends to be delayed until early adulthood, the majority of women report engaging in serious, long-term sexual relationships [64,72].

Of women trying to conceive, pregnancy is reported in only 66%; of these, 56% resulted in live delivery [73]. Gestation is complicated by preterm labor in 40% and malpresentation in 25%, while miscarriage or stillbirth affect over 40% [73,74]. These pregnancies are high risk to the mother as well, with reports of ureteral transection, ureteral obstruction, fistula formation, and hemorrhage [73]. Bedrest is often necessary in the later stages of pregnancy in most patients [75–77]. Additionally, over 50% develop uterine and/or vaginal prolapse requiring sacrocolpopexy [78]. Fixation of the uterus to the anterior abdominal wall can be prophylactic.

Psychological outcomes

Given the chronic cosmetic, sexual, and urological issues affecting these children, patients with bladder exstrophy are susceptible to increased psychosocial pressures/disorders [79]. Numerous studies demonstrate decreased quality-of-life parameters in children and parents when compared with matched controls [80–82]. Quality-of-life scores were associated with

functional outcomes [83]. Suicidal ideation was reported in 14.9% of patients [84]. Girls tended to score higher than boys. In contrast, affected children tend to score high on academic and professional indices [85]. Early psychiatric screening and counseling are recommended to continue throughout the life of these patients [64].

Conclusion

Following bladder closure, patients with bladder exstrophy are susceptible to complications throughout their lives. These patients need lifelong screening for urological, renal, sexual, reproductive, orthopedic, and psychological complications.

DOS AND DON'TS

Do

- Refer these patients to a center of excellence prior to surgical intervention
- Assess the glans following reapproximation of the symphysis. The pubic anastomosis may need to be repositioned or osteotomies performed (if not done) if there is evidence of penile venous congestion
- Assume that all children have vesico-ureteral reflux following closure; place children on antibiotic prophylaxis following initial closure
- Evaluate these children for evidence of renal insufficiency and obstruction as part of their routine evaluation

Don't

- Attempt to close a bladder with exstrophy-epispadias unless you and your medical center have substantial experience with this operation
- Forget to address psychological concerns of these patients, particularly during adolescence

CLINICAL PROBLEM/CASE: DISCUSSION

- 1 Reoperation should be delayed at least 6 months so that edema may resolve. Additionally, up to 25–100% resolve within 7.5 months.
- 2 (a) Cystoscopy is necessary to rule out a distal obstruction, presence of foreign bodies. (b) Renal and bladder ultrasonography to evaluate renal and bladder anatomy. (c) Urine for urinalysis and culture if indicated.
- 3 Assuming urodynamics confirm adequate compliance and acceptable bladder capacity, initial management options include observation until the child and family express interest in the child being continent. Options include bladder neck reconstruction with or without augmentation cystoplasty depending on urodynamic findings.
- 4 Continence following bladder neck reconstruction is often delayed up to 14–22 months. Observation is warranted in this case. Anticholinergic medications may be useful if the child has urge incontinence. It is critical to assess whether the child is completely emptying the bladder; partial urinary retention is a common finding in this setting and may require the initiation of clean intermittent catheterization (CIC) or placement of suprapubic drainage. Children should be psychologically prepared to perform CIC following continence procedures.

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CHAPTER 13

Umbilical and urachal anomalies

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KEY POINTS

- The urachus is an embryonic remnant of the allantois
- The urachal anomaly subtypes are reflective of the location of the incomplete involution of the urachus
- The medial umbilical ligaments are vestigial urachal structures and useful landmarks for identifying the ureters
- Ultrasound and/or sinography are useful diagnostic modalities for the majority of urachal anomalies

Introduction

The urachus, or median umbilical ligament, is a cord-like structure that is continuous with the anterior dome of the bladder inferiorly and extends in an extraperitoneal fashion to the umbilicus superiorly. The urachus is a normal embryonic remnant of the primitive bladder dome and may be affected by disorders related to the arrest of its normal involution. There are four clinical entities relating to the incomplete involution of the urachus during embryogenesis. These urachal anomalies include a patent urachus, urachal cyst, urachal sinus and vesicourachal diverticulum (Figure 13.1).

Prevalence

Urachal anomalies are rare. In a large pediatric autopsy series, the historical incidence of a patent urachus is 1 in 7610 cases and the incidence of a urachal cyst is 1 in 5000 [1]. Another example of the infrequency of urachal anomalies includes a report of 315 cases accumulated over a 40-year period [2]. The most frequent urachal abnormalities that typically present are either a urachal sinus or a urachal cyst [3–5]. There is generally

a 2:1 incidence of urachal anomalies in males compared to females and urachal anomalies usually present in early childhood but may be clinically silent and remain unrecognized until adulthood.

Embryology

The urachus is an embryonic remnant of the allantois [6]. The allantois in embryos of reptiles, birds, and some mammals has a respiratory function and may act as a reservoir for urine during embryonic life. The allantois remains very small in human embryos but is involved with early blood formation and associated with development of the urinary bladder [6]. As the bladder enlarges, the allantois becomes the urachus and is represented in adults as the median umbilical ligament. The blood vessels of the allantois become the umbilical arteries and veins. The obliterated umbilical arteries or medial umbilical ligaments are important landmarks to help locate the underlying ureters when performing surgery on the bladder and ureter (e.g. extravesical ureteroneocystomy).

The allantois arises from the yolk sac and extends to the cloaca – the precursor to the bladder (urogenital

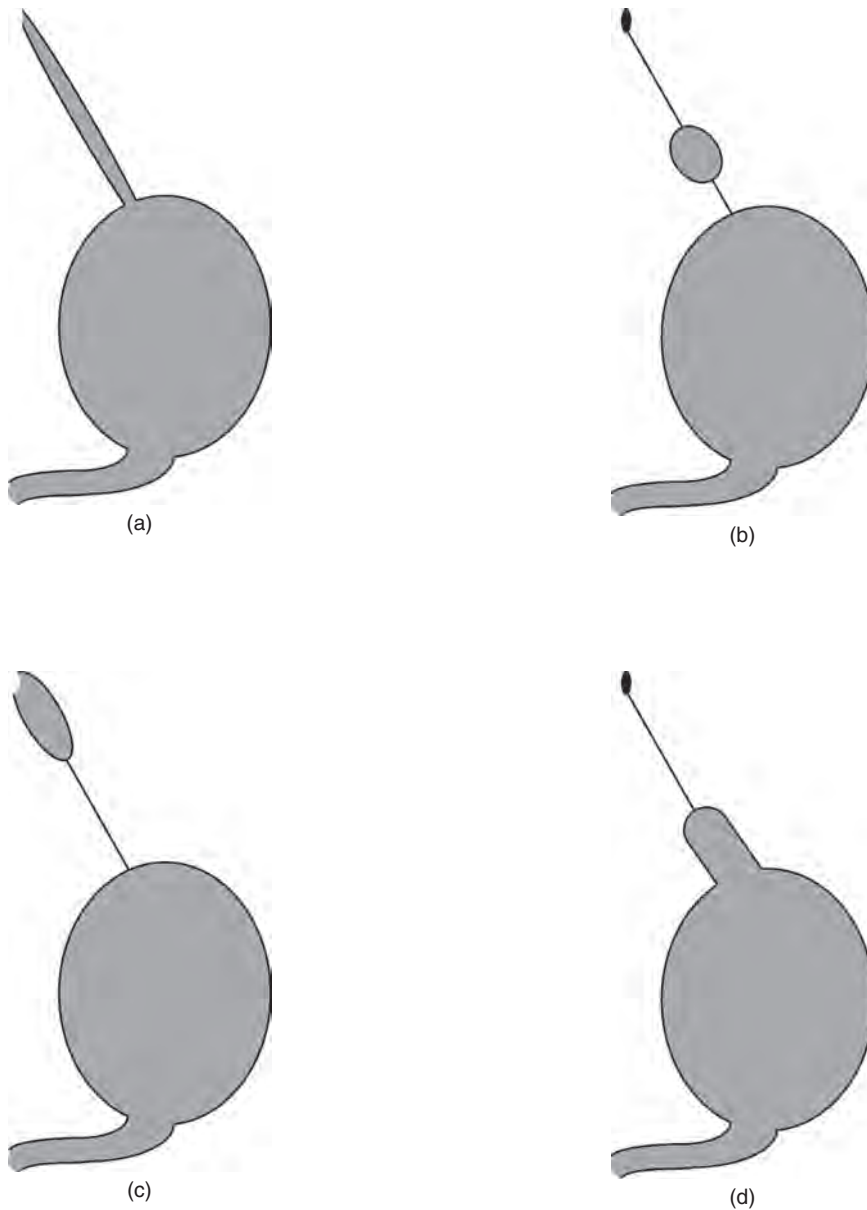


Figure 13.1 Urachal anomalies: (a) patent urachus; (b) urachal cyst; (c) urachal sinus; (d) vesicourachal diverticulum.

sinus) and the rectum (hindgut). With further development and division of the urogenital sinus and hindgut by the urorectal septum, the allantois is initially continuous with the bladder but soon constricts and becomes

a thick, fibrous cord called the urachus. During the fourth and fifth months of development the urachus narrows to a small caliber tube lined by transitional epithelium [7].

Anatomy

The urachus lies within the space between the peritoneum posteriorly and the transversalis fascia anteriorly that is known as the space of Retzius [8]. The urachus is bounded by the umbilicovesical fascia which extends laterally to each umbilical artery. Inferiorly, the fascial layers spread out over the dome of the bladder to the hypogastric artery posteriorly and to the pelvic diaphragm anteriorly. Thus, a potential pyramidal-shaped space is created that is completely self-contained and separate from the peritoneal cavity. These fascial planes act to limit the spread of a urachal infection or neoplasm. Knowledge of this anatomy becomes important in the diagnosis and treatment of urachal diseases [9].

Clinical urachal anomalies

Urachal anomalies result from a failure of fibrosis and involution of the urachus during embryonic development. A variety of clinical urachal anomalies exist and are dependent on where the failure of involution occurs in the urachal tract between the bladder and the umbilicus (Figure 13.1).

Diagnosis

The diagnosis of a urachal anomaly is made from a combination of the presenting history, physical exam and imaging. Periumbilical discharge suggests either a patent urachus or a urachal sinus while a palpable umbilical mass suggests an umbilical cyst. Patients may present with abdominal, suprapubic or periumbilical pain. Periumbilical erythema and tenderness suggest an underlying infection and patients with urachal anomalies may present with dysuria, fever and a urinary tract infection.

A variety of imaging may be used to make the diagnosis of a urachal anomaly. The appearance of a fixed, midline, cystic, extraperitoneal swelling between the umbilicus and the bladder on ultrasonography (US) is suggestive of a urachal anomaly [10]. During a work-up of abdominal or pelvic pain, computed tomography (CT) may allow the diagnosis of a urachal anomaly (Figure 13.1b) [11]. Several studies have advocated that US is the test of choice if a urachal cyst is suspected with a periumbilical mass and sinography is the best modality

to identify a patent urachus or urachal sinus [3,4,12]. In a recent review, the positive predictive value for US is 83%, whereas the negative predictive value is 25%, making the sensitivity for diagnostic US quite good at 79% but the specificity is only 30% [13]. Other imaging modalities may diagnose urachal anomalies including a voiding cystourethrogram (VCUG). It is common for patients with prune belly syndrome to have urachal diverticulum on VCUG.

Analysis of the umbilical fluids may provide another means of diagnosing a urachal anomaly. Fluid analysis would include measuring the umbilical fluid for content of urea and creatinine. Injecting methylene blue transurethrally or indigo carmine intravenously and observing a color change in the draining fluid or, conversely, injecting indigo carmine into the fistulous tract and looking for a color change in the urine may also provide the diagnosis of a patent urachus [9]. Finally, cystoscopy has also been reported to assist in the characterization of urachal anomalies [7].

Outcomes and complications of urachal anomalies

Congenital anomalies of the urachus represent an arrest of the normal process of involution of the urachus; they generally present during infancy and childhood but may not present until adulthood. Common presenting symptoms are periumbilical discharge, umbilical mass, periumbilical pain, and dysuria [4]. Bladder prolapse or eversion has been reported in a patent urachus [14,15], mimicking an omphalocele on antenatal scans. Sepsis secondary to a patent urachus has also been reported [16,17]. Urachal cysts may become infected. Usually the infection is restricted to the space of Retzius but occasionally the cyst may rupture intraperitoneally with resultant bowel fistula formation [18]. A urachal sinus usually presents with symptoms and signs of localized sepsis. Occasionally intra-abdominal contents may be densely adherent to the inflammatory mass and may be injured during resection. A vesicourachal diverticulum rarely requires treatment unless it is large with poor emptying due to a narrow neck or paradoxical contraction.

In a report by Ueno and associates [19], the authors advocate that patients with asymptomatic urachal

remnants do not require follow-up, and urachal remnants, especially those under 1 year of age, do not require surgical resection unless the patient has multiple symptomatic episodes. Their conclusions were based upon the finding of only one patient out of 44 patients that developed recurrent symptoms during follow-up (maximum follow-up was 32 months). The authors also cite the spontaneous involution rate of the normal urachus during infancy [20] and found that nearly one-third of their asymptomatic patients had disappearance of their urachal remnant.

One of the concerns of leaving a urachal remnant untreated is the potential risk of developing adenocarcinoma within these urachal anomalies. Urachal carcinoma is rare in children (0.01%) [21] and accounts for 0.034% of all bladder cancers [22,23]. The most common type is adenocarcinoma although other histological types have been reported [24–29]. The patients usually have a poor prognosis due to late presentation with local invasion. In a histologic review of 23 urachal remnants removed over a 10-year period, Upadhyay and Kukkady found normal urothelial lining in 17 urachal remnants whereas six (25%) showed abnormal epithelium. This abnormal epithelium included colonic epithelium, small intestinal epithelium and squamous epithelium which suggest concern for malignant degeneration. Given this potential risk of malignancy along with the minimally invasiveness of laparoscopy, a case may be made for laparoscopy as a treatment modality for asymptomatic urachal remnants [30]. In fact, laparoscopic resection of urachal abnormalities has been suggested as the first-line surgical modality for management [31].

Diagnosis and management of urachal anomalies

Patent urachus

A patent urachus usually presents itself at or soon after birth when the umbilical cord is ligated and urine drains from the umbilicus. Historically, lower urinary tract obstruction has been considered a contributing factor in its pathogenesis [32], but this is not seen in the majority of cases. In fact, urethral tubularization occurs after the urachal lumen obliterates during fetal development [33]. Subsequently, it suggests that

infravesical obstruction has little influence on urachal development.

As previously mentioned, the diagnosis is frequently confirmed after injecting the patent urachal opening with contrast during a sinogram (Figure 13.2b) or by analyzing the umbilical fluid. Other conditions that may present in a similar fashion with a wet umbilicus include anomalies of the omphalomesenteric duct (completely patent omphalomesenteric duct, omphalomesenteric duct sinus, vitelline cyst, Meckel's diverticulum) or an umbilical granuloma [34].

In the management of a patent urachus, observation may be indicated in young infants without symptoms because the involution of the urachus is not complete at birth and spontaneous closure can occur in the first few months of life [20]. If there is an associated bladder outlet obstruction, management of the bladder outlet obstruction is often adequate for involution of the patent urachus to occur in these patients.

When drainage is persistent, complete excision of the urachus with a small cuff of bladder by an extraperitoneal approach is recommended because of the concern for malignancy [7] (Figure 13.2).

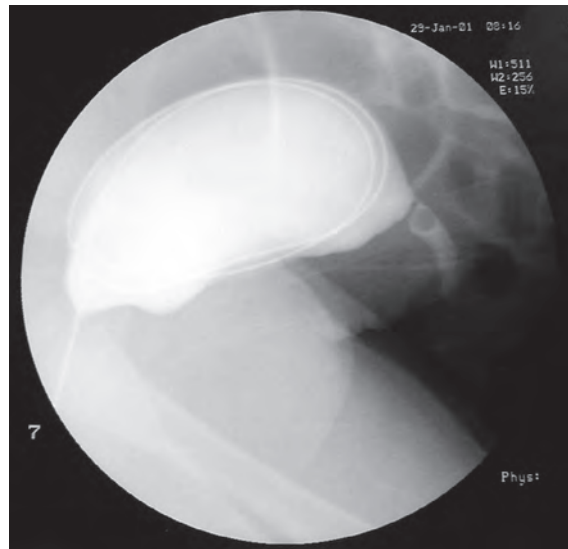
Urachal cyst

The majority of urachal cysts develop in the lower third of the urachus. Most urachal cysts go undetected unless they become infected or enlarge to a size that causes mechanical symptoms [35]. Following enlargement of the cyst, symptoms include lower abdominal pain, a feeling of heaviness and urinary frequency. If the urachal cyst becomes infected, it may develop into a urachal abscess. The majority of these urachal abscesses are infected with *Staphylococcus aureus* [3,4] and these generally manifest in adulthood. US is the most common diagnostic modality to identify urachal cysts [4] (Figure 13.3a). CT scan is beneficial when there is a large cystic abscess or there is severe periumbilical cellulitis which may cause misinterpretation on ultrasound [12] (Figure 13.3b).

Treatment of urachal cysts involves complete excision. However, when infection is present, management by perioperative drainage and antibiotics followed by subsequent elective excision may represent the most effective surgical option [36–38]. Excision of the urachal cyst may be done openly or may be performed laparoscopically [39,40].



(a)



(b)



(c)

Figure 13.2 Patent urachus. (a) Patent opening inferior to umbilical cord. (b) Patent urachus visualized on cystogram and sinogram. (c) Operative dissection of patent urachus.

Urachal sinus

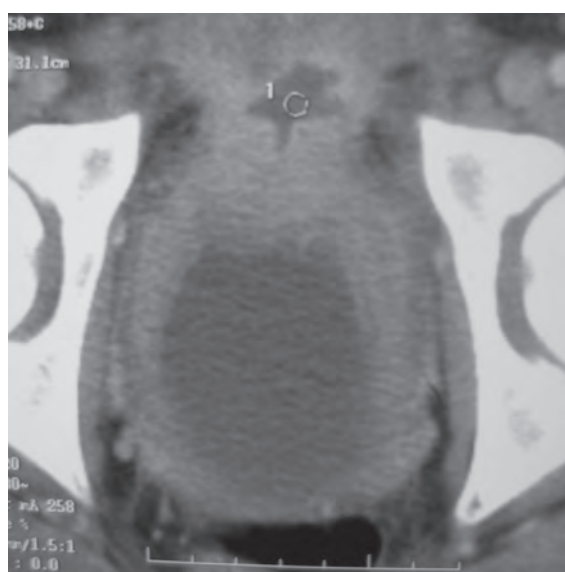
A urachal sinus most likely represents a urachal cyst that becomes infected and dissects to the umbilicus (Figure 13.4). Additionally, a urachal sinus may drain into the bladder or it may drain into either the umbilicus or the bladder and is termed an alternating sinus. These patients typically present in childhood with periumbilical pain and tenderness and may have umbilical erythema, excoriation or reactive granulation tissue. A fistulogram is usually diagnostic and will help delineate the extent of the sinus tract [4,12]. After treatment of the acute infection, surgical excision of the sinus tract is recommended.

Vesicourachal diverticulum

A vesicourachal diverticulum is frequently seen in a child with prune belly syndrome. A vesicourachal diverticulum may be seen in the setting of lower urinary tract obstruction (e.g. posterior urethral valves) but may also be discovered incidentally during an imaging work-up (e.g. VCUG for evaluation of vesicoureteral reflux). Patients that have this urachal anomaly are usually asymptomatic. A vesicourachal diverticulum is thought to occur when there is incomplete obliteration and closure of the lower portion of the urachus and the bladder apex. No treatment is usually necessary since this anomaly is primarily morphological and bears no functional consequences.



(a)

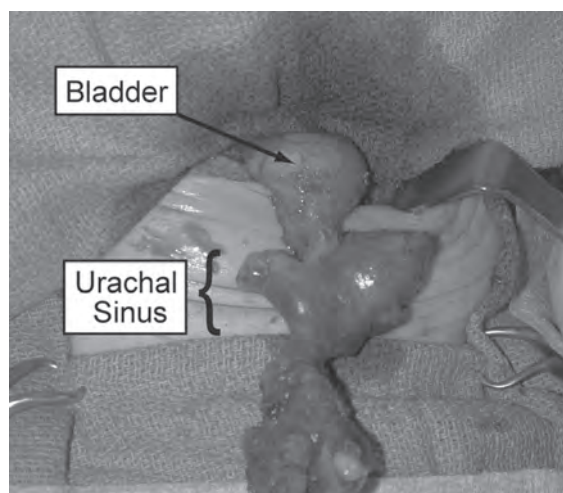


(b)

Figure 13.3 Urachal cyst. (a) Ultrasound image of urachal cyst, note proximity to bladder (BL). (b) CT scan evaluation of abdominal pain revealing infected urachal cyst.



(a)



(b)

Figure 13.4 Urachal sinus. (a) Urachal sinus presenting as a mucosal, protuberant umbilical mass with drainage. (b) Operative dissection of alternating urachal sinus. Note connection to bladder and umbilicus.

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PART V

Endoscopic Surgery of the Urinary Tract

CHAPTER 14

Cystoscopy and cystoscopic interventions

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KEY POINTS

- Primary endoscopic resection of both anterior and posterior urethral valves is the preferred treatment option in the majority of cases
- Routine second look procedures ensure completeness of resection and de-obstruction
- With modern instrumentation and good surgical technique, complications directly related to endoscopic manipulation are rare, e.g. iatrogenic urethral strictures
- Endoscopic treatment of pediatric urethral strictures is associated with a cure rate of 50%, with best results achieved for the short segment idiopathic bulbar strictures
- Endoscopic ureterocele puncture is an effective method of producing upper tract decompression and can be curative for the single system intravesical ureteroceles
- JJ stenting as the initial treatment for primary obstructive megaureter is curative in up to 50% of cases but is associated with a high morbidity (up to 70%)

CLINICAL PROBLEM/CASE

A male infant was noted to have antenatal left hydronephrosis on ultrasound. Prophylactic antibiotics were commenced at birth. He and his parents attend your clinic. Representative postnatal ultrasound images are shown below.

- 1 What do the ultrasound images show?
- 2 What are the usual associations?

- 3 How can you classify the structure seen on the ultrasound images?
- 4 How might these present clinically?
- 5 How should the patient be evaluated prior to intervention?
- 6 What are the goals of intervention?
- 7 How would you approach this problem cystoscopically?



Introduction

Pediatric endoscopy has come a long way in the past two decades, and advances and refinements in fiberoptic technology, along with miniaturization of equipment, allow endoscopic visualization of almost the entire urinary tract. Miniaturization has allowed the endoscopic appraisal of most neonatal urethras including those of preterm babies weighing >2 kg. There is a wide array of instruments available today which allow for safe assessment and intervention with minimal complications provided one remains within the limitations of the available equipment.

Posterior urethral valves

Posterior urethral valves (PUV) remain the most common cause of lower urinary tract outflow obstruction in male infants with an estimated incidence of 1:5000 live births. The majority are suspected on antenatal ultrasound screening and referred to specialist centers at birth. Modern endoscopic equipment, along with long-term outcome data, has dramatically changed the surgical approach to valve treatment. In the past, many newborns were treated with a vesicostomy primarily because of the relatively large instrumentation available. Supra-vesical diversions were in vogue due to their undisputed short-term benefits on renal function; however, the long-term outcomes of these diversions show no benefit for renal function and raise concern regarding the effect on outcome on bladder function. This section covers the various approaches to the endoscopic ablation of PUV, their complications, and an approach to avoiding these complications in the 21st century.

Surgical techniques and outcomes

In 1972, Whitaker et al. [1] reported the results of 112 patients in whom valves were ablated endoscopically with an infant McCarthy panendoscope, using either a bugbee or a loop electrode to destroy the valve membrane. In situations where the urethra (meatus and submeatal region) would not accommodate the instrument, a small perineal urethrostomy was made by cutting down upon the tip of a small sound placed in the urethra, and the panendoscope was introduced via the perineum just distal to the external sphincter.

The valve membrane was ablated using coagulating diathermy current. Many authors have expressed concern about iatrogenic stricture disease with this technique. Whitaker does not report on the incidence in his series; however, Myers and Walker [2] reported a 50% incidence of strictures in his series of valves ablated in infancy with a 25% incidence in the group as a whole.

Whitaker and colleagues [1] reported a 33% incidence of continuous incontinence with a further 55% stress incontinence in their series of 112 patients. It is difficult to attribute a cause to incontinence as some of their patients had additional bladder neck surgery, and the noncompliant valve bladder had not been ruled out. An addendum to that publication reported a dramatic improvement in daytime incontinence in five boys following treatment with imipramine.

In 1973, Williams and associates [3] advocated ablating the valves with a diathermy hook electrode under radiological control. This technique avoided a perineal urethrostomy and was successful in several cases, but complications arose when the hook engaged adjacent tissue like the bladder neck or verumontanum. Whitaker and Sherwood [4] subsequently modified the hook, which minimized the risk of adjacent tissue injury but did not completely eliminate it.

Hendren [5] was a proponent of valve ablation under direct vision and in the narrower urethra passed the insulated wire electrode (3F) alongside an 8F endoscope, a technique he described as "a little awkward". He ablated the valves using cutting diathermy current and reported no strictures with this technique. Several boys in his series required further resection of the valves, and once again the continence outcome was muddled by synchronous or metachronous bladder neck surgery.

In 1986, Diamond and Ransley [6] described the Fogarty balloon catheter ablation technique and noted that, while not universally successful, they achieved satisfactory results in nine of ten carefully selected patients. The bladder is filled with contrast material via a small feeding tube, which is then removed. A no. 4 Fogarty balloon catheter is passed and the balloon inflated within the bladder. The catheter is then slowly withdrawn under fluoroscopic control until the balloon engages the valve. A sharp tug causes rupture of the anterior membrane. The balloon is then deflated and the catheter is removed. Cromie and associates [7] describe a similar procedure using a modified venous valvulotome in which, under fluoroscopic control, the

valvulotome is used to engage and cut the obstructing membrane. They reported completeness of ablation in 13 of 15 patients treated. Both reports describe a 0% stricture incidence with their respective techniques.

Other described techniques include Mohan's [8] urethral valvotome in which the valves are engaged and destroyed without any fluoroscopy control and under local analgesia, relying on the instrument design to "catch" the valves. Mohan reports good results in eight boys treated using his instrument based on symptoms and improved radiological findings on repeat voiding cystourethrogram (VCUG).

Percutaneous, endoscopic, antegrade diathermy valve ablation has been popularized by Zaontz and Firlit [9] with a subsequent description by Gibbons and associates [10] of endoscopic antegrade diathermy valve ablation through a vesicostomy.

A bugbee or insulated wire electrode passed through an infant (5–7.5F) cystourethroscope is a useful technique in very small infants. Similarly, laser ablation using the potassium titanyl phosphate (KTP) (or Nd-YAG) laser via a small fiber passed through the working channel of the infant cystoscope has been reported as a safe technique in newborn infants with no stricture formation up to 3 years follow-up. This technique relies on coagulative necrosis of the membranous tissue and the tip of the electrode or fiber is placed in direct contact with the valve tissue and the current is applied. The process is repeated at multiple sites.

Videoendoscopy using a 8.5F (5° lens) to 10F (0° lens) resectoscope and a cold knife hook working element allow safe and effective ablation of urethral lesions without risk of thermal injury or significant bleeding and can be employed in infants as small as 2 kg. At the present time, this is the safest and most effective technique for ablation of membranous lesions of the infant

urethra and is the gold standard against which other techniques will be compared. The membrane is cut at the 5 and 7 o'clock positions until its connection with the verumontanum is lost. Some surgeons advocate additional routine incision at the 12 o'clock position.

The bladder is drained via a 6–8F feeding tube for 24–48 hours and subsequently removed.

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 and a 0% incidence of urethral stricture disease.

The instrumentation available for valve ablation at the authors' institution includes:

- 1 A 6–7.5F graduated Wolff cystourethroscope with a 3F working channel.
- 2 A choice of 10, 11, or 13.5F resectoscopes with a cold knife hook working element.
- 3 Bugbee electrodes, Colling's knife, and resectoscope loops.

In situations where the available instrumentation was too large to be safely accommodated in the infant urethra, valve ablation was deferred to a later date. In the interim, the urethra was serially and passively dilated using increasing caliber urethral catheters changed on a weekly basis.

Complications

Urethral stricture (Table 14.1)

Dense strictures following diathermy ablation are on the decline and most recent series report a ~5% incidence of strictures. Diagnosed early, they may be amenable to dilatation or visual internal urethrotomy. Recurrent strictures or long segment strictures will require definitive anastomotic or augmented urethroplasty.

Table 14.1 Incidence of urethral stricture following ablation of posterior urethral valves.

Author	Age at valve ablation (years)	Number (n)	Follow-up (years)	Stricture (%)
Myers and Walker [2]	<1	14	1–10	50
Myers and Walker [2]	>1	14	1–10	0
Nijman and Scholtmeijer [11]	< and >1	85	5–19	0
Lal et al. [12]	0–15	82	1–21	3.6
Crooks [13]	1.5–?	36	4–?	8
Churchill et al. [14]	<1 to >1	173	?	12

Incomplete valve ablation

Completeness of valve ablation must be confirmed on follow-up VCUG and/or cystourethroscopy. Incidence of incomplete ablation varies and in our review was seen in 21% of cases. It is more likely following bugbee or insulated wire ablation through a small cystoscope, as the vision is limited due to relatively poor flow of irrigation fluid.

It is prudent to re-scope the urethra within 6–12 weeks of the initial ablation to check completeness of resection, as the consequences of persistent obstruction are potentially disastrous.

Obstructive oliguria or anuria

True uretero-vesical junction (UVJ) obstruction is rare in boys with PUV.

Temporary UVJ obstruction, presumed to be due to entrapment of the UVJ by the thickened detrusor following decompression of the bladder, is not an uncommon occurrence in clinical practice.

The oliguria, or in severe cases anuria, typically lasts for 24–48 hours and resolves spontaneously.

There may be associated peri-renal urinary extravasation and nephrostomy drainage of one or both kidneys may occasionally become necessary; however, most cases can be managed with a policy of watchful waiting.

Sarkis et al. [15], Noe and Jenkins [16], and Jordan and Hoover [17] have all reported individual cases and Sarkis recommends use of non-self-retaining tubes to drain the bladder to minimize the risk.

Urinary tract infection

It is not uncommon for these children to have impaired renal function, making these neonates particularly vulnerable to developing urine infection following urethral instrumentation, VCUG, and valve ablation. In addition, they may acquire drug-resistant organisms and, with prolonged antibiotic treatment, secondary fungal infections which are extremely difficult to treat.

Renal impairment

The condition is associated with considerable morbidity and accounts for 16% of children with end-stage renal failure and for 25% of all children with end-stage renal disease (ESRD) who undergo renal transplantation, according to the UK Transplant Registry 2005. Early renal failure is attributed to inherent renal dysplasia. The etiology of late onset renal impairment is more complex,

and dysplasia, bladder dysfunction, and urinary tract infections are all implicated.

The fragile renal function in the neonatal period is exquisitely sensitive and vulnerable to fluid imbalance and urinary infection. Adequate precautions along with input from nephrological colleagues help to maintain optimum function and nadir creatinine levels along with a formal glomerular filtration rate (GFR) value at 1 year of age helps predict long-term outcome in these children.

Urinary incontinence

At the present time, urinary incontinence as a consequence of valve ablation and associated sphincter damage is rare. In the past, a high incidence was reported: 33% continuous and an additional 55% stress incontinence in a series of 34 patients who had both valve ablation and bladder neck surgery [1]. The subsequent description of the valve bladder and the abnormal urodynamic patterns observed in these children clarified the etiopathogenesis of incontinence in the majority of these patients.

Parkhouse and associates [18] reported daytime urinary incontinence at 5 years of age to associate with poor long-term renal outcome. A further compounding factor, which may be responsible, is polyuria, secondary to impaired renal function.

Recent studies, which have prospectively looked at the development of bladder function in these children, have shown a high incidence of bladder dysfunction (up to 70%) [19]. Serial invasive urodynamic studies have documented a changing pattern over time with a different etiology for incontinence at varying time points [20].

Preventing complications

Urethral stricture

The neonatal urethra is extremely delicate and forced or over-sized instrumentation will inevitably result in narrowing. It is good practice to prophylactically dilate the meatus and submeatal region prior to introduction of the endoscopes. Generous lubrication and gentle manipulation will minimize trauma. It is essential to have an array of instruments available, which will allow safe and satisfactory valve ablation.

The new pediatric resectoscopes have a blunt metal rounded tip compared to the Bakelite sharp beaks

found on older instruments. This design feature helps to minimize injury during introduction.

A cold knife incision is neat, specific, and not associated with surrounding tissue damage. Diathermy ablation has the potential to injure adjacent structures, and the current can penetrate to the deeper tissues, which has the potential to promote scarring. A pure cut setting on the diathermy is less damaging than coagulating current and the smaller surface area of the Colling's knife is more precise compared to the resectoscope loop.

Limiting operative time to a minimum, ensuring adequate visualization of the important landmarks during resection, and minimizing postoperative bladder drainage all contribute toward lowering the risk of stricture formation.

It was felt that a "dry urethra" following valve ablation promotes stricturing; however, Lal et al. [21] found no evidence to support this hypothesis in his study.

In the uncommon situation where the available instrumentation is found to be too large, deferring valve ablation or a temporary vesicostomy further reduce the risk of complications and improve outcomes.

Obstructive oliguria or anuria

Decompressing the bladder via suprapubic or urethral catheter usually results in a period of post-obstructive diuresis. Paradoxically, draining the bladder can sometimes result in a temporary obstruction at one or both vesicoureteric (VU) junctions, producing obstructive oliguria, anuria with or without calyceal rupture and urinomas. Sarkis et al. [15] have attributed this to the balloon of the urethral catheter obstructing the ureteric orifices in a small thick-walled bladder.

An alternative explanation is that the hypertrophied detrusor clamps down on the intramural part of the lower ureter, temporarily kinking the lumen to occlude the flow of urine. We have observed this phenomenon both pre- and post-valve ablation and we do not use balloon catheters to drain the bladder.

Avoiding balloon catheters, minimizing the length of tubing within the bladder when using feeding tubes, and avoiding repeated insertions by draining the bladder suprapubically (5F Cystofix® Minipaed) are measures that may minimize the risk. Slow decompression of the chronically distended bladder is recommended in adults; however, this is difficult to achieve in the neonate.

Incomplete valve ablation and sphincteric incontinence

Adequate visualization during valve ablation is the key to successful and complete resection. A selection of instrumentation should be available to allow safe and satisfactory ablation in the majority of cases. Routine second look appraisal of the urethra within 6–12 weeks will minimize complications as a consequence of incomplete resection. Avoid concomitant bladder neck surgery as proposed recently [22] because in the past this approach has been shown to be associated with high morbidity.

Urinary tract infections and renal impairment

Routine prophylactic antibiotic and antifungal cover around the VCUG and valve ablation minimizes the incidence of infections in the neonatal period. Antibiotic prophylaxis in infancy and ensuring a good fluid intake along with elective circumcision offered at the time of second look check cystoscopy are measures that will reduce the risk of urinary infections in infancy.

Summary

Videoendoscopic valve ablation using the hook cold knife offers a safe and effective way to ablate the valves in the neonatal period. A lesser number will require ablation using bugbee or insulated wire electrodes.

Prophylactic dilatation of the urethra, generous lubrication, avoiding forcible use of over-sized instruments, and gentle manipulation minimize complications.

Routine second look procedures ensure completeness of resection.

Careful monitoring and recruiting nephrological colleagues in the care of these children optimizes outcome. Routine periodic follow-up with appropriately timed investigations to address aberrations in urinary continence and renal function ensure optimal long-term outcome in these children.

Anterior urethral valves or syringocele

Anterior urethral valves (AUV) were first described by Watts in 1906 as a cause of urethral obstruction in boys. The origin is attributed to a cystic dilatation (syringocele) of the main bulbourethral glands described by Cowper in 1705.

The paired Cowper's glands lie dorsal to and on either side of the membranous urethra. The ducts from these glands are 2–3 cm long and enter the urethra individually or fuse proximally to enter as a single orifice. These mucus-secreting glands begin to function by 4 months gestation, and their secretion acts as a lubricant and transport medium for sperm during ejaculation.

The common location of the lesion, approximately 1 cm proximal to the fossa navicularis, suggests a faulty union between the glanular and penile urethra as the cause. Other plausible explanations suggested include congenital urethral stricture, abortive urethral duplication, and transient urethral obstruction *in utero*.

Rupture of the syringocele causes the distal lip to lift up and abut against the anterior wall of the urethra during voiding, acting as a flap valve resulting in urinary outflow impairment. The diagnosis is made on a VCUG.

Clinical presentation is variable and includes prenatal hydronephrosis, penile swelling, urinary tract infections, and voiding symptoms like poor urinary stream and dribbling.

Surgical techniques and outcomes

Transurethral incision or fulguration of the valve is the procedure of choice in the majority. Provided a significant defect in the corpus spongiosum has been eliminated on clinical exam and VCUG, no further intervention is necessary.

McLellan and associates [24], in a series of nine cases over 40 years, found incision to be successful in 75% of cases, with one child requiring a subsequent urethroplasty.

Van Savage et al. [25] in their series of 17 cases found this technique successful in all six cases treated with a transurethral incision of the valve leaflet.

A urethroplasty or diverticulectomy is recommended in cases where the spongiosal defect is large resulting in poor urethral support. The urethroplasty may be performed as either a one- or two-stage procedure and the published results of two of the series mentioned earlier show a success rate approaching 100%.

A vesicostomy as the primary treatment has been recommended in neonates and infants with associated high-grade bilateral vesicoureteral reflux (VUR). Subsequent management will depend on the degree of the spongiosal defect and may include correction of VUR.

In general, the effects of intravesical obstruction due to AUV are less severe than those due to posterior urethral valves (PUV). AUV cause few long-lasting upper tract

radiographic changes and are associated with a chronic renal failure incidence of 0–5% compared to up to 60% in children with PUV. Similarly, bilateral VUR is not associated with a poor prognosis in children with AUV, suggesting majority of reflux is secondary and resolves after treatment of the AUV.

In van Savage et al.'s [25] series, patients with anterior urethral valves were found to be continent of urine and free of urinary tract infections and obstructive symptoms at long-term follow-up.

Complications and management

Urinary extravasation

Urinary extravasation has been described in isolated cases following transurethral incision. In these patients, the subcutaneous tissue and corpus spongiosum is attenuated and allows for urinary extravasation. On removal of the catheter and subsequent voiding, urine leaks to track along the penile shaft and scrotum producing a boggy swelling. Reintroduction of a urethral catheter and a further period of drainage usually resolves the problem. Aspiration or drainage may be indicated if the urinoma is infected and, in rare cases, a subsequent urethroplasty may be necessary.

Urethral fistula

Urethral fistulae may occur following one- or two-stage urethroplasty and are managed in the same way as those occurring following a hypospadias repair.

Vesicoureteral reflux

Persistent VUR following successful relief of obstruction due to AUV may require intervention if symptomatic. Treatment options include endoscopic correction and surgery. Bladder dysfunction is rare in children with AUV.

Summary

AUV is a rare cause of male urethral obstruction; the majority can be treated endoscopically and the condition is associated with good long-term outcome for both renal and bladder functions.

Urethral strictures

Pediatric urethral strictures are uncommon and their etiology can be divided into "inflammatory," traumatic, and idiopathic. The inflammatory category includes

nonspecific urethritis, lichen sclerosis balanitis xerotica obliterans (BXO), and urethralgia posterior. Trauma is commonly iatrogenic and attributed to instrumentation or surgery but also includes the more dramatic fall astride and road traffic accident (RTA) injuries. The majority of the idiopathic strictures occur at the junction of the proximal and middle sections of the bulbar urethra in adolescents or young adults with no previous history, suggesting a “congenital” etiology.

In the pediatric population, the most common cause of strictures is previous hypospadias repair which in one series [26] was responsible for 40% of cases. Other etiologies include idiopathic in 18%, post-instrumentation in 12%, traumatic in 10%, and the remaining following surgery for anorectal malformation (ARM), pelvic radiation, balanitis xerotica obliterans (BXO), and posterior urethral valves (PUV) ablation.

Surgical techniques and outcomes

The time-honored method of treatment is urethral dilatation, which stretches the stricture and, more commonly, disrupts it. An alternative treatment with an equally long history is a urethrotomy. Initially this was performed blindly but with the development of endoscopic instrumentation is now carried out under vision. It is now widely accepted that both dilatation and urethrotomy are equally effective and can cure up to 50% of short bulbar strictures when first used (Table 14.2). Alternatives such as laser urethrotomy, indwelling urethral stents, or intermittent self-catheterization are not curative and, of these, only self-catheterization is occasionally helpful.

If instrumentation is required more frequently or is complicated then a urethroplasty is the only curative option.

Table 14.2 Outcomes of visual internal urethrotomy for urethral stricture.

Author	Number (n)	Success (%)	Follow-up (month)
Hsiao et al. [25]	40	50	24
Stormont et al. [27]	199	68	42
Pansadoro and Emiliozzi [28]	224	32	60
Heyns et al. [29]	163	39	24
Duel et al. [30]	37	21	?

Table 14.3 Incidence of complications of visual internal urethrotomy.

Author	Number (n)	Complication rate (%)
Hsiao et al. [25]	40	4
Stormont et al. [27]	199	18
Smith et al. [31]	?	27
Inversion et al. [33]	937	6.7

Excision and end-to-end anastomosis or anastomotic urethroplasty is the best option for short strictures, 1–2 cm long of traumatic or idiopathic origin in the bulbar or membranous urethra. For all other locations or recurrent strictures, 2 cm in length, a substitution urethroplasty is the procedure of choice. A stricturotomy and patch graft (foreskin, posterior auricular Wolfe graft, or buccal mucosa) is more successful than excision and circumferential repair, and in situations where excision of the stricture becomes necessary, a two-stage repair is a more successful and reliable option [27–30].

Complications of visual internal urethrotomy

Well-recognized complications of internal urethrotomy (Table 14.3) include:

- Bleeding.
- Fever.
- Epididymitis.
- Incontinence.

In the adult literature, serious complications like erectile dysfunction, rectal perforation, and chordee have been described [31–34].

Summary

Visual internal urethrotomy is a minimally invasive procedure associated with a cure rate of approximately 50% in carefully selected cases.

The best results are achieved in short-segment idiopathic bulbar strictures with a low-associated complication rate.

If a patient develops a recurrent stricture following a urethrotomy, however long the interval, further instrumentation is rarely curative. Similarly, penile or long-segment strictures are rarely cured by urethrotomy. In

these situations it is best to proceed to a urethroplasty early.

Endoscopic management of ureterocele

The aim in ureterocele management is prevention of renal damage secondary to obstruction (with or without the associated comorbidity of VUR and urinary infections). 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.

It is generally accepted that endoscopic puncture of the ureterocele is a definitive procedure in children with a single non-refluxing system with an intravesical ureterocele.

The management of children with ectopic ureteroceles and ureteroceles associated with duplex systems is controversial, with management strategies ranging from periodic follow-up to open ablative and reconstructive surgery.

Surgical techniques and outcomes

Montfort and associates [35] described endoscopic ureterocele incision in 1985 and found that a small incision was less likely to cause reflux compared to the previous practice of de-roofing the ureteroceles.

In 1994, ureterocele puncture was described which has supplanted incision to become the mainstay in the endoscopic management of ureteroceles.

The success rate of endoscopic management when assessed by the rate of decompression and subsequent urinary tract infections has been shown to be high although outcomes from some groups suggest that it is ineffective in preventing urinary infections. Endoscopic management is most successful in patients with intravesical single system ureteroceles. In children with ectopic ureteroceles associated with duplex systems, endoscopic puncture does not appear to be definitive, but it is successful in achieving decompression and minimizing the risk of urinary tract infections.

The conclusion of a recent meta-analysis of surgical practice in the endoscopic management of ureteroceles

by Byun and Merguerian [36], where the outcome measure was secondary operation was that:

- 1 Reoperation rates were significantly greater in patients with ectopic compared to intravesical ureteroceles.
- 2 There was a greater rate of reoperation in patients with duplex versus single system ureteroceles.
- 3 The presence of reflux preoperatively was associated with a significantly greater risk of reoperation.

The drawback of this meta-analysis is that the outcome measure of reoperation includes surgery for persistent reflux and a residual non-functioning decompressed upper moiety of a duplex system. The management of these clinical situations is varied and does not necessarily mandate further surgery.

Complications and their management

Inadequate decompression

Ureterocele management has moved from the extensive de-roofing to the current precise puncture of the lesion in a dependent position. This approach can result in an inadequate opening, particularly in thick-walled ureteroceles, causing persistent obstruction. Antibiotic prophylaxis and a repeat ultrasound assessment 4–6 weeks following the procedure will minimize the risk and will help determine the need for further puncture.

Vesicoureteral reflux

Ipsilateral or contralateral VUR into the lower moiety may persist following ureterocele decompression and occasionally VUR into the decompressed moiety may develop following endoscopic intervention. In 2011, Frimberger and colleagues reported the “watering-can” puncture as a technique for decompression; it resulted in successful decompression (91%), improved hydronephrosis (82%), but less *de novo* VUR compared to standard techniques (36% vs. 75%) [37].

Asymptomatic VUR is not in itself an indication for further intervention and may be a reflection of the developmentally distorted anatomy at the trigone. Persistent VUR in association with recurrent breakthrough urinary tract infections or voiding dysfunction will determine the need for further surgery.

Voiding dysfunction

Voiding dysfunction occasionally develops following ureterocele puncture, particularly in association with large thin-walled ureteroceles. The distal lip can act as a

flap valve to obstruct the bladder outlet during voiding. Treatment usually involves excision of the ureterocele, repair of any defect in the bladder wall with or without ureteric reimplantation.

With the more complex cecoureteroceles there may be associated bladder neck deficiency which results in urinary incontinence. This tends to occur in females and the trigone is grossly abnormal. Surgical intervention in such cases is indicated for continence and, in addition to ureterocele excision and repair, some form of bladder neck surgery is necessary in order to improve continence.

Summary

Endoscopic ureterocele puncture is an effective treatment modality for producing upper tract decompression and minimizing the risk of urinary tract infections. It is curative in the majority of single system intravesical ureteroceles and effectively decompresses a significant proportion of obstructed upper moieties of duplex systems so that no further interventions are necessary in carefully selected cases.

JJ stenting for primary obstructive megaureter

The majority (up to 80%) of perinatally detected primary obstructive megaureters resolve spontaneously, hence conservative management with watchful waiting is considered a safe initial approach for this condition. An increasing number of these cases are detected on routine antenatal screening and can result in a clinical dilemma when associated with ipsilateral reduced renal function. The need to temporize bladder/trigone surgery before 1 year of age because of the fear of jeopardizing evolving bladder function further compounds the problem. External urinary diversion is a well-established temporizing measure but is not without problems. Stenosis, inadequate drainage, and the need for further surgery suggest the need to look for alternative treatment modalities. JJ stenting of the vesico-ureteric junction (VUJ) is one such minimally invasive alternative to achieve temporary internal drainage.

Surgical techniques and outcomes

JJ stenting for symptomatic VUJ obstruction in infancy was reported in 1999 [38]. The limitation to

endoscopic insertion in infancy is the caliber of the urethra and available instrumentation. Ureteric JJ stents of 3F/12 cm length are available (Rusch International, Germany) which are suitable for endoscopic insertion.

Indications for stenting include distal ureteric dilatation 10 mm, reduced differential renal function (~40%), a drop in differential function, and an obstructive curve on diuretic renography.

The ureteric orifice and distal end of the ureter can be difficult to negotiate and conversion rates to open cystotomy, dilatation, and open insertion (of a larger 5F or 6F) of the stent are high.

In cases of bilateral megaureter, a single longer length (20–24 cm) stent can be used, with the looped ends up the ureter and a short length of the straight segment forming a bridge between the ureteral orifices. Open insertion was necessary in 50% of cases in Castagnetti et al.'s [39] series and can be higher.

The stents are left in position for 6 (recommended) to 9 months and the urinary tract is periodically assessed with ultrasonography and isotope renography during this period. Antibiotic prophylaxis is recommended. Fifty percent of cases treated in this manner require no further intervention [39]. Morbidity associated with this approach has been reported as high as 70% due to stent-related complications.

Complications

- Breakthrough urinary infections.
- Stent blockage/encrustations.
- Bladder spasms.
- Hematuria.
- Fungal urinary tract infection.

Management of complications

Careful monitoring with monthly ultrasonography for the first 3 months following stent insertion is prudent. Consideration must be given to synchronous circumcision with stent insertion in male patients. Ensuring a minimum amount of free tubing within the bladder will minimize bladder spasms. A high index of suspicion must be maintained when interpreting findings on ultrasound of debris within the collecting system, and urine must be examined for hyphae to rule out fungal infections. In the event of stent-related complications, early removal followed by either a diversion or reimplantation is warranted in the presence of ongoing obstruction.

Summary

JJ stent insertion across the VUJ can allow effective drainage in primary obstructive megaureters and in a proportion (up to 50% [40]) is curative.

There is some evidence to suggest that in those requiring subsequent reimplantation of the ureter, the need for ureteral tapering is obviated but the numbers are small.

The technique is associated with a high morbidity rate (up to 70% [40]) and a high rate of conversion to an open insertion method.

Given the high spontaneous resolution rate in primary obstructive megaureter and the high complication rate associated with stenting, its use should be restricted to carefully selected cases that are very critically monitored following placement of the stent.

The technique's potential to decrease the need for ureteral tapering needs further evaluation.

Botox® injections for neurogenic bladder dysfunction

Neurogenic bladder and bowel dysfunction is present in a large proportion of children with neural tube defects and caudal regression syndrome. Detrusor overactivity, impaired bladder compliance, and detrusor sphincter dysynergia are responsible for deterioration in renal function and early management in these patients remains controversial. It is generally agreed that the aim of urological management in these children is preservation of the upper tracts in infancy and early childhood and the subsequent attainment of urinary continence in later life. The management of the hostile bladder is varied, ranging from anticholinergic medication with or without clean intermittent catheterization (CIC), urinary diversion (vesicostomy), to bladder augmentation with a catheterizable channel to aid bladder emptying. Of these, anticholinergic medication is the least invasive and reversible of the options and is the preferred first choice by both patients and physicians.

However, problems arise when nonsurgical treatment fails. Approximately 10% of patients are non-responders to anticholinergic medications and a further proportion develop side effects to these drugs even when administered intravesically. Restoring

safe bladder dynamics in these patients has thus far been achieved by bladder enlargement surgery until the publication of encouraging reports describing the beneficial effects of Botulinum-A toxin into the hyper-reflexive detrusor in adults with spinal cord injuries.

Surgical techniques and outcomes

Botox® in the dose of 10 IU/kg (maximum 300 IU) or Dysport® in the dose of 40 IU/kg (maximum 1200 IU) is injected submucosally into the detrusor muscle, at multiple sites sparing the trigone. The effect usually lasts for 9–12 months and can be repeated if necessary.

Evaluation of outcomes in the limited studies published in children so far suggests significant subjective and objective benefits as evidenced by:

- Decrease in detrusor overactivity.
- Increase in bladder compliance.
- Increase bladder capacity.
- Reduction in detrusor voiding pressures.

In addition one study has noted a significant improvement in bowel symptoms in 66% of the patients treated [40].

Complications

Early reports suggest that this is a safe treatment with a small risk of hematuria and urinary infection. Urinary retention is a theoretical possibility and patients are counselled regarding the need for intermittent catheterization.

Summary

Preliminary results suggest 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 [41]. There is some suggestion that this may have beneficial effects on bowel function and needs further validation.

The unanswered question is whether or not repeat injections provide long-term relief without the development of an antibody response to repeated exposure and, if so, whether this treatment holds promise in carefully selected cases. If shown to be safe in the long term, an extended application will be in the treatment of resistant non-neurogenic bladder dysfunction.

DOS AND DON'TS**Do**

- Prophylactically dilate the meatus and submeatal region prior to introduction of endoscopes in the neonatal urethra (to avoid damage through over-sized instrumentation)
- Use generous lubrication and gentle manipulation during endoscopic procedures to avoid trauma
- Remember that good visualization during valve ablation is the key to successful ablation in the majority of cases
- Confirm the completeness of valve resection on follow-up VCUG and/or cysturethroscopy
- Ensure routine prophylactic antibiotic and antifungal cover around the VCUG and cystoscopic interventions to minimize incidence of infections in the neonatal period

Don't

- Be over-aggressive in resecting the posterior urethral valves at the first sitting. It is better to assess the completion as described above in the do's section and resect further if required
- Force the instrumentation if despite dilatation the cystoscope or resectoscope cannot negotiate the urethra. It is better to divert the urine urethrally or suprapubically till the baby is bigger. Serial urethral dilatation by weekly change of urethral catheter especially in pre-term neonates

CLINICAL PROBLEM/CASE: DISCUSSION

- 1 The study demonstrates a left-sided intra-vesical ureterocele. Both kidneys have normal echotexture and appear to have simplex configuration. The left kidney has moderate calyceal dilatation and has intermittent dilatation of the ureter along its length (not shown). The right kidney has no pelvi-calyceal dilatation.
- 2 Ureteroceles may be associated with either duplex systems (occurring in approximately 0.02% of individuals, of whom 80% are female, the ureterocele is associated with the upper-pole ureter) or simplex systems (more common in boys and almost always lie entirely within the bladder).
- 3 Ureteroceles may be intravesical or extravesical. Intravesical ureteroceles lie entirely within the bladder and above the bladder neck. The most important subtype to recognize is the caecoureterocele, in which the opening of the affected ureter lies within the bladder, but the cavity of the ureterocele extends beyond the bladder neck into the proximal urethra.
- 4 Ureteroceles may be detected on prenatal scanning (the majority are detected in this manner), as an incidental finding on imaging, or may present with urinary tract infection, voiding difficulty or urinary retention due to obstruction to bladder outflow or as a cystic mass prolapsing through the urethra (confined to girls).
- 5 Patients should undergo an ultrasound scan and VCUG prior to intervention. The ultrasound scan should provide the anatomical diagnosis. The VCUG assesses the bladder and the ureterocele will normally appear as a filling defect within the bladder. With further bladder filling the ureterocele may evert due to the underlying defect in the detrusor and appear as a diverticulum. The VCUG will also assess the urethra and presence or absence of ipsi and/or contralateral VUR. Functional imaging (MAG3 or DMSA) is useful in duplex systems to guide intervention (puncture vs. hemi-nephrectomy).
- 6 Goals of intervention are to: preserve renal function; eliminate infection, obstruction and reflux; and to maintain urinary continence.
- 7 Cystoscopic examination should examine the urethra, the bladder neck, and trigone in relation to the ureterocele. The position of other ureteric orifices should be noted (it may not be possible to identify the opening of the affected ureter). Appearances of the ureterocele will change with bladder filling; consequently, it is best to start with little filling and increase bladder volume slowly. The lowermost portion of the ureterocele needs to be noted as this is probably the best site for endoscopic puncture (best appreciated with minimal filling of the bladder). It should also be noted if the ureterocele extends down into the urethra or bladder neck (indicating a caecoureterocele) as puncture could cause problems with obstruction.

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CHAPTER 15

Interventional procedures of the genitourinary tract

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KEY POINTS

- Ultrasound guided biopsy has been shown to be a safe and effective method to obtain tissue diagnosis in different organs and locations
- Conventional angiography should be used for diagnostic purposes when the non-invasive methods (US, CT angiography, MRI angiography) haven't been able to provide the information required. Its therapeutic role (e.g. embolization in acute bleeding) cannot be replaced by these other techniques
- Nephrostomy tubes should be considered early in an infected and obstructed urinary collecting systems, and may be life-saving in severe septic complications
- Central venous lines should be used in a rational way in children. The aim is to use the smaller devices that will suffice to minimize chronic venous complications

Introduction

Pediatric interventional radiology (PIR) has been a rapidly evolving sub-specialty in recent years, becoming an increasingly important unit in most pediatric hospitals. The development of new technologies, devices and the improvement of the imaging equipment have promoted image-guided interventions that are safer and effective.

The imaging modalities that are utilized in PIR are fluoroscopy, ultrasound (US), computed tomography (CT) and magnetic resonance imaging (MRI). The interventional unit ideally should have integration of all these different imaging modalities and also utilize the potential for their use in an operating room environ-

ment, in order to perform combined cases. If the patient is exposed to radiation, the lowest dose as possible needs to be considered in order to decrease its inherent risks.

The collaboration between the interventional radiologist and the pediatric urologist is increasing every day. They are frequently seen working together to provide combined solutions to the patient's problems. This collaboration brings significant benefits to the patients and families.

This chapter uses case studies and discussions to describe typical PIR procedures performed in patients with genitourinary conditions. The procedures are explained in a very simple fashion, emphasizing potential complications and their management.

Clinical problem/case 15.1

A 5-year-old girl, previously healthy, presents with acute renal failure and mild hydronephrosis. JJ stents were placed and a US-guided renal biopsy was requested to assess the renal parenchyma. She presented with tachycardia and a drop of hemoglobin levels (40 g/l) a few days after the biopsy. An US showed a significant perinephric hematoma that expanded over 12 hours (Figure 15.1). An emergency angiogram demonstrated two areas of active contrast extravasation (Figure 15.2). A coil embolization was performed obtaining successful control of the bleeding (Figure 15.3).

- 1 What is the current role and technique utilized in image-guided biopsies of the genitourinary tract?
- 2 What are the potential complications of a renal biopsy?
- 3 What is the management of an acute post biopsy bleeding that does not respond to conservative management?
- 4 How is an arterial embolization performed for acute bleeding?



Figure 15.1 Longitudinal sonographic view of the left kidney (k) showing a large perinephric hematoma (h).

Discussion

- 1 Histopathology is still the gold standard for definitive diagnosis of many renal parenchymal diseases [1]. Therefore image guided “random” renal biopsies are fundamental in the diagnosis and treatment of patients with renal conditions. Biopsies are also essential in the diagnosis of solid tumors.

Kidney biopsies (native and transplanted) and tumor biopsies are the ones most frequently performed in the genitourinary tract. The majority of these procedures are performed using US guidance and automated biopsy devices, technique that had shown to provide better sample quality with fewer complications [2]. In cases in which US visibility is limited (e.g. obesity), cross-sectional image guidance may be required (CT, MRI);

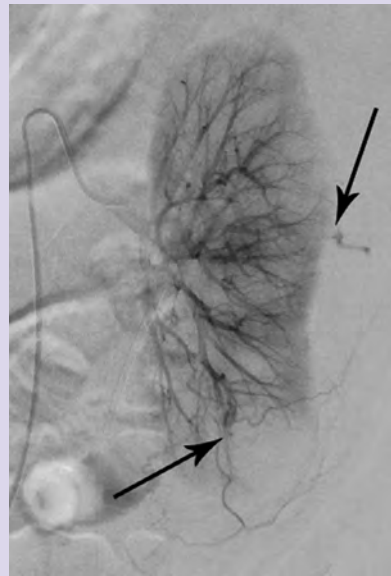


Figure 15.2 Selective angiogram of the left kidney showing two areas of contrast extravasation (arrows) compatible with active bleeding.

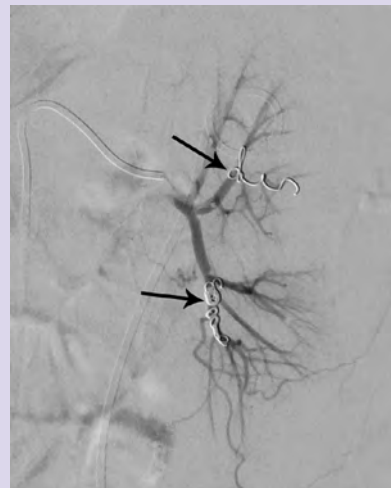


Figure 15.3 Post-embolization selective left renal angiography showing successful coil embolization (arrows) of the bleeding arterial vessels.

18 gauge is the needle size used in our institution. The length of the core of tissue can be adjusted according to the target area, with a 2.2 cm core being the most frequently utilized.

Multiple tissue cores are often needed in the cases of tumor biopsies to increase the diagnostic yield of the procedure. In these cases, a coaxial technique is used, in which an outer 17 gauge needle is advanced into the periphery of the lesion. Using this needle as a guide, multiple cores of tissues are obtained with the 18 gauge biopsy needle loaded in the automated device. After the samples are obtained, the tract is embolized through the coaxial needle using gelfoam slurry or the embolization material preferred by the operator, to decrease the bleeding risk [3].

- 2 Ultrasound-guided renal biopsies are safe procedures, with mortality rates reported between 0.02% and 0.1% [1] but with bleeding complications reported to be between 7.4% and 25% [4]. It is important to assure that coagulation parameters are normal or have been corrected, and that blood pressure is maintained within normal limits to decrease the risks of the procedure.

The most frequent complication of biopsies is bleeding, with asymptomatic hematoma being the most common finding [3]. Macroscopic hematuria can occur in up to 5–7% of the cases [1] and is usually managed in a conservative manner.

Arteriovenous (AV) fistulas and pseudoaneurysm are other potential complications of a biopsy. The majority of these lesions is clinically silent and resolves spontaneously [1]. Rarely angiography and embolization is required [5].

Hydronephrosis has been considered by some authors a contraindication for renal biopsy [6]. In cases with dilatation of the collecting system the decision to perform a biopsy should be taken in a case-by-case basis and obstruction should always be excluded as the cause of the renal dysfunction. In cases of hydronephrosis there may be a higher risk of gross hematuria and the potential of a urinary leak, which may require a surgical procedure.

Injury to abdominal organs related to biopsies (e.g. bowel) is rarely seen in these days due to the routine use of image guidance.

The management of a severe bleeding which requires an angiogram and embolization is described later in this chapter.

- 3 It is frequent to see bleeding from the biopsy tract in real time color Doppler US just after the biopsy needle is removed. Small to moderate perinephric hematomas are common and most of the time are self-limited. Routinely we perform a hemoglobin level check 6 hours after the procedure to exclude significant bleeding, keeping our patients under careful observation and bed rest during this time. In cases of suspected bleeding, with a hemodynamically stable patient, an abdominal US is performed to assess the site of the bleeding, size of the hematoma and the potential for associated lesions (AV fistula, pseudoaneurysm). If the bleeding persists or if the patient is hemodynamically unstable, there is then an indication for an angiography and embolization. Surgical nephrectomy should be considered the last treatment resource.
- 4 The steps of an angiography and embolization in the post-biopsy bleed are as follows. Access is obtained into the common femoral artery with a vascular access needle (using sonographic guidance or clinical landmarks). A vascular sheath is placed and a diagnostic catheter along with a guide wire is advanced into the abdominal aorta. An abdominal angiogram is performed to define the anatomy of the abdominal vessels. After deciding the vessel of interest, the diagnostic catheter is advanced into it. Selective angiograms are performed to demonstrate the area of bleeding, which can be seen as a localized puddle of contrast material or as an area of active contrast extravasation. If the bleeding is in a small vessel, a micro catheter can be used to get as close as possible to the bleeding site. In cases of post biopsy bleed, coils are usually the first choice of embolization material. In cases of tumoral or postsurgical bleed, pseudoaneurysm (PSA) or AV fistula, a combination of embolization materials can be used (including coils, gelfoam slurry, cyanoacrylate). The embolization materials are selected and used according to the operator preference and experience.

Clinical problem/case 15.2

A 4-year-old girl, previously healthy, presented with history of a palpable abdominal mass. A CT of the abdomen showed a large left renal mass with invasion of the left renal vein. The patient was scheduled for resection of a suspected Wilms tumor after one cycle of chemotherapy. A pre-surgical embolization of the kidney was performed

(Figure 15.4) and the surgery was successful with a minimal amount of blood loss. One week after the procedure the patient presented with a pulsatile palpable mass in the right groin (Figure 15.5). Due to its size, it was successfully treated with an US-guided thrombin injection (Figure 15.6).



(a)



(b)

Figure 15.4 (a) Selective angiogram of the left kidney showing paucity and abnormal architecture of arteries in the upper pole, secondary to a large mass. (b). Post-embolization angiogram shows successful embolization of the entire left kidney. A combination of polyvinyl alcohol particles and coils was used.



Figure 15.5 Sonographic examination of a palpable mass in the right groin. It shows an anechoic rounded structure, located anterior to the femoral artery with an internal swirling pattern of blood flow in the color Doppler study. These findings are typical for a PSA.

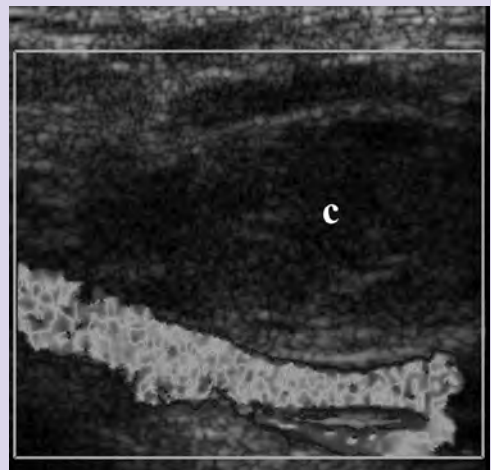


Figure 15.6 Right groin PSA after successful treatment with US-guided thrombin injection. The lumen of the PSA is entirely filled with blood clot (c) and there is no internal flow in the color Doppler study.

- 1 Is a pre-surgical embolization of the left kidney useful in this case?
- 2 What are the potential complications of angiography?
- 3 What is the most likely etiology of the pulsatile mass at the puncture site? And what is the preferred management?

Discussion

- 1 Embolization of renal tumors prior to surgery is one of the areas in which the collaboration between the interventional radiologist and the oncologist and surgeon is encouraged. Clearly in most suspected Wilms tumor patients, biopsy, at least in North America, is not done as it mandates upstaging the patient and hence necessitates

more toxic therapy. Renal or abdominal tumors with prominent blood supply or invasion of vascular structures may benefit from this intervention. The placement of a central venous line for chemotherapy can be done concurrently when the histopathological diagnosis of the tumor has been verified. The angiographic technique is the same described in the previous case. The decision of the extension of the embolization should be taken together by the interventional radiologist and surgeon, based on the surgical plan. The embolization material used depends on the preferences and experience of the operator. The author's preference is a combination polyvinyl alcohol (PVA) particles followed by coils in the main arterial branches. If a branch cannot be embolized, the angiographic study allows a clearer road map for the surgeon.

- 2 The published complication rate of angiography in children has been as high as 7–10% [7]. However, the improvement of vascular sheaths and catheters has made this procedure safer and possible to be performed even in neonates. The safest approach is using the common femoral artery [8]. IV heparin (75–100 U/kg) is usually given after obtaining femoral access to prevent thrombosis [7]. In our center, in acute bleeding and pre-surgical embolization we routinely do not use heparin; however, we minimize the procedural time. Coagulation parameters and renal function should be checked prior to an angiography and our maximum dose of contrast is 6 ml/kg.

Arterial spasm is a potential complication of angiography and can be treated in a conservative manner or using focal catheter-directed nitroglycerin injection (1 microgram/kg) [7]. Arterial dissection can be man-

aged with angioplasty and arterial perforations with balloon tamponade and/or placement of a stent [7]. Air embolism should be prevented, always keeping the catheters and sheaths isolated from the external environment. Local complications at the puncture site include: hematoma, pseudoaneurysm, AV fistula and nerve injury. A pseudoaneurysm is a false arterial aneurysm, with turbulent flow inside, contained by a single layer of fibrous tissue, usually secondary to arterial wall disruption. An AV fistula is an abnormal communication between arterial and venous vessel.

- 3 The most likely cause of this pulsatile mass is a femoral pseudoaneurysm, which can be diagnosed with an US of the area. The most important etiologic factor described is incomplete hemostasis. The management has evolved with time, being initially surgical. The treatment of choice is now a minimally invasive approach. The estimated incidence of PSAs ranges from 1% to 3% in diagnostic angiograms [9] and up to 7.6% in endovascular interventions requiring large sheaths [10]. Some authors had suggested expectant management in small lesions (1.8–3 cm) [11,12]. We had used this approach in small lesions and with early signs of thrombosis, obtaining acceptable results. If the expectant management is not successful or the lesion grows, US-guided compression can be performed. The success of this method depends on the cooperation of the patient and persistence of the operator. If this fails, US-guided thrombin injection into the PSA sac is the treatment of choice [13]. The thrombin is injected within the sac, as far as possible from the PSA neck. In our experience, this is a safe and effective treatment of femoral PSAs in pediatric patients.

Clinical problem/case 15.3

A 5-year-old boy presents with history of left flank pain and fever. Ultrasound of the abdomen showed a moderate left hydronephrosis and dilatation of the proximal ureter. An echogenic structure was seen within the ureteral lumen compatible with a ureteric stone (Figure 15.7). A combined approach was attempted by the pediatric urologist and the interventional radiologist. Cystoscopy initially was performed and attempts to advance a guide wire resulted in extravasation of contrast due to mucosal/wall injury of the ureter (Figure 15.8). A nephrostomy catheter was placed, showing a filling

defect in the proximal ureter consistent with a stone (Figure 15.9). After the infection subsided, the stone was surgically removed, a JJ stent was placed, and the nephrostomy was removed.

- 1 What is the best therapeutic option for this patient with an obstructive hydronephrosis in whom it is technically not possible to advance a JJ stent in a retrograde fashion?
- 2 How is a nephrostomy tube insertion performed?
- 3 What are the potential complications of a nephrostomy tube?

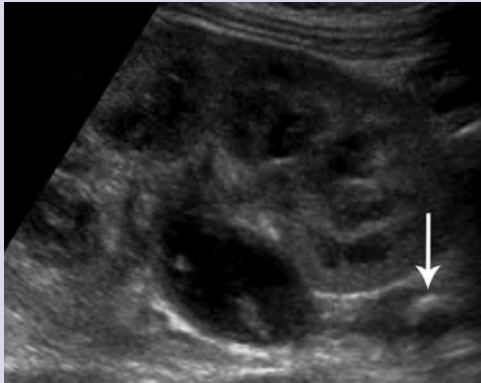


Figure 15.7 Longitudinal sonographic view of the left kidney showing a moderate hydronephrosis associated with dilatation of the ureter. There is an echogenic structure in the lumen of the proximal ureter highly suggestive of a stone (arrow).

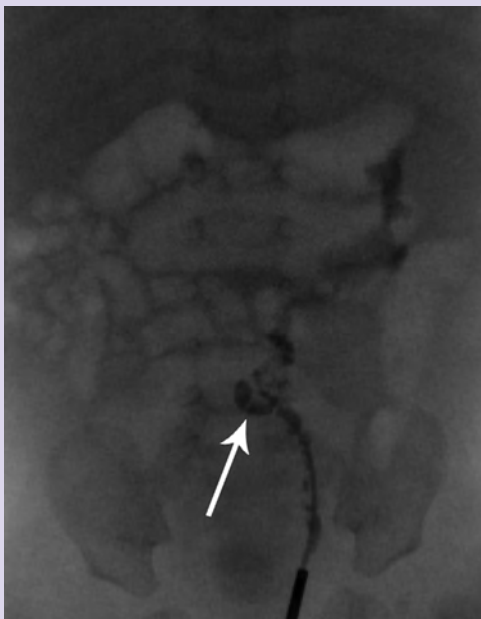


Figure 15.8 Retrograde injection of contrast into the left ureter showing extravasation of contrast material (arrow).

Discussion

- 1 This patient presents with an obstructive uropathy and urosepsis requiring emergency intervention. In this instance, an image-guided percutaneous nephrostomy is a safe and effective way to treat this emergency [14]. Dissection of the uroepithelium of the renal pelvis or ureter/ureteric wall damage is a known potential complication of the use of a guide wire whilst performing



Figure 15.9 Contrast study through a recently-placed left nephrostomy tube. It shows a filling defect in the proximal ureter compatible with a stone.

a percutaneous nephrostomy or ureteric endoprosthesis [14,15]. As soon as the infection subsides and time has been allowed for healing of the ureteric wall, an attempt to advance a JJ stent in an antegrade or retrograde fashion can be made. In this case, the nephrostomy tube is indicated to treat the infection and partially divert the ureteric urine flow in an attempt to minimize the potential urinary leak from the area of mucosal/wall injury.

The most frequent ureteric interventions performed by the interventional radiologist are antegrade JJ stent insertions and ureteric stenosis dilatations. The placement of an antegrade JJ stent requires a nephrostomy to be present. The author prefers to do this procedure when the nephrostomy tract is mature (after 2–6 weeks), to decrease the risk of losing access to the collecting system. A sheath is placed in the nephrostomy tract and a guide wire is manipulated using guiding catheters from the renal pelvis to the urinary bladder. After this is achieved, the JJ stent is advanced over the wire and left in place. The nephrostomy catheter is reinserted and it is clamped for 24 or 48 hours to make sure that the JJ stent is serving its purpose, before the nephrostomy is removed.

In cases of ureteric stricture, with a congenital, acquired or postsurgical etiology, a balloon dilatation can be performed. A guide wire needs to be advanced across the area of stricture in an antegrade or retrograde fashion. A contrast study should be performed to assess the diameter of the ureter. An appropriate balloon is selected and it is advanced into the area of stenosis and repeatedly inflated as many times as needed in order

to dilate the narrowed segment. The procedure can be repeated as many times as required [16,17].

- 2 The procedure is performed with the patient in the prone position. Deep sedation or general anesthesia is utilized. Using a sterile technique, access is obtained into the collecting system using sonographic guidance. Different types of access needles have been described. The author usually utilizes an 18 gauge needle, advancing a 0.035 in guide wire into the renal pelvis or proximal ureter. A subcostal approach is preferred and in the avascular plane between the anterior and posterior division of the main renal artery, if possible (plane of Brodel) [14,18]. The tract is dilated, a multipurpose draining catheter is advanced into the renal pelvis and the guide wire is removed. The position and function of the tube is tested using contrast. A trocar technique can be utilized in the case of a small renal pelvis.
- 3 A nephrostomy tube insertion can be an extremely difficult procedure to perform in neonates and infants due to the small renal size and the kidney movement during the procedure. Bleeding is a known complication of

nephrostomy, with self-limited hematuria a frequent finding. Severe bleeding requiring blood transfusions $\pm$ angiography and embolization have been described in 1–3% of patients [19]. Septic complications should be prevented by the use of prophylactic antibiotics, prompt decompression of the collecting system, minimal manipulation of guide wires and catheters and rational use of contrast [14,18]. Pneumothorax is a potential complication if an intercostal approach is used and accidental puncture of adjacent structures (e.g. colon) had been described [14,18].

Leakage of urine around the catheter can be seen in the postoperative phase. If it is significant, a contrast study is required to ensure that the catheter is in a satisfactory position and patent. If it is in a good location and the leakage persists, the catheter can be exchanged over a wire for a bigger one to improve the drainage.

If signs of infection at the nephrostomy tube site are identified, local care with frequent dressing changes and appropriate antibiotic therapy (when indicated), is suggested.

Clinical problem/case 15.4

A 15-month-old boy on hemodialysis due to end-stage renal disease is referred for evaluation due to problems with his central venous dialysis line that had recently been placed. During routine hemodialysis, the responsible nurse was able to infuse normal saline but unable to withdraw blood from one of the central venous catheter lumens. A radiograph of the chest was obtained showing that the tip was in an adequate position (superior vena cava/right atrial junction). A linogram was requested (Figure 15.10).

- 1 What are the findings of the linogram and what is the management?
- 2 Two months after the function of the line was reestablished, the patient presents with high fever and redness in the right chest. How is a central venous line (CVL) infection diagnosed? And what is the management?
- 3 What are other potential complications of a CVL insertion?

Discussion

- 1 The linogram shows a filling defect in the tip of the catheter and retrograde flow of contrast around the distal portion of the line. These findings are compatible with a tip clot associated with a fibrin sheath which explains the line malfunction (one-way valve mechanism). Fibrin sheath formation has a reported incidence between 13% and 57% in hemodialysis



Figure 15.10 Linogram of one of the lumens of a hemodialysis line. There is a filling defect in the distal portion of the line (white arrow) and retrograde flow of contrast around the distal portion of the catheter (black arrow).

catheters [20] and represents the most common cause of line malfunction.

If a CVL is not working adequately, the first step is to perform a chest radiograph to assess the location of the tip. If the tip is in a good position, tissue plasminogen activator (tPA) is used in our institution (with a dose and

protocol according to our hospital guideline), as tip clots and fibrin sheaths can respond to fibrinolytic therapy. If this does not work, re-establishing catheter function can be attempted with the use of a guide wire.

If the line shows partial function acting such as in this case, where infusion only is possible due to one-way valve mechanism, a linogram frequently shows a fibrin sheath. The therapeutic options include thrombolytic infusion through the line, fibrin stripping, use of an angioplasty balloon to disrupt the fibrin sheath, catheter exchange or line replacement [21].

- 2 CVL infection is a common and potentially serious complication. The incidence varies according to the type of the device installed, being the lowest for implanted ports at 0.1 per 1000 catheter days, tunneled CVL at 1.6 per 1000 catheter days and the highest incidence is in non-tunneled lines at 2.7 per 1000 catheter days [22]. Clinical symptoms range from low-level fevers to septic shock. It is important to differentiate exit site (local) infections to systemic catheter infections. Site infection usually presents with redness, pain and discharge at the site. Findings consistent with a cellulitis can be seen in severe cases. An exit site culture needs to be obtained, and appropriate antibiotics should be started and daily change of dressing performed. These superficial infections usually respond well to this management.

If a systemic infection of the line is suspected, blood cultures are obtained from the catheter and from a peripheral vein. In our institution we diagnose a catheter-

related bloodstream infection if the culture from the line is positive and the peripheral is negative or if both are positive and other sources of infection are excluded. Antibiotics are started and it must then be determined as to whether the catheter needs to be removed, according to the pathogen isolated from the cultures. In cases of aggressive organisms, we keep the patient without a line for at least 48 hours and, when possible, we perform blood cultures prior to the new catheter insertion to document that they are negative. Patients with end-stage renal disease are dialyzed just before the removal of the infected line and as soon as the new catheter is placed. Antibiotic covered lines, antibiotic lock solutions and topical application of antimicrobials had been used to prevent line infections [21].

- 3 Arrhythmias are a potential complication of central line insertion. They can be stimulated by the use of a guide wire or by the line itself. The suggested position of the tip of a line is at the superior vena cava/right atrial junction; however, lines in good position can also be causative for arrhythmias. Most of the time, the arrhythmias are asymptomatic; however, in some cases a pharmacological intervention is required (e.g. use of adenosine to control a supra-ventricular tachycardia). If the arrhythmia is persistent, adjustment of the position of the catheter tip is often required and occasionally the line needs to be removed [21].

Blood vessel or heart damage, nerve palsy, catheter fracture and air embolism are also potential complications of CVL insertions, but are infrequent.

DOS AND DON'TS

Do

- Carry out a careful ultrasound assessment prior to performing a biopsy. Choose a site with a low density of blood vessels and, as far as possible, no vital structures (e.g. main renal artery)
- Gently manipulate the guide wires inside the ureter and renal pelvis and choose guide wires with floppy tip. This may prevent perforations
- Carry out a detailed assessment of a central venous line that is not working properly and be familiar with various options in solving the problems that are frequently encountered (e.g. fibrin sheath)

Don't

- Perform a biopsy if the blood coagulation parameters and blood pressure are not optimal
- Over-distend or perform excessive manipulation of an infected and obstructed collecting system. After a nephrostomy is placed, a diagnostic contrast study can be performed when the infection has subsided
- Remove a central venous line due to presumed diagnosis as many infections and other reasons for malfunction can be precisely diagnosed and treated. Once a line is removed, that vein may not be able to be used again

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CHAPTER 16

Minimally invasive interventions for stone disease

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KEY POINTS

- The incidence of stones in children varies enormously geographically and is approximately 5–20 per 100,000 children aged under 18 years
- The pronounced incidence of underlying metabolic abnormalities results in a high rate of recurrence; diagnosing causative factors is essential
- Ultrasound is the first choice imaging modality. Urine microscopy and culture is mandatory before any treatment
- Treatment options include ESWL (extracorporeal shock wave lithotripsy), URS (ureterorenoscopy, if extending into the pelvicalyceal system also referred to as RIRS or retrograde intrarenal surgery), PCNL (percutaneous nephrolithotomy), PCCL (percutaneous cysto-lithotomy), laparoscopic surgery and open surgery
- Choice of treatment modality depends on stone burden, composition, location and size, symptoms and complications, as well as patient factors

CLINICAL PROBLEM/CASE

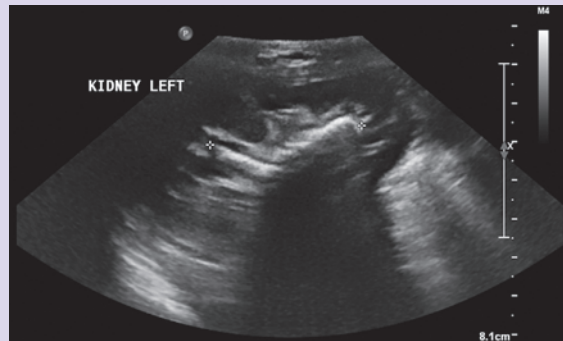
- 1 What further investigations are necessary?
- 2 What would be the most appropriate management?

Case 16.1

A 4-year-old girl has recurrent urinary tract infections. The plain film (a) and renal ultrasound scan (b) show a stag-horn calculus of the left kidney (see following images).



(a)

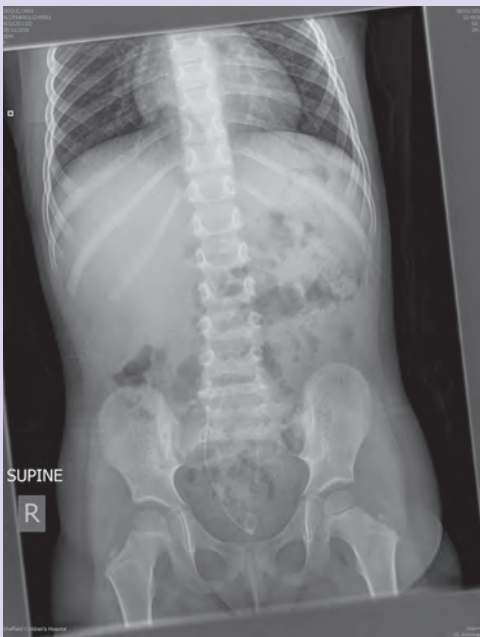


(b)

Case 16.2

A 2-year-old girl presents with hematuria and colicky right-sided abdominal pain. A plain film (a) and ultrasound (b) in the referring hospital shows a distal right ureteric stone 1 cm in length with minimal proximal dilatation.

- 1 What are the initial management options?
- 2 What would be your technique of choice for managing this stone?



(a)



(b)

Introduction

Stone disease in children, as in adults, shows marked geographical variation. The incidence of pediatric urolithiasis has been documented as 5 per 100,000 children in Iceland, almost 20 per 100,000 children in South Carolina in the USA and is endemic, for instance, in Turkey and Pakistan [3–5]. Interestingly, pediatric stone disease seems to be growing; indeed, in South Carolina the incidence of stones in children younger than 18 years appears to have doubled from 1996 to 2007, attributed to obesity and dietary factors [3]. While boys were previously found to be more commonly affected [6], an increase in urolithiasis particularly in adolescent girls, is thought to be responsible for an emerging female preponderance [3].

A key difference between adult and pediatric stone disease is its recurrent nature. Identifying any underlying metabolic abnormality, reported in 33–93% in the literature [3], is therefore essential. 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%) [6]. Infection is the second most common etiology.

In children, ultrasound can give exquisitely detailed information on the size, location and extent of stones. X-ray increases the detection rate particularly of ureteric stones. Low dose non-contrast computed tomography (CT) is occasionally indicated but intravenous urogram (IVU) is rarely needed.

Minimally invasive treatment of pediatric stones has advanced rapidly over the last few decades with open surgery now rarely used [7]. The minimally invasive techniques most commonly employed are extracorporeal shock wave lithotripsy (ESWL), percutaneous nephrolithotomy (PCNL), ureterorenoscopy (URS), percutaneous cystolithotomy (PCCL), cystoscopy and laparoscopy. The aim of any intervention is to render the child stone-free, whilst minimizing the impact of the procedure(s) on the child. Complications relate mostly to infection, bleeding, iatrogenic injury, urine leak, pain, obstruction, recurrence of stones and loss of renal function. Children with complex co-morbidities – such as, for instance, cerebral palsy and immobility, or gastro-intestinal failure – are at increased risk of stone formation and can pose additional management challenges in view of, for example, cardio-respiratory compromise and skeletal deformities. Choice of treatment

modality therefore depends on stone burden, composition, location and size, symptoms and complications, as well as patient factors.

Infection is one of the most important complications of any form of stone treatment and can lead to overwhelming sepsis or even death. Meticulous pre-operative checking of the urine for bacteria and appropriate timely antibiotic treatment will help reduce this potentially lethal complication. In a study by Coward et al., infection was identified in over 50% of children with stone disease, including in those who were subsequently diagnosed with a metabolic abnormality [6]. 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, and hence the term “insignificant residual fragment” is a misnomer in this situation. Indeed, one third of small residual fragments will regrow over a period of 1–4 years [8,9].

The current recommendations by the European Society for Pediatric Urology (ESPU) and European Association of Urology (EAU) in 2012 for the interventional management of stone disease are given in Tables 16.1 and 16.2.

Extracorporeal shock wave lithotripsy

This technique was first performed in 1980. Pressure waves shatter the stones into smaller fragments which can subsequently pass spontaneously. ESWL is widely used with good success (overall successful in <80%), especially for children with low stone burden [10,11]. It is the treatment of choice for single renal pelvic stones up to 20mm, lower-pole stones up to 10 mm and proximal ureteric stones [1,2,12–14]. A recent study with follow-up extending to over 10 years has negated previous concerns regarding its impact on immature organs, showing no effect on renal growth or on the incidence of hypertension and diabetes [15].

Stone clearance rates are dependent on stone size and composition [16]. Where a CT scan has been performed, Hounsfield units (HU) of density have been shown to be an independent predictive factor of clearance with stones <600 HU having a success rate of 80% compared to 20% for stones of >600 HU [17]. Cystine stones, calcium oxalate monohydrate and calcium phosphate dihydrate

Table 16.1 European Society for Pediatric Urology (ESPU) 2012 recommendations for interventional management of pediatric stones.

Stone size and location (excl. cysteine and uric acid stones)	Primary treatment option	Level of evidence	Secondary treatment options	Comment
Staghorn stones	PCNL	2b	Open/ESWL	Multiple sessions and accesses with PCNL may be needed, combination with SWL may be useful
Pelvis <10 mm	ESWL	1a	RIRS/PCNL	Multiple sessions of ESWL may be needed, PCNL has a similar recommendation grade
Pelvis 10–20 mm	ESWL	2b	PCNL/open	
Pelvis >20 mm	PCNL	2b	ESWL/open	Multiple sessions of ESWL may be needed
Lower-pole calyx <10 mm	ESWL	2c	RIRS/PCNL	Anatomical variations are important for complete clearance
Lower-pole calyx >10 mm	PCNL	2b	ESWL	Anatomical variations are important for complete clearance
Upper ureteric stones	ESWL	2b	PCNL/URS/open	Additional intervention need is high with ESWL
Lower ureteric stones	URS	1a	ESWL/open	
Bladder stones	Endoscopic	2b	Open	Open is easier with less operative time for larger stones

From Tekgul et al. 2012 [1]. Reproduced with permission of EAU.

ESWL, extracorporeal shock wave lithotripsy; PCNL: percutaneous nephrolithotomy; RIRS, retrograde intrarenal surgery; URS, ureterorenoscopy.

Table 16.2 European Association of Urology (EAU) 2012 guidelines for treatment of stones of different sizes and in different locations.

Stone size and location	Primary treatment option	Secondary treatment options	Comment
Renal <10 mm	ESWL	FlexiURS, PCNL	Spontaneous passage of fragments is more likely in children, hence ESWL can be used for stones up to 20 mm in the renal pelvis
Renal 10–20 mm	ESWL	FlexiURS or PCNL	
Renal >20 mm	PCNL	ESWL, flexiURS, open/laparoscopic	
Proximal ureter <10 mm	ESWL or URS	PCNL	
Proximal ureter >10 mm	URS or ESWL		
Distal ureter <10 mm	URS or ESWL		
Distal ureter >10 mm	URS		
Bladder stones	Endoscopic		
		PCCL/open	

Immediate complications: urinary tract infection/sepsis, bleeding, iatrogenic injury, urine leak, pain/colic.

Late complications: residual fragments/recurrence, obstruction due to stone or stricture, loss of renal function.

From Turk et al. 2013 [2]. Reproduced with permission of EAU.

ESWL, extracorporeal shock wave lithotripsy; PCNL, percutaneous nephrolithotomy; RIRS, retrograde intrarenal surgery; URS, ureterorenoscopy.

show poorer response to ESWL. In contrast to adults, no significant difference in stone clearance following ESWL has been observed for different stone locations, including the lower-pole calyces in children [11,19].

Whilst older children can tolerate ESWL under the cover of simple analgesia, young children usually require general anesthesia: the depth of the kidney from the skin is typically only a few centimeters and hence the skin area for shock wave entry (coupling) is very limited, about the size of a coin, making it much more painful. The child needs to lie still for the duration of the treatment, around 30 minutes, to enable the shock waves to be focused consistently onto the stone avoiding neighboring lung and intestine.

A large review by D'Addessi et al. [16] totaled complications in 336 of 1866 patients (18%). The most frequent complications were hematuria which occurred in 150 (8%), colic in 57 (3%), infection with or without fever in 41 (2%), and obstructing ureteric calculi (also known as Steinstrasse, stone street) in 50 (2.7%).

Hematuria

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

Some pain may occur in a high proportion of patients although colic is reported in less than 20%. In most, pain control is achieved by oral analgesia and antispasmodics. Persistent pain should trigger further investigation to exclude steinstrasse obstruction.

Infection

Urine should be tested preoperatively and bacteruria appropriately treated prior to ESWL. Stone fragmentation will release bacteria where these are contained within the stone. Particularly in those with UTI, or a history thereof, antibiotic cover for ESWL is advised. Postoperative fever should be investigated and appropriate antibiotics commenced based on previous sensitivities and local protocols.

Steinstrasse (stone street)

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. Incidence has been reported to be between

1–17%. [18,19]. JJ stents may be inserted prior to treatment in cases of large stones to prevent this complication. If the condition does not resolve conservatively, JJ stenting or ureteroscopy is required to relieve the obstruction and 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 KUB X-ray following ESWL (Figure 16.1a).



Figure 16.1 Complications following ESWL. (a) KUB X-ray showing (right) steinstrasse (stone street) distal ureteric obstruction in a bladder exstrophy patient. (b) Axial ultrasound view of right kidney demonstrating a subcapsular hematoma.

Bruising/ecchymosis and haematoma

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 [20]. Occasionally an intrarenal, subcapsular or perirenal hematoma may occur [21] (Figure 16.1b). This is usually self-resolving without any lasting effects.

Injury to other viscera

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.

The authors have once observed pancreatitis and hepatic transaminitis in an infant with severe comorbidities after ESWL. The effects on the kidney, liver and pancreas were transient and resolved spontaneously but necessitated supportive care for a period of 10 days.

Uretero(reno)scopy

URS is the first line treatment for distal ureteric stones and a popular treatment modality for proximal ureteric stones in children. Approximately 90% stone-free rates at first treatment can be achieved. The complication rates have reduced significantly with the development of newer and smaller instruments affording better visualization. The Turkish Pediatric Urology Society recently reported their combined experience in a retrospective multi-institutional study, documenting complications in just 8.4%, most low-grade and self-limiting [22]. Of note, prolonged operative time was an independent predictor of complications.

The technical principles of ureteroscopy are the same as for adults: it is recommended that a safety wire is used at all times so that if an intraoperative 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. 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.

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 significant ureteral injury [23]. A recent study comparing complication rates of ureteroscopy with or without access sheath in children documented a higher intraoperative complication rate with the use of an access sheath (15% vs. 2%, $P = 0.02$, including perforation/extravasation, submucosal wire and stent migration) although there were no long-term sequelae [24]. Post-procedural JJ stenting is not always required [25].

Technological advances are continuing at a rapid pace. Today small flexible ureteroscopes allow access to the pelvicalyceal system for retrograde intrarenal surgery (RIRS), although at maximal deflection stone baskets and laser fibers cannot always be deployed, so that stones in the lowermost calyces frequently remain inaccessible for retrograde endoscopic interventions.

Hematuria

Postoperative hematuria is common (27%) but usually resolves in a matter of hours with conservative management [26]. Profuse hematuria is rare (<1%) and requires prompt evaluation [22].

Infection

As for any stone intervention, preoperative checking for and treatment of bacteruria is mandatory. Despite this and the use of antibiotic prophylaxis at induction, UTI, pyelonephritis or septicemia do occur following ureteroscopy in 3–19% [22,26].

Failure of access

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 edema have improved, a second ureteroscopy allows the stone to be treated. Using this approach, the need to convert to an open ureterolithotomy is rare [22].

Other series, however, continue to report open conversion as a result of failed access, as a result of vesicoureteric junction edema and stone impaction, in as many as 10–20% [26,27].

Ureteric injury/extravasation

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 guide wire 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. Ureteric perforation occurs in about 1–2% in most large series [22,26]. In this situation, the procedure must be terminated and a JJ stent placed. The procedure can 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. Ureteric avulsion has not been reported in the pediatric literature but if it does occur necessitates open or laparoscopic repair.

Stricture

Stricture formation is rare and may be a consequence of stone impaction, its transureteric erosion or displacement, or iatrogenic injury (Figure 16.2). Post-procedural ureterovesical junction obstruction necessitating ureteric re-implantation is observed in 0–2% [22,27,28]. It has

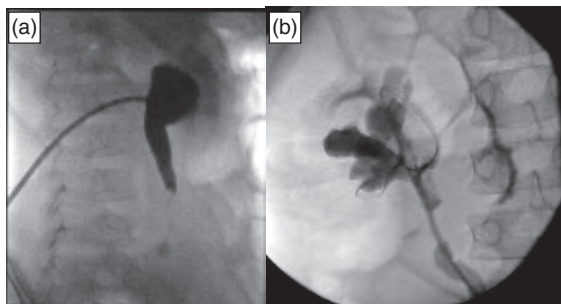


Figure 16.2 Ureteric strictures resulting from stone impaction. (a) Nephrostogram demonstrating a 3 cm mid-ureteric stricture and an impacted calculus at its distal aspect, just above the pelvic brim. (b) Retrograde study and stent insertion across a 1.5 cm proximal ureteric stricture. The stone has been dislodged back into the renal pelvis.

been postulated that this relates to active dilatation of the ureterovesical orifice and can be avoided by a period of “passive” dilation using a JJ stent [29].

Vesicoureteral reflux

VUR following ureteroscopy has been reported in the literature in as many as 17% using 8.5/9.5/11.5F ureteroscopes [30]. But this is transient, low-grade and clinically insignificant. Postoperative cystography is not required.

Stone retropulsion

This is a nuisance and occurs in 1–2% even in high-volume centers [22]. Laser lithotripsy is associated with less retropulsion than pneumatic or electrohydraulic lithotripsy. Various devices – such as for instance stone cones and parachutes, specialized 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 [31]. If stone fragments are lost into the kidney, or indeed the whole stone, then flexible URS may be utilized 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 fiber passed alongside the stone and the fragments fragmented using the laser fiber. 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 vision is obscured by bleeding and a safety wire is in place then a stent may be placed and the procedure halted.

Stent migration

The stent may migrate into the ureter/renal pelvis or indeed fall into the bladder and voided out per urethra. This occurs in approximately 1% [28] and usually relates to inappropriate size of stent, inaccurate placement, or a markedly dilated ureter.

Forgotten stent

Timely removal of a stent is paramount, and a register of patients with a JJ stent may help avert disaster.



Figure 16.3 KUB X-ray demonstrating a severely encrusted right JJ stent and bilateral renal stones in a cerebral palsy patient.

Forgotten stents may be the cause of severe encrustation, recurrent stone formation and UTIs, renal deterioration and litigation (Figure 16.3). 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-renaloscopy, to remove encrustation to enable safe stent extraction [32].

Percutaneous nephrolithotomy

Percutaneous nephrolithotomy (PCNL) was first described by Fernstrom and Johannsen in 1976 [33]. Almost 10 years later, Woodside et al. [34] reported the first pediatric series of seven children aged 5 to 18 years. Today, PCNL has been shown to be equally effective in preschool children, and age or weight is no longer a limitation [35]. On the contrary, minimal access techniques are preferable to open surgery in children in view of the recurrent nature of kidney stones and high risk of requiring multiple future treatments. PCNL is the first-line treatment modality for Staghorn stones, renal stones >20 mm, lower-pole stones >10 mm, cystine or struvite stones [1]. Where residual stones persist after

PCNL, these can be cleared subsequently by ESWL, second-look PCNL and/or uretero-renaloscopy [35–38].

The Turkish Pediatric Urology Society reported their combined retrospective experience of PCNL in 1157 children (1205 renal units) in 2013 [39]. It is the largest multi-center pediatric PCNL series to date: complete stone-free rates of 82% were observed in a single PCNL session in 96%. Complications occurred in 28%; bleeding 11%; requiring transfusion 10%; fever 12%; and sepsis 2% (including one death, 0.1%); renal pelvic laceration/extravasation 1%; pleural injury 1%; bowel injury 0.3%; and stricturing 0.2%. Various surgical and stone factors have been associated with a higher incidence of complications, for instance, positive preoperative urine culture, stone burden and location, number, site and size of tract [39]. At least in part, some of the associations may simply reflect a greater complexity of cases rather than a direct cause of complications, such as pre-operative CT scan, previous urolithiasis, operative and hospitalization time, and residual stone after PCNL. Also, the literature is contradictory on the importance of, for example, the size and number of PCNL tracts and size of instruments on adverse events [38–41]. Nevertheless, specialized pediatric nephroscopes with a working channel able to accommodate 5F instruments are widely available, enabling smaller tracts. Additionally, flexible scopes can be deployed via the tract to reach stones in neighboring calyces without torquing the kidney.

Hemorrhage

Blood should be checked for clotting disorders and grouped prior to PCNL. Bleeding requiring transfusion is reported in 0.4–24% [35–39,42,43]. Bleeding can result from the tract, or from undue manipulation of the sheath and scope causing torque of the kidney. Indeed, if the stone is inaccessible, an additional tract should be made rather than distort the kidney by zealous maneuvering the scope onto the stone. Paradoxically, tract size and number have been both associated and not associated with bleeding and the need for transfusion [38–41]. 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 tract and the procedure discontinued. The same tract may be utilized at another time for a re-look procedure [36]. 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. Thereafter, hemostatic sealing agents can be injected into the tract. Angiography and selective embolization or open surgery are rarely required.

Late bleeding after PCNL, typically after one week, may indicate a vascular injury [44]: an intrarenal pseudoaneurysm or arteriovenous fistula may rarely develop after PCNL and is managed by angiography and selective embolization.

Fever

Transient fever following PCNL is common (12–15%) and often resolves spontaneously [37,39,43]. However, fever may also indicate life-threatening urosepsis or dislodgment of the nephrostomy tube and a perinephric collection [37,42,45].

Infection

Up to 65% of patients requiring PCNL have positive urine cultures [45]. Treatment of positive urine cultures prior to PCNL and antibiotic prophylaxis perioperatively is strongly advised. Despite this, postoperative UTI/urosepsis may still occur, the management required ranging from parenteral antibiotics [38] to intensive care support [39,45]. The surgeon must have the greatest respect for the dangers of urosepsis as this can be fatal; indeed, there was one death (0.1%) from sepsis and multi-organ failure in the combined Turkish experience [39].

Hypothermia

Hypothermia is a significant risk in pediatric 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 waterproof sticky drapes [46]. The length of procedure time (operative and anesthetic induction) is also relevant, with no incidence of hypothermia in cases <150 minutes in one series [47].

TUR syndrome

The risk of absorption of irrigation fluid causing hyponatremia and electrolyte imbalance can be minimized by using a low-pressure irrigation system and ensuring that warm isotonic solution (e.g. 0.9% sodium

chloride) is used. The procedure should be completed in the shortest time possible and electrolytes checked postoperatively.

Urine leak, urinoma or extravasation of fluid

Extravasation, urine leak or perinephric collections do occur following PCNL [37,39,43], and may present as postoperative fever. They usually resolve with conservative management but may need percutaneous drainage if large, especially if infected.

Hydrothorax and pleural injury

Puncture above the 11th rib significantly increases the risk of intrathoracic complications [37]. Here a postoperative chest radiograph is advised to look for complications. Most cases of hydrothorax are asymptomatic and resolve spontaneously. However, a chest drain is likely to be required for a large hydrothorax [37].

Intestinal injury

This complication is rare, particularly in the prone position and if ultrasound is used to guide the puncture as the colon is readily visualized and easily avoided. If an intraperitoneal bowel injury is suspected intraoperatively, diagnostic laparoscopy may help delineate the injury before the onset of peritonitis [39]. If the diagnosis of a retroperitoneal urinary-intestinal fistula is made post-procedure, ureteric stent placement and gradual withdrawal of the nephrostomy has been described [43].

Steinstrasse (stone street)

Stone fragments migrating down the ureter are a well-recognized 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 unrecognized, these fragments may accumulate in the distal ureter, potentially causing obstruction. Ureteroscopy and stenting may be required. Successful treatment by ESWL has been reported [45,47].

Tract scarring/calcification

This is in fact uncommon. A series of 72 PCNLs using tracts up to 30FR in children aged 9 months to 16 years observed no scars on subsequent DMSA studies [36].

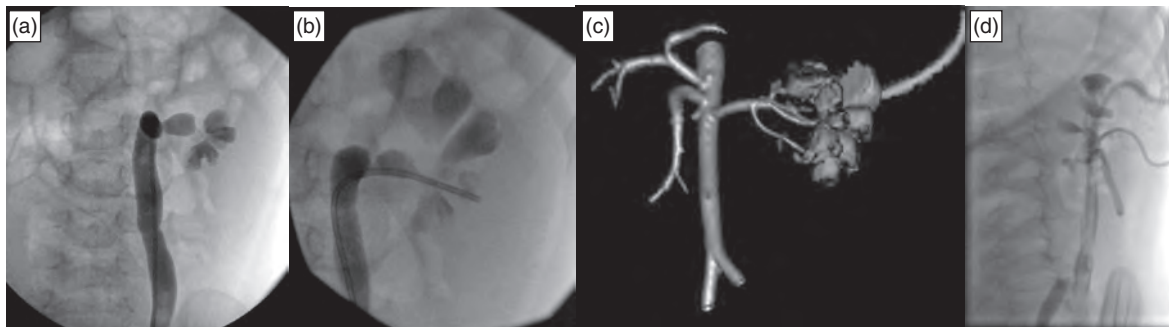


Figure 16.4 Pelvic calyceal stricture after PCNL for an obstructive staghorn calculus. (a, b) Retrograde uretero-pyelogram demonstrates the stricture extending from each of the calyceal necks to the extra-renal pelvis. (c) CT reconstruction of the arterial supply and calyceal system prior to anastomosis of the ureter to each of the calyces. (d) Nephrostogram via one of three nephrostents via the upper, interpolar and lower-pole calyces following the uretero-calyceal anastomoses.

Pelvic calyceal scarring and stricture

Many advocate antegrade pyelography to exclude obstruction prior to removal of the post-procedure nephrostomy/pigtail catheter [36,39]. Persistent urine leak via the tract can be an indicator. However, scarring of the renal pelvis or pelvic calyceal junction can occur after the immediate postoperative period. 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. Endourological approaches can be considered for a short stricture; however, extensive or persistent stricturing will require open uretero- or pelvic calyceal anastomosis [48] (Figure 16.4).

Minimally invasive cystolithotomy

Bladder stones can be dealt with in a variety of ways, including open cystolithotomy or by minimally invasive techniques either per urethra or percutaneously (PCCL) [50–52]. Endo-urological procedures afford a reduced length of stay and similar procedural time as compared to open cystolithotomy. Complication rates range from <1% to 10% [50–52] and were found to be greater for endo-urological approaches in one comparative study [50].

Infection

Fever was reported in 11% after PCCL and treated by parental antibiotics for 2–3 days [52].

Hematuria

This is rarely a significant issue. Injury to bladder mucosa due to inadvertent crushing within the jaws of the stone punch can result in bleeding which may necessitate catheterization and bladder wash-outs. In one study, gross hematuria was observed in 4% but spontaneously resolved in 24 hours [52].

Extravasation and leak

Percutaneous suprapubic puncture should be performed with a moderately full bladder so as to avoid a transperitoneal course and intestinal injury [52]. Preventing vesical overdistension will help avert extravasation and rupture. Placement of a urethral as well as a suprapubic catheter at the end of the procedure, with removal of the urethral catheter 24 hours after removal of the suprapubic catheter usually affords sufficient time for the percutaneous tract to close [51]. A recurrent or persistent suprapubic leak generally settles with placement of an appropriately sized urethral catheter over a matter of days. In contrast, intraperitoneal leaks may require drainage and operative repair [50,51].

Urethral stricture

Repeated instrumentation of the small caliber pediatric urethra, especially where the stone size is large, can lead to stricture formation. Optical urethrotomy and intermittent dilatation have been described [50], but if the stricture persists a formal repair is indicated.

Laparoscopy

Laparoscopy has a limited role in pediatric urolithiasis [53]. It is primarily indicated for nephrectomy of non-functioning moieties, concomitant correction of, for instance, 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 endo-urological procedures as an alternative to open surgery. Reported stone-free rates range from 70–100% for laparoscopic pyelolithotomy and approach 100% for laparoscopic ureterolithotomy. Operative time for laparoscopic pyelolithotomy is almost double that of PCNL in comparative studies [54,55].

Conversion to open

Even in the hands of experienced laparoscopic surgeons a need to convert to open occurs in around 12% (range 0–27%) [53–55], as a result of dense perirenal/ureteric inflammation/fibrosis, bleeding or stone migration.

Stone migration

Laparoscopic ultrasound may be needed to help locate a stone that has migrated into the calyces [54]. A stone may be flushed out using irrigation or retrieved by laparoscopic graspers or flexible nephroscopy. Where a stone remains hidden, ESWL or PCNL can be used to achieve stone clearance at a later date [53]. Care must be taken not to lose a stone in the peritoneal cavity on extraction.

Hemorrhage

Blood loss appears to be similar to PCNL [54,55].

Leak

Urine leak is primarily reported after laparoscopic ureterolithotomy. This is managed by JJ stenting with or without placement of a drain in the first instance [53].

Ureteric stricture

Strictures may result from stone impaction in the ureter or as a complication of the procedure (Figure 15.2). In an early series of laparoscopic ureterolithotomy in 14 adults by Keeley et al. [56], ureteric strictures were incised at the time of stone retrieval in three, with subsequent dilatation in two. A persistent stricture may require resection and uretero-ureterostomy [57].

Conclusion

The incidence of pediatric urinary stone disease is increasing. Advances in technology and instrumentation means that very few stones need to be managed by open surgery. Minimally invasive techniques reduce hospital stay and morbidity for the child and should be considered as the gold standard for the management of urinary stone disease in children.

DOS AND DON'TS

Do

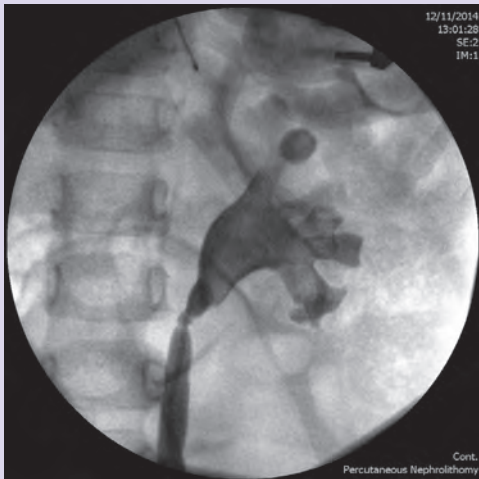
- Use a multidisciplinary team for management of urinary stone disease in children
- Ensure the same theatre team provides the support for the stone interventions to improve efficiency and safety
- Use the smallest instruments which will allow for complete stone clearance
- Ensure health and safety requirements are met when using the laser
- Use ultrasound to localize the stone where possible

Don't

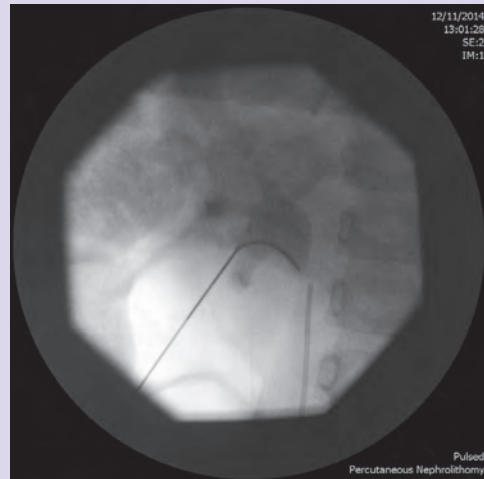
- Rush when doing any stone intervention!
- Be an occasional stone surgeon
- Use CT as first line investigation of urinary stones in children

CLINICAL PROBLEM/CASE: DISCUSSION**Case 16.1 discussion**

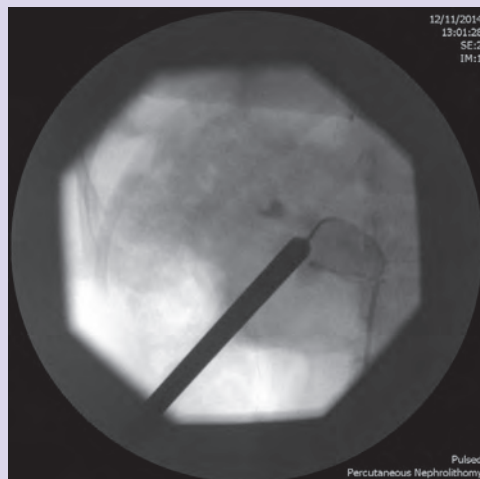
A DMSA scan is necessary to determine the baseline renal function. An IVU is recommended by some surgeons via the same cannula to allow visualization of calyceal anatomy and to plan the puncture. Considering the size of the calculus, this would best be managed by a percutaneous nephrolithotomy, preferably the mini PCNL technique. Literature suggests better outcomes with smaller sheaths (15 or 16F). The images show a preliminary retrograde ureteric catheterization and contrast study outlining the calculus, a posterior lower pole calyx puncture and the 16F sheath in position.



(a)



(b)



(c)

Case 16.2 discussion

The initial management would be to ensure the child is stable and manage pain with appropriate analgesia. Literature suggests a better outcome in terms of stone clearance and ureteric injury with initial stenting followed by ureteroscopy and laser lithotripsy. This patient had an initial stent inserted prior to referral and 6 weeks later had laser lithotripsy with complete clearance.

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CHAPTER 17

General laparoscopy

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KEY POINTS

- Laparoscopy is safe in trained surgical hands
- Attention to equipment setup and ergonomics improves task performance
- Energy sources must be fully understood and used judiciously
- Unrecognized bowel perforation is a catastrophic complication and must be considered in the septic postoperative patient

Introduction

As surgery progresses there is no doubt that the modalities of visualizing target organs are increasing whilst the trauma related to wound access is decreasing. Laparoscopy is a step in this progression that is likely to be superseded by open MRI intervention and robotic surgery. Caution is required with laparoscopy and a well-trained surgeon is essential. Laparoscopy is now widely used in pediatric surgery and urology for the removal of solid tissue (dysplastic, malignant or infected), reconstruction (i.e. pyeloplasty, orchidopexy), and diagnosis (i.e. intersex trauma). This chapter documents basic endoscopic techniques whilst focusing on the complications of laparoscopy in general.

History

Initial visualization of the abdominal cavity by papyrus was attempted by early Egyptians. Insufflation followed the introduction of the Veress needle in the 1930s and, finally, in 1982 Kurt Semm performed the first laparoscopic appendectomy. Improved equipment and the development of instrumentation enabled widespread introduction of laparoscopy in the 1990s. Laparoscopy

and endoscopic surgery are now considered routine in pediatric surgery and urology.

Procedure

Basic laparoscopy involves insertion of a primary optic trocar by open technique, usually in a transumbilical fashion, establishment of a pneumoperitoneum, visualization via a rigid telescope, and the insertion of secondary trocars for instrumentation. The diameter of the ports, telescope size and instrumentation is extremely variable and based on the availability of equipment at the institution and surgical expertise. It is inappropriate to prescribe a rigid diagnostic laparoscopy formula. Retroperitoneal or lateral access requires insertion of a balloon device to create a working space [1].

General anesthesia and full muscle relaxation is essential. The insufflation of carbon dioxide in the initial phase should generally be at 0.5 l/min. The surgeon, the target organ, and the monitor should be in line to facilitate ergonomics (Figure 17.1). Failure to position the patient correctly and orient the target organ monitor and surgeon in an ergonomic fashion results in major complications. There is no question that task performance is severely compromised by obtuse operating

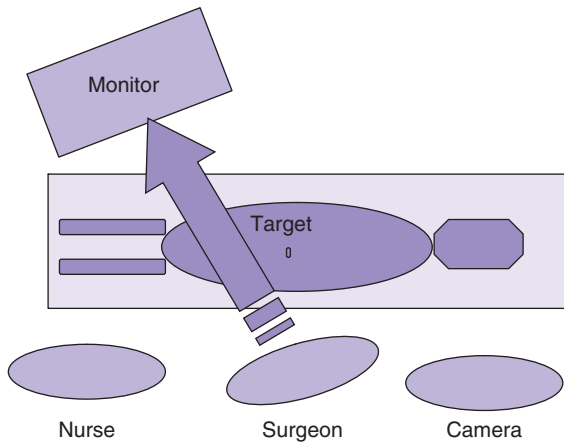


Figure 17.1 Alignment of surgeon, target organ and monitor to maximize ergonomic performance.

angles, and the failure to observe basic ergonomics is likely to result in poor task performance. These factors need to be considered prior to the commencement of any laparoscopic case.

The surgeon must be completely familiar with all associated equipment, including the insufflation machine, the camera processor, the light source and carbon dioxide. It is the surgeon's responsibility to be able to troubleshoot and rapidly identify problems with the equipment. In-depth training on each individual device is essential prior to commencement of any endoscopic surgery. Failure to achieve this level of competency can result in serious errors and can be a factor in a major complication. Familiarity with the equipment is essential [1,2].

The use of the Veress needle is contraindicated in children due to the risk of inadvertently injuring a hollow viscus such as the bowel or blood vessels. It is accepted that this risk is small; however, perforation may be unrecognized and catastrophic [3–5]. The initial port should be placed under direct vision, generally transumbilically. The exception to this is the blind balloon insufflation/port introduction of the retroperitoneal technique. Secondary instrument ports are always introduced under direct vision. Radially dilating ports (i.e. those with an expandable sheath inserted over a needle) improve port fixation and reduce CO₂ leakage. These ports are preferred for complex and difficult procedures.

Complications

Complications in laparoscopy can be summarized under the following headings:

- Port site.
- Insufflation.
- Inadvertent injury.
- Tissue approximation.
- Other patient-related complications.
- Surgeon-related complications.

Port sites

Port site herniation is a surprisingly common complication of laparoscopy in infants and children. Herniation can occur even through a 3 mm port site. To prevent this the following points are noted:

- 1 Most port site herniations occur from a midline trocar site, particularly the umbilicus. There is usually omentum entrapped within the port site, possibly exacerbated by a rush of CO₂ at the time of trocar withdrawal. All port sites should be closed if possible. Meticulous attention, particularly to the umbilical port, is essential to avoid this complication.
- 2 A port site herniation is usually identified by omentum appearing in the wound in the first 24 hours of surgery. The patient needs to be returned to the operating theater, have the omentum ligated and removed, and the port site reclosed.

Insufflation pneumoperitoneum

Carbon dioxide is a relatively inert gas and is well tolerated as a pneumoperitoneum agent. The potential complications are as follows:

- High-pressure pneumoperitoneum will lead to diaphragmatic splinting, reduced tidal volume, and retention of CO₂. Whilst end tidal carbon dioxide often remains elevated during laparoscopy, experienced anesthetic support is required to minimize this complication.
- Gas embolism is possible, particularly if an open viscus has been perforated. Fortunately, this is extremely rare in pediatric surgery. Immediate conversion to laparotomy is required if this is suspected. If gas embolism occurs, immediate insertion of a central venous catheter and aspiration of affected gas is required by the anesthetist. Gas embolism is most likely to occur by a misplaced Veress needle and, as discussed previously, this should not be used.

- Hypothermia can occur as a result of excessive CO₂ utilization. This is best avoided by utilizing an insufflation device that warms the gas, particularly in lengthy procedures.

Leakage of carbon dioxide into adjacent body spaces

Carbon dioxide pneumoperitoneum may escape into the subcutaneous space (subcutaneous emphysema), pleural space (pneumothorax), and mediastinum (pneumomediastinum).

Small pneumothoraces and leakage of air into the chest cavity can be treated conservatively and the operation can be continued. A major pneumothorax requires insertion of an intercostal drain and adequate consideration as to the cause. Subcutaneous emphysema is often a result of the gas spreading between the abdominal wall and the skin and will resolve in the next 24 hours. The pneumoperitoneum disappears within 24 hours on an erect abdominal X-ray. It is not the cause of air under the diaphragm after 24 hours and a viscus perforation should be suspected if this sign is present [4].

Inadvertent injury

Blood vessel perforation

Perforation of a major blood vessel requires immediate laparotomy and hemostasis. Urgent vascular surgical opinion should be requested.

Bowel perforation

If a perforation is recognized during laparoscopy it may be repaired endoscopically at that time. Open laparotomy is not essential; however, if the surgeon has minimal tissue approximation skills, or if the leak cannot be identified laparoscopically, then converting to open surgery is recommended. Delayed peritonitis due to viscus rupture may be treated either laparoscopically or by open surgery based on the experience of the surgical team. In most instances, the perforation should be sutured, extensive saline lavage performed, and triple antibiotic therapy instigated [6].

Unrecognized energy delivery

There is now a wide variety of instruments for dissection and energy delivery. It is important that each surgeon becomes familiar with the equipment available.

Specific training on energy sources is required and all surgeons performing laparoscopy surgery should have full training as to the risks of electrosurgery and associated energy sources, prior to the commencement of any procedure. Full knowledge and training is the key to avoiding complications in this area.

Diathermy

Damage to adjacent organ structures by electrosurgical equipment is a common problem in advanced endoscopic surgery and a source of major complications.

Diathermy injury is best avoided, utilizing the following points:

- 1 The operating surgeon must control the foot pedal of the affected device and not the assistant and/or scrub nurse.
- 2 Short bursts of electrosurgery utilizing direct tissue contact are essential.
- 3 An electrosurgical monitoring device should be fitted to each diathermy machine to detect electrosurgical leakage along the shaft of the diathermy instrument.
- 4 All operating centers should have a program of testing and maintaining the electrosurgical hook equipment on a regular basis.
- 5 The surgeon must understand and avoid capacitive coupling prior to the commencement of any laparoscopic procedure.

Alternative energy sources

There is now a variety of energy sources involving high frequency oscillating devices. The instrument delivering this type of energy often remains at temperatures in excess of 100°C, particularly at the tip of the instrument. Inadvertent damage to adjacent structures from direct contact with a heated tip is common and must be avoided. The surgeon must fully understand the basic physics and ergonomics of any alternate energy source prior to its utilization. Prior training on inert tissue is recommended.

Unrecognized injury to adjacent structures is an acknowledged complication of laparoscopic surgery. The emergence of this complication may be delayed for several days and can even lead to unrecognized intra-abdominal infection and subsequent death. The child may recognize a delayed viscus rupture. This involves the sudden occurrence of a warm sensation in the abdominal cavity followed by significant pain and then a

feeling of unwellness. Early recognition of these symptoms by the treating clinical team is essential.

This complication is best avoided by utilizing the following techniques:

- 1 The surgeon must be fully trained in the use of energy sources and instrumentation prior to the performance of any surgical procedure.
- 2 The surgeon must be able to relax within his level of competence and perform a tissue manipulation in a slow, smooth, and ergonomic fashion.
- 3 Clear visualization, an adequate working space, and a careful, meticulous dissection technique are essential.

Tissue approximation

The ability to join tissue structures is an essential skill of any advanced endoscopic surgeon. In most cases, suturing of the affected tissues is required, generally using interrupted technique. There are associated techniques including fibrin glue and endoscopic staples.

The following recommendations are made in order to prevent leakage from tissue approximation:

- 1 The surgical team should be fully trained in endoscopic suturing, and this is best performed by training on both simulators and on laboratory bench top. Mastery of this skill must be achieved in the skills laboratory prior to any surgeon undertaking this on a human. It is inappropriate to train a surgeon in suturing on live human tissue without full competence being achieved in a skills laboratory setting.
- 2 Clear visualization, an adequate working space, and excellent ergonomics all contribute to accurate tissue approximation. The combination of these factors can influence the outcome of the procedure and the surgeon must be fully trained in these aspects of surgical care prior to commencing the task.
- 3 In advanced tissue approximation procedures such as laparoscopic pyeloplasty a mentoring approach is essential, with an experienced surgeon accompanying the surgeon through the first few cases.

Leakage from endoscopic stapled anastomosis can be prevented by:

- 1 Ensuring the operator is completely familiar with the stapling device, and understands the staple depth and the maximum tissue thickness.
- 2 Cautious usage in inflamed bowel (particularly inflammatory bowel disease).
- 3 Accurately closing the enterotomy sites with interrupted sutures.

Other complications

Severe pain

Shoulder pain is a common complaint after laparoscopy, particularly with longer procedures. Occasionally, children undergoing laparoscopy demonstrate unexpectedly severe pain following the procedure unresponsive to narcotic administration. The likely cause of this pain is rapid pneumoperitoneum with associated high intra-abdominal pressure, resulting in distension and triggering of pain fibers. This complication results in not only significant shoulder tip pain, but generalized abdominal pain as well. Narcotic and other associated analgesic infusions including Ketamine may be required to control pain. The estimated incidence of this is one to two per thousand laparoscopies.

Tissue retrieval

The complications of tissue removal are as follows:

- 1 Tumor spillage into the peritoneal cavity and port site, resulting in implantation and metastasis. This is best avoided by ensuring all potential tumors are appropriately placed in a retrieval bag and removed through a generous port site incision.
- 2 Bag rupture during specimen removal: It is essential that the surgeon is fully familiar with the usage and operation of a retrieval bag. The surgeon must ensure that the specimen retrieval bag is opened fully and any rolled-up portion of the bag is fully unfurled prior to placement of the specimen within the bag. A port site must be appropriately enlarged to enable easy withdrawal of the specimen to prevent rupture [7].
- 3 If rupture does occur then widespread lavage and collection of all visible specimen is required. This is particularly of significant risk during removal of the spleen where splenic implantation may result in ongoing hemolysis.

Hematuria

Isolated hematuria has been reported after a variety of laparoscopic procedures, including appendectomy and Meckel's diverticulectomy. This complication appears to be self-limiting and usually occurs for two to three days after the procedure. A renal and bladder ultrasound is essential to ensure that no underlying secondary pathology such as a PUJ obstruction has been missed. The mechanism of the hematuria is uncertain, is probably related to pneumoperitoneum, and it usually self-resolves.

Complications specific to retroperitoneal surgery

Retroperitoneal surgery is often required for advanced urological surgery. The following table outlines the complication and the proposed methodology for overcoming it (Table 18.1).

Injury and trauma to the operating surgeon

There is no question that laparoscopic surgery is a demanding skill. Significant injuries to surgeons have been documented over the last decade [8–10]. The following complications can occur:

- 1 Radial nerve injury, particularly involving the thumb. This can occur by placing the thumb continually through the looped end of the needled holder and squeezing tightly. Digital pressure on the radial nerve may result and surgeons are advised to develop a relatively tension-free grip on instruments so that this does not occur.
- 2 Shoulder strain and rotator cuff injury. This injury is reasonably common and results from the surgeon holding his or her shoulders in an abducted position for prolonged periods of time. This has resulted in significant loss of function, neurapraxia, and muscle pain, particularly if there is associated lateral rotation of the spine during the procedure. This complication is best avoided by lowering the patient's position, ensuring the monitor is in a gaze-down position, and avoiding prolonged periods of fixed stance.
- 3 Anterior osteophyte formation and spinal degeneration: It is becoming increasingly clear that surgeons adopt fixed neck positions during endoscopic surgery

and this may result in associated vertebral damage. Once again, the ability to relax and achieve mastery with advanced endoscopic surgery is essential.

Endoscopic surgery does place significant stress on the operator. During prolonged cases it is recommended that two competent surgeons be present and that they rotate as the main operator during the case. In our practice, we have found that to achieve complex difficult surgery, particularly during the early stages of our careers, it made an enormous difference to have two surgeons who could reflect difficulties on each other and exchange as the main operator. It certainly reduces the physical damage to the surgeon.

Optimizing laparoscopy

We believe that laparoscopic surgery is enhanced by creating a team atmosphere in the operating room, and being aware of when one's limitations as a surgeon have been reached.

Surgical team approach to improving overall laparoscopic performance

Surgical performance is best improved by taking notice of the ten golden rules as outlined below. Any unit undertaking laparoscopic surgery must have a meticulous attitude to training and preparation for this type of surgery. A team approach is recommended with two surgeons of similar standing and competence being involved, particularly in difficult cases. The surgical team must have the ability to clearly question the dissection

Table 17.1 Retroperitoneal complications.

	Cause	Action
Failure to achieve access	Balloon expands into muscle or subcutaneous space	Enlarge wound, separate muscle with deep retractors, and place balloon deeper
Peritoneal perforation	Instrument or trocar damage	Advise anesthetist, increase flow rate to 3–4 l/min, widen tear to allow peritoneal space to equilibrate, continue task. Convert to intraperitoneal procedure if poor visibility
Duodenal perforation	Excessive diathermy dissection, failure to recognize renal structures (often confused with a small multicystic kidney)	Nasogastric tube, triple antibiotics, direct endoscopic suture if recognized and possible. Laparotomy if anatomy uncertain or closure difficult
Poor exposure/visibility	Small working space and inadequate dissection, possible CO ₂ leak from trocar	Check trocars, increase pressure and flow rate, blunt dissect peritoneum to create larger space, insert additional port if possible

moves of the main operator and challenge any operative decision at any point. To achieve excellent outcomes, it is important to avoid significant time pressure and never rush complex endoscopic surgery. Tired surgeons, particularly with fatigue from lack of sleep, result in worse outcomes. There is clear evidence that these team approaches and combined attitudes influence decisions.

Elective versus emergency conversions

Converting a procedure from a laparoscopic to an open one can be a difficult clinical decision. We firmly feel that a surgeon should be able to determine that on a particular day, with a particular patient, with that surgeon's particular ability, if they are unable to achieve completion of the task laparoscopically then an elective conversion to an open procedure should ensue. Whilst we do not place firm time limits on this occurring, one must recognize that after two hours of advanced endoscopic operating, fatigue is likely to have set in. The outcome for a patient who has been electively converted to an open procedure is excellent.

Emergency conversion in the face of significant bleeding, major perforation or adjunct organ damage represents a more difficult situation. Emergency conversions usually occur after a long series of errors including poor vision, poor port placement, failing instrumentation, and unclear anatomy. This scenario needs to be recognized that conversion occurs at an elective stage rather than at emergency. The outcome for emergency conversion has been documented as worse in several large adult series [10,11].

In summary, multiple complications can occur from laparoscopic surgery. Paramount to preventing these is a well-prepared and trained surgical team. Competent surgeons who approach these tasks in a methodological fashion with adequate surgical skills training are unlikely to commit major errors.

Ten golden rules for safe laparoscopy

- 1 Achieve complete training in all endoscopic equipment and energy sources.
- 2 Understand the ergonomics of task performance.
- 3 Always use an open initial port insertion technique.
- 4 Correct any deterioration of visibility or working space early.
- 5 Work as a team, in a team environment. Encourage other surgeons and nurses to question/comment on the dissection.
- 6 Do not aim to complete the operation laparoscopically at all cost. Remember that elective conversions are far safer than emergency conversions.
- 7 Proceed to advanced surgery with another colleague, share the operating time and work together.
- 8 Plan your surgery well, do not apply list pressure or operate fatigued.
- 9 Recognize that, to achieve mastery, a calm cautious approach is required.
- 10 Remember the ego of the surgeon must always be subservient to the patient's welfare.

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CHAPTER 18

Complications of laparoscopic surgery of the upper urinary tract

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KEY POINTS

- Ensure preoperative huddle/timeout and postoperative debriefing
- Develop a standard approach for surgeries that are performed, modifying as needed for each individual case
- Create a minimally invasive surgical team with operating room personnel who have greater familiarity with the instruments that are used and to also facilitate the flow of surgery
- If assimilating a new procedure into existing surgical repertoire, start with more "straightforward" cases first or have access to a more experienced surgeon on standby should difficulties arise
- Have ready access to images for intraoperative consultation as needed
- Have low threshold for stent or drain in difficult cases or where risk for urine leak is high
- Have low threshold for intraoperative consultation for injury to adjacent structures, or repair primarily if small and comfort level is adequate

CLINICAL PROBLEM/CASE

A 3-year-old boy is referred for refractory hydronephrosis after open pyeloplasty performed in the periphery. Ultrasound and excretory renography are shown in Figure 18.1.

The patient undergoes a left redo laparoscopic pyeloplasty. Intraoperatively, the anastomosis is difficult due to scar and fibrosis from the previous surgery. A missed crossing vessel was identified. Attempted JJ stent placement was unsuccessful with the stent unable to traverse the ureterovesical junction (UVJ). A stentless procedure was elected. A Penrose drain was placed at the end of the case.

1 In a case which is intraoperatively challenging, and where insertion of a JJ stent is desired, what options exist when the stent cannot be negotiated through the UVJ?

Postoperatively, the patient becomes febrile and complains of nausea and vomiting. There is no increased output from the Penrose drain. An ultrasound is shown in Figure 18.2.

2 What are the options in this ill child with the findings shown in Figure 18.2?

The surgeon elects to insert a percutaneous nephrostomy tube. The images of the antegrade nephrostogram are shown in Figure 18.3.

Once inserted, a thick purulent effluent is collected. The patient's condition improves.

3 Which options exist at this point in this child's care?

After a period of observation and convalescence, the child returns to the clinic and the surgeon elects to repeat the antegrade nephrostogram to check for patency and drainage, once anastomotic edema has resolved. The nephrostomy tube is capped for a period of time and a repeat nephrostogram is then performed. The nephrostogram is shown in Figure 18.4.

4 What is the next step in the management of this child?

The surgeon elects to insert a JJ stent after dilating the stenosed UVJ. The stent remains in place for several weeks. The patient tolerated clamping of the nephrostomy tube once the JJ stent was removed. The patient remains symptom free and a follow-up ultrasound is shown in Figure 18.5.

Repeat ultrasounds on subsequent follow-up show resolution of hydronephrosis and stability of upper tracts.



Figure 18.1 Ultrasound and excretory renography.

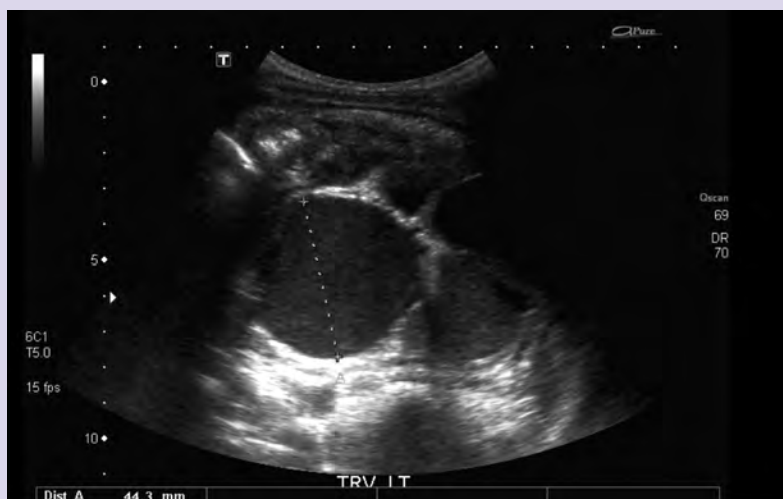


Figure 18.2 Ultrasound.



Figure 18.3 Antegrade nephrostogram.



Figure 18.4 Nephrostogram.

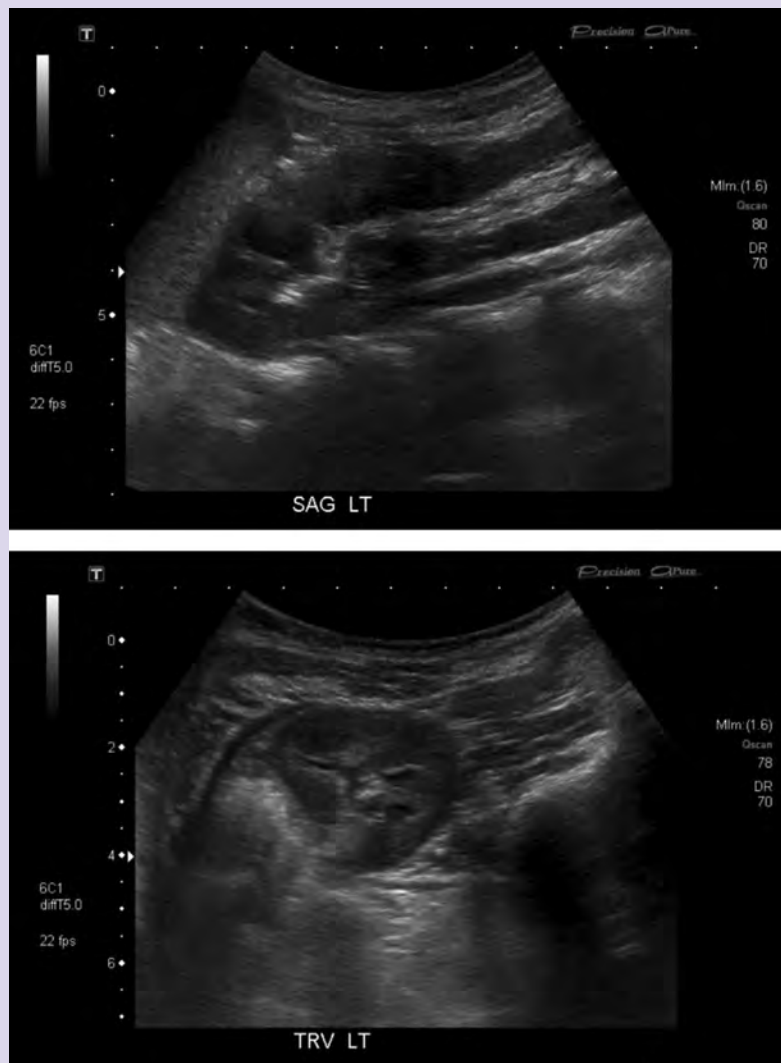


Figure 18.5 Ultrasound.

General operative considerations for optimizing execution of laparoscopic surgery

Essential to the successful completion of any minimally invasive surgery, especially multistep complex surgeries, is effective communication with an understanding by the entire surgical team. Each case

should inevitably begin with a preoperative huddle, or timeout, and postoperative debriefing. This will ensure the smooth running of the case with minimal disruption, while promoting collaboration among all OR staff [2,4–9]. Moreover, adequate time should be set aside to educate patients about possible complications and risks to surgery, especially prior to foreseeably challenging cases.

As difficulty can arise with intraoperative anatomy, especially with renal surgery, it is imperative to review preoperative imaging and have it available in the operating room for quick and easy access. In the rare event that visual cues are inadequate to proceed safely with the case, intraoperative ultrasonography can be advantageous and help with progression, especially for extirpative procedures.

In the modern era of laparoscopy, there exist few true contraindications to laparoscopic surgery. However, most would agree that true contraindications to reduce the risk of foreseeable intra- or postoperative complications include untreated infected access sites, uncorrected coagulopathy and significant hypovolemic shock [10–13]. With experienced anesthetists and consideration with regard to patient co-morbidities, many of the previously prohibitive physiologic patient factors can be balanced.

Surgery-specific complications of laparoscopic surgery of the upper tract

Laparoscopic partial nephrectomy

Laparoscopic partial nephrectomy (LPN) remains one of the most challenging operations with established risks to bleeding, potential adjacent organ injury and fluid extravasation. Especially in the modern era of robotic surgery, adult literature has convincingly demonstrated the feasibility of LPN for a wide range of benign and malignant presentations with several extirpative approaches described to minimize damage to residual renal parenchyma with minimal risk of complication [14–19].

The strength of the supporting literature in the pediatric population, although not as robust, is rapidly expanding, showing comparable results to experienced adult counterparts. For heminephrectomy, it has been associated with shorter hospital stay and better cosmetic results without significantly increasing operative time [12,20].

In experienced hands, complication rates have been reportedly low [5–9,21–24]. Although LPN has been performed for malignant disease [12,25,26], in the more common setting of nonmalignant indications for surgery in the pediatric population, adhering to certain key principles will help achieve success whilst preventing unwanted adverse events.

One of the most important aspects to any LPN is case selection. Surgeons must take into account both patient and disease-specific factors. Size of the kidney, degree of hydronephrosis, amount of residual parenchyma, age and size of patient, associated anomalies and side of lesion are all important and will help determine approach, as well as number of trocars needed to facilitate surgery. Thorough understanding of the limited working space, especially for younger children during transperitoneal or retroperitoneal surgery is crucial [2,14,16,18]. And although the working space can be limited, benefits of the transperitoneal and retroperitoneal approach vis-à-vis improved vascular exposure and pedicle control with reduced need for renal mobilization, are well described [12,27].

Beyond the apparent risks of bleeding, chance of infection and anesthetic-related complications, risk of complication can often depend on the approach and side of the lesion, taking into account patient and disease-specific factors as well.

For right-sided procedures, consideration should be given to preventing liver and bowel injury, as well as unwanted damage to great vessels, all of which can be devastating if managed poorly. Identifying the adrenal gland, along with its accompanying vessels, particularly venous, should be a consideration in upper-pole surgery, especially in bilateral extirpative renal surgery, as inadvertent removal of adrenal glands can be devastating, especially if unrecognized. The liver occupies 40% of the peritoneal space at birth and is most at risk for injury whilst obtaining access. Its size may also impact exposure and insertion of an accessory port for liver retraction early in select cases where surgery can be challenging, may help achieve optimal exposure and facilitate dissection [25,28]. Superficial solid organ injury can usually be controlled with pressure, the use of cautery or with laser coagulation, such as the argon-beam coagulator [10–13]. Clear identification of the duodenum during exposure of the kidney is also paramount as an unrecognized injury can be devastating. Early identification with primary, intraoperative repair has been shown to be most advantageous [14–19,29,30]. In the accompanying video (see Video 18.1), we illustrate a standard technique for suturing the bowel after an inadvertent serosal tear during upper tract surgery. During surgery for left-sided pathology, care must be taken to avoid any splenic or pancreatic injuries. Injuries to the superior mesenteric

artery and celiac trunk are rare, but have been reported [12,30,31].

Intraoperative consultation after early identification of major vascular, splenic or pancreatic injuries should be obtained where appropriate, especially when complete staging of the injury is unclear. Untreated injuries can be difficult to manage in the postoperative setting, especially with an unrecognized pancreatic injury, where prolonged drainage and diet modification may be necessary, both of which coming at a price to the patient. Significant splenic injuries often result in splenectomy [5–9,21–24,32].

Finally, depending on the extent of the extirpation, some patients may be at increased risk for postoperative urine leak, especially with larger or incomplete resections, and some have reported urine leak from a refluxing distal ureteric stump [7,12,25,26]. In those patients with a high risk of urine leak and no intraoperative identifiable defect, prophylactic drain placement through one of the trocar sites may aid in postoperative management. With adequate drainage and no distal obstruction, most will close spontaneously without further intervention. With inadvertent entry into the adjacent collecting system within a duplex kidney, closure can be performed primarily with an absorbable suture. Consideration for intraoperative JJ stent insertion in the event of a large defect may also help to encourage antegrade flow of urine and promote closure of the defect. In this situation, insertion of a Foley catheter may be of benefit as well. Should an unrecognized urine leak develop, image-guided drainage may be necessary, especially after cyst-decortication or heminephrectomy, where the collecting systems are separate and a stent will unlikely be successful.

Need for completion nephrectomy, especially in the context of benign disease in the pediatric population, is regrettable but thankfully quite rare. Most series have reported extremely low rates of conversion to total nephrectomy; however, loss of the remaining moiety has been described in a handful of cases, accounting for 1–6% of laparoscopic heminephrectomies on postoperative imaging [5,7,11,28,33,34]. Cyst formation after heminephrectomy has also been described, and is not ubiquitously reported as a complication in all series [5,10,11]. It most likely reflects a small urinoma from unresected urothelial tissue that is urine producing. Most often asymptomatic, they can be left alone without the need for intervention [7,8,29,30,35,36].

Converting to open, although occasionally regrettable, is never a failure when necessary. Although rare, this is usually associated with technically difficult cases or early surgeon experience. Several series show higher conversion rates to open early on in their experience with a significant reduction in need to convert as surgical volume grows [31,33].

LPN is a novel technique that has gained significant momentum in pediatric urology for management of poorly functioning moieties. Advantages of its use include minimal blood loss, optimal magnification with better parenchymal and distal ureteral visualization, as well as faster recovery with improved cosmesis [7,20,32,36–38]. It is feasible with low conversion rates to open surgery and should be considered for most extirpative pediatric renal surgery [7,34,39,40].

Laparoscopic total/radical nephrectomy

The first laparoscopic nephrectomy (LN) was reported by Clayman in 1991 for malignancy in an elderly woman and by Ehrlich in 1992 in a child with a non-functioning cystic kidney [5,7,11,33,34,41,42].

Common indications for nephrectomy in children differ widely from those for adults. The majority of nephrectomies in children are for nonmalignant diseases, most often for reflux nephropathy, ureteropelvic junction obstruction (UPJO), multicystic dysplastic kidneys and congenital dysplasia [5,43,44]. There is, however, substantial literature advocating for LN in the setting of childhood malignancy [7,8,35,36,45–52]. Its feasibility has also been shown in children who are less than 1 year of age or smaller than 10kg [33,53,54].

Since first reported, LN in children has been performed with rare adverse events. Complications mirror those previously discussed in the section covering laparoscopic partial nephrectomy with regards to dissection and exposure. Complications usually arise with anatomic unfamiliarity or aberrancy. This is particularly important in the setting of small dysplastic renal units where visualization of the renal unit may be difficult. Appreciation for larger, hydronephrotic kidneys may influence location for port placement and approach to maximize ease of surgery and reduce risk of unwanted collateral damage in these severely dilated kidneys. Drainage of the dilated renal unit may be of benefit.

In a systematic review, Kim et al. showed low complication rates for both laparoscopic trans- and retroperitoneal

nephrectomy. Of 634 patients undergoing LN, only two had vascular and three had bowel injuries [1,7,20,36–38]. All of these were for nonmalignant disease.

Similar to LPN, it is critical to have preoperative imaging readily accessible. As previously discussed, major complications with injury to bowel, adjacent solid organs, vascular structures, or with control of the renal pedicle are rare, but can pose serious problems and a systematic approach to management when identified is vital.

Difficulty with bleeding when controlling the vascular pedicle can be difficult to manage. Small vessel venous bleeding can usually be controlled with pressure for several minutes.

Conversion to open surgery is rare and in some reports is not listed as a complication [7,34,39,40,55].

Laparoscopic pyeloplasty and uretero-ureterostomy

Surgery of the upper collecting system in pediatric renal surgery has developed significantly over recent years. Intraoperative complication rates are rare, varying from 0–2% [4,41,42,56–58]. Regarding approach, some favor a transmesenteric approach for laparoscopic pyeloplasty (LP) with excellent reported results; however, surgeons should have a low threshold to formally mobilize the large colon and optimize exposure as needed, especially in challenging cases [21–24].

As previously noted, vascular, bowel, splenic and hepatic lacerations can occur with instrument exchange or with dissection but can usually be repaired primarily if there is no gross spillage in bowel injuries, or controlled with coagulation, use of hemostatic agents or minimal suturing [10,11,32,43,44].

In LP or uretero-ureterostomy (LU-U), whether performing the case with traditional laparoscopy or with robotic assistance, the approach to ensuring success is similar. A widely patent, tension-free, watertight anastomosis is ultimately the desired result, with or without an intraluminal stent left in place. Disease-specific factors may be a hindrance, especially in the context of a nondilated or intrarenal pelvis, or with a long segment defect during LU-U. To minimize complications in difficult scenarios, it is important to have a standard approach for the uncomplicated ones.

The use of hitch stitches during LP and LU-U or the appropriate use of a third arm in robotic-assisted cases to stabilize the kidney or ureter can be crucial in es-

tablishing exposure and performing the anastomoses. Prior to dismemberment, if the renal pelvis is found to be intrarenal or nondilated, further mobilization of the ureter with possible renal descensus can be performed. Increased risk to vascular compromise of the ureter will limit mobilization and should be weighed. Nephropexy may be necessary in this setting to prevent renal malrotation. Should tension persist with these mobilization maneuvers, non-dismembered pyeloplasties, such as a Foley Y-V plasty can be attempted [4,45–52]. Several reconstructive options exist to address these issues and the critical surgeon should be familiar with several options to address length discrepancies. And although the need of these lengthening maneuvers are rare and most cases will proceed smoothly if done so in a systematic manner, they inevitably add to one's surgical armamentarium for dealing with difficult cases and result in a decrease in postoperative complication and improved success.

The role for intraoperative stent is controversial; however, several series have shown success with stentless procedures or by using an externalized pyelo-ureteral stent to reduce the need for a subsequent general anesthetic [53,54,59,60]. This type of stent also circumvents occasional difficulty with negotiating the ureterovesical junction distally, should the problem arise. The role of stenting should be assessed by the surgeon on a case-by-case basis, reflecting overall surgeon comfort and case performance.

Postoperative complications occur in less than 2% of cases [3,4,61–65]. Should urine leaks occur, most are small and usually resolve with time as anastomotic edema decreases, should a drain be left in place. When no drain is left postoperatively, treatment of any distal obstruction, either with stent insertion (with or without a urinary catheter) or nephrostomy tube (which can be challenging in the postoperative setting) will usually be successful.

Late failures are also rare, reported to be up to 7% of cases LP [18,66–68]. There is no standard approach for repair. Some favor a redo pyeloplasty, while endoscopic management has been shown to be successful in select cases [67,69]. There is some literature for the role of ultrasound early postoperatively to help predict risk of future obstruction after open pyeloplasty in antenatally detected ureteropelvic junction obstruction [70].

Conclusion

Minimally invasive surgery has been established as a feasible approach to upper tract surgery. Literature has shown that complications are rare, with high success

rates for the vast majority of procedures currently being performed by pediatric urologists. A systematic approach to any surgery will afford all pediatric urologists the ability to perform these surgeries with increasing ease and with fewer complications.

DOS AND DON'TS

Do

- Have access to more experienced surgeons for assistance should the need arise
- Leave stent or drain in difficult cases when risk of urine leak is high
- Repair intraoperative injuries primarily when possible, or seek consultation when needed
- Have imaging readily accessible for intraoperative consultation when needed

CLINICAL PROBLEM/CASE: DISCUSSION

- 1 In certain cases when passage of a JJ stent is desired and the UVJ cannot be negotiated easily with transperitoneal antegrade attempts, either a nephrostomy tube, an externalized pyeloureteral stent or a retrograde JJ stent can be left in place [71]. Externalized tubes will address any anastomotic edema and allow for antegrade diagnostic tests, should they be required, without manipulation of the anastomosis. Another added benefit of externalized drainage is reduction in bladder symptoms that may occur with the presence of a JJ stent, as well as the ease with which removal occurs, as subsequent need for general anesthetic, which is often required in small children, can be circumvented. In this situation, a drain was not helpful as there was no anastomotic leak.
- 2 Urgent decompression in an unwell child is warranted. Initiation of supportive measures with antibiotics should not be delayed. Insertion of a nephrostomy tube would be ideal while anastomotic edema settles. In the early postoperative setting, with the presence of a fresh anastomotic suture line, although some might be reluctant to do so, retrograde insertion of a JJ stent can still be attempted if image-guided therapy is not readily available. In the same vein, insertion of an antegrade nephro-ureterostomy tube can also be attempted.
- 3 Similar to question 2, a period of convalescence while anastomotic edema settles may prove beneficial. Proper staging of the upper and lower urinary tract can then take place and any issues can be addressed.
- 4 Antegrade studies are highly suggestive of UVJ stenosis. Attempt at dilation and stenting would be the most logical with repeat studies to confirm antegrade flow. The error in this case would be to prematurely remove the nephrostomy tube prior to confirmation of patency of both ureteropelvic and UVJ. In this case, dilation of the UVJ proved sufficient to ensure adequate drainage. Follow-up imaging is reassuring for stability.

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CHAPTER 19

Complications of laparoscopic surgery of the lower urinary tract

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KEY POINTS

- Minimally invasive surgery of the lower urinary tract should aim at reducing morbidity and hospital stay and give at least comparable outcomes to open surgery
- Ureteral reimplantation, excision of urachal remnants, and urinary tract reconstruction such as augmentation and appendicovesicostomy lend themselves to minimally invasive techniques
- Ureteral reimplantation can be done with robotically assisted extravesical or transvesical approaches. Early identification and minimal dissection of the ureter is key to extravesical reimplantation
- It is important to measure the length of the appendix for appendicovesicostomy to determine its suitability for the procedure

Introduction

Lower urinary tract surgeries in children have classically been performed in an open approach. However, with recent advances in technology and improved training in both conventional laparoscopic and robotic surgical techniques, minimally invasive approaches are becoming more common, more practical and increasingly in demand. Assuming similar clinical outcomes, when presented with the choice, both patients and parents appear to prefer minimally invasive surgical approaches [1]. Additionally, surgeons are increasingly expanding their use of these minimally invasive techniques as their comfort with them grows.

The ultimate goal of performing any procedure in a minimally invasive manner is decreased morbidity and decreased complications with at least comparable outcomes to the respective open procedure. This chapter reviews the procedures that may be performed in a minimally invasive manner on the lower urinary tract, with specific emphasis placed on the potential complications and techniques available to avoid complications.

Laparoscopic/robotic ureteral reimplantation

The diagnosis and management of a child with an initial or recurrent febrile UTI remains controversial and without consensus on treatment among health care professionals. Current options for treatment of vesico-ureteral reflux (VUR) in children include observation, endoscopic injection of bulking agents, and ureteral reimplantation. In those patients whose families elect surgical treatment, open surgical repair remains the gold standard [2–5]. However, laparoscopy, and specifically robotic-assisted laparoscopy, is being utilized more frequently as a surgical tool to treat this condition whilst minimizing the morbidity associated with open surgery [6].

Robotic-assisted extravesical ureteral reimplantation

Intra-abdominal access and insufflation is obtained in the surgeon's preferred manner (Verres or Hassan). An 8 or 12 mm camera port is placed in the umbilicus. Two

robotic working ports are used, one on each side of the camera port. An assistant port may be added if needed or preferred by the surgeon. Identification of the ureter with as little dissection as possible is a key to minimizing the risk of postoperative urinary retention for bilateral procedures [7]. Sound knowledge and attention to anatomic landmarks are important to identify the ureter. The ureter is reliably found crossing over the iliac vessels. In males, the vas deferens is identified and mobilized cephalad to avoid injury. In females, mobilization of the uterus, uterine ligament, and vessels posteriorly may be needed to identify the ureter. The ureter is then isolated and mobilized to the level of the bladder. The bladder is filled by a preplaced Foley catheter. A 5:1 ratio (tunnel length : ureteral diameter) detrusor tunnel is made over the bladder urothelium. This length is generally 2.5–3.0 cm. A stabilizing hitch stitch(es) may be placed during the dissection or detrusor reconstruction to prevent sliding of the ureter within the detrusor tunnel, in a manner similar to the open technique [8]. The detrusor tunnel is then closed over the ureter with running or interrupted sutures per surgeon preference.

In a large series with conventional laparoscopic ureteral reimplants, Bayne et al. described overall reflux resolution of 93.5%. In that series, urinary retention was present postoperatively in 6.5% (3/46) patients with bilateral reimplants and in no children (0/52) with unilateral reimplant [9]. A series by Chalmers et al. reported a 90.9% resolution of reflux without significant complications [10]. In a series comparing open vs. robotic extravesical reimplants, Smith et al. demonstrated an overall success rate of 97% in the robotic group vs. 100% in the open group. Mean operative time was significantly longer (12%) and mean length of stay (33 vs. 53 hours) and pain medication usage were significantly shorter for the robotic group. Of note, three of eight patients with bilateral robotic reimplants experienced transient retention [6]. In a large series by Kasturi, in which 150 patients underwent bilateral robotic reimplantation, overall success rate was 99.3%, and no patient experienced any voiding issues postoperatively [11].

The main complication specific to the bilateral procedure is urinary retention. Caution should be taken with several studies showing increased likelihood of postoperative retention for bilateral reimplantation compared to unilateral reimplantation [6,9]. Lateral dissection should be avoided to limit damage to pelvic nerves. The nerves to the bladder may be exposed by retracting the

ureter medially, whilst adjacent tissue located medially and caudally is pulled laterally. This should reveal the pelvic plexus, which may be avoided for the remainder of the procedure [7,11,12]. Whilst not clinically reported in the literature, one should also be aware of the potential complication of ureteral obstruction from making the detrusor tunnel too tight or kinking the ureter. The ureter should be buttressed by the detrusor to prevent VUR when the bladder is full, but not obstruct the flow of urine into the bladder.

Laparoscopic/robotic transvesical reimplantation

For conventional laparoscopy, a 5 mm trocar is placed into the bladder under direct visualization with a cystoscope within the bladder through the urethra. The anterior bladder wall is then sutured to the anterior abdominal wall under direct visualization. This helps maintain the operative working space. Two additional working trocars (3 or 5 mm) are placed under direct visualization. Bilateral ureteral catheters are commonly placed, as well as a urethral catheter. The ureteral catheters are sutured in place. At this point, the procedure is similar to the open Cohen cross-trigonal reimplantation [4]. The ureters are mobilized, a submucosal tunnel is made across the trigone of the bladder, and the ureterovesical anastomosis is performed with intracorporeal suturing.

Chung et al. reported their results of 48 cases of bilateral transvesical reimplantation. Overall, three cases were converted to open. Overall VUR resolution rate was 96.4%. Mean operative time was 155.6 minutes. No significant complications were reported [13]. Cannon et al. compared vesicoscopic cross-trigonal and open reimplantation. Overall success rate was 91% vs. 97% vesicoscopic vs. open respectively. Mean OR times were 199 and 92 minutes respectively. No complications were seen in the open group. In the vesicoscopic group, one patient had urinary leakage, one child developed bladder stones, and one child had bilateral ureteral obstruction [14]. An additional study from France had similar outcomes, with an overall 92% reflux resolution rate [15].

Minimally invasive transvesical ureteral reimplantation is a technically demanding procedure and should be undertaken by experienced laparoscopic surgeons. Whilst no direct comparison study has yet been published, the additional component of performing an intravesical ureterovesical anastomosis makes the procedure

more challenging than the extravesical approach. The anastomosis adds the potential for ureteral obstruction. Placement of ureteral stents may minimize this risk. The added element of urinary leakage may only be avoided by watertight closure of the laparoscopic ports at the conclusion of the procedure. Additionally, several studies have raised the possibility of injury to the upper GU tract by pneumovesicum [16,17]. The potential for visceral (bowel) or vascular injury exists, which would not be expected with an open procedure.

Complicated urachal remnants

Failure of timely closure of the allantois may lead to persistence of urachal remnants (URs). While some persistent URs may close spontaneously during the first year of life, it is thought that URs present beyond 6 months of age are less likely to close, and surgical treatment may be considered [18]. URs that fail to resolve spontaneously may lead to infections and have the potential for formation of malignancy (adenocarcinoma) later in life. URs may include urachal sinus, urachal cyst, vesicourachal diverticulum, and patent urachus.

Historically, the gold standard for treatment of URs has been open surgical excision of the urachus from the bladder to the umbilicus. Whilst most series are small, secondary to the rarity of the URs, the successful use of laparoscopy, and more recently robotic surgery, for removal of URs has been increasingly reported in the literature [19–22]. An added benefit of minimally invasive surgery for URs is that the entire pathologic process can usually be viewed easily from the intra-abdominal access, and decisions can be made by direct visualization. In this manner, laparoscopy allows for simultaneous accurate diagnosis and treatment.

Given the unique nature of each UR, each case must be approached on an individual basis with appropriate preoperative planning in terms of number and position of port placement. It is imperative that a good view of the UR pathology be achieved so that intraoperative decisions can then be made. It has been proposed that an acute angle of the trocars can aid in dissection near the umbilicus [19] and lateral port placement may assist with complete view of the UR tract [23]. There is controversy regarding the need to excise the dome of the bladder if the UR is not a diverticulum. If part of the UR remains, persistent infection or urinary leak may

result. When the UR is in direct contact with the bladder lumen, care should be taken to perform a watertight closure of the bladder lumen. An indwelling urethral catheter may be left for a period of time to allow complete healing of the bladder defect.

Complex lower urinary tract reconstruction (appendicovesicostomy and bladder augmentation)

Reconstructive surgery of the lower urinary tract is arguably the most challenging and technically demanding aspect of pediatric urological surgery. Reconstructive procedures in this realm include bladder augmentation, creation of continent catheterizable channels, bladder neck sling, and bladder neck reconstruction. Gastrointestinal augmentation cystoplasty and appendicovesicostomy have been performed with minimally invasive techniques. Only a few centers are performing these with minimally invasively surgery. The procedures have been demonstrated to be technically feasible in patients, whilst the unique challenges presented and potential complications are becoming clearer.

Famakinwa et al. reported on 18 children who underwent robotic-assisted appendicovesicostomy. Mean overall operative time was 494 minutes. Overall, there was 94.4% continence rate with one stomal infection noted and two patients requiring stomal revision [24]. Nguyen et al. reported on 10 patients with robotic appendicovesicostomy. Mean operative time was 323 minutes. Urinary leakage was present in one patient, requiring open revision, and minor incontinence in two patients, with one case managed by Deflux injection. Of note, one procedure was converted to an open procedure secondary to a short appendix [25]. Gundeti et al. reported on six patients with spina bifida who underwent robotic ileocystoplasty with or without simultaneous appendicovesicostomy. Average operative time was 8.4 hours. One patient was converted to an open procedure secondary to failure to progress. Three of five patients with a channel required stomal revision at the skin level. One patient had a wound infection, one patient had a lower leg deep vein thrombosis, and one patient had transient lower extremity paresthesia. At the time of publication, all patients had day and nighttime continence, no UTIs, and reasonable bladder capacities

[26]. Other complications may include the same problems seen with open surgery. Specific to the patient population undergoing these procedures, laparoscopy has been shown to be safe in patients with ventriculoperitoneal shunts [27]. Follow-up is premature for assessment of late complications.

Whilst no publications directly compare open and laparoscopic ileocystoplasty, several describe its feasibility on a limited basis [28–30]. Similar to the open technique, of particular concern is the potential risk of bowel leak causing leakage of stool into the abdomen with resultant infection and abscess formation. A large series comparing intracorporeal and extracorporeal urinary diversion following robotic cystectomy in the adult population found no difference in overall complications between the two groups, no difference in reoperative rate, and overall significantly less gastrointestinal complications [31]. Whilst not specific to urological literature, several pediatric surgical publications have demonstrated at least equal outcomes with similar complication rates with an intracorporeal bowel anastomosis [32,33].

Aside from the benefits of laparoscopic surgery previously mentioned, it has been proposed that minimally invasive techniques may lead to less adhesions and intra-abdominal scar formation [34]. This may potentially be important not only to reduce the probability of small bowel obstruction, but to decrease the complexity of future surgeries that these patients may undergo. Clearly, longer follow-up and larger studies are needed to directly compare minimally invasive reconstructive surgery with open surgery. When considering which technique to perform, surgeon comfort, experience, and skill with laparoscopic and robotic surgery are key to performing these complex surgeries safely and successfully.

In order to minimize, recognize, and avoid complications, several important factors need to be taken into account. It is vital to accurately measure the length of the appendix when performing appendicovesicostomy. This may be achieved by passing premeasured suture, vessel loops, or other measuring devices into the abdomen. If the appendix is not of adequate length, the surgeon may consider early conversion to an open surgery in order to perform an ileal Montie or other intestinal channel. Once an adequate length appendix is identified, it is important to make an adequate length detrusor tunnel in order to prevent leakage through the channel with a full bladder. A 4 cm detrusor length is thought to be sufficient to prevent incontinence [35].

Conclusion

As comfort with laparoscopic and robotic surgery increases, the applicability of minimally invasive surgical techniques for the lower urinary tract grows. The shorter hospital stays, decreased analgesic requirements, and improved cosmesis are appealing to patients, parents, and surgeons alike. The demand for such procedures is likely to grow. While few long-term and prospective studies exist comparing traditional open to minimally invasive techniques, there is a growing body of literature highlighting the feasibility and success of these procedures. Surgeon understanding of the potential complications with the minimally invasive approach to the urinary tract is paramount to helping avoid, identify, and treat complications should they occur.

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CHAPTER 20

Complications of robotic surgery of the urinary tract

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KEY POINTS

- Awareness, recognition, and a plan of management will help the surgeon decrease adverse events in any surgery, but especially when beginning a pediatric robotic practice
- With knowledge, complications are preventable

Introduction

In the last decade, robotic surgery has changed many urologists' practices. As media attention continues to increase, urologists in all disciplines have become more enthusiastic about the robot and its use. Its advantages are numerous – including high resolution 3D magnification of the visual field, increased dexterity and rotation, tremor control, and thus increased precision – making it particularly well suited to pediatric urology. Continued surgical innovation has now seen commonplace laparoscopic procedures such as pyeloplasty, ureteroureterostomy, and ureteral reimplantation transitioned to robotic-assist, which may be the standard of care in the future.

As robotic surgery becomes more commonplace in pediatric urology, the understanding and discussion of possible complications is a priority. There is a paucity of information available in all robotic surgery, but particularly in pediatric urology. Because of this, we have to rely partially on the laparoscopic literature due to its similarity. However, there are also unique robotic issues that the operative team should be aware of and ready to address. Recognition of the possible problems that might be encountered is critical in the prevention of adverse events. With this awareness, efficient and

quick management of complications during any surgical procedure is more likely and the best way to improve patient outcomes.

It is beneficial to note that utilizing robotic surgery in children comes with special considerations due to their widely varying body size. Even the most robotically adept operating rooms and surgical staff have to be flexible and have a clear understanding of the setup and size-appropriate instruments that are necessary for an efficient and safe procedure when beginning a pediatric robotic program. This awareness and careful planning will help ready the entire operating room for any situation.

This chapter is presented as a surgeon would progress through the procedure. It begins with preoperative considerations, continuing to intraoperative ones, procedure-specific complications and, finally, postoperative concerns.

Preoperative complications

From the time that robotic surgery is offered to a patient, especially a child, the surgeon must consider the patient's past medical history and other disease processes. This careful consideration continues throughout the preoperative period.

Positioning

In every procedure, safe and correct positioning is of the utmost importance. This is no different in robotic surgery. Prevention begins with understanding that there are several special matters to consider due to the robots' arms and their movement in regards to positioning. Following the standard of care for all surgical procedures, the patient's pressure points should be adequately supported. Some surgeons prefer to use a gel pad in addition to the standard operative bed. Due to reports of compartment syndrome, it is important to be aware of lower extremity positioning if using stirrups, but also to be mindful of gluteal compartment syndrome. One patient in a recent series developed gluteal compartment syndrome during robotic assist pyeloplasty [1].

During positioning the surgeon should be in constant communication with anesthesia. Their needs should be established prior to the patient's entrance into the operating room. Easy access to the child's head as well as IVs, Foley catheter, blood pressure cuff, and any other lines or tubes should be a priority. Anesthesiologists sometimes recommend placement of central and arterial lines for increased monitoring capabilities including blood gases and blood pressure [2]. In general, there is no need for this level of invasive monitoring. Recognize at the beginning and throughout the procedure if the child's position is not as planned or the robotic arms are too close to maintain safety. Management in this case is immediate repositioning of the child or robot, increased padding, and a recheck once this is done.

The physiologic effects of robotic surgery are similar to those of pediatric laparoscopy. As surgeons learn robotics, increased operative times increase the amount of absorbed CO_2 . The surgical team should be aware of the effects of prolonged operative times, especially in children with complicated medical histories. Children with known cardiopulmonary, neurologic, or cerebrovascular disease who would not be considered candidates for laparoscopic surgery should be considered for robotic surgery with extreme caution [2]. Because of the increased amount of CO_2 that is absorbed, the risk for CO_2 embolus is increased, especially in those with cardiac insufficiency [3]. When patients have a sudden decrease in end-tidal CO_2 levels and a mill-wheel murmur, the robot should be rapidly undocked and they should be placed in Trendelenburg in the left lateral decubitus position. This is followed by aspiration of the embolus via the central line. This occurrence is most

commonly seen with initial access and indicates insufflation into a major vein.

After assuring anesthesia's access, the patient needs to be secured to the table. We suggest always performing a tilt test in every direction focusing on those that the table might be adjusted to when the robot is docked.

Docking the robot

The surgeon and the operative team should assess clearance and positioning of the robotic operative arms in relation to the patient when docking, and then anesthesia and the surgical team assist in relation to port placement and positioning. Painful injuries are possible. The team should take care that as the robot is advanced towards the patient, the robotic arms do not graze or hit the patient's extremities. Placing extra padding around the legs, arms, head and shoulders prior to prepping and draping the child is one precaution that the surgical team and anesthesia can take. However, this should not be done in place of careful scrutiny of the robotic arms' positioning and the space between the arm and the patient.

Operative complications

Preparedness for intraoperative complications and knowledge of their management is of the utmost importance.

Complications of access

Port placement and closure

The insertion of ports is no different than in standard laparoscopy. In 2004, it was reported that there was a two-fold increase in major complications when comparing utilization of the Veress technique versus the open Hasson. An open technique, however, increased the amount of time in port placement [4]. If using a Veress needle, avoid past-pointing and follow the standard for laparoscopy with the saline drop test aspirate and irrigate prior to insufflation. Significant complications with port insertion include bowel, bladder, and vascular injury with a rate of 1.18% [5]. The most common complication in pediatric laparoscopy was that of pre-peritoneal insufflation and subcutaneous emphysema with a rate of approximately 5% [5]. It is important to note that although injury is reduced



Figure 20.1 Avoid past-pointing by holding the cannula firmly with the index finger near its tip; it is less likely to past-point as the instrument is passed through the fascia. The preplaced fascial box suture provides for counter-traction in placing the trocars. (Inset) The point of the trocar tenting up the peritoneum. Access should be under direct vision.

with the open technique it is not completely eliminated. As surgeon experience increases with port placement, complications decrease. When placing trocars the surgeon should take into account the child’s body habitus, anatomical anomalies, and past surgical history when determining the angle and depth of insertion. Obese children will have less periport mobility, while thin children and small infants will have a greater risk of injury of peritoneal structures.

To facilitate port placement, we place a box-stitch of absorbable braided suture under vision for the first umbilical port as well as the working ports (see Video 20.1). This provides a way to offer resistance to the placement of the port, as well as making port site closure efficient (Figure 20.1). It can also serve to control any port site bleeding.

The chosen method of access also leads to different concerns. Retroperitoneal access has its own specific complications with the most important being inadequate development of the surgical field (Table 20.1) [6]. The retroperitoneal working space can be developed by balloon insufflation or by blunt dissection. When using blunt dissection, especially in smaller children, the peritoneum may tear, which will lead to insufflation of that space [7].

At the conclusion of the case we recommend port site closure in all pediatric patients regardless of size. We close all 5 and 3.5 mm port sites. The ratio of port incision to abdominal wall becomes larger as the patient gets smaller. Many have experienced postoperative port site hernias in pediatric surgery and this phenomenon is well reported [8–10].

Visualization of instruments and the tissues

Proponents of robotic surgery in pediatric patients cite increased visualization of the surgical field as a benefit. With this in mind, having constant visualization of instruments is a necessity in robotic surgery. With an absence of haptic feedback, inadvertent damage to a nearby structure may occur without the operator being aware. Attention must be paid to which instruments are connected to what kind of cautery (monopolar and bipolar). Inappropriate wiring of the monopolar forceps, thus turning them into bipolar cautery, is a mistake that can be easily avoided with a check. It is also important to know which part of the instruments will become “hot” when using them actively. In addition, the surgeon and the assistant should only use clips, coagulation, and cutting when they have direct vision of the tissue that they are acting on.

Table 20.1 Complications of access.

Complication	Signs	Management
Parietal hematoma	Bleeding, loss of visual field	Localize bleeder and control
Visceral injury	Inadequate field, absence of normal landmarks	Control bleeding, localize injury, Robotic repair if possible; if not, convert to open, pending visualization, presence of and amount of gross spillage
Peritoneal insufflation ^a	Loss of working field, bulging peritoneum	Vent to peritoneal cavity, window peritoneum, convert to transperitoneal procedure

From Caione et al. 2003 [6]. Reproduced with permission of Springer.

^aRetroperitoneal access only.

Bowel injury

Bowel injuries are devastating when missed intraoperatively. They can occur at the time of access as well as during the procedure. Prevention of this is accomplished by awareness of anatomy, the space in which the arms are working, and careful use of cautery. Specifically with access, the Hasson or open technique as discussed above has fewer complications including bowel injury [4]. Take special precautions with children who have undergone prior surgery or have unusual anatomy. Any available images should be reviewed prior to the surgery with special attention given to the bowel's relation to the fascia. Intraoperative prevention's focus is on awareness. Care and attention should be paid to the working arms including the possibility of arcing. They should remain in the surgeon's field of vision at all times. Recognition's mainstay is careful inspection of bowel and operative field at the end of the procedure and also during if the anatomy is not straightforward. As a rule, if the anatomy or the field look abnormal it should be inspected closely, including all nearby viscera.

Finally management is usually intraoperative repair after a thorough assessment. Bowel injuries can usually be repaired primarily and without difficulty. For serosal injuries, oversewing for reinforcement with one suture layer is usually adequate. Rare cases such as a sizable segmental injury or devitalization could require bowel diversion [11–13].

Intraoperative bleeding

Control of bleeding is an important skill for the robotic-assist laparoscopic surgeon to have in his or her armamentarium. Prevention of intraoperative bleeding is very similar to prevention of bowel injury; awareness of anatomy and visualization of the working arms are mainstays. Bleeding can be caused at the time of access, during the procedure itself, and at the time of closure. Recognition here relies on hypervigilance and continuous appreciation of proximity to the great vessels and/or the renal hilum. In the event of significant bleeding exposure and vision are key to successful management. One of the robotic arms will likely be used to maintain exposure and should not be moved. A suction-irrigating device that allows for maintenance of pneumoperitoneum can be used, but visualization of the bleeder is of primary importance. In cases where the bleeding is too swift for this, apply pressure to the area with one of the instruments and, if still inadequate, control conversion

to open may become necessary. When controlling bleeding in a minimally invasive fashion most use cautery if possible and then clips, although staples have also been reported [14]. Cautery should be used judiciously with full vision. We suggest that two endoscopic clip applicators are in the room prior to the beginning of surgery. Keep in mind that bleeding can be perceived as greater than it is due to the magnification of the scope.

Surgery-specific complications

Upper urinary tract

Currently, upper urinary tract procedures such as partial nephrectomy, pyeloplasty and ureteroureterostomy are the most widely-adopted pediatric urologic robotic surgeries. These are performed via transperitoneal and retroperitoneal approaches (Table 20.2).

Pyeloplasty and ureteroureterostomy are similar in regards to complications. Urine leak is a possible complication to most urologic robotic surgeries and can present with leakage from the port site, drainage via the Penrose, ileus, pain, fever, or infection. This can be managed with ureteral stenting or nephrostomy placement pending the etiology of the urine leak. Ureteral urine leaks have been reported due to migrated and occluded ureteral stents [15]. Other complications include anastomotic stricture. In 2011, Minillo et al. published a long-term follow-up on 155 patients who underwent robotic-assisted pyeloplasty. They reported a complication rate of 11%. They re-operated on 3.2%, a total of five patients, due to recurrent obstruction. Two patients had a persistent ureteropelvic junction (UPJ) obstruction. One was found to have retroperitoneal fibrosis, and the other two had intrinsic stenosis [16]. Prior to that Olsen had reported a 67 case series with an 18% complication rate with complications including transient hematuria, urinary tract infection, conversion to open due to lack of space, and obstruction. In these two studies, three patients were returned to the operating room and found to have missed crossing vessels [17]. All vascular anatomy should be closely inspected prior to performing the reanastomosis.

When faced with an obstructed kidney after pyeloplasty in a child, most favor initial conservative management via retrograde or antegrade ureteral stenting. Balloon dilation as well as laser endopyelotomy of anastomotic stricture has been reported as a successful, less invasive option for management [1,17]. Success of these

Table 20.2 Complications of robotic surgery by procedure.

Procedure	Complication	Cause	Management
Pyeloplasty	Urine leak	Unrecognized non-watertight closure intraoperatively, obstructed stent, migrated stent	Initial management: ureteral stent vs. percutaneous nephrostomy tube
	Stent migration	Latrogenic, poor intraoperative stent placement	Replace vs. remove stent
	Persistent UPJ obstruction	Anastomotic stenosis, kinking ureter, redundant pelvis	Redo pyeloplasty (robotic vs. open)
Ureteroureterostomy	Ureteral obstruction	Stent migration, anastomotic stenosis	Ureteral stent [27]
Partial nephrectomy	Bleeding (immediate vs. delayed)	Anticoagulant therapy, vigorous movement	Assure hemostasis intraoperatively Supportive care followed by ablation, then if necessary reoperation
	Urine leak	Unrecognized leak intraoperatively, obstructed stent	Drainage, ureteral stent placement
	Urinoma	Unrecognized collecting system leak or residual renal parenchyma	Closure of renal polar defect
Antireflux surgery	Urinary retention (bilateral reimplant)	Likely neurogenic in origin	Usually transient: intermittent catheterization, Foley placement followed by voiding trial
	Bladder leak	Unrecognized intraoperative leak	Bladder drainage and percutaneous drain [22,28]
	Persistent reflux: low grade	Inadequate ureteral sheath	Observation unless urinary tract infection
	Persistent reflux: high grade	Inadequate ureteral sheath, bladder dysfunction	Assess bladder dynamics Redo robotic reimplant

approaches is somewhat limited, however. If simple balloon dilation and stenting is unsuccessful, the author prefers definitive operative management, with robotic reoperation. This approach has become a viable option for prior open and laparoscopic cases, and may be more efficient than open surgery [18,19]. A principle advantage is that most open pyeloplasties are performed retroperitoneally, and with a transperitoneal approach the area of most scarring is avoided. The robotic approach also permits thorough mobilization of the ureter as needed, as well as performing a ureterocalycostomy if the pelvis is too densely scarred. This has not been necessary in the authors' experience of over 20 reoperative pyeloplasties, but has been used more commonly by Casale [20].

Antireflux surgery

Extravesical reimplantation is the most widely-documented in the literature in regards to robotic antireflux surgery, although an intravesical approach is an option. Prior to embarking on this procedure it is suggested that

the patient has a bladder capacity of 130 cm³ or more [21]. In 2004, Peters reported a series of 24 extravesical reimplants with a complication rate of approximately 5%. One patient experienced a bladder leak that was treated with a percutaneous drain; three had persistent reflux (one of whom was a bilateral repair); and one had a transient ureteral obstruction which was treated with a ureteral stent [22]. In 2011, 37.5% of patients (three) had trouble voiding postoperatively after bilateral reimplant. One of these underwent straight catheterization, one had an in-dwelling catheter placed on postoperative day 2, and the third had a catheter placed on transfer to the floor. One of the 17 unilateral reimplant patients had difficulty voiding postoperatively and had to be straight catheterized. Also one of the bilateral reimplant patients had unresolved reflux that was reduced [23].

Nephrectomy and partial nephrectomy

There are a limited number of reports of nephrectomy and partial nephrectomy using robotics in children,

although these procedures can be enhanced using robotics. In a report of nine partial nephrectomy cases from 2002 to 2005, two complications were reported: urine leak and umbilical port site infection [24,25]. When performing a partial nephrectomy, if the collecting system is entered it is important to close, and it is suggested that a drain be left. The authors prefer to close the renal defect using mattress sutures of an absorbable suture placed over a fat pad. This may limit the occurrence of urinomas that have been reported in laparoscopic pediatric partial nephrectomies. It is important to consider that urinomas can be asymptomatic and also resolve on their own as reported by Lee et al. [25]. Conservative management should be attempted if feasible [25]. Follow-up with renal ultrasound at 4–6 weeks is also recommended [26]. Concern should be raised for the possibility of injury to the vessels of the remnant renal pole, which has been described in pediatric laparoscopic partial nephrectomy. This likely represents over-enthusiastic mobilization of the remnant pole and can be avoided by limiting mobilization to the renal moiety to be removed. The renal vessels are particularly sensitive in infants and it may be useful to have injectable papaverine available for the procedure.

Unique complications

Stent placement

Errors of stent placement are an avoidable complication. When placing the initial wire to cannulate the ureter, the surgical team should be certain that the floppy tip of the wire is passed first. The stiffer side of the wire can lead to aberrant placement and urinoma or possibly even visceral injury. Prior to completion of the procedure, an on-table KUB or intraoperative ultrasound can be obtained to ensure proper placement [29]. If a stent is misplaced, it can be removed and replaced either in an antegrade or retrograde fashion.

In the event of equipment failure or an event that mandates conversion to open, the operating room should have an established plan for a safe, efficient, and rapid undocking of the robot. It is suggested that this has been practiced in a simulated emergency training session, although limited data are available to validate this approach. When the patient is prepped and draped for the procedure, he or she should always be prepared for

the possibility of conversion, including full preparation of the abdomen.

Postoperative complications

Postoperative complications often stem from events that should have been recognized intraoperatively. They include: hemorrhage; intraperitoneal injury; and urine leak as previously discussed. Bleeding usually occurs at the site of the operation or the abdominal wall. High volume blood loss and patient instability often mandates return to the OR. Other less significant bleeding can be managed with close observation, labs and supportive care. Intraperitoneal injury can be elusive intraoperatively and is best corrected with careful examination of the operative field and meticulous inspection of violations of the peritoneum if the case was performed retroperitoneally. It can present with a variety of symptoms that range from ileus to florid sepsis. Free air as a normal finding has been documented over seven days after laparoscopic surgery [30], and therefore is not a reliable indicator of intraperitoneal injury. Instead, one should use the patient's clinical picture to make determinations about intervention.

Important clinical indications of an intestinal injury include development of a low-grade fever, depressed white blood cell count and port-site pain. While not definitive, they should prompt careful assessment in the child who is not recovering rapidly. Abdominal CT imaging is usually the best first step when this is a significant concern.

Robotic surgery in the management of failed surgery

Providers might tend to be wary of using robotics as a method for repairing previous repairs, but due to increased precision and visualization it should be considered as a viable option. All redo surgery comes with the inherent property of being more technically challenging secondary to postoperative adhesions and abnormal anatomy [31]. Patients should be selected carefully and the open technique should be used to gain port access. As with open surgery, in both redo pyeloplasty and redo reimplant, one must take care to inspect the ureter for ischemia and integrity at the beginning of the repair.

Reoperative pyeloplasty

Using robotic assist to complete redo pyeloplasty is also becoming more common and has been reported to be effective with up to a 100% success rate [18]. One center reported 94.1% resolution in 17 patients as compared to the same center's primary robotic assist pyeloplasty resolution rate of 96%. No significant difference was observed in regards to complications between the two cohorts. In addition, they reported no unique complications to those who were undergoing redo pyeloplasty [1]. In 2008, Hemal et al. reported the use of robotic assist redo pyeloplasty status post initial open pyeloplasty with resolution of the UPJ obstruction in all nine patients [32]. Reoperative pediatric pyeloplasty was described by Passerotti et al., with approximately 90% success in these challenging cases [19]. More recently in 2012, Lindgren et al. [18] reported a series of 16 patients who underwent redo pyeloplasty (13), and redo ureterocalycostomy (3). Of these, 12 had a previous endoscopic procedure, one case was converted to open, two experienced transient postoperative hypertension, and one had stent discomfort and ileus. The patient who was converted to open had a horseshoe kidney and required a transfusion. All of the patients who presented with symptoms have had resolution [18]. Ureterocalycostomy is an option for reoperative pyeloplasty when the renal pelvis is severely scarred and reported success rates, including some primary cases, is 100% [19]. In the authors' experience, ureterocalycostomy has not been needed in over 25 reoperative cases. It has been reported that up to 8% of patients will require redo-pyeloplasty and robotic-assist is an effective option [33].

Reoperative antireflux surgery

Robotic redo-reimplantation is an excellent consideration regardless of prior procedure, previous robotic or laparoscopic repair, open surgery, or endoscopic management with injection. Lopez reported redo laparoscopic reimplantation of 43 ureters in 30 patients after initial open repair. Their complications were found to be no different than previously reported, with one ureteral leak and one urinary retention requiring temporary catheterization [34].

Robotic antireflux surgery has proved to be effective in children who have had prior endoscopic therapy with Deflux, and have persisting or recurrent reflux. The dissection of the distal ureter is made somewhat more complex but, with care, the ureter can be fully mobilized. The Deflux pseudocyst is opened and the

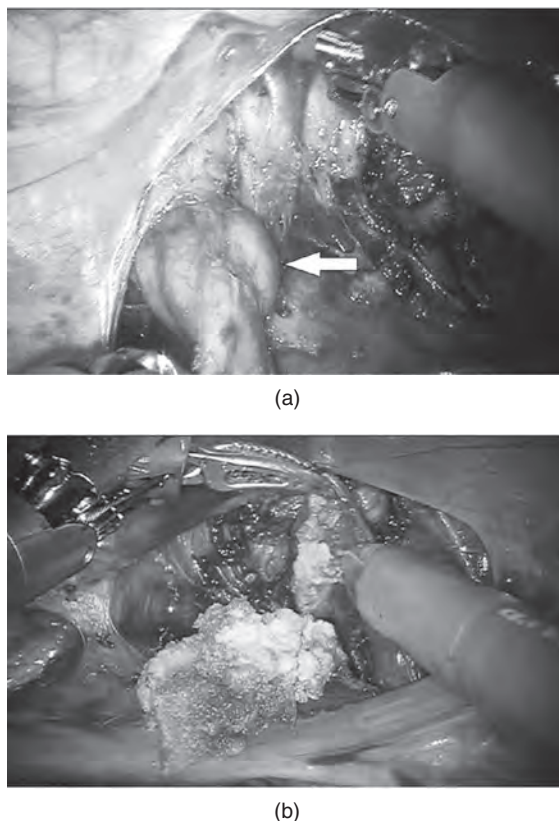


Figure 20.2 (a) Appearance of bulging distal ureter during robotic extravesical ureteral reimplantation following failed Deflux injection. The arrow indicates the collection of the bulking agent. (b) The Deflux paste is extruded from the pseudo-capsule surrounding it within the adventitia of the ureter. This does not usually present a major difficulty in performing the procedure, but the bulking agent should be removed.

material removed by suction. In all cases seen, it is encapsulated and has not appeared to be degraded. The excess wall of the pseudo-capsule is removed, but not adjacent to the ureter. The remainder of the procedure is identical (Figure 20.2).

Conclusion

As robotic surgery continues to gain favor because of increased dexterity, visualization, and surgical precision its adoption will increase. Awareness, recognition, and an educated management plan will help pediatric urologists avoid and methodically resolve complications that occur in the operative suite.

DOS AND DON'TS**Do**

- Be aware of the management of possible procedure-specific complications prior to beginning a case.
- Take care that the patient's extremities are not too close to the robotic arms.
- Use the saline test when gaining access via a Veress needle.
- Develop an adequate working space when using retroperitoneal access.
- Have a plan with your operative team for equipment failure and possible conversion to open.
- Ensure when placing an intraoperative ureteral stent that the wire is oriented correctly.
- Consider intraoperative on table KUB if the wire's position is in question.
- Carefully inspect the operative field, including all adjacent structures, prior to closing.
- Think about common intraoperative errors and recognize them before removing ports
- Close all port sites.

Don't

- Deviate from standard of care when positioning and padding your patient.
- Pass-point with trocar or Veress needle.
- Forget to disarm the bladed trocar during open insertion.

CLINICAL SCENARIOS**Occult vascular injury at access**

- 1 During intraperitoneal procedure the light seems to dim and the field seems smaller.
- 2 Inspect the entire field for hemorrhage – large pool of blood discovered in pelvis.
- 3 Aspirate and identify source.
- 4 Pressure if active bleeding.
- 5 Assess arterial vs. venous and vessel involved.
- 6 Assess whether you are comfortable managing lap/robotically.
- 7 With pressure proximally, expose vessel in retroperitoneum.
- 8 Oversee injury.
- 9 Open with pressure on injury if not reparable endoscopically.

Bowel injury at access

- 1 Veress needle access transumbilically.
- 2 Aspirate with brown fluid – what to do?
- 3 Withdraw needle until no aspiration of intestinal contents.
- 4 Insufflate and scope to identify puncture site.
- 5 Widen umbilical site and bring bowel up through umbilicus.
- 6 Clean, inspect, repair.
- 7 Replace bowel and continue procedure if no gross spillage.
- 8 Antibiotics, observation overnight.

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PART VI

Genitalia

CHAPTER 21

Hernia and hydrocele repair

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KEY POINTS

- Recent evidence and balance of risk and benefit do not support contralateral groin exploration when performing an open inguinal herniotomy
- Laparoscopic inguinal hernia repair may be safely performed in infancy
- Laparoscopy is a safe option in the management of large or recurrent inguinal hernias or hernias which were previously incarcerated
- A prospective randomized study showed no difference in recurrence rates, testicular atrophy, iatrogenic testicular ascent, patient satisfaction or cosmesis between open or laparoscopic inguinal hernia repair at 2-year follow-up
- Risk of recurrence is between 0.8% and 8% and is higher in premature infants, large hernias or where the sac is inadvertently opened at open surgery
- The risk of testicular atrophy is higher in previously-incarcerated hernias
- Repair of congenital hydroceles may be delayed until 2–3 years of age; post-operative hydroceles may also resolve spontaneously

CLINICAL PROBLEM/CASE

Case 21.1

You are called to review a 36-week gestation infant on the special care unit who underwent bilateral inguinal herniotomies the previous day. He was known to have had a previous left incarcerated hernia. His left hemiscrotum is swollen, erythematous and tender.

- 1 What is your differential diagnosis?
- 2 What investigation would you perform in order to confirm your diagnosis?
- 3 How would you manage this situation?

Case 21.2

A 3-year-old boy presents with a right-sided groin swelling 4 weeks after undergoing a right orchidopexy. It is difficult to examine him and you are not sure whether a testis is palpable in the hemiscrotum.

- 1 What is the differential diagnosis and what investigations would be appropriate?
- 2 What risks would you tell the parents about preoperatively?
- 3 What surgical approach would you use and why?

Introduction

Inguinal hernias and hydroceles in children result from a patent processus vaginalis (PPV) which fails to obliterate, allowing the passage of bowel or peritoneal fluid,

respectively down the inguinal canal and often into the hemiscrotum. This developmental arrest explains the increased incidence in premature infants. The incidence of pediatric inguinal hernia is 10–20 per 1000 live births and is highest during the first year of life. One-third of

children undergoing surgery for hernia are younger than 6 months of age. Premature infants have an even higher risk of developing inguinal hernia with reports of incidence of up to 25% [1,2]. Inguinal herniotomy in infancy is a challenging operation and awareness of potential complications and their management is of utmost importance. Hydrocele repair, on the other hand, may be delayed until 2–3 years of age and is a less technically-demanding procedure.

Inguinal hernia repair

A number of approaches have been described for repair of inguinal hernias but the essential principles are the same. In the open procedure the hernial sac is identified, dissected off the surrounding structures (testicular cord in males, round ligament in females) as proximally as the deep inguinal ring, assessed for contents that are reduced, and then transfixed and ligated. The wound is closed after ensuring that the testis (in the male) is pulled down into the scrotum without tension. Inguinal herniotomy in the neonate and premature infant can challenge the most experienced pediatric surgeon. The tissues are thin, often edematous and friable, especially if the surgery is performed less than 48 hours after a reduced incarcerated hernia. The surgical approach is usually the same as for a hernia in an older child although it has to be remembered that the internal and external ring may overlies each other and that the tissue planes may not be as easily defined due to edema.

The indications for contralateral inguinal exploration to avoid the 7% incidence of metachronous hernias has been debated at length [3], and a recent meta-analysis has shown that 18 contralateral explorations would need to be performed to prevent the occurrence of one hernia [4]. Niyogi et al. explored an alternative method to view the contralateral deep ring during an open approach by inserting a 70 degree telescope via the hernial sac, detecting an open ring in 13% of cases [5].

The laparoscopic approach has increased in popularity over the last few years and may be safely performed in babies. In a study of 67 infants weighing 3 kg or less, of whom 22% presented with incarceration, recurrence rate was 4.4% with no cases of testicular atrophy. Twelve babies were subsequently noted to have high testes, although only four eventually required

an orchidopexy [6]. The transperitoneal technique involves a purse-string or Z-stitch suture closure of the open deep ring, carefully avoiding the testicular vessels and vas in boys. Alternatively, the peritoneum at the deep ring could be circumferentially-divided and closed proximally with a purse-string suture [7]. An advantage of the laparoscopic approach is the identification of a contralateral open deep ring, which could be closed under the same anesthetic, although there is no strong evidence to support this. Identification of an open deep ring via a 30 degree umbilical scope has been shown to be more effective than inspection using a 70 degree scope via an inguinal approach [5]. Laparoscopy also facilitates the identification of a “sliding” hernia, whereby intra-abdominal structures such as the appendix, the fallopian tube, or even the bladder wall can be intimately associated with one wall of the hernial sac such that it is impossible to separate the two, making the open approach more challenging [8]. Similarly, large neonatal hernias (defined as a hernia extending to the bottom of the scrotum associated with and diameter of open internal ring >2 cm) may also be repaired safely laparoscopically [9].

Surgical management of the incarcerated inguinal hernia that has failed manual reduction can be a real test of surgical skill. The open preperitoneal approach is well-documented [10]. An open transperitoneal approach with ligation of the internal ring avoids tackling the cord, allows inspection of the gut, and can aid hernial reduction [11]. The laparoscopic approach to incarcerated inguinal hernias has been shown to be feasible and safe [12] and has the advantage of bypassing all edematous tissue and cord structures, as well as easier inspection of reduced bowel. In a study comparing the two surgical techniques, the open approach was associated with a higher incidence of complications (vas transection, iatrogenic testicular ascent, testicular atrophy and recurrence) than the laparoscopic approach (equivalent rate of recurrence only) [13].

Hydrocele repair

Repair of the congenital hydrocele, or hydrocele of the cord, involves identification and ligation of the PPV via an inguinal approach. Secondary hydroceles, or those arising following varicocelectomy, are not usually associated with a patent PPV and may be treated

via a Lords or Jaboulay procedure [14]. The large “dumbbell” hydrocele or abdominoscrotal hydrocele is rare and was first described by Dupytren in 1834 as “L’hydrocele en bissac.” Surgical management involves complete excision of the abdominal element with an additional Jaboulay or Lord’s procedure for the scrotal element. A laparoscopic-assisted approach may aid the diagnosis and ensure adequate closure of the internal ring [15].

Outcomes

The overall success rate for inguinal herniotomy, with success being defined as one operation with no complications, is over 90%. The main factors that determine outcome are size/gestational age at surgery, previous incarceration and approach.

- 1 Age:** Inguinal hernia repair in the neonate is known to be more challenging, although a study of open inguinal herniotomy in babies <5 kg showed an acceptable recurrence rate of 2.3%, a 2.7% testicular atrophy rate and a 2.7% incidence of acquired high testes [16]. A study following laparoscopic hernia repairs in infants <3 kg revealed a 4.4% rate of recurrence, 6% iatrogenic high testes but no cases of testicular atrophy [6]. Baird et al. showed that prematurity was an independent risk factor, with the highest incidence of complications occurring in babies below 26 weeks gestation [17].
- 2 Previous incarceration:** Previous incarceration increases the risk of complications following open but not laparoscopic repair. Nah et al. showed that time to full feeds and length of hospital stay were equivalent whether the surgery was performed laparoscopically or open, but that the open group had a higher incidence of complications including vas transection, acquired ascending testes and testicular atrophy, which did not occur following a laparoscopic approach. The recurrence rate in both groups was similar (2.9 vs. 3.6%) [13].
- 3 Surgical approach:** Koivusalo et al. carried out a prospective single-blinded, randomized comparison of laparoscopic versus open hernia repair in children aged 4 months to 16 years and showed equivalent recovery times but a higher requirement of postoperative analgesia in the laparoscopic group. At 2-year follow-up, recurrence rates were equivalent (lapa-

roscopy 6%, open 5%), and there were no cases of testicular atrophy or ascent in either group. Cosmesis and patient satisfaction were also not significantly different [18].

Complications and their management

The overall operative complication rate associated with hernia and hydrocele surgery is 1–8%.

Bleeding

Bleeding due to the severing of small veins in the superficial fascia or from the pampiniform plexus may be controlled by gentle pressure or very cautious use of bipolar diathermy. Only after adequate hemostasis should the wound be closed in order to avoid a postoperative hematoma. A hematoma usually does not need to be explored unless it continues to enlarge or becomes infected. Scrotal elevation is encouraged and analgesics administered.

Wound complications

Infection occurs in less than 2% of cases and can be prevented by meticulous hemostasis and asepsis. Treatment is with antibiotics. Hypertrophic scars and keloid formation can be prevented by keeping the incision within Langer’s lines/skin creases, utilizing the knife rather than the diathermy for making the incision, and avoiding wound infections. Evidence supports silicone sheeting, pressure dressings, and corticosteroid injections as first-line treatments [19].

Ilioinguinal nerve injury

Hypesthesia and neuropathic pain can result from nerve entrapment or injury, and has an estimated incidence of 5% in children [20]. The ilioinguinal nerve and the genital branch of the genitofemoral nerve may easily be damaged on incising the canal. Localization of the nerves before dissection, diathermy, or ultimately closing the canal reduces possible damage risk.

Injury to the vas deferens

Damage to the vas is reduced if it is not grasped between forceps and only dissected off using one blade of a non-toothed forceps if necessary. Bilateral injury or injury to the vas in a solitary testis results in infertility. Vasal injury identified at the time of surgery may be

repaired using microscopic anastomosis or delayed to a later stage, in which case the vasal ends should be marked with a permanent suture to perform vasovasotomy after puberty [21–23].

Recurrence

Factors that contribute to recurrence following inguinal surgery include: failure to ligate the sac proximal enough at the deep ring; a large/ dilated internal ring; injury to the floor of the inguinal canal resulting in a direct hernia; weakness due to co-morbidity such as malnutrition or collagen disorders; and increased intra-abdominal pressure such as the presence of a VP shunt. The recurrence rate after inguinal herniotomy is reported to be between 0.8% and 6%. More than 50% occur in the first year postoperatively and more than 90% by 5 years post-operatively. Vogels et al. specifically looked for predictors of recurrence following open inguinal herniotomy and suggested that inadvertent opening of the hernial sac during dissection, a larger size of hernia, and gestational age (60% of recurrences occurred in babies <37 weeks gestation) but not previous incarceration were the greatest predictors of recurrence [24]. A prospective randomized study did not show a significant difference in recurrence rates between open and laparoscopic approaches [18]. Recurrences are best dealt with sooner rather than later and methods include high ligation of the recurrent sac, snugging the internal ring (McVey repair), and preperitoneal repair for multiple recurrences [8]. Laparoscopic repair of recurrent inguinal hernias was shown to be quicker without increased incidence of testicular atrophy or further recurrence, possibly due to avoidance of a fibrotic inguinal canal [25].

Postoperative hydrocele

A residual hydrocele occurs in less than 5% of cases following an inguinal herniotomy. Davies et al. showed that a postoperative hydrocele may resolve spontaneously and does not indicate a residual PPV [26]. However, if the hydrocele does not resolve spontaneously after one year, reoperation is indicated.

Testicular atrophy

Testicular atrophy occurs where the blood supply to the testis is compromised, either by an incarcerated hernia prior to the surgery, or as a result of intraoperative injury to the testicular blood supply. Injury may be avoided

by not grasping the vessels, not stripping all the tissue off the vessels, and only prudently using the diathermy, if at all. No intervention is required once a testis has atrophied.

Iatrogenic testicular ascent

Iatrogenic testicular ascent occurs following 1.3% of open inguinal herniotomies, with a higher incidence, up to 6%, in babies <5 kg [16]. It is usually caused by insufficient dissection of the sac to the deep ring or failure to make sure that the testis is pulled down into the scrotum at the end of the operation, causing snagging of the cord under the external oblique layer of the canal and subsequent adhesion to it. Attention to these points reduces the risk of ascension. An orchidopexy after previous groin surgery may be challenging due to fibrosis in the inguinal canal, and a scrotal approach has been advocated with 96% success rate [27].

Bladder damage

Damage to the bladder is rare and usually occurs if an incision is too low and medial, confusing the surgeon with the bulge of the bladder mimicking a hernial sac [28]. Management involves urethral catheterization and a two-layer repair of the bladder wall.

Iliac and femoral vessel damage

This complication is also rare and tends to occur in older children. It results from the sutures of the canal closure at the level of the lower border of the external oblique being placed too deep. Management involves appropriate exposure and proximal and distal control of the vessel and repair with non-absorbable sutures.

Complications specific to laparoscopic herniotomy

One of the largest personal series of laparoscopic inguinal hernia repair in children has shown a recurrence rate of 3.7%, a hydrocele rate of 0.7%, and a testicular atrophy rate of 0.2% [29]. In this series the hernial sac was not transected and was only closed from inside using a purse-string type of “N” stitch with non-absorbable suture laparoscopically. The recurrence rate was lower in the last 100 cases than at the beginning, suggesting a learning curve. Evidence suggests recurrence rates are not increased by using absorbable sutures, although a different series of 972 repairs using the laparoscopic percutaneous extraperitoneal closure (LPEC) revealed

a recurrence rate of 12.5% with absorbable sutures and no recurrences with non-absorbable sutures [30].

Conclusion

Inguinal hernias and hydrocoeles are common pediatric surgical problems. Inguinal hernias may be repaired

using an inguinal or laparoscopic approach with comparable results. Laparoscopy has been shown to be safe in neonates and in the repair of previously-incarcerated or recurrent hernias. A positive outcome may be expected in over 90% of procedures, and complications may be avoided by detailed knowledge of the anatomy of the inguinal canal and meticulous surgical technique.

DOS AND DON'TS

Do

- Use bipolar diathermy sparingly, in particular in the vicinity of the vas or ilio-inguinal nerve
- Avoid opening the hernial sac at open surgery
- Ensure the testis is in the scrotum and the inguinal cord lies without tension following hernia and hydrocele repair

Don't

- Attempt an open inguinal herniotomy less than 48 hours after manual reduction of an incarcerated hernia
- Use laparoscopy following incarceration or recurrence

CLINICAL PROBLEM/CASE: DISCUSSION

Case 21.1 discussion

- 1 Possible diagnoses include ischaemia of the left testis consequent on previous incarceration or as a complication of surgery (damage or diathermy injury to the testicular vessels, compression by tight closure of the inguinal canal) or epididymitis.
- 2 A scrotal ultrasound Doppler. In this case, the investigation revealed a well-perfused normal left testis and a congested epididymis suggestive of epididymitis (Figure 21.1).

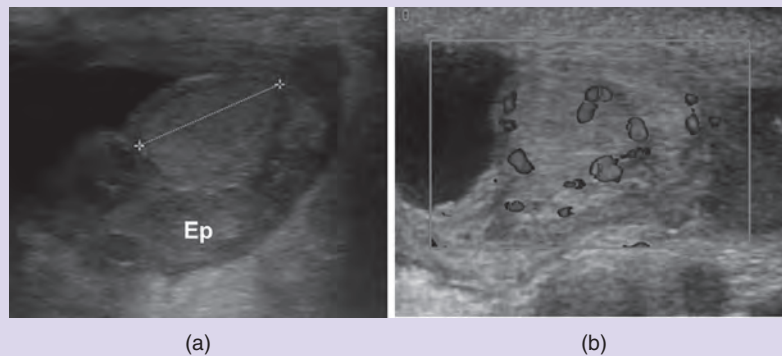


Figure 21.1 Ultrasound Doppler of left testis showing edematous epididymis (Ep) suggestive of epididymitis (a) and a well-perfused testis (b).

- 3 Epididymitis was treated with a 10-day course of co-amoxiclav. The ultrasound Doppler was repeated 3 months after the event to ensure testicular viability (Figure 21.2).

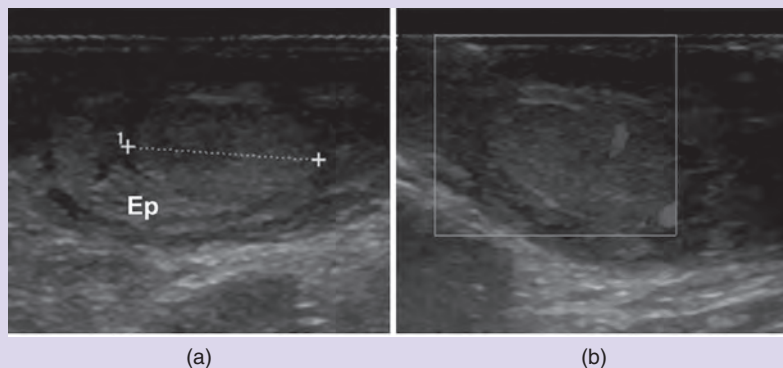


Figure 21.2 Repeat ultrasound Doppler 3 months later showing resolution of the epididymitis (a) and good perfusion of the testis (b).

Case 21.2 discussion

- 1 The differential diagnosis is that of any groin lump: hernia, iatrogenic ascending testis, epididymo-orchitis, torsion, hydrocele. Investigations would include a urinalysis and groin/scrotal ultrasound.
- 2 Risks of surgery include vessel/cord damage, bleeding, wound infection, testicular loss, recurrence.
- 3 Depending on the extent of swelling and edema either a direct groin approach or an initial laparoscopy to determine whether or not a hernia containing omentum or bowel is present. A direct approach with the presence of edema raises the risks of cord damage and/or inadvertent opening of any hernial sac. If the diagnosis is an ascending testis postsurgery, then risks of atrophy and cord damage are higher than at primary surgery.

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CHAPTER 22

Orchidopexy and orchidectomy

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KEY POINTS

- Congenital undescended testis is a common condition with palpable testes managed via open surgery with an inguinal or scrotal approach
- Laparoscopic orchidopexy is becoming the gold standard for impalpable testes with a success rate >90%
- Initial position of the testis affects surgical outcome – higher complication rates with intra-abdominal testes
- Most common complications are testicular re-ascent and atrophy
- Repeat orchidopexy is technically demanding and aided by adequate exposure, good lighting, optical magnification and a clear understanding of inguinal anatomy
- Acquired undescended testis is an increasingly recognized condition, and may be managed nonoperatively
- Altered body image following orchidectomy in childhood may lead to the request for a testis prosthesis later
- Orchidectomy following acute torsion may result in subsequent infertility

CLINICAL PROBLEM/CASE

A 13-month-old boy is referred by his GP with an impalpable testis. The child is otherwise fit and well. He appears healthy with no syndromic features. Genital examination shows a palpable left testis positioned in the scrotum, of normal size and consistency. The right hemiscrotum is empty.

- 1 What is the diagnosis for this child?
- 2 If the right testis was impalpable, what would be the investigation of choice?

- 3 This child had a right testis palpable in the inguinal region. What operation should be offered?
- 4 What surgical tricks might facilitate orchidopexy and avoid recurrence?
- 5 What are the postoperative risks following orchidopexy?
- 6 What is the risk of infertility for children with undescended testis and retractile testis?
- 7 Six months after orchidopexy the right testis is found in the inguinal region again and cannot be manipulated to the scrotum. What further management should be offered?

Introduction

The undescended testis (UDT) is a common problem in pediatric urological practice with 3–5% of newborns affected, although the majority descend in the first few months of life resulting in an incidence of 0.8–1.1% at 1 year of age [1]. Treatment options are hormone manipulation and surgery. Surgical procedures for the palpable testis include the standard inguinal and more

recently described scrotal orchidopexy. The impalpable testis can be managed by open surgery with testicular vessel preservation or an open one- or two-staged Fowler–Stephens procedure. Microvascular transfer of the intra-abdominal testis with anastomosis of the testicular artery and vein to the inferior epigastric vessels is a viable option for successful orchidopexy. Increasingly, however, the intra-abdominal testis is being managed laparoscopically via a one-stage orchidopexy

or, if necessary, a laparoscopic Fowler–Stephens procedure performed in one or two stages. Recently, a novel laparoscopic-assisted two-stage elongation technique, incorporating traction whilst preserving the testicular vessels, has been described with preliminary encouraging results [2,3]. This chapter will not cover these laparoscopic techniques as they are discussed in Chapter 23. Controversies in the management of cryptorchidism include age at operation, surgical approach, management of complications, and follow-up. Orchidectomy is performed for small, dysplastic UDTs or for nonviable testes at exploration for acute torsion. The aim of this chapter is to look at the outcomes of these surgeries, to document the range of surgical complications that occur, and provide advice on how to prevent and manage these complications.

Outcomes for orchidopexy

The success of orchidopexy can be measured from an anatomical or functional perspective. In the former, the surgeon aims to relocate the testis in a dependent position within the scrotum, with preservation of testicular volume indicating a lack of testicular atrophy.

Most of the present literature describes outcomes in these terms and relates success to initial testis position and operative technique. In the latter, the objectives are to maximize and preserve the future spermatogenic and endocrine functions of the testis. These data are often difficult to acquire, as patients must be followed through adulthood and long-term prospective, randomized and controlled studies to determine the optimal age and technique of orchidopexy, from both an anatomical and functional standpoint, which have yet to be published.

Outcome by testis location

The excellent review by Docimo in 1995 summarizes the importance of preoperative testis position on successful orchidopexy [4]. Of over 300 articles and book chapters, a final assessment was limited to 64 reporting 8425 orchidopexies which contained sufficient data for analysis. Of 2491 testes, where a preoperative position and postoperative result were reported, a successful outcome was noted in over 90% located beyond the external ring and increasing failure rates observed with progressively higher testes (Figure 22.1). These results are expected when related to the increased complexity of surgical techniques required to bring peeping and

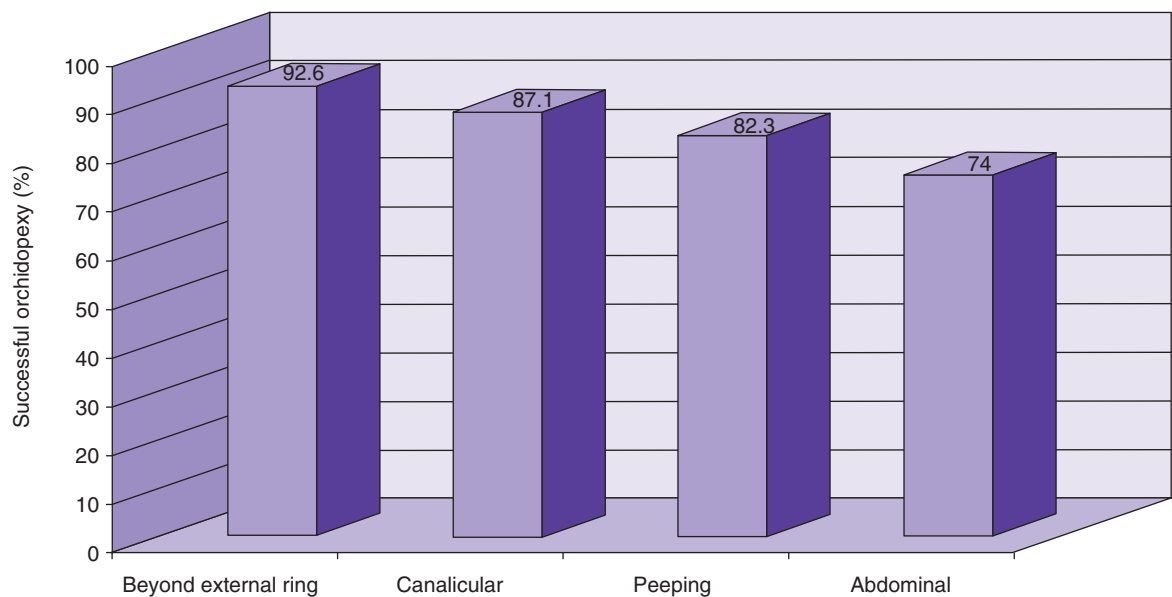


Figure 22.1 Historical success rates for orchidopexy in relation to preoperative testis location. (Data from literature review by Docimo 1995 [4]. Reproduced with permission of Elsevier.)

intra-abdominal testes to the scrotum. Contemporary publications report that success of orchidopexy has increased to >95% for inguinal testes and >85–90% for abdominal testes [5,6].

Fertility potential after orchidopexy in unilateral ectopic, canicular and emergent testes, as long as surgery is performed in early childhood, is good (>90%) and fertility for most cases of unilateral intra-abdominal testis and patients with unilateral anorchia or vanishing testis is expected, whereas the majority of patients with bilateral intra-abdominal testes are infertile [7].

Outcome by age

The age at orchidopexy has been decreasing steadily over the years with most surgeons now recommending surgery at 6–12 months of age. At present in England, only 1 in 5 boys are operated on at an “evidence-based” age and although the trend is towards more boys receiving age-appropriate surgery, levels of early surgery remain unacceptably low. The reasons are likely multifactorial but probably partly explained by the phenomenon of testicular ascent [8].

A drive for earlier orchidopexy has come from histological data documenting germ cell degeneration during the second year of life [9,10] and findings of delayed and defective prepubertal maturation of germ cells in cryptorchid testes [11]. To address possible concerns of earlier orchidopexy, and specifically any increased risk to testicular vessel integrity, Wilson-Storey et al. retrospectively reviewed their results in 100 orchidopexies under and 100 over 2 years of age [12]. Results were similar in both groups with an atrophy rate of 5%. A more recent study assessing the outcome of 349 orchidopexies concluded there was no increase in testicular atrophy in patients operated on less than 13 months of age [13].

Although it is too early to know if orchidopexy in the first year of life will improve long-term outcomes, a few studies suggest earlier surgery may be beneficial. In a randomized controlled study of 149 boys, 70 were randomized to surgery at 9 months of age and 79 to orchidopexy at 3 years of age. Over the first 24 months of life testes operated on at 9 months showed statistically significant better growth, as assessed by ultrasound measured testicular volumes, than non-operated boys [14]. In another study of 86 patients between the ages of 1 and 9 years undergoing orchi-

dopexy, only boys operated within two years of birth showed significant recovery of testicular volume at follow-up – so called testicular “catch up” growth [15]. Hadziselimovic et al. have published data on infertility in 218 cryptorchid men correlating testis biopsy findings and age at orchidopexy with total sperm counts. If transformation into Ad spermatogonia (the adult stem cell pool) had occurred, age-related differences in fertility outcome were observed, with earlier surgery resulting in higher sperm counts. Age at surgery had no effect in the group of cryptorchid men having no type A dark spermatogonia at the time of orchidopexy and this failure of germ cell maturation predicted infertility and azoospermia [16].

When orchidopexy is delayed to later childhood, subclinical decreases in Leydig cell function have been documented [17]. Surgery before the age of 2 results in higher inhibin B levels in adulthood, implying better-preserved Sertoli cell function [18]. Outcomes for palpable testes.

Inguinal orchidopexy

Saw et al. reported on 1057 palpable testes operated via an inguinal approach and 943 (89%) were in the bottom of the scrotum at the end of surgery, a further 67 (6%) in the middle, 41 (4%) in the top of the scrotum, one in the groin, and five boys underwent orchiectomy [19]. In Docimo’s literature review inguinal orchidopexy was successful in 88.6% of cases (Figure 22.2) [4].

Scrotal orchidopexy

The Bianchi transscrotal orchiopexy has gained popularity as an alternative to the traditional inguinal orchiopexy in the management of palpable UDTs [20]. The advantage of this technique is said to be reduced postoperative pain due to the single incision used, improved cosmesis as the incision is placed in the inguinoscrotal crease or a high rugosal fold of the scrotum, and reduced operative time compared to a standard inguinal approach as there is only one incision to make and close. Exposure of the external ring through the same incision can be achieved, allowing access to the inguinal canal for high ligation of the sac when indicated. Should conversion to an inguinal incision become necessary for additional mobilization of the spermatic cord, the patient is no worse off than with a traditional inguinal orchiopexy.

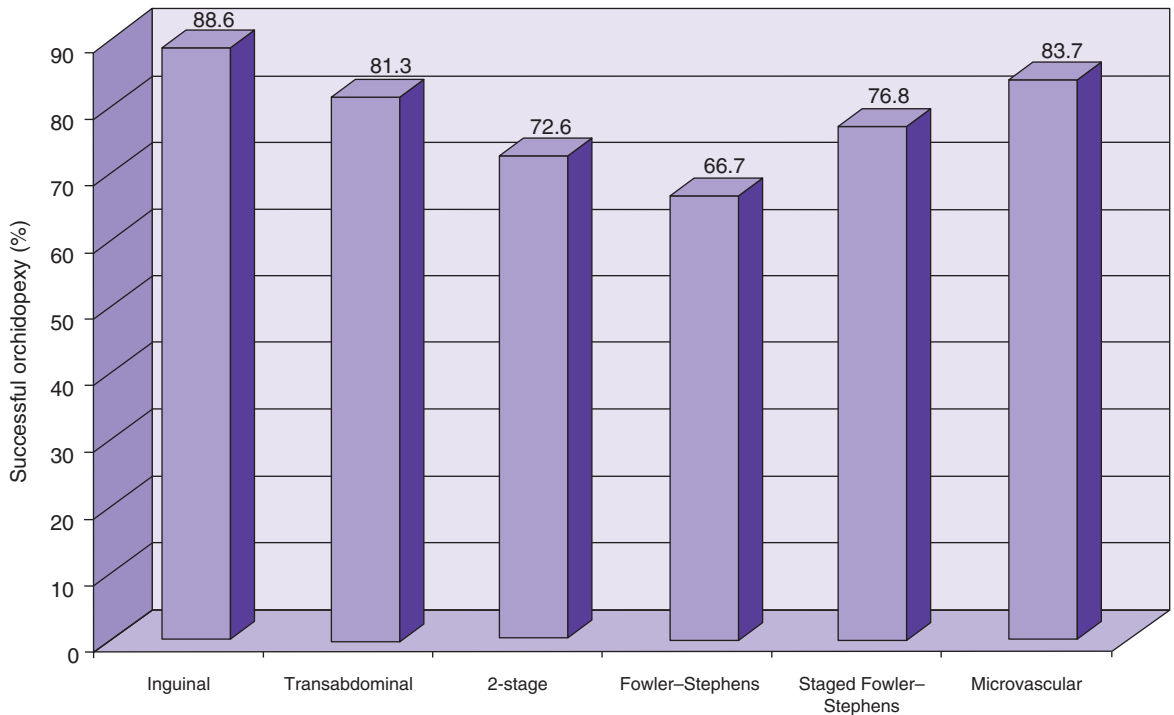


Figure 22.2 Historical success rates for orchidopexy in relation to type of orchidopexy. (Data from literature review by Docimo 1995 [4]. Reproduced with permission of Elsevier.)

A recent systematic review analyzed 1558 orchidopexy surgeries initiated with a transscrotal incision. Patient' ages ranged from 5 months to 21 years. Of the 16 studies assessed, 13 used high scrotal incisions and three performed surgery via a low scrotal incision. In 55 cases (3.53%) an inguinal incision was necessary to complete the procedure. Recurrence was observed in nine cases, testicular atrophy in three, testicular hypotrophy in two, surgical site infection in 13 and reported success ranged from 88–100% [21]. The success of scrotal orchidopexy has been subjected to a prospective randomized controlled trial with results comparable to the standard inguinal approach [22].

Outcomes for impalpable testes

Inguinal approach and preperitoneal orchidopexy

In a number of series, management of the nonpalpable testis with open surgery has proved successful. Kirsch et al. reported on 1866 boys with UDTs of which 447 (24%) were impalpable [23]. Surgery

was successful in identifying testis location or blind-ending vas and vessels in 100% of cases. Of 91 intra-abdominal testes, 33 were managed with inguinal orchidopexy and transperitoneal mobilization of the vas and vessels without vessel transaction. Results were excellent (good scrotal position and size) or acceptable (palpably normal testis in the high scrotum) in 32/33 (97%). A more recent but smaller series of 23 boys operated for nonpalpable testes underwent 28 orchidopexies (5 cases bilateral) via a standard inguinal approach, with no intraoperative complications and all testes normal in size and within the scrotum after 6 months follow-up [24]. Youngson and Jones published on 90 boys with an impalpable testis utilizing a muscle-splitting preperitoneal orchidopexy for 28 intra-abdominal and 42 canalicular testes with success in 66 (94%) at 1 year [25]. However, on late follow-up after a mean of 11 years (range 6–16 years) this figure had reduced to 81%, with only 57% of the testes normal in size. Gheiler et al. have described initial laparoscopy and a subsequent open Jones

orchidopexy for intra-abdominal testes with success in 18 of 19 (94%) cases [26].

Staged orchidopexy

In Docimo's analysis of the literature, staged orchidopexy was successful in 180/248 (72.6%) testes [4] (see Figure 22.2). However, staged orchidopexy can result in high failure rates. Corbally et al. reported on 33 boys who had two orchidopexies on the developing testis and documented a high failure rate for intra-abdominal and canalicular testes with testis atrophy in 40% and a mean volume loss of 46% in the majority of remaining testes [27]. Although the role of staged orchidopexy has become largely historical, as successful one-stage techniques for transfer of the high intra-abdominal testis have emerged (e.g. laparoscopic orchidopexy, Fowler–Stephens orchidopexy (FSO), and microvascular transfer), modified techniques of staged orchidopexy have been reported with gratifying results. In a series of 38 cases, 45 testes (7 bilateral) that were intra-abdominal or peeping underwent a first stage mobilization with testicular vessel preservation and wrapping of the cord with polytetrafluoroethylene pericardial membrane (Preclude®), followed by second stage orchidopexy after a 9–12 months interval. All testes were in a satisfactory scrotal position with stable or increased testicular volume at 1- to 8-year follow-up [28].

Fowler–Stephens orchidopexy

In some cases, the high intra-abdominal testis may not be amenable to either extensive mobilization or staged orchidopexy because of an extremely short vascular pedicle. In these circumstances, the testis can be mobilized on a vasal/peritoneal mesentery containing collateral circulation having divided the testicular vessels well above the body of the testis. The testis continues to be perfused by branches of the deferential and cremasteric arteries as described by Fowler and Stephens [29]. Some authors have suggested an advantage to staging this operation with testicular vessel ligation with minimal testicular handling as a first procedure and gonadal vessel transaction, testicular mobilization, and orchidopexy several months later following robust collateral vessel development [30].

A systematic review of the FSO analyzed a total of 61 articles. Nine (204 testes) included the success of single-stage FSO, 36 (751 testes) including success of two-stage FSO, and 16 that discussed success rates of

single and two-stage FSO (232 high intra-abdominal testes managed by single stage and 218 by two-stage FSO). There were no randomized trials and most studies were cohort or case series, with a mix of open and laparoscopic procedures. The success of two-stage FSO was higher than a single-stage approach (85% vs. 80% with an odds ratio 2 in favor of two-stage FSO). There was no difference between open or laparoscopic techniques [31]. In cases with a long loop vas an open approach to the second stage of a two-stage FSO is advisable because of reported high atrophy rates (83%) following laparoscopic orchidopexy [32].

An interesting technical variation incorporating low spermatic vessel ligation, straightening of the looped vas and preservation of collateral circulation has been reported by Koff and Sethi [33]. In their series of 33 patients with intra-abdominal testes or testes visible at the internal inguinal ring, low vessel ligation resulted in successful orchidopexy in 38 of 39 testes (97%) examined at 1 month and 25 of 27 (93%) at 1 year follow-up.

Microvascular orchidopexy

From a logical point of view, testicular autotransplantation should, by maintaining a full blood supply to a high inguinal/intra-abdominal testis, maximize the potential of future testis development and avoid the significant testicular loss and atrophy rates seen with staged and FSO. First described by Silber and Kelly in 1976 [34], the technique of microvascular transfer is technically demanding, requires specialized instrumentation, and is time-consuming (total operative time 2.5–3h). These factors have undoubtedly played a role in limiting its widespread application.

Occasionally, the vas deferens is also short and a scrotal position cannot be achieved despite microvascular transfer. A novel technique of vasal mobilization and testicular inversion has been described to overcome this pitfall [35]. In addition, in cases where the establishment of arterial inflow proves difficult, the testis will often survive on collateral circulation provided a successful venous anastomosis is performed – the “refluo” technique [36].

Most large series report good results with an adequately sized testis in the scrotum in 80–90% of patients postsurgery (see Table 22.1, Figure 22.2 and references [4,37–41]). Less successful results could reflect a learning curve for this technically demanding surgery.

Table 22.1 Results of microvascular orchidopexy including laparoscopic testicular autotransplantation.

Author	Year	Patient age (mean) in years	Number of testes	Number successful (%)
Wacksman et al. [37]	1982	1.9–20 (9.7)	7	6 (86)
Upton et al. [38]	1983	2–18	10	6 (60)
Bianchi [39]	1995	2–15	51	47 (92)
Boeckx et al. [40]	1998	3.25–15.75 (7.9)	25	24 (96)
Tackett et al. [41]	2002	0.5–13 (3.6)	17	15 (88)

Complications of orchidopexy

Intraoperative

- Failure to achieve a dependent position in the scrotum.
- Tearing of the hernial sac.
- Injury to vas and/or testicular vessels.
- Inadvertent torsion of spermatic vessels during testicular tunnelling.
- Tension on vascular pedicle.
- Avulsion of testicular vessels.
- Ilio-inguinal nerve injury.

Early postoperative

- Pain.
- Bleeding.
- Hematoma.
- Local edema.
- Wound separation.
- Wound infection.

Late postoperative

- Testicular malposition or re-ascent.
- Testicular atrophy.
- Torsion of testis.
- Inguinal hernia.
- Hernia alongside peritonealized vas after FSO – rare complication reported only as isolated case report.
- Ureteral obstruction due to vasal compression after FSO – rare complication reported only as isolated case report.
- Impaired spermatogenesis and infertility.
- Testicular malignancy.

Preventing complications of orchidopexy

The complications of orchidopexy can be prevented by appropriate case selection, choosing the right surgical

procedure for the individual patient, and by adhering to a philosophy of gentle tissue handling and meticulous surgical technique.

Preoperative considerations

Retractile testis, congenital UDT and acquired UDT are three different entities that should be differentiated clinically and managed accordingly [42–44].

Retractile testes

It is important to make an accurate diagnosis and correctly identify retractile testes as the majority end up in a satisfactory position long-term without surgery [45] and, as reported by Puri and Nixon [46], these patients have normal fertility following conservative treatment. The concern with retractile testes is the risk of ascent, which is reported to occur in 7–32% of cases. Boys with retractile testes should be kept under annual review until beyond puberty, reserving orchidopexy for testes that can no longer be brought down into the scrotum [47].

Nonpalpable testes, contralateral hypertrophy, and initial scrotal exploration

In patients with nonpalpable testes, lubricating the examining hands with liquid soap may assist in identifying a difficult-to-feel testis and avoid further investigation or inappropriate laparoscopy [48]. Most pediatric urologists and surgeons prefer laparoscopy for the assessment of the impalpable testis. However, in cases with a nonpalpable testis and contralateral hypertrophy, an initial scrotal incision may be more appropriate as 90–100% of patients have features consistent with the “vanishing testis syndrome” and perinatal torsion [48–51]. In the study by Hurwitz and Kaptein [48] in patients with a unilateral nonpalpable testis, hypertrophy with a testis length 1.8 cm or greater predicted monorchia with an accuracy of approximately 90%, and in the series

published by Belman and Rushton [51] of 22 boys with a left nonpalpable testis and hypertrophied right testis 19 (86.4%) were found to have scrotal nubbins on initial scrotal exploration. Laparoscopy was reserved for three cases where scrotal exploration was negative, with two vanishing testes and one intracanalicular testis found.

Snodgrass et al. [52] have taken this scrotum-first approach for the nonpalpable testis further with the suggestion that laparoscopy be reserved for cases where a scrotal nubbin is not identified and in patients where a patent processus vaginalis is found. In their series of 40 boys with a unilateral impalpable testis managed with an initial scrotal incision followed by laparoscopy, the scrotal exploration revealed 22 (55%) scrotal nubbins, four (10%) extra-abdominal testes and six (15%) patients with a long looping vas associated with an intra-abdominal testis. Laparoscopy documented 13 (32.5%) intra-abdominal testes and one intra-abdominal vanished testis. Interestingly, laparoscopy falsely diagnosed an intra-abdominal vanished testis in six (15%) boys who had scrotal nubbins [52]. Callewaert et al. have adopted this scrotal incision approach for both palpable and impalpable UDTs and of 194 scrotal orchidopexies, 36 of the 46 impalpable testicles (78%) could be diagnosed and treated accordingly, with 11 (73%) of 15 abdominal testes amenable to scrotal incision orchidopexy, due to an associated open patent processus vaginalis [53]. Not all palpable testes will be felt in the outpatients and a careful examination under anesthesia, and prior to a surgical procedure, should be performed as 18% of impalpable testes become palpable with the patient asleep [54].

The ascending testis

The entity of acquired UDT, where a previously normal scrotal testis retracts into an ectopic position, is a recently described phenomenon [55–57] with a prevalence of 1.2% age 6, 2.2% age 9, and 1.1% age 13 [58]. There is no consensus on etiology or correct management for these cases although recent data support a conservative approach. Sijstermans et al. followed 139 boys with 164 acquired UDT and documented spontaneous descent at puberty in 76% of testes (early puberty in 71.4% of these, 26.5% mid puberty, 2.1% late puberty) [59], with their expectant policy for the ascending testis reducing orchidopexy rates in their hospital by 61.8% [60]. Interestingly, long-term testicular

growth following spontaneous pubertal descent or pubertal orchidopexy is reported to be within the normal range [61], whereas cases undergoing orchidopexy at diagnosis have significantly reduced testicular volumes on follow-up [62], so appropriate management of the acquired UDT is unclear. A randomized controlled trial assessing treatment of acquired UDT with orchidopexy at diagnosis vs. a conservative approach would be helpful. Whether conservative treatment will affect future sperm counts, fertility or malignancy risks in boys with acquired UDT is yet to be investigated.

Benefit of preoperative investigation

Ultrasound and standard magnetic resonance imaging (MRI) are unreliable for investigating the impalpable testis. Two different groups of investigators have, however, documented the accuracy of gadolinium (Gd)-enhanced MRI, with sedation, in localizing intra-abdominal testes, canalicular testes, hypoplastic/atrophic testes and vanishing testes, with a sensitivity between 96% and 100% [63–65]. One article looking at a cost/risk analysis suggests that with MR angiography and observation of testicular nubbins, a substantial number of boys could forego operative intervention with minimal additional risk and no increased health care costs [66]. So far, although clearly Gd-MRI can be very reliable in experienced hands, it has yet to replace laparoscopy as the investigation of choice for the impalpable testis.

Operative considerations

Surgical technique

A good understanding of the operative principles for orchidopexy is required to prevent complications. The technique of inguinal orchidopexy was first reported by Bevan [67], with modifications described by Gross and Jewett [68], and subsequently Koop and Minor [69]. The history of cryptorchidism and its early surgical correction is elegantly described in an article by Tackett et al. [70]. For a contemporary description of orchidopexy, the reader is referred to a major operative pediatric surgery textbook [71].

Complications can be prevented by:

- Careful positioning of skin incisions to provide optimum cosmetic results. In the groin a skin crease incision is used and in the scrotum, if the incision is placed along a rugosal fold, subcuticular skin closure

makes the subsequent scar virtually invisible (see Videos 22.1 and 22.2). The author advises a two-layer closure of the scrotum to remove any chance of wound disruption and possible testis extrusion post-operatively (see Video 22.2).

- Early identification of the testis, once Scarpa's fascia is opened. This will prevent inadvertent testis injury and can often be achieved by passing an index finger down into the scrotum in preparation for the future subdartos pouch. On removing the finger the palpable UDT usually pops directly into view within the operative field (see Video 22.1).
- Division of all attachments, including the gubernaculum, the cremasteric fibers, and the lateral spermatic fascia (see Videos 22.3 and 22.4).
- Adequate retroperitoneal dissection, division of lateral bands with identification of the patent processus vaginalis in the anteromedial surface of the cord, and performing a high ligation. The sac/processus is usually twisted prior to transfixion and division, and it is important not to trap the vas or vessels (see Video 22.5).
- Gentle handling of the vas and gonadal vessels. On no account should they be held or picked up with forceps.
- Prevention of tension on the cord structures which is likely to lead to ischemia or re-ascent.
- Creation of a subdartos pouch and, if possible, avoidance of suture fixation (see Video 22.6).
- Careful assessment of the orientation of the vascular pedicle prior to testicular tunneling to prevent torsion and subsequent ischemia (see Video 22.6).
- Positive identification of the ilio-inguinal nerve just beneath the external oblique and its preservation.

Full mobilization of the testis with division of the hernia sac or processus and adequate retroperitoneal dissection are key to a satisfactory outcome. Davey [72] studied the relative importance of each step in 313 orchidopexies and found that sac/processus division accounted for 60% of increased cord length, whilst the remaining 40% was related to dissection within the internal ring and division of tethering lateral bands. A thorough knowledge of inguinal and retroperitoneal anatomy is required to prevent complications, and failure to achieve a satisfactory dependant scrotal position for the testis is often related to inadequate retroperitoneal mobilization. The reader is referred to the articles published by Hutcheson et al. [73] and Redman [74] on the applied anatomy of this region. If, despite full

mobilization, there is still insufficient length, the cord structures can be redirected medial to the inferior epigastric vessels for a shorter route to the scrotum – the “Prentiss” maneuver [75–77].

In cases being assessed for a FSO, the procedure should be avoided when major ductal anomalies are present. Any case with absence or atresia of a segment of the vas or non-union of the vas and testis is likely to have inadequate collateral testis blood supply following testicular vessel transection. In addition, the Fowler–Stephens procedure should be avoided in cases with an intrinsically short vas [78] and in reoperative cases [79].

With intra-abdominal testes it can be difficult to decide which procedure would be best suited to achieve a dependant scrotal position. Banieghbal and Davies [80] have used testicular mobility assessed at laparoscopy as a guide to management, and of 20 intra-abdominal testes that could be stretched to the contralateral internal inguinal ring a successful conventional orchidopexy was achieved. Other surgeons have predicted a successful orchidopexy without vessel division when the testis lies within 2 cm of the internal ring. When a small dysplastic intra-abdominal testis is present, orchidectomy is the best option. For microvascular orchidopexy a major problem can be the size discrepancy between the larger deep inferior epigastric artery and testicular artery. This can be overcome with accurately placed mattress sutures [40] or by creating an arteriovenous fistula between the inferior epigastric artery and vena comitans, which increases run off and prevents microanastomotic thrombi within the transplanted testis [81].

The creation of a subdartos pouch provides the best form of testis fixation [82,83]. In infants, when performing the orchidopexy through a small 1.5–2.0 cm inguinal skin crease incision, there may be little space to pass an index finger down to the scrotum for pouch preparation without compressing and traumatizing the cord structures in the corner of the wound. A novel and commercially-available testicular tunneler (Surgical Innovations Limited, UK) can assist in creating a direct path to the scrotum (Figure 22.3 and [84]). Once the tunneler has been passed from the inguinal incision and out through the scrotum, the testis is guided into position following the placement of a suture between the gubernaculum and end eye of the tunneler (see Video 22.6). The testis is then placed in the prepared subdartos pouch. Re-ascent may be further prevented by narrowing the subdartos fascia at the scrotal neck,



Figure 22.3 Lambert testicular tunneler.

but care is required not to constrict the testicular vessels [85] (see Video 22.7). On occasion, the surgeon may deem it necessary to suture the testis to the midline scrotal septum to prevent re-ascent although there may be inherent risks.

Coughlin et al. [86] have reported a link between suture fixation at orchidopexy and infertility in previously cryptorchid men (relative risk 7.56; 95% CI, 1.66, 34.39) and experimental studies performed by Bellinger et al. [87] in rats have documented inflammatory and necrotic changes after suture fixation. It is unclear, however, if these findings can be extrapolated to humans as follow-up imaging studies have noted very minimal changes despite suture fixation at orchidopexy. Ward et al. [88] performed testicular ultrasound on 22 men operated in childhood with suture fixation and showed a single tunica albuginea calcification (1–2 mm) in seven (32%), and a further three (14%) patients with a single hypoechogenic subtunical cyst (1–2 mm). The remaining 12 (54%) patients had normal scans and no difference was noted in testis size between the normally descended and operated testes in any patient. Theoretically, breaching the tunica with a suture may lead to disruption of the blood/testis barrier and antibody formation, but in a study by Mirilas et al. [89] on 22 pubertal males (aged 12.1–17.7 years) operated for cryptorchidism before puberty sera were negative for anti-sperm surface antibodies in all patients. If a suture is required, PTFE may be ideal due to its softness and specific handling characteristics [90]. Fibrin glue as

an alternative to sutures has been described in a rat model [91].

Postoperative considerations

There are no specific precautions in infants. In older boys, most surgeons advise restricted physical activities (riding a bike, kicking a football) for several weeks after orchidopexy until healing has fixed the testis in the subdartos pouch.

Managing complications of orchidopexy

Initial

A torn hernial sac requires careful identification and dissection free from the other cord structures prior to proximal transfixion. Micromosquito forceps are invaluable in holding the edges of delicate sac while control is achieved. Inadequate closure or failure to spot a torn sac may lead to a subsequent inguinal hernia. Although a transected vas is a rare event, the correct management is primary microsurgical vasovasostomy. If a testis cannot be placed in the scrotum, despite full mobilization and a Prentiss maneuver, it should be positioned as low as possible with a planned second procedure after 6–12 months. Small wound hematomas are likely to settle with conservative treatment and wound infection will respond to antibiotics and, if necessary, wound drainage.

Definitive

If there has been a vascular insult at the time of orchidopexy resulting in atrophy, the testis is lost. Some boys decide to have a testis prosthesis at puberty for cosmetic concerns. Postoperative hernias are uncommon and managed by herniotomy and herniorrhaphy if there is a direct component. Testis torsion after previous orchidopexy is rare but requires emergency exploration with de-torsion and repeat fixation or orchidectomy if the testis is nonviable [92].

Re-ascent of the testis requires a redo orchidopexy, which can be performed via the original inguinal incision [93,94] or with a scrotal approach [95–98]. Surgery is usually made difficult because of scar tissue, and careful dissection is required to prevent vas or testicular vessel injury (see Videos 22.3 and 22.5). In both techniques early identification of the testis

is important, with retrograde dissection of the cord structures to gain adequate length. When an inguinal approach is used, a strip of external oblique aponeurosis overlying the cord may be left attached, thus avoiding difficult dissection between the scarred external oblique, related to previous incision and closure, and the anterior aspect of the spermatic cord [94,99,100]. The previously divided hernia sac needs to be separated from the vas and vessels, the peritoneum swept away and retroperitoneal dissection completed. If scar tissue around the deep inguinal ring makes dissection problematic, opening the peritoneum above the ring and dissecting down from above may avoid potential vas or vessel injury [94,99]. A Prentiss maneuver may be required to achieve a dependant scrotal position and good operative exposure, excellent lighting, optical magnification and tension-free placement within a scrotal subdartos pouch are important in achieving a satisfactory outcome.

Outcome of redo orchidopexy

Most papers on repeat orchidopexy include patients that had initial surgery for inguinal hernia, hydrocele, or cryptorchidism and results for these different groups are often amalgamated. A successful result has been documented in 89–100% of cases [93–104] (Table 22.2). In most series, the length of follow-up has been short or not stated. In the report by Pesce et al. [102] of 20 boys followed beyond puberty, 65% had significantly reduced ultrasound-determined testicular volumes (when compared to controls, $P < 0.005$, although volumes were respectable, study group mean 12.7 ml SD 3.96; controls mean 15.4 ml SD 3.11). With regard to fertility, seven (35%) of the 20 had slightly impaired and three (18.7%) severely impaired sperm analysis [102].

Orchidectomy

The removal of a testis in childhood is usually for non-viability after acute torsion, for small dysplastic UDT, for testicular nubbins related to a nonpalpable testis, or for atrophy documented at exploration for a previously failed orchidopexy. Immediate complications following orchidectomy are related to wound hematoma and infection. In cases of torsion the contralateral testis is fixed using a subdartos pouch or suture fixation

to prevent metachronous torsion. Opinion is divided whether the contralateral testis should be fixed in other cases requiring orchidectomy. Implantation of a testicular prosthesis is available at puberty for boys with a solitary testis and cosmetic concerns or psychological issues [105]. Complications include infection, hematoma, extrusion, unsatisfactory size or positioning, and implant rupture. A groin incision is usually preferred for insertion because of lower risks for infection and extrusion. Saline-filled prostheses can deflate but appear safe and well tolerated [106]. Testicular torsion in adolescents and young adults is complicated by abnormalities of spermiogenesis and infertility with semen analysis normal in only 5–50% of patients on long-term follow-up [107]. In contrast, although data are limited, torsion in prepubertal boys does not seem to affect subsequent fertility [108].

Neonatal testicular torsion can be an indication for orchidectomy but the correct management of this condition is not entirely clear – a conservative approach is adopted by some, as most torsion events are prenatal and the testes nonsalvageable. A survey of consultant pediatric surgeons and urologists working in the United Kingdom and Ireland showed 82 surgeons (74.5%) explore the scrotum, and 59 (71.9%) perform ipsilateral orchidectomy and contralateral orchidopexy of the “healthy” testis, with few surgeons undertaking emergent exploration [109]. A pragmatic approach to the need for emergent surgery, depending on the time of presentation (not present at birth) or the presence of bilateral involvement seems reasonable [110].

Conclusion

Considerable operative expertise is required for successful infant orchidopexy. Inguinal or scrotal approaches to the palpable UDT are successful in >95% of cases. The impalpable testis can be managed by a variety of techniques with success in >85–90% of cases. Testis atrophy is avoided by careful dissection, gentle tissue handling, and meticulous surgical technique. Although uncommon, the failed orchidopexy is successfully salvaged by redo surgery in >85% of cases. In boys where an orchidectomy is performed, the possibility of testicular prosthesis insertion should be discussed.

Table 22.2 Results of redo orchidopexy.

Author	Year	Patient age in years	Number of operated testes	Follow-up in years	Complications (%)	Successful redo surgery (%)
Maizels et al. ^a [93]	1983	1–15 (median 6)	36 (2 orchidectomies)	Not stated	0	34 (100)
Cartwright et al. ^a [94]	1993	1.4–11	25	≥ 0.33	1 (4) re-ascent 1 (4) hypoplasia	23 (92)
Cohen et al. ^a [99]	1993	2–12 (mean 7.25)	27	0.5–1.5 (mean 0.74)	0	27 (100)
Palacio et al. ^a [100]	1999	4–16 (mean 7.2)	29	0.67–3.2 (mean 1.75)	0	11 (100) ^b
Redman [101]	2000	mean 4.8	13	1–5.1 (mean 2.6)	0	13 (100)
Caruso et al. ^{a,c} [95]	2000	2–14 (average 9)	15	0.12–1	1 (6.7) testicular infarction	14 (93.3)
Pesce et al. [102]	2001	6–15 (mean 9.3)	41 (7 orchidectomies)	2–15	1 (2.9) testis atrophy	33 (97.1)
Rajimwale et al. ^{c,d} [96]	2004	–	25	0.12–1	1 (4) re-ascent	24 (96)
Ziylan et al. [103]	2004	Mean 6.8	32 (1 orchidectomy)	1–7 mean 3.8	2 (6.5) high scrotal	29 (93.5)
Tong et al. ^e [104]	2009	2.5–13 (mean 5.5)	35	0.5–2.7 (mean 1.8)	4 (11.4) atrophic	31 (88.6)
Karaman et al. ^{a,c} [97]	2010	3–14 (mean 6.7)	16	0.25–3 (mean 1)	0	16 (100)
Dudley et al. ^{a,c} [98]	2011	1.3–16.2 (mean 6.4)	27	0.02–5.8 (mean 1)	1 patient (4.2) high testis 1 patient (4.2) fungal skin rash	27 (100) after redo/redo surgery in 1 patient

^a Study included patients with an UDT following previous orchidopexy, inguinal herniotomy or division of patent processus vaginalis for hydrocele.

^b Results reported only for patients undergoing cordopexy (suturing of retained external oblique aponeurosis on anterior surface of cord to the pubic bone or tendinous part of the gracilis muscles).

^c Surgery performed via a scrotal approach. Inguinal conversion required in 6–20% of cases.

^d Paper included 85 patients undergoing 100 'Bianchi' scrotal orchidopexies with mean age 3.2 years, age of patients with secondary trapped testes not provided separately, 3/25 (12%) of trapped testes required inguinal conversion for success.

^e Some patients operated with laparoscopy-assisted surgery.

DOS AND DON'TS**Do**

- Identify the testis early during dissection
- Carefully separate any hernia sac from vas and vessels, with transfixion at the deep ring
- Divide all cord attachments including the lateral spermatic fascia
- Perform adequate retroperitoneal dissection
- Utilize techniques such as the Prentiss maneuver to reduce tension on the cord structures
- Maintain adequate hemostasis prior to wound closure
- Use micro-mosquito forceps where applicable

Don't

- Cut or damage the ilio-inguinal nerve
- Pick up the vas or vessels with forceps
- Ensnare the vas and/or vessels while transfixing the sac
- Traumatize the cord structures whilst using a finger technique to create a subdartos pouch
- Overlook a twist in the testicular vessels prior to tunneling
- Suture the testicle unless absolutely necessary
- Place the testis in the scrotum under tension

CLINICAL PROBLEM/CASE: DISCUSSION

- 1 An empty hemiscrotum could be due to a palpable UDT, ectopic testis, impalpable UDT, a vanishing or retractile testis. A two-handed examination technique should attempt to milk any testis toward the scrotum. If there is difficulty then lubrication using soap or gel may reduce friction and allow a previously impalpable high testis to be appreciated. A retractile testis is a normal-size testis that can be brought fully to the base of the scrotum without difficulty, with no tightness on the cord structures that remains in position until stimulation of the cremasteric reflex. Ectopic testes (outside the normal line of descent) and palpable UDT may be manipulated into the scrotum, in some cases, but readily return to the groin when released. Don't forget to inspect and palpate the suprapubic region, femoral triangle and perineum when the testis is impalpable, so not to miss rare ectopia to these sites.
- 2 Many diagnostic modalities are mentioned in the literature to locate the impalpable testis including ultrasound, magnetic resonance imaging and technetium scans. Most pediatric urologists and surgeons, however, do not rely on radiological studies due to concerns over false negative rates, and recommend diagnostic laparoscopy as the procedure of choice for investigating impalpable testes.
- 3 The child should have an examination under anesthesia. Most surgeons would perform a scrotal orchidopexy if the testis is easily manipulated to the scrotum, a mobile testis, and advise an inguinal orchidopexy for a more fixed testis. There is no role for laparoscopy where the testis is palpable.
- 4 Division of the lateral spermatic fascia and gubernaculum, separation, transfixion and division of the patent processus vaginalis, adequate retroperitoneal dissection, and division of tethering lateral bands is usually enough to bring a palpable UDT to the scrotum. If despite full mobilization there is still insufficient length, the cord structures can be redirected medial to the inferior epigastric vessels, for a shorter route to the scrotum – the Prentiss maneuver. A careful subdartos pouch with narrowing of its proximal limit around the cord structures can help prevent retraction and re-ascent.
- 5 Injury vas/vessels, testicular atrophy/ascent, postoperative hernia and reoperation. Although testicular torsion is very rare this still can happen. The relative risk of malignancy is increased 5–10 times in children with UDTs in comparison to the normal population. Positioning the testis in the scrotum will facilitate easier diagnosis of malignancy if this happens.
- 6 Fertility potential after orchidopexy in unilateral ectopic, canalicular and emergent testes, as long as surgery is performed in early childhood, is good (>90%) and fertility for most cases of unilateral intra-abdominal testis and patients with unilateral anorchia or vanishing testis is expected, whereas the majority of patients with bilateral intra-abdominal testes are infertile. Patients with retractile testes have normal fertility after conservative management.

- 7 In cases of recurrence a redo orchidopexy is warranted via an inguinal or scrotal approach, by an experienced pediatric surgeon/urologist. In some cases, a missed hernial sac or patent processus vaginalis is found and division provides considerable increased cord length. Early identification of the testis is important and retrograde dissection may incorporate a strip of external oblique aponeurosis, to avoid dense scar tissue between this and the cord structures. Sometimes an intraperitoneal approach from above facilitates finding the correct anatomical plane, allowing subsequent dissection to proceed smoothly.

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CHAPTER 23

Laparoscopic orchiopexy

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KEY POINTS

- Laparoscopy is the gold standard for localizing the nonpalpable testis
- Primary laparoscopic orchiopexy is successful in 97% of cases; Fowler–Stephens orchiopexy success rates are reduced compared with primary laparoscopic orchiopexy but are still successful in three-quarters or more boys
- Laparoscopic orchiopexy is very safe, with testicular atrophy being the most common complication
- Compliance of the pediatric abdomen increases risk to intra-abdominal structures; therefore, careful attention to landmarks and the reduced working space of the pediatric abdomen is necessary to avoid complication
- Complication rates are low for laparoscopic orchiopexy. Large series approximate 3% major and 2% minor complication rates, which are significantly influenced by both surgeon experience and ongoing case volume

CLINICAL PROBLEM/CASE

Case 23.1

You are lecturing to residents and while preparing your talk you wish to determine:

- 1 How many syndromes are presently associated with cryptorchidism?
- 2 What are the driving factors of testicular descent, and when do these occur during fetal development?

Case 23.2

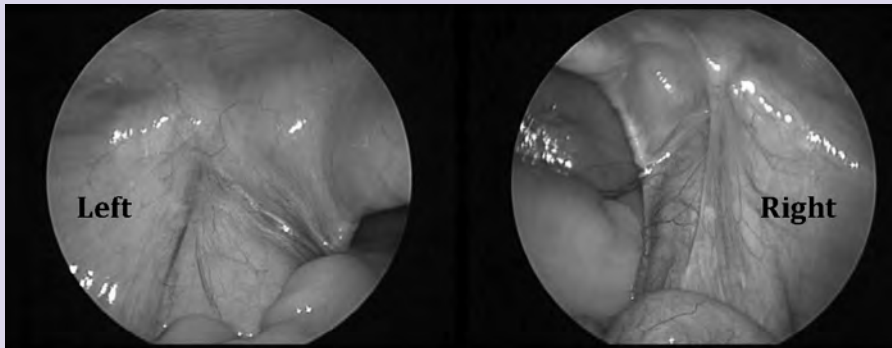
A non-syndromic, obese, prepubertal 9-year-old boy with bilateral nonpalpable testes is referred from Endocrinology with the following testing. Bilateral inguinal ultrasound does not clearly identify testes. His parents are uncertain if the testes were ever palpable. He has a normal prenatal history and 46XY karyotype.

An hCG stimulation test using 1500 IU delivered IM twice weekly for 3 weeks has the following results, though the testes remain nonpalpable:

	LH (mIU/ml)	FSH (mIU/ml)	Testosterone (ng/dl)	DHT (ng/dl)
Pre-hCG	0.40 (N)	4.33 (N)	8 (N)	<5 (N)
Post-hCG	0.19 (N)	4.01 (N)	220 (H)	30 (H)

(N) represents a normal value for age and pubertal status.
(H) represents a high value for age and pubertal status.
DHT, dihydrotestosterone; FSH, follicle stimulating hormone; hCG, human chorionic gonadotrophin; LH, luteinizing hormone.

- 1 What surgery do you recommend to parents, and what do you expect to find based upon the hCG stimulation test? Intraoperative findings of both internal inguinal rings are depicted in the following images:



- 2 What is the next surgical step?
- 3 A testis biopsy was performed at the time of orchiopexy. What would you expect to find histologically?
- 4 Parents question if you can give a prognosis for future fertility. What do you answer?

Case 23.3

A healthy 2-year-old male has undergone first-stage Fowler–Stephens laparoscopic right orchiopexy for a high intra-abdominal testis 6 months ago. He has a normal, descended left testis. At the time of his second-stage orchiopexy, you divide the spermatic vessels and carefully

mobilize the collateralized vascular pedicle between vas and vessels. You note persistent tension on the pedicle after delivery of the testicle into the scrotum. Tension remains even after desufflating the abdomen.

- 1 What surgical steps may alleviate tension on the already mobilized vascular pedicle?
- 2 If the testicle achieves adequate scrotal positioning but there is still concern for *mild* tension, what temporary maneuver can allow the cord to stretch?
- 3 If there is too much tension on the pedicle for the testis to achieve an examinable scrotal position, what is the alternative?

Introduction

Cryptorchidism is present in 30.3% of premature infants and 3.4% of term infants. Rates of cryptorchidism in neonates are also inversely proportional to birth weight. By 1 year of age, 0.8% of boys have persistent cryptorchidism, with nonpalpable testes accounting for 20% of this group [1,2]. Diagnostic laparoscopy is less invasive than open exploration, and the most accurate test available for determining the position of a nonpalpable testis. Laparoscopic orchiopexy is the natural sequel to diagnostic laparoscopy for simultaneous management of the testis, with advantages of improved cosmesis, reduced postoperative pain and shorter recovery when compared with open techniques [3–7] (Figure 23.1). Laparoscopic orchiopexy is safe and highly successful, with testicular position reportedly superior to open series [8] (Table 23.1). Historically, laparotomy was performed via Pfannenstiel or high inguinal (i.e. Jones) incision to localize an intra-abdominal testis or diagnose blind-ending vessels [9]. It is now standard practice to proceed with diagnostic laparoscopy when the testicle is nonpalpable.

Cortesi first reported diagnostic laparoscopy for the evaluation of a nonpalpable testicle in 1976 [10]. In 1991, Bloom first reported the feasibility of Fowler–Stephens laparoscopic orchiopexy in 10 boys [11], with Jordan reporting primary laparoscopic orchiopexy in 1992 [12]. Since that time, laparoscopy has become the gold standard in localizing nonpalpable testes with reports of its superiority to even open exploration. Bargawi and colleagues reported a series of 18 patients with missed intra-abdominal testes or remnants, one with seminoma, from 27 laparoscopic explorations after prior inconclusive inguinal exploration [13].

Terminology

An *ascended testis* is one that has been documented as descended in the first years of life, but is later found to be undescended as the result of primary or secondary/iatrogenic cause. A *retractile testis*, which appears undescended initially, can be brought into the dependent scrotum without a sense of tethering to ectopic

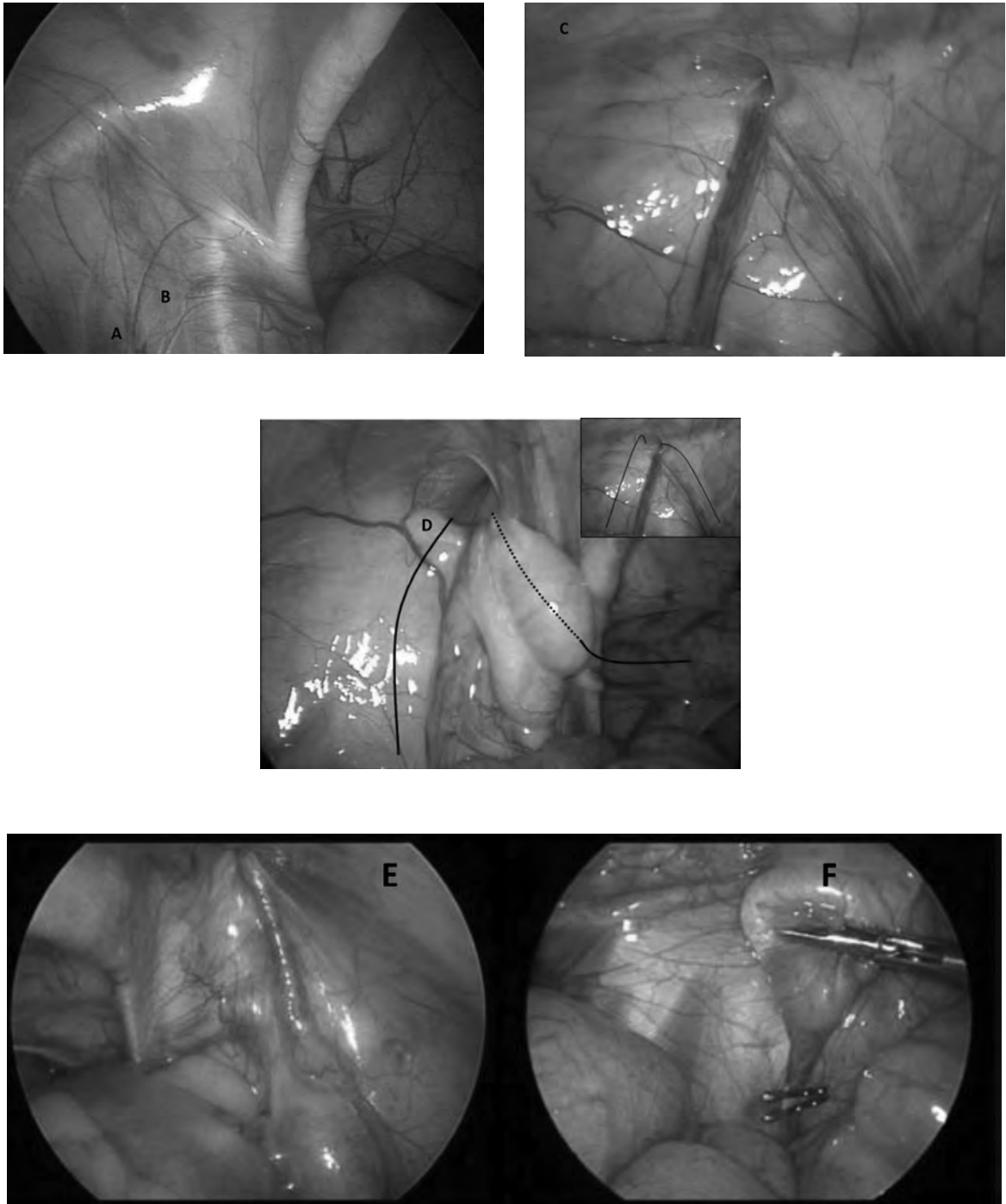


Figure 23.1 Laparoscopic findings with a nonpalpable testis. Atretic vessels with a blind ending vas are seen with a vanishing testis (A, B); both spermatic vessels and vas entering the internal ring suggest intracanalicular gonad (C). A low intra-abdominal testis near the internal ring (or peeping testis) can be intermittently palpable (D). A high intra-abdominal testis, further from the internal ring (E), typically requires a Fowler–Stephens approach with first-stage vascular clipping depicted (F).

Table 23.1 Summary of the largest series and meta-analysis orchiopexy success rates for intra-abdominal testes.

Technique	Primary	One-stage F-S	Two-stage F-S	Complications
Open [8]	81.3%	66.7%	76.8%	N/A
Microvascular [8]	83.7%	N/A	N/A	N/A
Laparoscopic [22,44,45]	91–100%	78–84%	86–93%	Major: 3% Minor: 2–5%
	Atrophy: 0–2%	22.2%	10.3%	
	Malposition: 0–9%	7.4%	1.7%	
SILS [34]	96%	N/A	N/A	0
Needlescopic [66]	100% (<i>n</i> = 8)	N/A	N/A	0
Robotic [28]	100% (<i>n</i> = 1) ^a	N/A	N/A	0

^aPrimary laparoscopic.

F-S, Fowler–Stephens orchiopexy; SILS, single-incision laparoscopic surgery.

structures. It is thought to be the result of the cremasteric reflex, a function of the genitofemoral nerve, which is most active in 3–9-year-old boys. An *ectopic testis* is an undescended testis that is located outside the external inguinal ring, in the superficial inguinal pouch, and can include femoral, perineal or contralateral scrotal locations. *Undescended testes* are generally categorized as intra-abdominal, intracanalicular (proximal or distal), or prescrotal in location, which is an important component to include in the surgical record. *Peeping testes* are those intra-abdominal gonads located at the internal inguinal ring, which slide between the abdomen and inguinal canal and may be variably palpable or nonpalpable on exam. Finally, *vanishing testes* are absent secondary to fetal or neonatal torsion of the spermatic vessels and appear as blind-ending spermatic vessels but normal vas deferens near the internal inguinal ring; this is more common than the rare and difficult to prove case of testicular agenesis. In *vanishing testes syndrome* (bilateral vanishing testes), endocrinologic follow-up is necessary for testosterone replacement therapy to facilitate pubertal development once FSH and LH levels begin to rise [14].

Testicular location

Diagnostic laparoscopy will yield the following scenarios (Figure 23.1):

- Blind-ending testicular vessels with visible vas deferens located proximal to the internal ring (vanishing

testis) are found in 21–50% of cases. This requires no further intervention [15–17].

- A normal-appearing vas deferens and testicular vessels exiting a closed internal ring suggests an intracanalicular testis or nubbin, which requires groin exploration. A nubbin is present in up to 30% of cases. If a nubbin is identified, controversy exists regarding the need for removal. While seminiferous tubules may be found in up to 14% of nubbins, warranting removal by some, only one case of intratesticular germ cell neoplasia has been identified indicating very low malignant potential [18,19]. If a nubbin is present, the internal ring is generally closed. If, on the other hand, the internal ring is open, a high canalicular testis may be withdrawn or “milked” into the abdomen for primary laparoscopic management [20].
- A blind-ending vas may be noted without the presence of testicular vessels indicating gonadal disjunction. Diagnostic laparoscopy should continue with emphasis on identifying the testicular vessels as they will lead to the gonad [21].
- Finally, in 50–60% of nonpalpable cases, an intra-abdominal or peeping testis is identified. With a viable testis, one can expect 44.8% to be >2 cm from the internal ring, 30.6% to be within 2 cm of the ring, 17.4% to be peeping, and 7.1% to be far from the ring and ectopically located [22].

Ultimately, surgical goals include preservation of fertility, reducing risk and monitoring for testicular malignancy, psychological wellbeing, hernia management

and prevention of spermatic cord torsion [14], which is of reportedly higher risk in cryptorchid testes.

Surgical technique

Once the decision is made to proceed with laparoscopic orchiopexy, the length of the vessels will determine if primary laparoscopic orchiopexy is possible and, if not, the surgeon must determine to perform a one- or two-stage Fowler–Stephens orchiopexy. Success of these laparoscopic orchiopexy techniques are assessed by correct scrotal positioning of the testis and lack of atrophy (Table 23.1).

The procedure begins with supine, frog-legged patient positioning with general endotracheal anesthesia and orogastric suctioning. A Foley catheter is placed. Muscle paralysis is typically recommended. The child is padded and secured to the table. The patient will be placed in slight Trendelenburg after port placement, and the table may be rotated as needed to elevate the ipsilateral abdomen for visualizing high intra-abdominal testes. Peritoneal insufflation can be achieved with open or blind techniques. Blind access with a Veress needle is less commonly used in the first few years of life [23] due to increased compliance of the pediatric abdominal wall, which enhances the risk of vascular and bowel injuries. It is the authors' preference to use the Bailez technique for open access [24], with the modification of a radially dilating trocar [25] (Figure 23.2). In our current technique, a 2-0 vicryl suture is first placed as a deep traction suture through the skin of the umbilicus. A 5 mm infraumbilical incision is made, and Metzenbaum scissors are used to spread and cut through umbilical fascia in a cephalad direction. Care is taken not to undermine the umbilical attachments to fascia, by angling the plane of the scissors at an approximately 20° angle over the plane of the abdomen. The goal is to enter umbilical fascia where it most closely approximates the peritoneum. Alternatively, the rectus fascia and underlying peritoneum may be entered sharply at 90° under direct vision. The abdomen is insufflated to 8–10 cmH₂O and a 5 mm 0° camera used to first inspect for trocar-related injury.

Mobilization of the spermatic vessels begins with a peritoneal incision overlying the gubernacular attachment to the testis at the internal ring. At this point, the entire path of the vas deferens should be identified to



Figure 23.2 External view of all working ports for laparoscopic orchiopexy. A Foley catheter is in place with the patient secured to the table. A 5 mm umbilical trocar, two 3 mm abdominal trocars and a 10 mm scrotal trocar have been placed. The area of intra-abdominal testis, near internal ring, is transilluminated with the camera.

prevent transection of a long looping vas, which can descend as far distal as the scrotum. The gubernacular attachments are transected and may be then used as a handle to prevent injury to the testis. Peritoneal incisions are extended lateral to the testicular vessels, then superior and medial to the vas deferens as it approaches the internal ring. Alternatively, one can first incise peritoneum lateral to the vessels and superior to the vas, then move toward gubernacular transection to mobilize an outlined peritoneal triangle [20]. The peritoneal triangle contains collateralized vasculature between the deferential and internal spermatic (testicular) arteries (Figure 23.1D). This entire pedicle is gently dissected free of the external iliac vessels, recognizing the majority of pedicle length will be achieved by mobilizing the peritoneum lateral to spermatic vessels. Adequacy of pedicle length may be assessed by extending the testis to the contralateral internal ring, which also aids in dissecting the peritoneum lateral to testicular vessels. If the length is still inadequate, a perpendicular “relaxing” incision can be delicately created in the peritoneum immediately overlying the testicular vessels at the superior extent of the dissection (Figure 23.3). Intra-abdominal testes in older boys, or those located proximal to the iliac vessels, are more likely to require a Fowler–Stephens procedure to obtain adequate length [26] and are probably best managed with a planned, staged approach (Figure 23.1E, F).

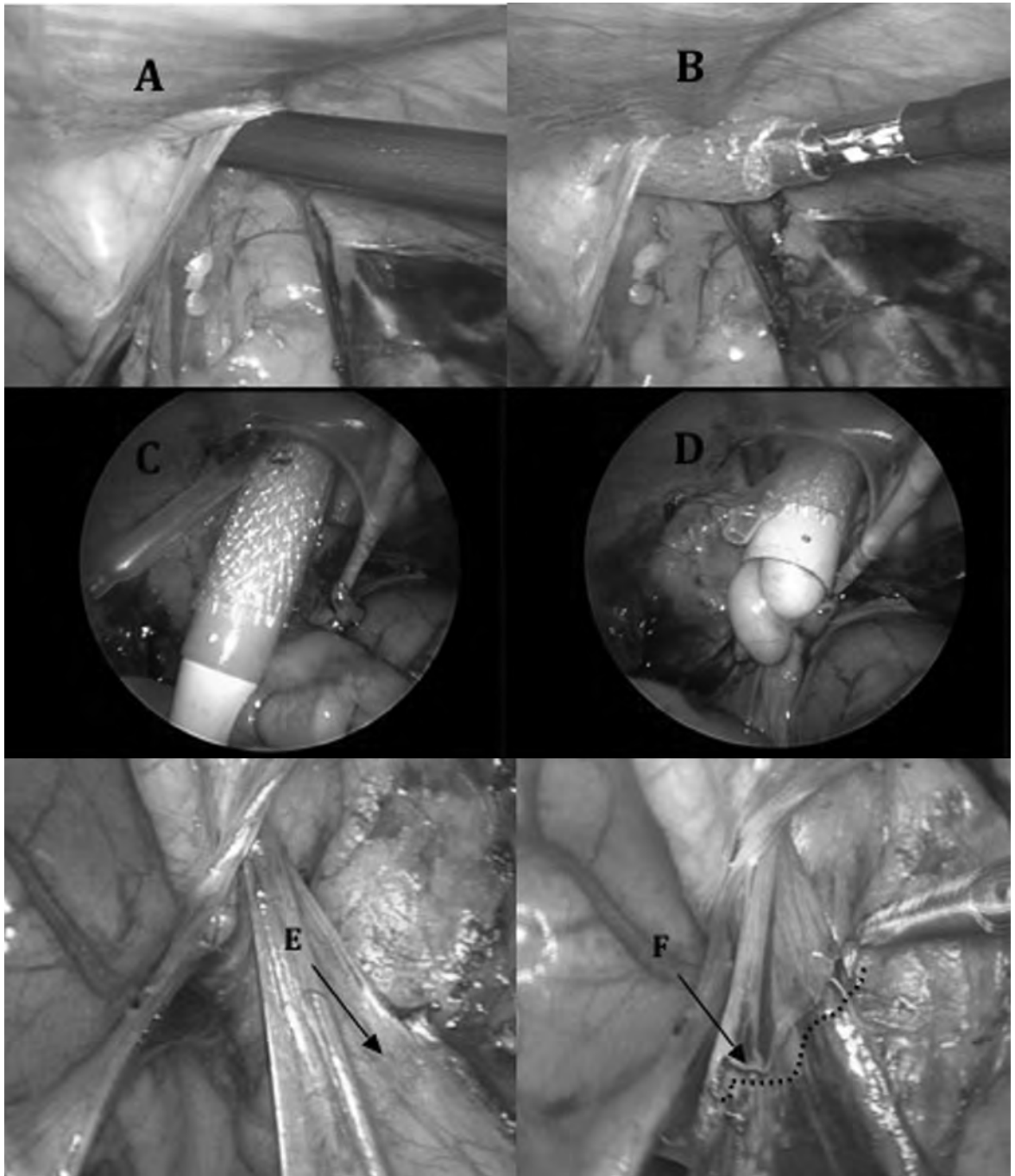


Figure 23.3 Neo-inguinal hiatus. Delivering the testicle into the scrotum requires developing a neohiatus (A, B) to facilitate passage of the testicle, epididymis and cord structures into the scrotum without resistance. Using a 10 mm scrotal trocar (C) opens a neohiatus with minimal resistance to minimize the risk of an avulsion injury and allow straight passage of the vessels (D, E). If there is residual tension on the cord, meticulous incision of the peritoneum overlying collateralized retroperitoneal vasculature can be performed (F).

Aggressive dissection of the vas generally does not add significant length and may risk injury to the bladder or ureter (Figure 23.4). If pedicle length is still not adequate, a Fowler–Stephens orchiopexy is performed by clipping the testicular vessels close to the testis to maximize surface area for collateralization, as described by Koff and Sethi [27]. This will require up-sizing one lateral trocar to 5 mm for introduction of the smallest clip applicator. The decision to perform a one-stage versus two-stage Fowler–Stephens orchiopexy is up to the surgeon; however, if a two-stage approach is planned, dissection of the peritoneal pedicle is generally not performed; the vessels are simply clipped, and the procedure ended with return in six months for the final stage.

Various methods to deliver the testicle into the scrotum have been described. The technique described by our group utilizes a radially dilating trocar to create the neoinguinal hiatus [25]. A 10 mm ipsilateral scrotal incision is first made and a subdartos pouch is created. A 2 or 3 mm laparoscopic grasper is placed through the ipsilateral 3 mm abdominal trocar and directed superior to the pubic arch between the medial umbilical ligament and epigastric vessels. The surgeon's free hand should palpate the pubic ramus, guiding the instrument through the scrotal incision. This antegrade technique is the authors' preferred method for creation of the neoinguinal hiatus. Such maneuvers reduce risk of injury to femoral vessels or the bladder (Figure 23.4). The instrument exiting the scrotum is now used to pull the trocar's sheath through the scrotal incision into the abdomen. A 10 mm trocar is then inserted, creating a smooth tunnel through the neoinguinal hiatus. A locking grasper is advanced through the scrotal trocar to grasp the gubernaculum and deliver the testicle into the scrotum under direct vision, monitoring for cord torsion and tension (Figure 23.3).

New technology

Robotic-assisted laparoscopy was first described for orchiopexy in 2009 [28] using the daVinci® surgical platform. Due to the ease and cost effectiveness [29] of pure laparoscopic management, robotic-assisted laparoscopy for the undescended testis is generally an unnecessary technological advantage. Single-incision laparoscopic surgery (SILS) and laparoendoscopic single-site surgery (LESS) have been slow to gain momentum in pediatric

urology given questionable added benefit over conventional laparoscopic orchiopexy. LESS orchiopexy was first reported by our group in 2010 [30] with other small series since [31–33]. Noh and Kalyanaraman [34] recently published a modification of this technique, using a single transumbilical trocar for the camera and conventional straight instruments introduced directly through stab incisions in the abdominal wall. Of 27 patients, only one atrophied testis was identified at 1 year follow-up. A related technique, multi-incisional transumbilical orchiopexy, involves the insertion of three separate ports within the umbilicus. Aside from concealed scars, no outcomes advantages have been identified despite slightly longer operative times (54.2 vs. 47.3 minutes) when compared with conventional laparoscopic orchiopexy [35]. The largest series of multi-incisional transumbilical orchiopexy [36] reports feasibility of both primary laparoscopic orchiopexy and the Fowler–Stephens technique, with 82% of testes in an ideal and 99% in an acceptable scrotal location.

Dedicated SILS platforms have been used in children, although they are designed for adult patients (Triport™ or Quadport™ systems, Olympus, Advanced Surgical Concepts, Center Valley, PA; Uni-X™, Pnavel Systems, Brooklyn, NY; SILS™ Port, Covidien, Dublin, Ireland; and GelPOINT or GelPort™, Applied Medical, Rancho Santa Margarita, CA). These universal platforms present logistic difficulty with clashing when using shorter pediatric instruments. The learning curve is also much steeper due to inability to triangulate instruments from a single insertion site. Flexible tip laparoscopes and curved laparoscopic instruments may reduce triangulation difficulty, but must be separately purchased. Though feasibility studies and aesthetic benefit are reported, any comparative benefit over standard laparoscopic orchiopexy remains to be seen.

Timing of surgery

Surgical timing is intended to prevent decline in fertility potential, reduce testicular malignancy (controversial), and minimize age-related anesthetic risks, including developmental delay. A short period of postnatal observation is recommended as spontaneous testicular descent has been noted postnatally at 4–6 months. This must be weighed against the decline in fertility potential, which is not due to obvious genetic defect, but is known to

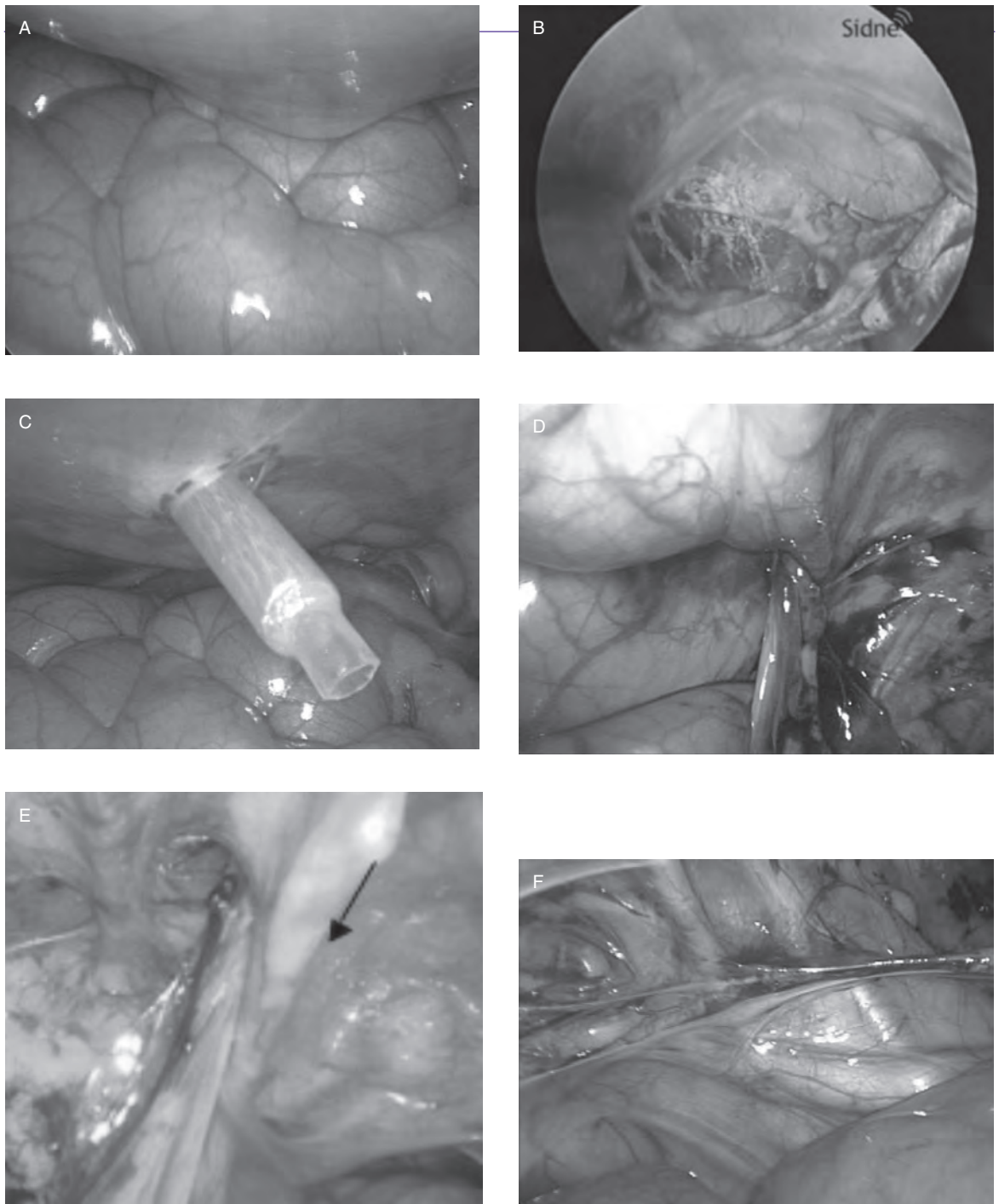


Figure 23.4 Common injuries. Despite maximal insufflation pressure, the great compliance of the pediatric abdomen makes trocar placement difficult and places underlying organs at risk for injury (A). The ligamentum teres can be injured and risk bleeding during umbilical and supraumbilical trocar placement (B). The vessels most prone to injury during port placement are the inferior epigastric vessels (C), though the aorta, vena cava, femoral and iliac vessels are also at risk. Tracking of the neo-inguinal hiatus medial to the medial umbilical ligament risks bladder injury (D), therefore the neo-inguinal hiatus is placed lateral to the medial umbilical ligament and medial to epigastric vessels (E). During elevation of the peritoneal triangle, one must be cautious of the underlying ureter and iliac vessels (F).

be present and irreversible by 15 months of age [37]. A prospective Swedish study recently randomized the timing of orchiopexy with testicular biopsy to either 9 months or 3 years of age [38]. This study demonstrated that, with time, testes will demonstrate reduced transverse tubular diameter, reduced testicular volume, and reduced interstitial tissue. Sertoli cell and germ cell number at 3 years were also reduced when compared with 9 months. Therefore, most experts recommend orchiopexy from 6 months to about 18 months of age [21].

Aside from surgical timing, bilateral undescended testes increase the infertility risk. Kraft and colleagues [39] recently published a modern series of fertility potential for cryptorchid testes, comparing testis histopathology at the time of orchiopexy to adult (age 18) semen analysis and hormone levels. The authors confirm the infertility association of bilateral cryptorchidism [40], noting that abnormal sperm count, sperm density, reduced motility and higher FSH levels occur in adults with a history of repaired bilateral cryptorchidism.

Outcomes

Docimo [8] performed a meta-analysis in which overall success rates of various open orchiopexy techniques were compared (inguinal 89%; one-stage Fowler–Stephens 67%; two-stage Fowler–Stephens 73%; transabdominal 81%; microvascular 84%). Data on success rates after 1985 document a modest improvement in outcomes [8], with significant room for improvement. This opened the door for alternative techniques, and facilitated greater utilization of the laparoscopic approach. Several multi-institutional reviews and numerous single institutional papers have reported improved success rates over an open approach (Table 23.1). Some authors advocate a single approach for all patients with either primary laparoscopic orchiopexy without division of vessels [41,42] or in two stages with division of vessels [43]. The majority of clinicians tend to manage each case on a more selective basis, determining their approach based on the ability to obtain sufficient length to place the testis in the scrotum [12–16]. Baker et al. completed a 10 center, multi-institutional analysis to evaluate the outcomes of laparoscopic orchiopexy. Primary laparoscopic orchiopexy was successful in 97.2% of cases. One- and two-stage Fowler–Stephens

orchiopexy were successful in 74.1% and 87.9% of cases, providing evidence for superiority over open techniques for management of the intra-abdominal gonad. Other studies refute these data, suggesting equivalent success rates for minimally invasive and open techniques, despite morbidity advantages of the former. One recent meta-analysis noted no difference in atrophy or malposition rates between laparoscopic and open orchiopexies for nonpalpable testes, though a significantly shorter hospital stay was observed [3]. In 2006, Abolyosr conducted a prospective, randomized study between open and laparoscopic orchiopexy for the management of 75 intraabdominal testes [4]. Primary orchiopexy was successful in 100% for both modalities, whilst the success rate was 85% and 90.5% respectively for open- and laparoscopic-staged Fowler–Stephens orchiopexy. A morbidity advantage was seen in the laparoscopy group, including shorter time to oral feeding, shorter hospital stay, shorter operative time, and markedly quicker return to normal activities (8.4 vs. 25 days) [4]. Such information is important when counseling patients prior to surgery.

Additional factors influencing surgical success are prior inguinal or testicular surgery. Prior orchiopexy attempt carries a high risk of failure for Fowler–Stephens orchiopexy, presumably related to compromise of collateral blood flow between the spermatic vessels and vas [26,44,45].

Finally, outcomes for laparoscopic orchiopexy of *palpable* undescended testes have been found to be similar to a conventional open inguinal approach. Docimo and colleagues proved feasibility for the high palpable undescended testis, with 100% success rates. Post-1985 meta-analysis data suggest that inguinal testes are successfully positioned and viable 85.3% of the time [8,20]. One recent series actively compared open to laparoscopic orchiopexy cohorts for palpable undescended testes and involved a prospective, randomized trial design [46]. This study was unable to demonstrate a difference in surgical success, though pain was reduced in the laparoscopic group. Laparoscopic orchiopexy for the palpable testis can result in conversion to open inguinal orchiopexy for improperly selected patients, such as those with low inguinal testes which reside outside the external oblique fascia [47]. Repeat exam under anesthesia should be performed prior to the procedure, if laparoscopic management is planned for a palpable undescended testis.

Complications

Overall, laparoscopic orchiopexy, even in a staged fashion, carries a low complication rate. In a large multi-institutional review [22], Baker and colleagues reported a major complication rate of 3.0% and a minor complication rate of 2.0%. Major complications were cardiopulmonary compromise, bowel obstruction or injury, severe wound infection or shunt complication, and injury to spermatic vessels, vas, major vessels, or bladder. It is important to note that complications during laparoscopic procedures are reflective of surgical experience. Lindgren and colleagues observed that at least 10 laparoscopic cases were required to flatten the learning curve for laparoscopic orchiopexy; though a survey of 153 practicing pediatric urologists found that complication rates did not significantly decline until after 100 laparoscopic cases had been performed [48,49]. Similarly, ongoing laparoscopic case volume and, to a lesser extent, access technique were predictive of complications in a large, retrospective review from the Children's Hospital of Boston [49,50]. Surgeons performing more than 12 total laparoscopic cases per year had significantly lower complication rates.

The prevention of complications associated with laparoscopy starts with patient preparation. Communication with the anesthesia team should include need for neuromuscular paralysis and gastric decompression. Proper positioning and padding are then performed to reduce the risk of neuromuscular injuries. The patient should be routinely taped and padded as if steep Trendelenburg and table rotation are required, since intra-abdominal testes may require colonic reflection. A Foley catheter is then placed to prevent bladder injury.

Testicular atrophy

Testicular atrophy is the most common long-term complication associated with laparoscopic orchiopexy. Etiologies include: injury to or tension on spermatic vessels resulting in ischemia, torsion of the spermatic vessels, or intentional ligation of the vessels as part of Fowler–Stephens orchiopexy [51].

The frequency of this occurrence is dependent on the initial position of the testicle, the chosen technique for repair, and underlying potential for testicular development as influenced by age at orchiopexy and *normal* organogenesis. Generally, primary orchiopexy has the lowest atrophy rate, ranging from 0–2% in larger se-

ries. Atrophy rates are moderate for two-stage Fowler–Stephens procedures at approximately 10%, and remain highest for a one-stage Fowler–Stephens procedure at 22% (see Table 23.1). Atrophy can be minimized by limiting manipulation of the vas, testis and its collateral, retroperitoneal vasculature [44]. Experience helps surgeons avoid unnecessary dissection in an attempt to do a primary orchiopexy, only to find that a two-stage approach is required. Finally, when transecting ligated spermatic vessels during a first- or second-stage Fowler–Stephens orchiopexy, one should avoid electrocautery to prevent thermal injury to the nearby collateral blood supply.

Atrophy of the delivered testicle is closely monitored at the time of follow-up, and most commonly done with orchidometers in reported series. Esposito and colleagues reported a small cohort of 12 retrospectively identified patients with between 7 and 17 years follow-up after unilateral two-stage Fowler–Stephens orchiopexy, who reported for further evaluation with echo color Doppler resistive indices and ultrasound-measured testicular volumes [52]. They were able to demonstrate that the resistive indices change from a low flow profile (for the testicular artery) to a high flow profile (for collateral vessels). In this series, the atrophy rate (reflecting completely infarcted testes) was 16.7%. The authors also point out all surgically treated testes were smaller at long-term follow-up compared with their descended contralateral counterparts, making a case for routine clinical follow-up until puberty along with interval ultrasounds for volumetric measurements. Follow-up protocols after laparoscopic orchiopexy remain highly variable, both in duration and objective assessment of outcomes. The majority of series evaluate testicular size following laparoscopic orchiopexy simply as a subjective assessment of atrophy, though some objectively follow with orchidometer measurements and even fewer with ultrasound dimensions using ellipsoid formulas for volume calculation [38,52,53].

Testicular malposition

Testicular retraction following laparoscopic orchiopexy is attributed to short spermatic vessels, inadequate mobilization of the vascular pedicle, or improper fixation of the testis in scrotum [51]. Intrascrotal location was achieved in 93–96% of all laparoscopic orchiopexies in two large series [22,44,45], with 3% requiring repeat surgery. It is important to note that a few testes

palpable near the pubis may be considered acceptable in location [44].

When malposition is sufficient to justify reoperation, it is the authors' preference to perform redo orchiopexy with a scrotal approach provided the scrotum can be brought to the testis [54,55]. Surgery for testicular malposition after failed orchiopexy or inguinal surgery can be challenging, and one must also recognize that testicular lie may not be normal. Often, the testis is flipped over a more dependent portion of the fixed cord, making the orientation an unreliable landmark [51].

Peritoneal access

The pediatric abdomen presents a unique challenge for peritoneal access due to increased abdominal wall compliance and limited working space. Certain urologic conditions such as Prune-Belly syndrome may further complicate the situation, but are not absolute contraindications to minimally invasive orchiopexy. Due to great abdominal wall compliance, the epigastric vessels, iliac vessels, and bowel all come into close proximity to access trajectories (see Figure 23.4). Immediate inspection of these structures before and after port placement is mandatory. Riquelme and colleagues reported that half (2 of 4) of their 13% complication rate involved injury to the epigastric vessels during scrotal trocar placement. This occurred early in their learning curve of the procedure [47].

Injury to the aorta and vena cava may occur during umbilical access, while injury to the major pelvic vessels may occur with introduction of the lateral trocars or distal mobilization of the testis on its peritoneal triangle. Upon removal of the trocar, brisk blood flow will be evident. The obturator should be replaced to tamponade the hemorrhage and guide the eventual repair. Immediate laparotomy is often indicated and prompt communication with anesthesiologist and vascular surgeon is prudent [56]. To avoid excessive downward pressure through umbilical fascia during peritoneal access, the authors prefer to place a 2-0 vicryl suture through the umbilical skin as a method of countertraction.

Bowel injury can occur during open and blind peritoneal access, instrument passage, and the use of electrocautery. Specifically, Veress needle puncture of the sigmoid colon, cecal volvulus, and small bowel injury have been reported [22,50]. Serosal tears or isolated bowel injuries may be repaired primarily by an experienced laparoscopist [44]. More extensive injuries may

warrant formal laparotomy. If bowel injury occurs or is suspected, one should "run" the entire circumference and length of bowel to ensure injuries are not missed. Cautery injury is a particular risk in the smaller working space of the pediatric abdomen. The surgeon should be aware that smaller instrument diameters increase current density, with injury at even higher risk due to a thinner abdominal wall to dissipate the electricity [57]. For this reason, we recommend limiting the use of electrocautery and reducing settings to the minimum needed.

Abdominal wall hemorrhage may be encountered. The epigastric vessels lie at the lateral border of the rectus sheath and are easily visualized in thin patients when placing assistant ports under direct vision. If significant bleeding occurs and the location of vascular injury cannot be visualized, various suture techniques may be utilized. Bhayani and Kavoussi describe obtaining circumferential control of the vessel using either the Carter-Thompson fascial closure device (Inlet Medical Inc., Eden Prairie, Minnesota) or a Keith needle that is passed percutaneously through the incision, then directed with laparoscopic grasper around the vessel [58]. The authors find it useful to close assistant port sites under direct vision to identify possible hemorrhage that may have been tamponaded by a trocar and prevent incorporation of bowel, omentum or epigastric vessels into the wound.

Omental herniation through a 3 mm umbilical trocar site has been reported [59]. However, this is a rare complication of laparoscopy with an incidence of 0.15% in over 5400 laparoscopic cases [60]. If feasible, we recommend fascial closure at all port sites, though it is not mandatory for 3 mm incisions. Additionally, removing the trocar or cannula over the laparoscope after desufflation may help to reduce omental or bowel herniation [59]. To further ease umbilical fascia closure at the end of the case, stay sutures through the umbilical fascia can be placed at the time of initial port placement and simply tied together after port removal.

Shunt complications

Modern ventriculoperitoneal shunts contain proximal and distal unidirectional valves, which regulate flow or pressure, and are not a contraindication to laparoscopy [61]. Infection-related complications must be

recognized [62], as they occur in 9.6% of pediatric laparoscopic bladder reconstruction involving bowel. The incidence of infection was not modifiable by bowel preparation [63]. Distal shunt failure has been reported with laparoscopy [61], though *in vitro* experiments with intra-abdominal pressures of 3–25 mmHg do not seem to demonstrate retrograde reflux [64]. Pneumocephalus in an adult with a 20-year-old Holter valve ventriculoperitoneal shunt has been reported, although insufflation pressures up to 50 mmHg were used, making this an unlikely pediatric occurrence. Nonetheless, peritoneally drained shunts risk infection, pseudocyst formation, or mechanical injury with any intra-abdominal surgery requiring careful attention by the pediatric urologist.

Testicular avulsion

Avulsion of the spermatic vessels due to excessive traction placed on the testicle has been reported [22,44,65]. The largest series, a multi-institutional study reported by Baker, suggests that avulsion of the spermatic vessels is a rare complication occurring in one of 310 testes [22]. Conversion to a Fowler–Stephens approach is necessary in this situation and testicular atrophy may result [65]. If it is possible to visualize the site of avulsion, then clipping each end of vessels may be performed laparoscopically. If hemorrhage ensues, open vascular ligation may be necessary.

Vasal injury

Transection of a looped vas deferens is a potential complication. To avoid injury to the vas, one must anticipate the possibility of the vas looping down into the inguinal canal, and take care to transect only gubernacular attachments to the testis that enters the internal inguinal ring. The vas deferens should generally be left on the peritoneum and rarely dissected free, as this maneuver will not improve length of the vascular pedicle. Overly aggressive dissection of the vas off the peritoneum can result in vasal injury, testicular atrophy and, rarely, ureteral obstruction [21,66,67].

Ipsilateral hernia

Metachronous hernia is felt to be an exceptionally rare occurrence, with sufficiently low incidence that formal laparoscopic hernia repair is not required at the time of laparoscopic orchiopexy. A case report of

metachronous ipsilateral indirect inguinal hernia was described by Metwalli and Cheng [68] following laparoscopic orchiopexy. In their series, this complication had a 4% incidence (1 in 25 patients), although this is not a consistent risk within the literature. A patent processus vaginalis ipsilateral to the undescended testis occurs is reported in 69% of cases, though in contrast to the prior series none went on to develop clinical hernia [69]. Up to 9% of undescended testes may be found to have a contralateral patent processus vaginalis. Though some authors advocate laparoscopic hernia repair during the contralateral orchiopexy [70], insufficient evidence exists to support that the patent processus vaginalis will become a clinical hernia. Therefore, it is the practice of the authors at the time of laparoscopic orchiopexy to ablate the ipsilateral peritoneal membrane at the level of internal inguinal ring in order to mobilize the testis. The contralateral ring is generally untouched.

Bladder injury

Injury to the bladder is most likely to occur when creating the neoinguinal hiatus (Figure 23.4D). Drainage of the bladder with a Foley catheter or feeding tube should be performed prior to creating the hiatus to minimize this risk. The anesthesia and nursing teams should also be instructed to alert the surgeon if gross hematuria is seen in the catheter bag, suggesting a bladder injury and subsequent need for repair of the intraperitoneal bladder defect. The risk of bladder injury is increased if the neohiatus is not formed superior to the pubis in a plane lateral to the medial umbilical ligament and medial to the epigastric vessels. Need for excessive pressure to create a neohiatus probably indicates misdirection of the laparoscopic grasper. Guiding the grasper over the pubis onto the tip of one's finger, which is placed through the scrotal incision, can reduce risk of collateral injury. Hsieh and colleagues report a 3.2% incidence of bladder injury (three cases in 93 patients with 101 undescended testes) in their retrospective review [67]. Two of the three bladder injuries were recognized intraoperatively due to presence of hematuria, and one was identified in a delayed fashion (without intraoperative hematuria) due to chemical peritonitis from the urine leak. The authors stress the importance of thorough bladder evaluation with either cystoscopy, irrigation of the

bladder under direct laparoscopic vision, or adequate cystogram at bladder capacity should hematuria, “gaseous distension of urinary bag,” or clinical picture suggest bladder injury. Bladder defects may be managed in an open or laparoscopic fashion.

Iatrogenic or delayed testicular torsion

Torsion of the testicular vessels following delivery to the scrotum has been reported [66]. As with inguinal orchiopexy, one must confirm normal straight, non-twisted spermatic vessels upon pulling the testis through its neo-inguinal ring into the scrotum. Also, intraperitoneal examination of the vessels is also important to ensure they are not kinked. Therefore, we typically leave all trocars in place after desufflation until the scrotal portion of the orchiopexy is complete.

Postoperative ileus

Ileus is uncommon after laparoscopic orchiopexy. Conservative management, consisting of limiting oral intake, intravenous fluids, and nasogastric suction when indicated, has resulted in resolution of symptoms [21]. Interestingly, intraperitoneal adhesions are not a frequent problem following laparoscopic orchiopexy [71,72].

Wolffian duct anomalies and testicular ectopia

If no testis is readily identified and ipsilateral renal anomaly is known (multicystic renal dysplasia, agenesis), an exhaustive search should be carried out, from bladder to the superior aspect of renal fossa investigating for testicular ectopia [73,74]. This may require additional port placement and exaggerated Trendelenburg and/or table rotation.

Unilateral vasal agenesis can be found on occasion along with an intra-abdominal testis; if primary laparoscopic orchiopexy is not possible, a Fowler–Stephens orchiopexy should not be successful in the absence of the vasal blood supply. In this situation, orchiectomy is warranted and follow-up renal ultrasound prudent to determine if ipsilateral renal agenesis or renal ectopia are responsible for congenital unilateral absence of the vas deferens (CUAVD), reflecting an early mesonephric duct anomaly. Inspection for a contralateral vas should be performed to ensure this is not a situation of congenital bilateral absence of the vas deferens

(CBAVD), in which case CFTR mutation testing should be performed in follow-up. Almost all males with cystic fibrosis have CBAVD; conversely, 70% of males with CBAVD but no clinical evidence of cystic fibrosis have an identifiable CFTR mutation [75]. Rarely, infertile adult males with CUAVD may be found to have a palpable scrotal segment of vas, which terminates or is absent more proximally; in situations of infertility, CFTR receptor mutation testing is also recommended [76]. The pediatric urologist must be prepared to deal with developmentally abnormal gonads and Wolffian ducts, given 41 of 252 patients in Baker and colleagues’ multi-institutional series had congenital anomalies (some of which included ambiguous genitalia, Denys Drash, multicystic renal dysplasia and persistent Mullerian duct syndrome) [22].

Finally, persistent Mullerian duct syndrome (PMDS) is a clinically rare diagnosis, which is almost always a surprise at the time of surgery in a male with normal external genitalia. PMDS may present with hernia uteri inguinale or transverse testicular ectopia, which can include cryptorchidism in association with the expected fallopian tubes, uterus, cervix and upper vagina [77–79]. Figure 23.5 is an example of transverse testicular ectopia as diagnosed at the time of planned laparoscopic orchiopexy, with one descended testis and the contralateral testis crossing midline. Karyotype is warranted and in PMDS will be 46XY. Mullerian structures persist as a result of lack of Sertoli cell synthesis or release of Mullerian-inhibiting substance (MIS, or anti-Mullerian hormone), or a mutation in the MIS receptor [80]. A genetic mutation (chromosome 12: MIS, chromosome 19: MIS type II receptor) can be identified in approximately 85% of cases [81,82]. Surgical management must factor in a short vas, tethered to the surface of the uterus, which should not be dissected free for risk of injury [83,84]. Options for laparoscopic orchiopexy therefore include splitting the uterus in midline to just above the vasal confluence, orchiopexy of both testes through a single neo-inguinal canal into separate subdartos pouches (personal communication, FX Schneck), single- or two-stage Fowler–Stephens orchiopexy to gain length (provided vas is not too tethered), or intentional division of vas and fallopian tube. If such maneuvers fail, orchiectomy is indicated [83]. Fertility is rarely reported in these patients [79, 84].

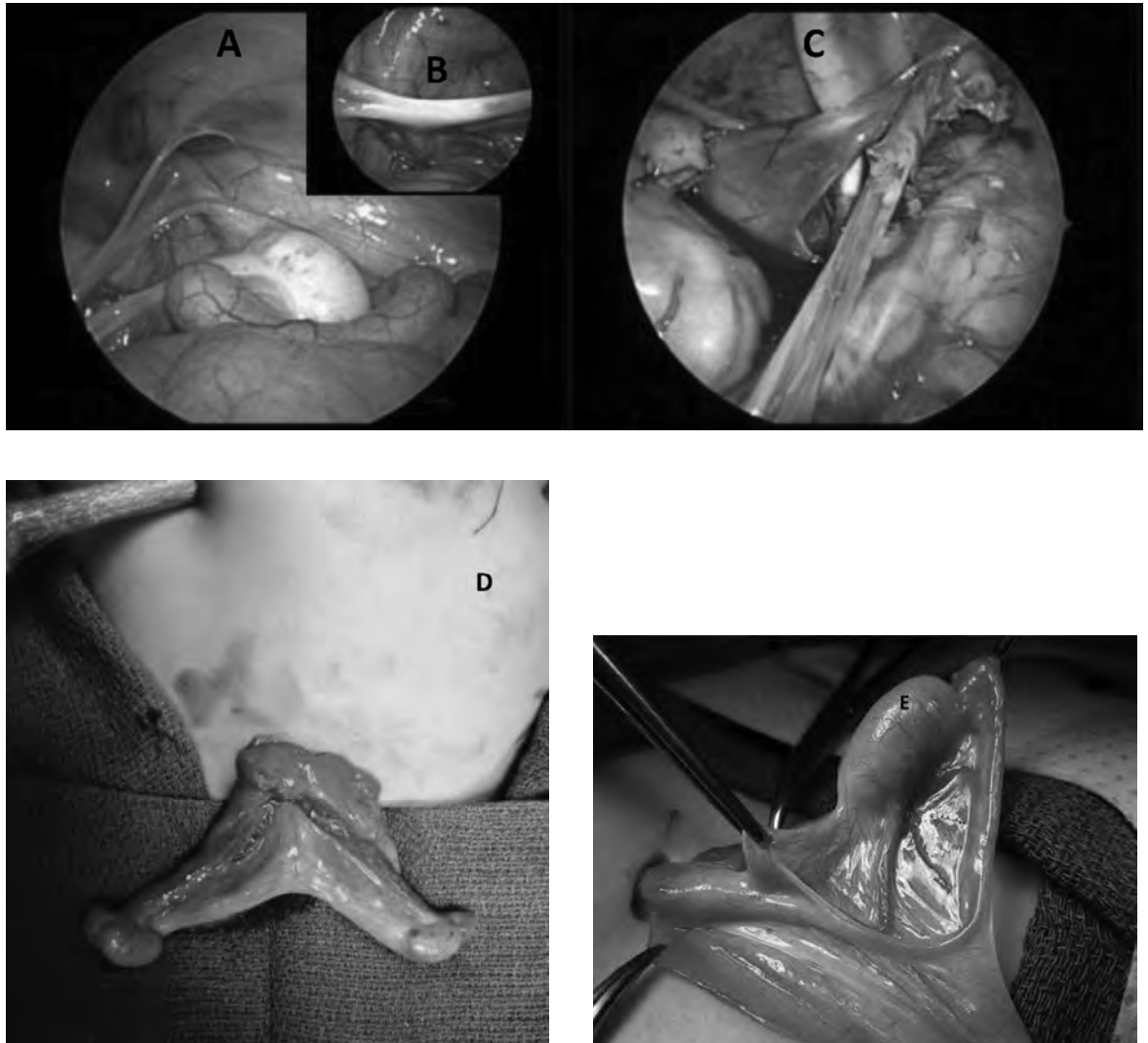


Figure 23.5 Persistent Mullerian duct syndrome (PMDS). PMDS was discovered at the time of laparoscopic orchiopexy; a left non-palpable testis was the only abnormal finding on external genital exam. Transverse testicular ectopia with midline urethra structure was seen (A, B). Laparoscopic orchiopexy is still the mainstay of treatment for the intra-abdominal testis in PMDS, though the vas should not be dissected away from Mullerian structures to prevent injury (C). In a separate patient with PMDS discovered at inguinal orchiopexy, the uterus and fallopian tubes can be seen terminating in a distal testis (D), with vasa running intimately along the fallopian tubes (E).

Conclusion

Advances in laparoscopic instrumentation have facilitated widespread adoption by pediatric urologists of all levels of expertise. Improved diagnostic accuracy, postoperative convalescence, cost effectiveness and superior outcomes have resulted in routine use of diagnostic laparoscopy and laparoscopic orchiopexy for

management of the nonpalpable testis. Minimally invasive orchiopexy poses rare risk of major complications (3%), which are predominantly related to the small intracorporeal working space or delicacy of tissues in the pediatric abdomen. Long-term follow-up for complications, testicular position and the presence of atrophy following minimally invasive orchiopexy is still essential to identify areas for ongoing improvement.

DOS AND DON'TS**Do**

- Ensure the child is appropriately padded and secured to the operating table to allow for steep Trendelenburg and lateral rotation. This will facilitate laparoscopic examination up to the renal fossa for very high intra-abdominal testes
- Use open techniques to establish pneumoperitoneum, given higher risk of intra-abdominal injury with needle access techniques
- Attempt to create the neo-inguinal hiatus lateral to the medial umbilical ligaments to minimize risk of bladder injury
- Preserve a distal triangle of peritoneum to have the option of vessel ligation available until it is clear that there is adequate length for scrotal placement

CLINICAL PROBLEM/CASE: DISCUSSION**Case 23.1 discussion**

- 1 Cryptorchidism may result from isolated conditions impairing the hormonal, mechanical, or neurological migration of the gonad. Such syndromes include disorders of sexual development, prune-belly syndrome, connective tissue defects, abdominal wall defects (i.e. bladder exstrophy), posterior urethral valves, myelomeningocele, genitofemoral nerve/CGRP neurotransmitter defects, and cerebral palsy. In addition, many chromosomal and malformation syndromes are associated with cryptorchidism, such as Robinow, Cornelia DeLang, ulnar-mammary, Noonan, LEOPARD, Rubenstein-Taybi, HITCH, and Aarskog (Aarskog-Scott) syndromes [85]. There are 373 syndromes or isolated genetic mutations presently described in association with cryptorchidism, as reported in OMIM (NCBI's Online Mendelian Inheritance of Man; www.omim.org). When another congenital anomaly is present, about 7% have chromosomal aberrations by karyotype alone [86]. Comparative genomic hybridization (microarray) now enhances detection of genetic alterations, revealing copy number variants in as many as 18.5% of children with cryptorchidism, after excluding those with defects in steroid synthesis or activity [87].
- 2 The transabdominal and inguinoscrotal phases of testicular descent occur at 10–15 weeks and 25 weeks gestation, respectively. The initial transabdominal phase involves the testis and Wolffian duct, lying anteromedial on the mesonephros, attaching to gubernaculum caudally. As the gubernaculum anchors to the anterior abdominal wall and swells under the influence of INSL3 (produced by Leydig cells), the cranial suspensory ligament regresses under influence of androgen and the enlarging testis approximates the future inguinal canal. The receptor for INSL3 is LGR8. No further descent occurs until about 25 weeks gestation when, over the following 10 weeks, the gubernaculum actively elongates all the way into the scrotum, forming the inguinal canal. Testicular migration for the inguinoscrotal phase appears to be more heavily influenced by androgens. Data from rodents show that androgen seems to masculinize and stimulate the genitofemoral nerve to release CGRP, which binds at the gubernaculum promoting cremasteric contractions and a chemotactic migration into the scrotum. CGRP may also be responsible for closure of the processus vaginalis [85].

Case 23.2 discussion

- 1 Laboratory values show normal prepubertal hormones before hCG stimulation. The hCG stimulation test is then performed at 100 IU/kg (max. dose 1500 IU) twice weekly for 3–4 weeks. Results of hCG stimulation for this 9-year-old boy help to determine presence of at least one hormonally functional testis since serum testosterone is >200 mg/dl within 24 hours of last injection. Shorter 3-day hCG stimulation tests (1500 IU daily) do not determine whether testes are dysgenetic, as does the full 3-week course [88]. It is essential to know dose, timing, and confirm administration of the hCG relative to bloodwork to avoid an iatrogenic inadequate response. Because there is always the potential for Leydig cell dysfunction with viable testes, an inadequate testosterone response means exploratory surgery is still performed. An alternative, sensitive serum test would be an AMH/MIS (anti-Mullerian hormone or Mullerian inhibiting substance) level, which reflects presence of Sertoli cells; if anorchia occurred, AMH/MIS levels would be undetectable. In this case, the recommended surgery is diagnostic laparoscopy, with possible bilateral laparoscopic orchiopexies, possible staged procedures, possible unilateral or bilateral inguinal exploration, and possible orchiectomy of nubbin (per surgeon preference).
- 2 Bilateral inguinal exploration is warranted.

- 3 When using parameters to predict fertility, semen analysis is gold standard. However, in the prepubertal child, testis biopsy information is all we have, so normal histology must be understood. In the normal prepubertal testis, one observes presence of adult dark (Ad) spermatogonia (upon neonatal testosterone surge) and primary spermatocytes (upon transient LH spikes at age 3–5). Primary spermatocytes are the meiotic precursor to form adult pale (Ap), then secondary spermatocytes, early and late spermatids then sperm [89]. Unilateral undescended testes do not seem associated with significantly higher rates of infertility compared with average men. However, studies do confirm that a history of bilateral cryptorchidism has a nearly 6-fold increased risk of infertility in adulthood [90]. Bilateral undescended testes tend to have reduced Ad or germ cells per tubule, which is clinically significant for the association with abnormal FSH and semen analysis parameters after age 18 [91].
- 4 While paternity rates are reportedly lower with bilateral versus unilateral cryptorchidism [92], one should still be cautious in future fertility counseling based upon testis biopsies. Undescended testis histopathology has yet to be definitively correlated with later paternity, although a small retrospective study by Cendron et al. suggests there is good correlation [93].

Case 23.3 discussion

- 1 Careful incision of the peritoneum overlying the collateralized retroperitoneal pedicle between the vas and divided vessels (see Video 23.1).
- 2 A scrotal button may be sutured to the testicle for up to a week.
- 3 If too much tension on the testicle remains after the second-stage procedure, then orchiectomy is warranted.

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CHAPTER 24

Varicocele

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KEY POINTS

- Diagnosis of varicocele is essentially clinical
- Treatment of varicocele for subfertility is controversial. Recent evidence is strengthening arguments favoring early intervention
- Intervention options include venous occlusion or surgical ligation
- Surgical ligation remains the preferred approach
- Nonartery sparing technique has better success than artery sparing approach
- Hydrocele formation is the most frequent complication regardless of approach
- Lymphatic sparing techniques are promising in reducing morbidity

CLINICAL PROBLEM/CASE

A 13-year-old boy presents to your clinic with left-sided scrotal swelling.

1 What clinical features will clinch the diagnosis of a varicocele?

2 What is grade 3 varicocele as per the World Health Organization (WHO) classification?

3 What is the incidence of varicocele in adolescence?

4 What is the role of a color Doppler ultrasound (CDU) scan?

5 What is testicular hypotrophy?

Introduction

Varicocele is the abnormal dilatation of the pampiniform plexus of veins within the scrotum. The plexus receives tributaries from the epididymal veins and lies posterior to the testes. The plexus then ascends around the vas deferens to the superficial inguinal ring, where venules coalesce to form 3–4 veins that lie within the spermatic cord in the inguinal canal.

In a unique study of over 4000 boys in Turkey, the prevalence of varicocele increased significantly with age, from 1% in under 10-year-olds compared to 7.7% in 11–14-year-olds and 14.1% in the 15–19-year-olds [1]. This suggests a probable causal relationship

between increased blood flow to the pubertal testis and varicocele. Recent studies support the clinical experience showing an inverse relationship between varicocele occurrence and body mass index [2,3].

The relationship between varicocele, male subfertility, and testicular atrophy is what underlies the current practice of correction of varicocele in prepubertal males [4]. Yet the empirical varicocelectomy as a remote treatment for subfertility remains controversial. Although the effect of increased temperature caused by a varicocele seems well established, this effect appears to be in both the testes and not in the ipsilateral gonad. Increased thickness of the lamina propria, increased nitric oxide levels in the venous plexus, and impaired transformation of the

myofibroblasts to fibroblasts are some of the abnormal findings reported from research studies [5–7].

New concepts in pathophysiology are emerging with increasing evidence that high levels of reactive oxidative stress combined with decrease in total antioxidant capacity impair the integrity of the DNA in the sperm nucleus. The imbalance causes DNA fragmentation although the sperm analysis is normal [8–11]. Therefore, there is a call for sperm DNA integrity as a better indicator of male fertility potential [12,13]. Surgical correction seems to significantly improve the DNA fragmentation index [14–16].

Diagnosis

Diagnosis of varicocele has historically been clinical. Characteristic symptoms are of scrotal discomfort and heaviness, which is worse when standing or walking. Physical examination demonstrates a classic “bag of worms” feel and appearance of the affected scrotum. The ipsilateral testis can often be palpated separate to the varicocele and may be reduced in size when compared with the contralateral testicle. Varicocele is left-sided in 90% and bilateral in 10%. A unilateral right-sided varicocele is exceedingly rare [1].

Traditionally, varicoceles have been classified clinically based on the classification suggested by Dubin and Amelaar adopted by the WHO [17]:

- Grade I – Palpable by valsalva maneuver.
- Grade II – Palpable without valsalva maneuver.
- Grade III – Visible before palpation.

Color Doppler ultrasonography (CDU) allows diagnosis of varicocele which may not be clinically apparent. The venous reflux into the venous plexus is diagnosed using the Doppler color flow mapping in the supine and upright position, at quiet respiration, and with valsalva maneuver. Continuous reflux pattern (venous rush) during quiet inspiration (without valsalva maneuver or deep inspiration) is the main criterion for diagnosis of varicocele [18]. CDU is gaining popularity and some centers now call for routine preoperative CDU assessment for both testicular volume discrepancy (hypotrophy) and venous rush [19,20]. This is particularly useful for subclinical varicocele especially on the right side which, according to some authors, is underreported [21]. Peak retrograde flow as measured by CDU could be a useful adjunctive tool in decision-making [22].

Testicular hypotrophy

Testicular hypotrophy may be determined by the atrophy index (AI: $[Vr2Vl/Vr]3100$; Vr and Vl volumes of right and left testicle, respectively) [23]. In adolescents and adults, testis size should approximately be equal on both sides with a standard deviation of not less than 2 cm³ on ultrasound examination, which uses the ellipsoid method of volume calculation (0.5233 HWL). Ultrasound estimation of testicular size has been shown to be more accurate than the physical estimation using orchidometers [24].

The criterion for defining testicular hypotrophy is inconsistent in various reports with atrophy index ranging from 10–25% [25–27]. Also absolute volume differentials vary from 2 cm³ in some reports to 3 cm³ in others [28,29]. Therefore, the reported incidence of hypotrophy in varicocele varies from 30% to 70% [30,31]. Recently, more evidence has been published with 10% asymmetry as a cut-off for intervention [32,33].

Testicular injury

Bach et al. reported that the Sertoli cell function was impaired even in young men with medium- to high-grade unilateral varicocele. By contrast, Leydig cell function seems to be undisturbed. They demonstrated that these men had an exaggerated GnRH test response with high-elevated FSH levels. The baseline FSH levels were elevated above 5.6 U/l. This could provide an easier way to predict the outcome of the GnRH test, which is labor intensive and expensive [34]. Other investigators have shown a mixed response to the GnRH test [29]. However, there is no evidence yet that GnRh test response correlates with fertility in the long term.

Recently, there have been calls for better indicators for assessment of testicular injury like sperm DNA integrity even when sperm analysis is normal as explained above.

Indications for intervention

Kogan [35] divided the indications for repair into the following groups:

1 Absolute:

- A small hypotrophic testis (2–3 cm³ smaller than the other).
- Additional testicular condition affecting fertility.
- Abnormal semen analysis.
- Bilateral palpable varicocele.

2 Relative:

- Large size (grade).
- Softer ipsilateral testis.
- Pain.
- Supranormal LH and FSH response to GnRH stimulation test.
- Patient or parental anxiety.

3 Minor:

- Abnormal scrotal appearance.

The most common and absolute criterion is a small testicle indicating growth arrest [31]. The testis remains small if conservatively managed, whilst catch-up growth has been reported by several authors [23,31]. Parrott et al. reported reversal of hypotrophy in 53–90% postintervention [25]. Kocvara et al. argue that the so-called reversal of growth may be due to edema associated with division of lymphatic vessels [36]. Pinto et al. report no correlation between testicular hypotrophy and fertility [4], but others have demonstrated the complete opposite [23,31]. The criterion of softer testis is very subjective and there is little written about it in the literature.

In our experience, pain and discomfort should be an absolute indication for intervention and surgery successfully alleviates the symptoms. Anxiety is another common feature in adolescent boys and intervention often helps to reassure them. GnRH stimulation test is intensive and expensive and is a weak criterion as an indication for intervention in the absence of correlation with fertility.

Age at intervention

The argument for early intervention lies in the observation that as the testicle is still growing, testicular atrophy associated with varicocele can be overcome during the pubertal growth spurt [37]. In adolescents, there are limitations in parameters that can influence decision-making as fluctuating levels prevent hormonal testing and testicular biopsy is considered too invasive. It is impractical to request semen analysis and normograms are not available. This leaves testicular volume as the sole parameter.

Studies have also demonstrated that varicocele-related testicular hypotrophy is a developing condition [1]. Hypotrophy was not present in boys younger than 11 years. In 11–14-year-olds, the incidence was 7.3% and in 15–19-year-olds it increased to 9.3%. These results suggest that varicocele-related testicular hypotrophy increased with puberty, supporting the idea that prepubertal intervention is ideal. However, RCTs

are heterogenous, poor in quality and do not contribute significantly to any conclusion on efficacy of varicocele treatment [32,38]. A collaborative review concluded that, although there is no evidence that varicocele treatment improves spontaneous pregnancy rates, it does improve sperm counts and morphology [39].

Intervention methods and outcomes

Broadly, the options of intervention in varicocele are either venous occlusion or surgical ligation. Venous occlusion is by either embolization or sclerotherapy. Sclerotherapy can be performed antegrade or retrograde. Surgical ligation can take place at various levels at, above, or below the inguinal region. Furthermore, it can be either by an open or by a laparoscopic approach and also can take the form of microsurgery (Figure 24.1). We will discuss the pros and cons of artery sparing and lymphatic sparing techniques.

Classification of methods of intervention in varicocele

- Venous occlusion:
 - percutaneous embolization/sclerotherapy
 - antegrade scrotal sclerotherapy.

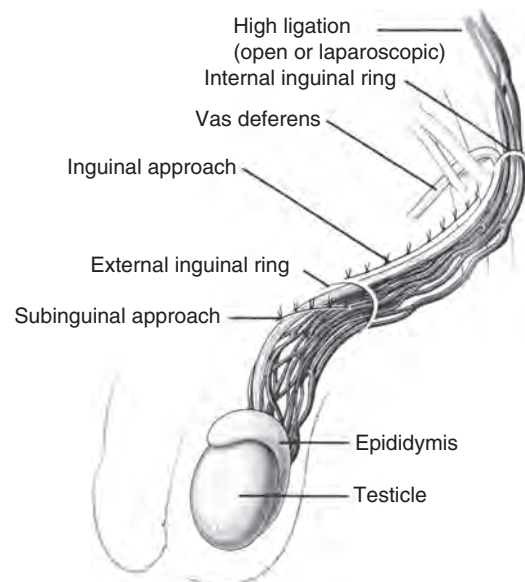


Figure 24.1 Anatomical details of different approaches.

- Surgical ligation – standard or microsurgical; artery sparing; and lymphatic sparing techniques.
- High (suprainguinal) – laparoscopic or open.
- Inguinal.
- Subinguinal.

Percutaneous embolization/sclerotherapy

This is an alternative to surgical ligation whereby the internal spermatic vein is embolized. The materials used include metal coil, spiders, plastics, brushes, detachable balloons, and sclerosing agents. A catheter is percutaneously inserted into the femoral vein under radiological guidance and the left renal vein is catheterized. The left spermatic vein is identified and the catheter is advanced into it far enough to not allow the distal end of the coil near the renal vein. Embolization is then performed using the chosen material; preferably the metal coil is used in children [40].

Advantages

This procedure is both artery and lymphatic sparing. Rivilla et al. perform this procedure under local anesthesia in children (mean age in their study: 11 years) with no complications of hydrocele or hematoma reported [40]. There is no mention of bilateral or right-sided cases in the study. Clarke et al. have reported 90% success with this procedure [41].

Disadvantages

This procedure will require the services of an interventional radiologist with expertise and this can be a limitation [41]. There is a risk of coil dislodgement, left renal thrombosis or pulmonary embolus [40,42]. Collateral veins from the left renal vein could lead to failure to cannulate the internal spermatic vein along with severe vasospasm [41] and the diameter of the coil could be larger than the target vein.

Antegrade scrotal sclerotherapy

Tauber et al. described this new technique of an antegrade approach to the internal spermatic vein combining surgery and sclerotherapy [43]. The procedure

involves an incision at the root of the scrotum and isolation and cannulation of the spermatic vein to the level beyond the internal ring. The sclerosing agent is injected mixed with air (2–3 ml agent with air in 3:1 ratio; air block technique). Patient cooperation is required to achieve increased intra-abdominal pressure by valsalva during injection under fluoroscopic control.

Advantages

This can be performed under local anesthetic and is artery and lymphatic sparing. Therefore, hydrocele formation is rare after this procedure. Tauber et al. [41] have reported a large series of 285 patients with 9% failure rate. Mazzoni et al. recommend this method as first line for recurrent varicocele, where they have had a success rate of 96% [44].

Disadvantages

An interventional radiologist with sufficient expertise is required and patient cooperation is paramount [44]. Therefore, this procedure can be of limited use in children. Complications noted include scrotal hematoma, epididymitis (3.8%), testicular atrophy, and left flank erythema [41,45]. Zaupa et al. report their experience with this technique, highlighting the fact that the procedure can take longer to perform in children due to difficulty in cannulation [46]. They warn that focal testicular necrosis can occur rarely despite an uncomplicated primary procedure.

Subinguinal (microsurgical) ligation

In a study involving infertile men undergoing microscopic varicocelectomy (mean age 32 years), Hopps et al. describe the number and relationship of internal and external spermatic arteries, veins, and lymphatics within the subinguinal portion of the spermatic cord and compare it to the inguinal approach. They found that this approach was associated with more internal spermatic veins, more external spermatic veins greater than 2 mm in diameter, and more total spermatic arteries per dissection compared to the inguinal approach [47]. Goldstein et al. were the pioneers of this approach through a subinguinal incision and identify the veins, arteries, and lymphatics with the help of an operating

microscope [48]. Schiff et al. have described the feasibility of this microsurgical approach in boys (18 years old) with a low complication rate [49]. Chan et al. report accidental ligation of the artery despite the use of an operating microscope in approximately 1% of cases [50]. They describe several reasons, including men who have small sized arteries, aggressive manipulation leading to spasm, and the close proximity of the arteries to the venae comitantes. To increase detection of the lymphatics, the use of isosulfan blue has been proposed [51].

Advantages

Artery and lymphatic sparing approach using the microscope reduces the incidence of recurrent varicocele and hydrocele formation. In theory, preservation of the artery should lead to improved spermatogenesis as demonstrated by Cayan et al., probably by a positive effect on Leydig cell function [52]. A recent collaborative review suggests that this repair has least complications and less recurrence rates compared to all other viable surgical options [39].

Disadvantages

Dissection can be tedious [26]; accidental ligation is a possibility, particularly in the pediatric age group, and it requires the availability of an operating microscope.

Inguinal ligation

The inguinal approach was described by Ivanissevich in 1918. It involves opening the inguinal canal and ligating the dilated veins whilst allowing preservation of the spermatic artery with minimal morbidity. The reported success rate is approximately 85% with hydrocele formation in about 15% [24].

Hopps et al. observed that the inguinal approach was easier compared to the subinguinal technique [47]. This approach is best avoided if the integrity of the inguinal canal has been breached with some prior procedure.

Advantages

This approach provides a familiar anatomy, easy access to cord structures, identification, and preservation of the artery without compromising success rates.

Disadvantages

This may not be suitable with previous inguinal surgery; hydrocele formation secondary to ligation of the lymphatics may occur although this can be overcome by the use of isosulfan blue.

High (suprainguinal, retroperitoneal) ligation

This approach, described by Palomo, is performed via a high inguinal incision and is very popular. It has also been successfully adapted to use by the laparoscope. The spermatic vessels are mass ligated well above the pampiniform plexus, where only a few branches require ligation. Attempts at modification to this technique by an artery sparing method led to a higher incidence of recurrence. Kass and Marcol [53] reported 89% success with artery sparing compared to 98% with the artery nonsparing original Palomo procedure. There is no evidence yet to prove that artery ligation affects testicular hemodynamics and function. Atassi et al. noted compensatory growth in both artery sparing and nonsparing groups with no detrimental growth effects such as testicular atrophy [54]. This technique spares the artery to the vas and therefore preserves the collateral blood supply to the testis. Secondary hydrocele formation varies from 3% to 36% in different series [55,56]. Oswald et al. [57] proposed the use of isosulfan blue to identify the lymphatics, and Riccabona et al. [58] demonstrated a low recurrence rate of 2% with the lymphatic sparing approach and no hydrocele formation in their series.

Advantages

There are extremely low recurrence rates, it is easy to perform, it allows adaptation by minimally invasive methods, and is very popular.

Disadvantages

Hydrocele formation is a problem due to lymphatic ligation but can be overcome using isosulfan blue to identify and preserve the lymphatics. Figure 24.2 shows the reported series in literature comparing lymphatic sparing and nonsparing approaches.

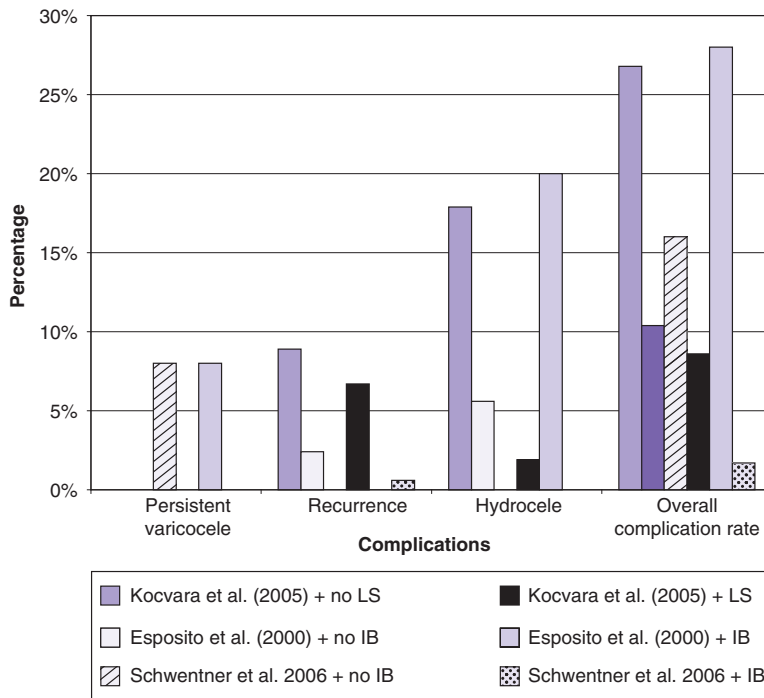


Figure 24.2 Comparing lymphatic sparing versus nonlymphatic sparing laparoscopic varicocele repair. IB, isosulfan blue; LS, lymphatic sparing.

Laparoscopic varicocele ligation

As explained earlier, the Palomo procedure has been adapted for use with the laparoscopic approach with similar recurrence rates to the open procedure, as well as low morbidity and complication rates [45,59]. Incidence of hydrocele formation due to ligation of lymphatics with this approach [45,60] remains high (11–23%). Various modifications, including the artery sparing and lymphatic sparing techniques have been described, similar to the ones explained earlier [61]. Kocvara et al. describe the laparoscopic microsurgical approach using 103–203 magnification to preserve the lymphatics [62]. This magnification is achieved by working quite close to the target. They showed that hydrocele formation and testicular hypotrophy occurred less with this technique, 1.9% and 2.9% respectively compared with 17.9% and 20.1% respectively in the conventional group, $P = 0.0003$. Poddoubny et al. have excellent results (99% success) with the laparoscopic artery and lymphatic sparing approach with no complications [63]. Recently, there is further evidence supporting the lymphatic sparing approach claiming lowest hydrocele rates [64].

Advantages

Minimally invasive Palomo procedure with similar results.

Disadvantages

Similar to the Palomo procedure.

Management and prevention of complications

Hydrocele

Regardless of the surgical approach, the most frequent complication following an operation for varicocele is the formation of a hydrocele. The incidence of hydrocele formation increases with extended follow-up and some recommend following-up for 2 years after surgery [64]. In a large multicenter study involving 278 children between 7 and 17 years of age, Esposito et al. [65] showed that the median incidence of hydrocele following varicocele surgery is about 12%. This incidence was higher (17.6%) with artery nonsparing procedures compared to artery sparing procedures (4.3%) and is well demonstrated in Figure 24.2. Esposito et al.

[65] have recommended noninvasive procedure like scrotal puncture for persistent hydrocele, which does not disappear after clinical observation. This successfully eliminated the hydroceles in 82% of the cases. The rest of the children (18%) required surgery to correct the hydrocele. Hassan et al. [60] reported an incidence of 22.8% after laparoscopic varicocele ligation with a statistically significant decrease in hydroceles when the internal spermatic vein is simply ligated rather than ligated and divided. This possibly suggests that more lymphatics get divided by the latter approach. The general consensus is that the ligation or obstruction of the lymphatics is the cause for hydrocele formation and lymphatic sparing procedures have successfully reduced the incidence of this complication (Figure 24.3) [51,57,58,61,64].

Recurrent varicocele

It is difficult to ascertain the true incidence of persistence or recurrent varicocele in comparison to different techniques with most authors claiming good success rates. However, some have suggested that efforts in trying to spare the artery result in an increased incidence of recurrent varicocele. Kass and Marcol [53] had better success with artery nonsparing Palomo procedure (98%) compared to artery sparing techniques (89%). Riccabona et al. [58] compared four different

techniques and concluded that artery sparing procedures resulted in more recurrence rates as shown in Figure 24.4. However, others have shown excellent results with artery sparing methods [63]. There is little information available as to the best way to treat recurrence. Mazzoni et al. report that antegrade sclerotherapy was more successful in recurrent cases than as a primary procedure. They recommend this method as the treatment of choice for recurrent varicocele, particularly if the internal spermatic vein has been completely occluded in the primary treatment (percutaneous retrograde sclerotherapy or open and laparoscopic ligation).

Other complications

Chrouser et al. reported transient numbness following possible nerve injury in 4.8% of cases, which appeared within 10 days of surgery and resolved at an average of 8 months [66]. This study was of boys with a mean age of 14 years who had these symptoms around the anterior part of the ipsilateral thigh consistent with injury to the genitofemoral nerve. They recommend that cautery or harmonic dissection of the peritoneum overlying the spermatic cord and excessive traction on the tissues surrounding the cord should be avoided intraoperatively. Isolated cases of partial testicular necrosis have been reported [50] while others like sigmoid serosal tear are due to laparoscopic misadventure or effects of a laparoscopic learning curve [59].

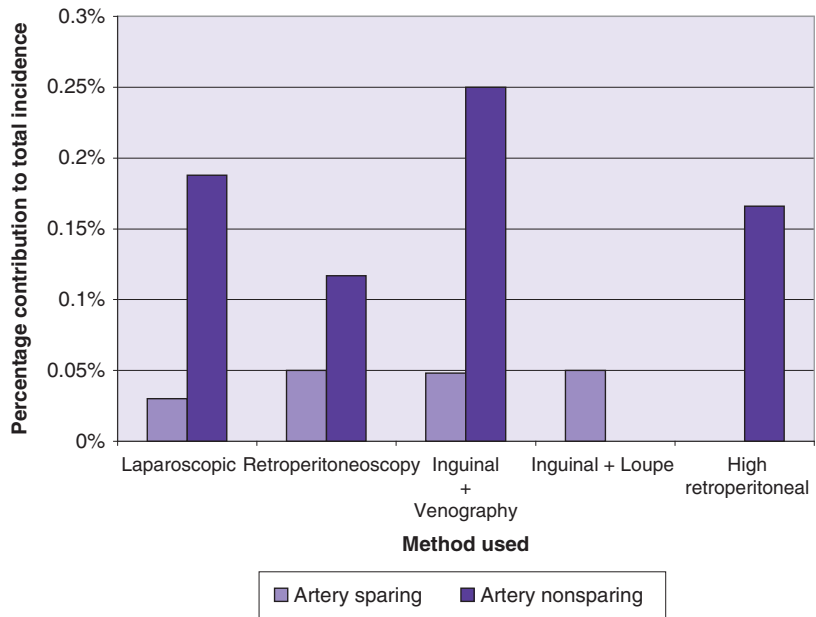


Figure 24.3 Incidence of hydrocele following artery sparing versus non-artery sparing technique.

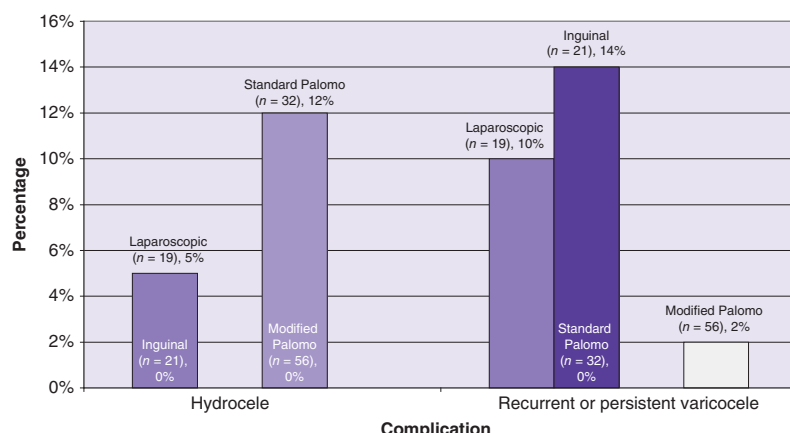


Figure 24.4 Optimizing the operative treatment of boys with varicocele: sequential comparison of techniques.

Conclusion

Various surgical procedures have been used to ligate or obliterate the internal spermatic vein including the laparoscopic or open surgical ligation in the retroperitoneum or the venous plexus inguinally or subinguinally. Microsurgical subinguinal approach seems to have least recurrence and complication rates but is not popular because of the need for a microscope and is tedious. The

standard open surgical or laparoscopic Palomo technique of nonartery sparing mass ligation remains popular with better success than the artery sparing approach. Hydrocele formation is the most frequent complication but lymphatic sparing measures seem to reduce its incidence. However, long-term studies with these techniques are needed to validate the results. Children who undergo varicocele surgery should be followed up for potential complications.

DOS AND DON'TS

Do

- Check for subclinical varicocele. Varicocele is usually clinical in diagnosis but subclinical varicocele is not uncommon and right-sided may have been under-reported until now
- Act early. Early intervention is known to give better outcomes. A Doppler ultrasound scan can be useful in diagnosis
- Follow up varicocele for at least 2 years as late occurrence of complications such as hydrocele is known

Don't

- Wait until testicular growth arrest has taken place to intervene

CLINICAL PROBLEM/CASE: DISCUSSION

- 1 Painless swelling that is restricted to the scrotum; able to reach the top of the swelling; bag of worms feel; testis felt separately and may be smaller in size compared to contralateral testis.
- 2 Traditionally, varicoceles have been classified clinically based on the classification suggested by Dubin and Amelaar adopted by the WHO [17]:
Grade I – Palpable by valsalva maneuver
Grade II – Palpable without valsalva maneuver
Grade III – Visible before palpation
- 3 Incidence [1] is approx 10% in adolescence (11–19 years; range 7.7–14.1%)

- 4 Color Doppler ultrasonography (CDU) allows diagnosis of varicocele which may not be clinically apparent. The venous reflux into the venous plexus is diagnosed using the Doppler color flow mapping in the supine and upright position, at quiet respiration, and with valsalva maneuver. Continuous reflux pattern (venous rush) during quiet inspiration (without valsalva manoeuvre or deep inspiration) is the main criterion for diagnosis of varicocele [18]. CDU is gaining popularity and some centers now call for routine preoperative CDU assessment for both testicular volume discrepancy (hypotrophy) and venous rush [19,20]. This is particularly useful for subclinical varicocele on the right side which, according to some authors, is underreported [21]. Peak retrograde flow as measured by CDU could be a useful adjunctive tool in decision-making [22].
- 5 Testicular atrophy means growth arrest. This is an absolute indication for surgery as it means that varicocele has already affected the testis significantly. The idea is to diagnose this as soon as possible. The CDU scan is useful in this respect even if it is not clinically apparent. A cut-off of 20% discrepancy in size is significant but recent evidence is that 10% asymmetry should be considered for intervention. Testicular hypotrophy may also be determined by the atrophy index [24–33].

In adolescents, there are limitations to parameters that can influence decision-making as fluctuating levels prevent hormonal testing and testicular biopsy is considered too invasive. It is impractical to request semen analysis and normograms are not available. This leaves testicular volume as the sole parameter.

Studies have also demonstrated that varicocele-related testicular hypotrophy is a developing condition [1]. Hypotrophy was not present in boys younger than 11 years. In 11–14-year-olds, the incidence was 7.3% and in 15–19-year olds it increased to 9.3%.

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CHAPTER 25

Complications of newborn circumcision

Prevention, diagnosis and treatment

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KEY POINTS

- Circumcision for the vast majority of infants and children is a safe and well-performed procedure
- In experienced hands, acute complications should occur in less than 1% of patients
- Recognizing genital anomalies and judicious patient selection is the key to preventing complications
- If there is questionable genital anatomy, circumcision should be deferred and the patient referred for consultation
- Parents should be provided with clear and complete post-procedural instructions

CLINICAL PROBLEM/CASE

Case 25.1: Delayed bleeding

Twenty-four hours following a routine circumcision performed in the operating room, a 1-year-old boy returns to the emergency department with oozing of blood from the dorsal suture line. The parents noted the problem 4 hours earlier but pressure has not stopped the bleeding. He is noted to have a swollen, purplish discolored penis. There are clots covering the distal shaft and glans and the diaper is bloody.

- 1 What should be included in the differential diagnosis and what is the most likely cause of bleeding?

Case 25.2: Glans injury/amputation

You are called to the NICU because of excessive bleeding following a Mogen clamp circumcision. The obstetrician is concerned that there may be a glans injury. Urologic evaluation reveals that the resected foreskin specimen contains the entire glans.

- 1 What immediate steps should be taken to deal with this crisis?
- 2 What is the approach to re-anastomosis in the OR?

Case 25.3: Trapped penis

The parents of a 1-month-old boy who had a neonatal circumcision have noticed that his penis looks as though it has disappeared. There is very little outside penile skin present and the penis is palpable inside the pubic fat pad. There is a tight constricting ring over the glans and the meatus is just visible. He has no difficulty voiding. The parents are very upset.

- 1 What is the problem?
- 2 What are likely causes?
- 3 What are the options of treatment?
- 4 What steps should be taken during the revision?

Case 25.4: Adhesions

Three-year-old twin boys are referred by their pediatrician because their circumcised penises don't look right. They are otherwise asymptomatic and voiding well. On exam, twin A's coronal margin is visible only on the right side. The coronal margin is covered by skin dorsally and on the left, and this looks like a simple adhesion. Twin B's coronal margin is also mostly obscured but has a different appearance because the adhesions are caused by dense bridges

of skin. Small openings can be seen on the sides of the skin bridges.

- 1 What causes these adhesions?
- 2 What are the treatment options for twin A?
- 3 What are the treatment options for twin B?

Case 25.5: “Bad circumcision”

Parents of a 1-year-old boy are upset because it looks to them that not enough skin was removed during his circumcision. On exam, the penile shaft skin overhangs the glans.

- 1 How do you determine if the correct amount of skin was removed?

- 2 If there is excess skin, can the child eventually “grow into it”?

Case 25.6: Urethral injury

A 2-year-old boy who had a neonatal circumcision is referred because his penis curves down, especially with an erection. On exam, his meatus is located on the glans, but he has a small urethrocuteaneous fistula, and his ventral skin is short. Insertion of an 8 French catheter for 2 cm shows no obstruction, but there is a paper thin urethra with fistula and moderate-to-severe ventral curvature.

- 1 What are the treatment options?

Introduction

Elective newborn circumcision remains a common and controversial practice. Increasingly, parents are faced with making this highly personal decision in a context of outspoken advocacy on both sides and with few sources of unbiased guidance. The most widely-used information resource, the Policy Statement and Technical Report produced by the American Academy of Pediatrics Task Force on Circumcision [1], recommends that parents be informed of the relevant risks and benefits and then be allowed to choose within a context of cultural, religious, and ethnic factors. It is important that physicians who counsel parents and surgeons who perform neonatal circumcision understand the incidence of complications, their impact, treatment and, most importantly, their prevention.

Epidemiology of complications

The true incidence of complications after newborn circumcision is unknown, in part due to differing opinions about what constitutes a complication. Adding to the confusion is the separation of acute complications, such as bleeding or infection, from late complications such as adhesions, meatal stenosis, and unacceptable cosmetic outcome [2].

Acute complications

Two large studies evaluating elective newborn circumcision performed by physicians in a US hospital setting reported a complication rate of <1%.

Wiswell and Geschke [3] in their classic study reviewed the records of more than 136,000 boys born in US Army hospitals. There were more than 100,000 circumcisions and the overall complication rate was 0.19%. The incidence of bleeding was 0.08%. Infection occurred in 0.06% including eight cases of bacteremia, too much or too little skin removed in 0.01%, and there were no deaths.

Christakis et al. [4] reviewed the hospital discharge records of over 350,000 boys born in Washington State from 1987 to 1996. This included over 130,000 circumcisions. They were able to identify a complication in 0.22% of circumcisions. The risk of bleeding was the most common with an incidence of 0.18%. A penile injury occurred in 0.04% whilst infection was identified in 0.0008%.

A significant limitation of both studies is reliance on hospital coding. Many minor complications may have not been captured, underestimating the actual incidence. Another limitation is that neither study was able to capture minor complications that may have been treated in an outpatient setting following discharge.

In a study of approximately 1800 boys who were circumcised in Atlanta over a 3-month period, the overall rate of complications was significantly higher at 3.1% [5]. Bleeding was the most common complication occurring in 2.1%. The higher incidence may reflect a more thorough review of the patient’s record including physician and nursing notes as opposed to coded discharge data.

Unfortunately, none of these studies report late complications of neonatal circumcision.

Acute severe complications including infections, glans amputation, urethrocuteaneous fistula, ablation of the

phallus, and death have rarely been reported [6–20]. Given their rarity, it has not been possible to determine their true incidence.

Late complications

Late complications may be recognized weeks, months or years after neonatal circumcision. These complications include excessive residual skin (incomplete circumcision), excessive skin removal, adhesions (natural and true skin bridges), meatal stenosis, phimosis (trapped penis), and epithelial inclusion cysts.

In a review of 468 consecutive boys presenting to a general pediatric practice whose consultation included a genital examination [21], late complications of circumcision included a redundant residual prepuce in 20.1%, adhesions in 25.6%, balanitis in 15.5%, a skin bridge in 4.1% and meatal stenosis in 6%. Ponsky [22] found that the rate of penile adhesions was related to patient age. Children less than 1 year old had a 70% incidence of adhesions. The rate of adhesion in those aged 1–5 years was 28%, those 5–9 was 8% and those over 9 had a rate of 2%. Blalock [23] reported a 2.9% incidence of phimosis following newborn Gomco circumcision.

Van Howe [24] reported a 7.3% incidence of meatal stenosis in Tanner stage 1 boys greater than 3 years old. However, this was based on appearance rather than function and therefore may represent an overestimation.

Comparison of neonatal circumcision techniques

There are three techniques most commonly used in the USA for newborn circumcision: the Gomco clamp, the Plastibell clamp and the Mogen clamp. The Gomco and Plastibell devices both have shields that cover and protect the glans during the circumcision and are more commonly used in hospitals by physicians. The Mogen clamp is mostly used by Mohels for religious circumcisions. In less experienced hands there is a greater potential for glans injury using the Mogen clamp. It is evident from the case reports of neonatal amputation that the majority, if not all, occur with the Mogen.

Prevention of complications

Complications of neonatal circumcision may result from poor surgical technique, incorrect postoperative care, or

unrecognized penile anomalies that predispose the circumcision to a poor result. Proper patient selection is critical to achieving a favorable outcome [25,26]. Optimal conditions during the circumcision are necessary to prevent complications. An experienced physician must perform or carefully supervise the procedure. Optimal lighting, a comfortable position, and readily-available assistance and supplies are required.

Following the circumcision, parents must receive explicit instructions about post op care including the frequent application of antibiotic ointment to all raw/denuded areas to prevent adhesions.

It cannot be overemphasized that neonatal circumcision is elective and, where anatomic contraindications are present, circumcision should be postponed until such time when it can be combined with the specific penile surgery that is needed. The foreskin is often needed to correct anatomic abnormalities; thus, its removal may preclude or reduce the available options for subsequent surgical correction.

A pre-circumcision exam must be done prior to applying surgical drapes to prevent a subtle anomaly from being obscured. The anatomic conditions that should alert the clinician to deferring neonatal circumcision may be divided into primary penile abnormalities, dartos fascia abnormalities, and penoscrotal anomalies and distortions.

Primary penile abnormalities

Newborns with a primary penile abnormality are most often easily recognized, and typical newborn circumcision techniques should be deferred when these anomalies are present. Some anomalies may be subtle and require a high index of suspicion to detect.

Hypospadias and chordee

Hypospadias and chordee without hypospadias usually have a dorsal hooded foreskin and variable degrees of ventral bending of the penis. In hypospadias, the urethral meatus is located proximal to its usual position on the glans (Figure 25.1a), whereas in chordee without hypospadias (a forme fruste of hypospadias), the meatus is in a near normal position, but the distal urethra may be dysplastic, inelastic, and it may be responsible for the chordee [27]. In both conditions, there is often a deficiency of the ventral shaft skin. Therefore, in these



(a)



(b)



(c)



(d)



(e)

Figure 25.1 Contraindications to circumcision: (a) hypospadias; (b) congenital buried penis; (c) megaprepucium; (d) penoscrotal fusion; (e) large hydroceles – relative contraindication.

conditions, the foreskin must be preserved because it may be needed for both urethral reconstruction and ventral penile skin coverage. Ten percent of boys with hypospadias have a normally-formed foreskin and no ventral curvature, thereby masquerading the defect [28]. This is most commonly associated with a very large meatus extending onto the distal shaft of the penis and referred to as megameatus intact prepuce (MIP) hypospadias. The abnormal urethral meatus is usually discovered after the dorsal slit of the circumcision is done or after the circumcision is completed. Fortunately, in the majority of these cases, the defect can be corrected without the foreskin.

Epispadias

Epispadias is a condition most commonly associated with exstrophy of the bladder, but it may also occur as an isolated entity. In epispadias, the urethral opening is on the dorsum of the glans or shaft, or it may end at the level of the pubis. The proximal forms are associated with sphincteric insufficiency and urinary incontinence. The entire foreskin is located ventrally, and there is a variable degree of dorsal penile curvature. In epispadias, as in hypospadias, foreskin preservation is critical for use in urethral reconstruction and skin coverage. Rarely, there are covered epispadias variants where the prepuce appears completely formed but with a concealed dorsally positioned meatus.

Micropenis

A micropenis is a normally-formed but abnormally short and slender penis that has a stretched length that is at least 2.5 standard deviations below the mean. In the full-term newborn, this corresponds to a stretched penile length smaller than 1.9 cm [29]. In extreme cases of micropenis, there is barely any shaft, and the glans appears to sit almost on the pubic skin. This should be distinguished, however, from the completely hidden or buried penis which may also have very little penile shaft evident above the skin level but have a normally palpated shaft below skin level. The child with micropenis should be evaluated endocrinologically for a chromosomal or systemic hormonal abnormality. Treatment of micropenis is primarily hormonal stimulation with variable degrees of growth response. Circumcision should be deferred in newborns with a micropenis. Performing a circumcision may create the illusion of making the penis appear even smaller.

Dartos fascia abnormalities

The dartos fascia is the subcutaneous layer of the penile skin that is normally fixed to Buck's fascia which in turn is fixed to the corpora of the penile shaft. This close application of the skin, dartos, and Buck's fascial layers is responsible for the usual snug fit of the penile skin to the shaft and the normal protuberance of the penis from the pubic region [30]. Dartos fascia abnormalities are often subtle and easily missed during a pre-circumcision exam. Too often the penis is viewed as just small and then circumcised when in fact it was a buried penis and circumcision should have been deferred.

Buried penis

A common anatomic variant that is often not recognized is that of a penis whose shaft, and often glans, lie just beneath the surface of the penoscrotal skin. This anatomic variant – known by the synonyms buried penis, concealed penis, inconspicuous penis, hidden penis, and engulfed penis – has abnormally loose or nonexistent dartos attachments to Buck's fascia, which allows the penile skin that would normally be fixed to the shaft to be pushed up and over the penile shaft (Figure 25.1b). In these cases, there appears to be a small “foreskin cap” on the surface with the penis sunken into the pubic fat. The “foreskin cap” is the foreskin and penile shaft skin combined, and if this is removed in a circumcision, the patient will have no outer penile shaft skin. Physiologic phimosis is invariably present. In addition, other deep dartos attachments commonly adhere to the Buck's fascia, keeping the penile shaft in a buried or trapped position and preventing it from assuming a more protuberant position after the circumcision. The outcome of a circumcision in newborns with this anatomic variation is often excessive removal of the outer penile shaft skin or insufficient removal of the foreskin and a relatively rapid formation of a secondary post-circumcision phimosis (trapped penis). This is an extremely important anatomic variant to recognize and requires circumcision deferral. If the parents prefer circumcision, it should be done simultaneously with the buried penis repair, which is usually deferred to beyond 6 months of age. Circumcision in combination with complete take down of all tethering dartos bands and tacking the base of the penile skin to the corpora can then be performed. This can be a complex penile skin reconstruction, and for patients with a heavy pubic fat distribution, it may be in their best interest to avoid circumcision until they outgrow or lose their excess pubic fat by weight loss.

Megaprepuce

Megaprepuce is a condition at the extreme end of the spectrum of buried, concealed, or trapped penis [31]. What differentiates megaprepuce is the marked trapping of urine inside a large pocket of massively redundant inner preputial skin (Figure 25.1c). Megaprepuce has never, to our knowledge, been detected *in utero*, and parents do not recognize it at birth. It presents gradually, rarely before 3 months of age, as a swelling of the upper scrotum that balloons during voiding.

Penoscrotal transposition

Penoscrotal transposition is a congenital anomaly in which the penis is engulfed in the middle of the scrotum. It may occur as an isolated anomaly but more often is seen in association with other genital abnormalities, such as hypospadias, ventral curvature, or various forms of urethral duplication. When correction is indicated, the reconstruction is complex and all available tissue is needed; therefore, circumcision is contraindicated.

Penile torsion

Penile torsion is a counterclockwise rotational defect that is commonly associated with lesser degrees of hypospadias. The ventrum of the penis (both glans and shaft) is directed toward the patient's left side, a counterclockwise rotation. The deformity is due to abnormal skin and/or dartos attachments, and not to any defect in the penile corpora. In isolated cases, repair is indicated when the degree of torsion is greater than 90°. A pediatric urology consult should be obtained for any rotation greater than 45°. Neonatal circumcision should be deferred if surgical repair is being considered and can be performed at the time of the repair.

Penoscrotal anomalies and distortions

Webbed penis

A webbed penis (penoscrotal webbing) is caused by an abnormally high scrotal attachment onto the ventral penile skin (Figure 25.1d). This condition occurs as a spectrum with mild forms requiring no treatment and posing no contraindication to circumcision. However, in the more severe forms in which the web extends to and is confluent with the distal ventral penile skin, neonatal circumcision should be deferred because it would

create an appearance of having no ventral penile skin with glans tethered to scrotum. Circumcision under these circumstances can also lead to secondary phimosis. If the parents prefer circumcision, this configuration should be corrected under general anesthesia after 6 months of age. It should be noted, however, that not all of the severely webbed penises require surgical correction, especially if circumcision is not desired. After the natural preputial adhesions resolve and the foreskin becomes retractable, the point of attachment of the web becomes more proximal with foreskin retracted.

Hydroceles

Large bilateral hydroceles, which are usually observed for spontaneous resolution during the first year of life if not associated with an inguinal hernia, may engulf the penis, pushing the penile skin upward, mimicking a buried penis. It may be difficult to judge how much skin to remove with this degree of distortion, especially if the penis is already on the small side (Figure 25.1e). If a circumcision is done, the penis may "sink back inside" and the actual circumcision line may extend beyond the glans and result in a trapped penis. Large bilateral hernias may cause similar distortions, but generally need to be corrected earlier.

Ambiguous genitalia and disorders of sex development

The finding of ambiguous genitalia in the newborn is a medical and social emergency. No surgical procedures should be considered except those related to making the correct diagnosis. All genital skin must be preserved for potential reconstructive procedures. Most often the ambiguity is evident. However, certain conditions in the apparent male should trigger a workup for a disorder of sex development (DSD), such as bilateral impalpable gonads, regardless of the degree of external virilization, or hypospadias, with a unilateral impalpable gonad [32].

Diagnosis and treatment of complications

Acute, non-catastrophic complications

Bleeding

Postprocedure bleeding is the most common complication of circumcision (Figure 25.2a). Most bleeding



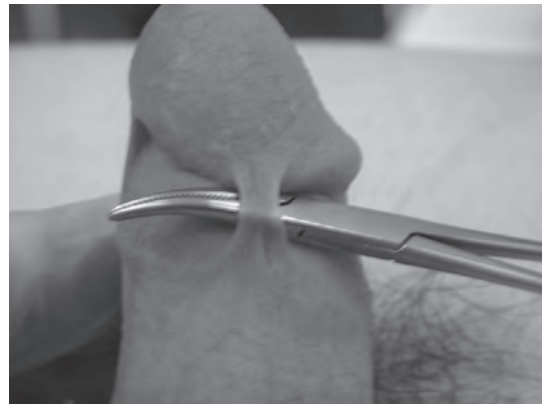
(a)



(b)



(c)



(d)



(e)



(f)

Figure 25.2 Non-catastrophic complications of circumcision: (a) postoperative bleeding; (b) meatal stenosis; (c) trapped penis; (d) penile skin bridge; (e) incomplete circumcision; (f) epithelial inclusion cyst.

occurs shortly after the procedure and its recognition is obvious. Parents should be instructed to inspect for bleeding, especially following the removal of any dressing. A blood-saturated dressing or diaper should be cause for seeking attention.

Minor bleeding can typically be controlled with manual pressure. The use of thrombostatic agents such as Surgicel or thrombin, can be quite helpful. For more significant bleeding, such as from the frenular artery, suture ligation is usually necessary. Caution can be used only with great caution due to risk of urethral injury. The need for blood transfusion is exceedingly rare.

Significant or prolonged venous oozing should suggest the possibility of a bleeding disorder and initiate an evaluation. Patients with suspected bleeding disorders or significant family histories should undergo pre-procedure evaluation. In rare circumstances where the family is adamant, circumcision can be safely performed with appropriate preparation with fresh frozen plasma or specific blood product.

Infection

Infection is the second most common complication of neonatal circumcision [33–34]. Infection can be characterized by fever, erythema, wound separation and discharge. Infection can almost always be adequately treated with local wound care, with or without oral antibiotics. Infection is best prevented by strict adherence to antiseptic technique. With the specific use of the Plastibell clamp, prolonged failure to separate can be associated with a greater risk of infection. Families should be instructed to seek attention if the ring has not fallen off within the normal 3–7 days.

Denuded penis

When an excess amount of skin is removed during circumcision, the skin edges may separate leaving areas of the shaft denuded. The denuded area can generally be left to safely heal by secondary intention with only local wound care. Primary closure is unnecessary and may leave a shortened phallus. Success has been described with full-thickness skin grafts; however, the cosmetic outcomes are usually substandard using an extragenital skin graft and it is unclear whether or when this is necessary [35,36].

Anesthetic complications

Circumcision has without question been shown to cause pain and physiological stress. There is no longer

any justification for withholding effective pain control. There are four different anesthetic techniques primarily used for circumcision, including: oral sucrose, locally applied eutectic mixture of local anesthetics (EMLA) cream, dorsal penile nerve block, and subcutaneous ring block. Complications from oral sucrose administration have not been described. A rare complication of EMLA cream is methemoglobinemia caused by one of prilocaine's metabolites oxidizing hemoglobin to methemoglobin, leading to life-threatening oxygen deprivation and central nervous system toxicity [37]. This complication is dose-dependent. The dorsal penile nerve block is administered by injecting 1 ml of lidocaine or bupivacaine without epinephrine at the base of the penis in the 10 and 2 o'clock positions using a 27 gauge needle. Aspiration prior to injection is important to prevent intravascular or intracorporeal injection. Bruising and hematoma formation at the injection site are the most common complications following dorsal penile nerve block and occur in approximately 1% of cases [38]. A subcutaneous ring block, which alleviates pain better than a dorsal penile nerve block, is achieved by injecting lidocaine or bupivacaine circumferentially at the midshaft or base of the penis, raising a wheal [39–41]. Temporary skin deformation and hematomas at the injection sites have been reported, but systemic toxicity has not [42–46].

Late, non-catastrophic complications

Meatitis and meatal stenosis

Meatitis occurs in up to one-third of circumcised boys [47]. It typically occurs when the child is still in diapers, and the urethral meatus, no longer shielded by the prepuce, is exposed to the ammonia on urine-soaked diapers. Bacterial enzymes produce the ammonia, which can lead to meatal inflammation even in the absence of infection. Meatal stenosis occurs when the inflamed ventral meatal edges fuse to form a web over the meatus (Figure 25.2b). There are no well-defined prevention strategies. Diagnosis is usually considered because of dysuria, a very thin or upwardly deflected stream, and/or the observation of a small-appearing meatus. Treatment is surgical incision either as a meatotomy or a more formal meatoplasty.

Phimosis

Post-circumcision phimosis can occur when the glans drops back inside the circumcision line which then

contracts distal to the glans (Figure 25.2c). This can create a trapped penis in which the glans cannot be fully exposed. In rare cases, voiding can be painful or compromised. Prevention includes careful patient selection and removal of the appropriate amount of inner and outer preputial skin. Parents should be instructed to maintain the incision proximal to the glans during the initial postoperative period. Initial treatment of post-circumcision phimosis can be with topical steroids which, if initiated early, can soften the cicatrix sufficiently to liberate the glans. If the phimosis is severe and fails to respond to topical therapy, a circumcision revision under general anesthesia is needed [48,49].

Penile skin bridges and adhesions

One of the most frequent sources of parental concern is the development of post-circumcision adhesions. It is important to understand that there are two distinct forms of adhesions: natural adhesions and penile skin bridges (Figure 25.2d).

Natural adhesions form as attachments between the former inner preputial skin, which is now the subcoronal collar, and the glans. These adhesions are very common and will resolve spontaneously over time. They are especially frequent during infancy when the abundant suprapubic and peripenile fat causes the penile shaft skin to push forward and places these raw skin surfaces in constant opposition. Diagnosis is made by clearly identifying the circumcision incision line as being safely away from the area of attachment. Treatment can be reassurance or with topical steroid ointments. Forceful manual separation is not necessary and should be discouraged.

Penile skin bridges are differentiated as true vascularized scars between the circumcision incision line and the glans. It is believed that their incidence can be reduced by careful attention in the immediate postoperative period. Unlike natural adhesions, these will not resolve and do require surgical lysis. If identified early they can often be incised in the office setting under topical anesthetic, simply relying on crushing of the tissue for hemostasis. More extensive bridging or a later presentation may warrant a formal lysis in the operating room with suture repair of the incised areas with or without circumcision revision.

Incomplete circumcision

An all-too-frequent concern of parents is having too much redundant prepuce remaining following

circumcision (Figure 25.2e). The majority of complaints occur during mid-infancy when the development of significant suprapubic and peripenile fat has the ability to advance loosely attached penile shaft skin distally over the glans. The majority will resolve with time as the child thins and the penis grows. However, for some children, clearly an inadequate amount of prepuce was removed. Diagnosis depends on a careful physical assessment with the fat pushed away from the base of the penis. If there is clearly too much residual prepuce and the family is unaccepting of the appearance, a circumcision revision can be performed. Any ambivalence should be treated with observation, because the decision becomes easier as the child thins and grows.

Epithelial inclusion cysts

Inclusion cysts can form along the circumcising line. They are believed to be caused by either implantation of smegma in the circumcision wound or by surgical inversion of the epidermis at the time of circumcision (Figure 25.2f) [50]. Epithelial inclusion cysts can enlarge or, rarely, become infected. Treatment includes conservative observation or surgical excision.

Catastrophic complications of circumcision

Urethrocuteaneous fistula

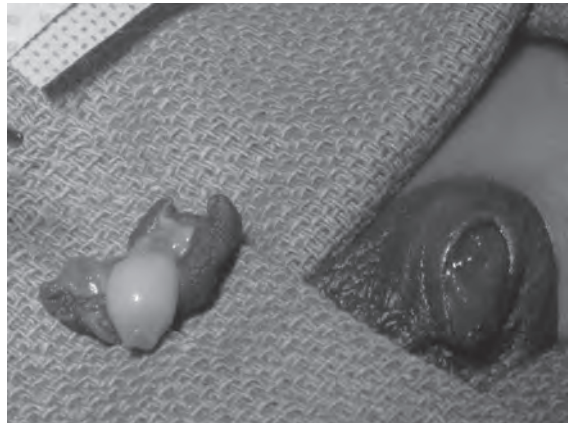
Urethrocuteaneous fistulas are rare but severe complications (Figure 25.3a). Fistulas result from injury to the urethra, typically during control of frenular bleeding, particularly with the use of cautery or overly aggressive suturing. Urethrocuteaneous fistulas require formal repair 6–9 months after the initial injury [52–54].

Glanular injury or amputation

Traumatic penile amputation is the most devastating complication associated with circumcision (Figure 25.3b). This occurs primarily with the Mogen clamp technique of circumcision during which a portion of the glans is inadvertently caught between the jaws of the clamp. An inappropriately sized clamp or a clamp that has become worn and unacceptably lax has been suggested as a cause of this complication. Prevention relies on appropriate adhesion release prior to circumcision, ensuring the width of the Mogan clamp is appropriately limited and that the clamp is carefully



(a)



(b)



(c)

Figure 25.3 Catastrophic circumcision complications: (a) urethrocutaneous fistula; (b) complete glans amputation; (c) necrotizing fasciitis and debridement.

placed. Diagnosis should be immediate and the specimen retained. It should be kept moist and placed in a plastic bag and placed on ice. Treatment is immediate reattachment of the amputated glans. If there is urethral involvement, the urethra is first anastomosed around an indwelling catheter followed by the glans-to-glans re-anastomosis, which can be performed with good results [53–57].

Sepsis

Severe infection following circumcision can, rarely, lead to overwhelming sepsis. These babies may have an underlying immunological disorder predisposing them to infections. Treatment includes broad-spectrum followed by culture-specific antibiotics, hospitalization, and supportive care. Necrotizing fasciitis is a rare but devastating, rapidly-spreading infection caused by gas

forming bacteria that usually requires immediate life-saving surgical debridement of all necrotic tissue (Figure 25.3c) [58–62].

Necrosis

When circumcision is performed with a Gomco clamp, the skin should always be excised with a scalpel. Penile necrosis is a rare and completely preventable complication caused by skin removal using electrocautery during a circumcision with a Gomco clamp. Electric current can then transmit throughout the surface of the metal clamp, leading to widespread cauterization necrosis [63]. Electrocautery should never be used during a Gomco circumcision.

Conclusion

Circumcision for the vast majority of infants and children is a safe and well-performed procedure. Complications should be kept to a minimum through attention to prevention. Along with careful technique and clear postoperative instructions, the key to prevention is judicious patient selection. Since circumcision is an elective procedure with minimum measurable benefit, it is always better to err on the side of caution. All questionable genital anatomy should be referred for pediatric urology consult. It is best to defer circumcision when there is an associated penile anomaly that will require future correction.

DOS AND DON'TS

Do

- Carefully assess the anatomy before initiating the circumcision
- Carefully assess the overall health of the child and look for red flags that may signal a bleeding disorder
- Carefully explain to parents the postoperative care you desire and provide written instructions

Don't

- Be afraid to defer circumcision and seek expert consultation
- Use a clamp device that appears worn or loose, or one where the parts do not fit tightly together
- Use an electro cautery with any metal clamp device

CLINICAL PROBLEM/CASE: DISCUSSION

Case 25.1 discussion

An underlying hematologic abnormality should be considered, but the most likely cause is bleeding from an artery or vein that has reopened spontaneously.

The management is immediate surgical exploration. Get an H&H before going to the OR. There may have been more bleeding than was apparent. A systematic approach to finding the bleeder should be used. Open the incision, evacuate the clot, and use copious irrigation. If a bleeder is not readily apparent, proceed with a four quadrant exploration looking for a retracted vessel. Sometimes, after hemostasis seems intact, bleeding may recur when the blood pressure rises associated with awakening. Therefore, after hemostasis is achieved and before awakening, have the anesthesiologist raise the blood pressure to a higher than normotensive level to assure security of the hemostasis. Reapproximate the skin and apply a mild compression dressing. A hematologic screening should be done that includes a family history of bleeding problems, PT, PTT, CBC, and platelets.

Case 25.2 discussion

- 1 Compress circumcision site to stop bleeding.
Place the specimen in very cold sterile saline.
Notify the operating room.
Meet with parents immediately with complete disclosure of the accident and the surgical plan.
Alert hospital "adverse events" team.

- 2 Time is of the essence; a microsurgical vascular repair is not indicated. Align the urethra over the catheter. Consider three separate anastomoses: urethra-to-urethra, glans to distal corpora, and skin-to-skin using fine absorbable suture.

Postoperative care: No dressing and no handling of the penis except for gentle application of antibiotic ointment to prevent desiccation and infection. Strict immobilization of the patient and intensive constant monitoring by NICU staff is absolutely necessary. This is a delicate composite graft and any inadvertent trauma could result in complete graft loss. Even with successful graft take, expect some atrophy.

Case 25.3 discussion

- 1 The glans has dropped back inside the circumcision line and the circumcision line has contracted, forming a dense cicatrix distal to the glans trapping the penis.
- 2 Incorrect patient selection (buried penis, megaprepuce), excessive inner preputial skin, and inadequate parental postoperative care instructions may be contributory.
- 3 The options include betamethasone cream initially for 4–6 weeks twice a day versus a circumcision revision. Parents can be told that there is about a 30% chance of resolving the phimosis with betamethasone cream, but a revision (some form of penile skin reconstruction) still might be needed to correct a buried penis or to remove redundant skin.
- 4 The revision should begin with a complete release of the penis from the tethering dartos. Try to remove the least amount of skin. It is important to tack the base of penile skin to corpora in four quadrants to prevent the penis from retracting again.

Case 25.4 discussion

- 1 Simple adhesions are caused by incomplete take-down of the natural adhesions during the circumcision or by readherence of the raw inner preputial skin to the raw glans surface in the immediate postoperative period. These adhesions are usually preventable by generous application of ointment until the raw surfaces have re-epithelialized. Skin bridges are formed when a separated portion of the circumcision line densely fuses to the raw glans.
- 2 Blunt lysis in the office is to be condemned. Even with instructions to apply ointment, readherence usually occurs; observation alone virtually always results in slow but complete resolution over time. Betamethasone cream is usually effective in hastening this process.
- 3 Surgical lysis is the treatment for skin bridges. If the bridges are few and thin, they can be divided in the office under EMLA cream local anesthesia. Otherwise they are treated in the OR. The bridges are crushed with curved hemostats and divided flush with glans. Fine sutures may be needed to close skin separations. Parents should be told that small skin tags may remain attached to glans.

Case 25.5 discussion

This is a common situation resulting from pubic fat pushing the base of the penile skin forward. In some cases, the skin and dartos are not properly fixed to Buck's fascia.

- 1 Push back on base of penile skin and stretch penis (simulating erection) to see if there is truly excessive skin.
- 2 Parents of infants need to be reassured that the majority will resolve with natural thinning of the suprapubic and peripenile fat and growth of the penis with time. It is rare to encounter this condition in adolescents. Asymmetric flaps and residual grossly excess wrinkly preputial skin merit revision for cosmetic reasons.

Case 25.6 discussion

A urethral injury could happen easily on a patient with this anomaly.

Were there any tips that there was an underlying dysplastic urethra in this patient with chordee without hypospadias?

A dorsal hooded foreskin or a deficient ventral foreskin should suggest the possibility of an underlying abnormal urethra. Other clues are obvious chordee or short ventral skin.

Urethral injury may result from excessive cauterization of the frenular artery. If cautery is necessary a fine tipped bipolar or thermal eye cautery should be used. Cautery should never be used with the Gomco clamp to excise the skin. Dislodgement of a Plastibell string that encircles the penis is another potential cause of urethral injury.

If a urethral fistula develops, delayed repair following resolution of the tissue trauma using formal urethroplasty/hypospadias principles should be employed.

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CHAPTER 26

Surgical complications of hypospadias and their management

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KEY POINTS

- Urethroplasty complications are more common after proximal than distal hypospadias repairs, and following reoperations compared to primary operations
- Glans size <14 mm also correlates with increased urethroplasty complications
- Various technical factors, including two-layer subepithelial urethroplasty and coverage flaps over the neourethra, have been shown to reduce complications in proximal tubularized incised plate (TIP) repair
- Age at repair is not an independent risk factor for complications, and evidence that preoperative androgens reduce complications is mixed

CLINICAL PROBLEM/CASE

This is a 9-month-old boy who was admitted electively for a hypospadias repair. His preoperative appearances are shown in the figure.

- 1 What factors are important in determining whether to perform a staged versus single-stage repair?
- 2 What are the determinants of a successful outcome in a primary TIP repair?



Introduction

This textbook opens with a description of surgical audits intended to inform surgeons of their personal results, as well as to suggest areas where practice improvement is possible.

The first author of this chapter (WS) began prospective data collection for all his hypospadias operations in 1999, and has continued to enter preoperative, intraoperative and postoperative information on the day of service into computer spreadsheets to the present time.

We have published various reports derived from review of these databases, including changes in surgical technique to reduce fistulas and glans dehiscence, our two most common complications.

But an added benefit of a large dataset of systematically-collected information on consecutive patients undergoing a standard operative algorithm is the ability to apply advanced statistical analyses to isolate factors impacting outcomes. Using training obtained while earning a Master's Degree in Clinical Sciences, the second author (NB) has used multiple logistic regression to assess the contribution of individual factors impacting results so that further practice changes can be made and then evaluated.

This chapter is written from these databases and their analyses.

Key factors predicting hypospadias complications

Multiple logistic regression analysis was done using a dataset with outcomes for 669 consecutive boys after tubularized incised plate (TIP) repair for distal ($n = 540$, 81%), midshaft ($n = 50$, 7.5%), and proximal hypospadias ($n = 79$, 12%), of which 73 (11%) were reoperations and the remainder primary repairs all done by WS. Overall urethroplasty complications (fistula, dehiscence, meatal stenosis, stricture) occurred in 11.5%. The only independent risk factors for these complications were:

- Reoperation: odds ratio (OR) 3.07 (95% confidence interval (CI) 1.54–6.13).
- Meatal location: OR 1.79 (95% CI 1.34–2.40).

Compared to patients with primary distal repairs, those with midshaft and proximal TIP had a 1.8× and 3.6× increased risk for complications [1].

Based on these data, it is apparent that the first repair is very important, especially in patients with proximal hypospadias, since reoperation carries an increased risk for additional complications and further surgeries. It is also apparent that proximal cases comprise only approximately 10% of hypospadias repairs. A recent report from the American Board of Urology stated the average number of proximal repairs done annually by full-time pediatric urologists in the USA was only two [2]. Given the small number of patients with proximal hypospadias and the added complexity of their surgical repair, our group created a subspecialization model with one surgeon (WS) designated to perform all proximal repairs, frequently working with a successor (NB) who will gain the experience needed to eventually assume that practice.

Distal TIP outcomes

We reported outcomes in 551 consecutive boys with distal hypospadias undergoing primary repair by WS. TIP was used in each case, with no contraindications. Follow-up of 426 patients at a mean of 8 months found urethroplasty complications in 4%, comprising 2% fistulas and 2% glans dehiscence. There were no short-term meatal stenoses and no urethral strictures [3].

Our prospective series and two other retrospective studies included characterization of the urethral plate as “deep,” “cleft” or “intermediate,” and “flat” or “shallow.” None found urethroplasty outcomes related to these descriptions of the urethral plate groove [3–5].

However, both these other studies reported urethral plate width <8 mm, found in 23–49% of patients, increased complications [4,5]. The series reported by Sarhan et al. [5] included midshaft patients who, on univariable analysis, were found to have greater complications than those with distal repairs. However, the series did not report multivariable analysis controlling for this variable, and it was unclear how many midshaft patients were included in the group with urethral plate width <8 mm. Furthermore, since urethral plate width was not the primary outcome of interest and the authors did not adjust for multiple statistical testing, the difference they report could have been found by chance alone.

Since 2012, we have measured urethral plate width in consecutive boys with distal hypospadias, finding 90% have plate width before incision <8 mm. Whilst we have not yet analyzed these data to determine if

urethral plate measurements correlate with outcomes, we have observed that small glans width ≤ 14 mm is an independent risk factor for urethral complications (OR 3.1, 95% CI 1.4–7). Multivariate logistic regression analysis with glans size as a continuous variable demonstrated that each 1 mm increase in glans diameter was associated with a decreased risk for urethral complications (OR 0.79, 95% CI 0.63–0.98, area under the curve (AUC) = 0.743) [6].

A review of 35 series published from 1994 to 2009 found complications reported in 0–24% after distal TIP, with 25 of the studies having <10% urethroplasty complications; mostly fistulas and meatal stenoses [7].

Proximal TIP outcomes

We reported outcomes in 59 consecutive patients undergoing proximal TIP by WS, with 53% complications in the initial 15, 25% in the subsequent 20, and 13% in the final 24 ($P = 0.02$). Fistulas comprised 44%, glans dehiscence 31% and meatal stenosis or urethral stricture 12.5% each. Technical modifications were made to reduce fistulas, including changes in urethroplasty from one-layer continuous epithelial chromic to two-layer subepithelial tubularization using interrupted polyglactin followed by continuous polydioxanone. All patients initially had a dartos barrier flap, to which spongioplasty was added in the second group. In the final patients, spongioplasty and a tunica vaginalis flap provided coverage layers, with dartos no longer used. There were no fistulas in the final 24 patients [8].

Alternative one- and two-stage proximal repair techniques

No series report technical modifications to reduce complications for other surgical techniques in proximal hypospadias repair. All published studies are retrospective and subject to selection bias for patients with varying degrees of hypospadias and the methods used to repair them. Generally, overall urethroplasty complications range from approximately 15% to over 50% for onlay or tubularized prepuce flaps, or staged flaps or grafts [9–15].

Potential risk factors for urethroplasty complications

Age at surgery

There is common belief that urethroplasty complications increase with increasing age at repair, based on

several retrospective studies. For example, an analysis of 316 distal repairs found that complications in the 194 patients undergoing TIP were less frequent in those below 6 months of age, although the same was not true after metal advancement and glanduloplasty (MAGPI) and glans approximation procedure (GAP) repairs [16]. Another report stated patients of mean age 1.6 years had fewer complications after TIP than those with a mean of 5 years of age [17]. Similarly, multivariable analysis in 391 boys after TIP found greater complications in those older than 4 years at repair [18].

We studied age as a potential risk factor using prospectively entered data from 669 consecutive boys undergoing TIP by WS. Mean age was 17 months (SD 22.5, range 3–144 months). Age at surgery was evaluated as a continuous variable as well as <6 months, >1 year, >5 years, by log age and by age quartiles, with none of these categories demonstrating increased risk for urethroplasty complications using multivariate analysis [1].

We next expanded our series to include 65 adults at mean age 21 years. Of these, seven had primary repairs while the others were reoperations. Multiple logistic regression showed no increased rate of urethroplasty complications in these older patients versus those who were prepubertal [19]. In both series, meatal location (proximal>midshaft>distal meatus) and reoperations (reoperation>primary repair) were independent risk factors for complications, not age. Boys who have more severe (proximal) hypospadias may undergo surgery at an older age secondary to other medical conditions more common in these cases or after awaiting natural or preoperative androgen-induced growth for a small penis. Since patients undergoing reoperation are by default older than they were at the time of their primary surgery, analyses that do not adjust for these factors may show a higher rate of complications in older boys simply due to the higher prevalence of proximal hypospadias and reoperations in the older age cohorts.

Sutures

There are limited data regarding outcomes related to sutures used for urethroplasty, glansplasty and skin closures.

We evaluated potential risk factors for glans dehiscence, comparing outcomes in primary TIP operations with glansplasty using 6-0 chromic suture versus subsequent glans repairs using 6-0 polyglactin. This change

was made when it appeared glans dehiscence occurred less often in published series doing glans closures with polyglactin, but multivariable analysis of our patients found no significant differences based on suture type [OR 1.33 (95% CI 0.52–3.01), $P = 0.4$] [6].

A prospective trial allocated 100 patients to distal TIP using polyglytone (rapid absorbing) versus polydioxanone (slow absorbing) in repairs done by a single surgeon. The primary endpoint was fistula development, which occurred in 6% with no difference between the two cohorts [20].

Another prospective study evaluated running versus interrupted suturing technique in 80 boys undergoing repair of distal or midshaft hypospadias using 6-0 or 7-0 polyglactin. The primary endpoint was not specified, but there were no differences noted in fistula, dehiscence and/or meatal stenosis, with five with continuous suture and six with interrupted suture having developed one or more of these complications ($P = 0.5$) [5].

Adjuvant hormonal stimulation

Preoperative hormonal stimulation has been shown to increase glans circumference and penile length. However, only two studies report urethroplasty outcomes following hormonal use, with conflicting results.

A trial randomized 75 consecutive patients mean age 33 months to preoperative 2.5% topical dihydrotestosterone (DHT) applied daily for 3 months versus no stimulation, followed by TIP repair. Hypospadias was coronal in 70% vs. 84%, penile in 24% vs. 16% and penoscrotal in 5% vs. 0. Fewer urethroplasty complications occurred in those with preoperative DHT (1/37 vs. 9/38, $P = 0.01$) [21].

Based on the observation that glans width in normal newborns is a mean of 14mm at median 5 weeks of age, and that glans width in infants with distal hypospadias is 15 mm at median age 9 months, we gave 2 mg/kg testosterone cypionate IM for 2–3 injections to patients with midshaft and proximal hypospadias for glans width <14 mm with the aim to achieve size ≥ 15 mm. When failure to achieve target glans width occurred, the dose of testosterone was increased to 4 mg/kg, and then to 8 mg/kg, etc., with remeasurement 4 weeks later until glans width was ≥ 15 mm. Mean glans width in those treated grew from 12 to 16.5 mm, and urethroplasty complications were compared for these patients versus others with mean glans width of 15.4 mm not receiving stimulation. Despite increasing glans size, complications

remained significantly greater in those receiving androgens, 34% vs. 11%, $P < 0.001$, with multiple logistic regression analysis finding adjuvant therapy an independent risk factor for complications [22].

Preoperative antibiotics

There are no reports concerning outcomes related to preoperative antibiotics in either primary or reoperative hypospadias repair.

Postoperative antibiotics

One trial reported TIP outcomes in 101 patients receiving postoperative oral cephalexin versus no antibiotic during 8 days of stenting. There were no differences in urethroplasty results, but febrile UTI was more common in those without antibiotics (3/52 (6%) vs. 12/49 (23%), $P < 0.05$) [23].

Urinary diversion

One randomized controlled trial (RCT) studied 64 toilet-trained boys median age 6 years undergoing distal TIP randomized at the end of surgery to diversion or not. There was no difference in urethroplasty complications (3/35 vs. 6/29, $P = 0.3$), but those not diverted had significantly increased dysuria, urinary retention and extravasation. Of 29 unstented patients, 12 (41%) had a catheter placed postoperatively [24].

There are few data regarding proximal hypospadias repair without bladder drainage, and no studies examining duration of urinary diversion.

Bandages

Two trials reported no differences in urethroplasty complications in patients receiving a transparent adhesive bandage versus no dressing [25,26]. There are no data supporting use of more complicated bandages.

Prevention and management of urethroplasty complications

Fistulas

Prevention

There is little evidence for the common belief that subepithelial urethroplasty covered by dartos flaps reduces fistulas. WS changed both sutures and suturing methods simultaneously to reduce fistulas in proximal TIP

repairs, and so cannot state which modification(s) resulted in the observed improvement. Since fistula rates as high as 25% occurred despite dartos flaps, he also decided to instead use tunica vaginalis as the neourethra coverage layer in proximal repairs.

One retrospective study of repair by urethral plate tubularization implied there were two similar cohorts, the first operated without a dartos flap and the second using one, with a reduction in fistulas from 22/82 (27%) to 2/57 (3.5%). However, other factors of known importance for urethroplasty complications including meatal location and primary versus reoperative repair, were not described for the two groups [27].

One prospective trial randomized boys undergoing distal TIP with prepucioplasty to have a ventral dartos flap over the neourethra versus no barrier flap. Fistulas occurred in 20/130 (15%), with more in those not covered by a flap (15/65 vs. 5/65, $P < 0.05$) [28].

There are also data indicating benefit to a barrier flap in reoperative TIP. We reported 5/10 (50%) cases done without a barrier flap developed fistulas versus 2/51 (4%) with a dartos or tunica vaginalis flap, $P = 0.008$ [29].

Correction

Generally accepted steps for successful fistula closure include assessment to exclude distal obstruction (meatal stenosis/stricture) or diverticulum, subepithelial urethral closure, and coverage with a barrier layer to avoid overlapping suture lines. Retrospective series report success in 65–96%, especially with fistulas $< 2\text{mm}$ in size [30,31].

The authors use a midline ventral incision to excise the fistula tract, repair the urethra with a one-layer subepithelial closure, and cover the defect with a ventral dartos flap.

Coronal fistulas can either be primarily repaired or require reoperative urethroplasty. The authors dissect under an intact glans to close the fistula and place a dartos flap without urinary diversion. However, when a fistula occurs in combination with partial glans dehiscence leaving the wings held together by only a band of skin, reoperative urethroplasty is done with urinary diversion (Figure 26.1).

Glans dehiscence

Glans dehiscence is complete separation of the glans wings resulting in a subcoronal meatus.

Prevention

We found this complication to occur significantly more often in proximal versus distal hypospadias repair despite glansplasty by the same surgeon using the same sutures and suturing methods. Initially, WS changed from 6-0 chromic to 6-0 polyglactin but analysis showed no difference in glans dehiscence. Then we used preoperative testosterone injection to grow the glans larger in those boys with midshaft and proximal hypospadias and glans width $< 14\text{ mm}$ as described above. However, successful increase in glans size did not decrease dehiscence.

Subsequently, WS observed surgeons in Japan perform glansplasty on small glans $< 14\text{ mm}$ in which the

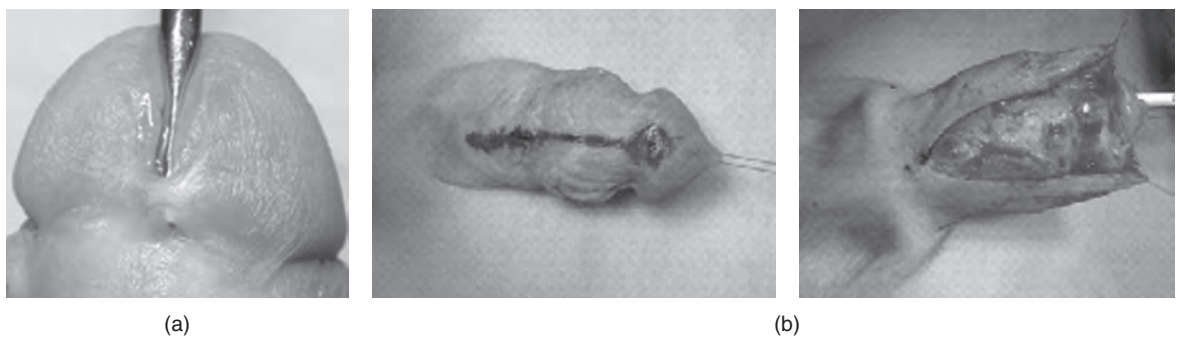


Figure 26.1 Coronal fistula repair. (a) Coronal fistula with partial glans dehiscence, the wings held together by a band of skin. Reoperative urethroplasty and glansplasty is needed to repair the fistula. (b) Coronal fistula with good fusion of glans wings. The incision line circumscribes the fistula and extends down the median raphe. The glans is lifted cephalad and there is excellent exposure to close the fistula and create a ventral dartos flap to cover the repair. Any excessive shaft skin can be trimmed to revise the circumcision with subsequent closure of the median raphe.

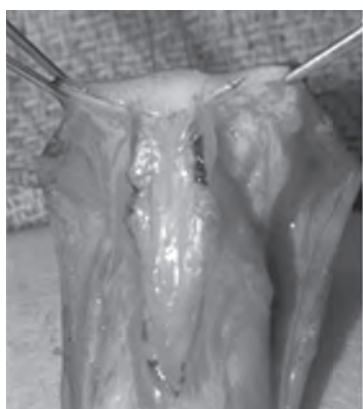
wings were mobilized laterally from the urethral plate, and then also from proximal to distal off the corpora at 3 and 9 o'clock. This additional dissection further reduces tension on the subsequent glans wings approximation, with dehiscence in <1% of proximal cases (Figure 26.2) [33]. We currently use this method for glans <14 mm, when approximation of the wings is under tension, and/or during reoperations for prior glans dehiscence.

Correction

Treatment is reoperative urethroplasty and glansplasty (reoperative TIP), and the authors now routinely use



(a)



(b)

Figure 26.2 Glans wings mobilization. (a) Standard lateral mobilization of the glans wings, used for glans ≥ 15 mm in diameter. (b) Extended glans wings mobilization done for glans <15 mm, or during reoperations for glans dehiscence or when approximation after standard glans wings dissection appears under tension. Additional mobility is obtained by dissecting along the corporal bodies proximally to distally at 3 and 9 o'clock.

the extended glans wings mobilization in cases with prior dehiscence. We do not yet have follow-up data to report outcomes.

Before this modification, we simply repeated TIP repair and reported reoperative glansplasty was successful in 84% (recurrent dehiscence in 18/111 (16%)). Third glansplasty failed in 5/8 (63%) patients undergoing a third glansplasty repair [34].

Meatal stenosis

There is no precise definition of meatal stenosis, and studies reporting its occurrence rarely state how it was diagnosed. This creates a significant impediment to comparing reported outcomes. For example, although meatal stenosis was a primary outcome in the systematic literature review by Wilkinson et al. [35], they stated papers did not define this complication, so they simply reported its occurrence as did the various studies realizing there likely was variation among them in its diagnosis.

We consider a neomeatus <8Fr 6 months after surgery to be stenotic regardless of symptoms, based on a minimum reported normal urethral caliber ≥ 8 Fr in boys 1–10 years of age [36]. Evaluation earlier than 6 months postoperatively may find transient meatal edema.

Prevention

We reported outcomes in 426 patients followed after distal TIP by WS, with 62% having calibration of the neomeatus. None had meatal stenosis by calibration or by obstructive voiding symptoms [3].

Based on those results we believe meatal stenosis most often indicates a technical error, such as failure to deeply incise the urethral plate, or tubularizing the plate too far distally. Plate incision should extend to near the surface of the corpora, and tubularization should end within approximately 3mm of the distal edge of the plate, leaving an oval, not rounded, opening (Figure 26.3).

Correction

Meatotomy is an option to enlarge a stenotic meatus, assuming it can be done without recreating hypospadias. We incise the meatus dorsally, not ventrally, to minimize risk for recurrent hypospadias. Otherwise, stenosis requires reoperative urethroplasty and glansplasty, by reoperative TIP, inlay grafting, or two-stage oral graft repair depending on the extent of distal fibrosis [29].



Figure 26.3 Urethral plate tubularization. Note tubularization of the urethral plate ends approximately 3 mm below the junction of the plate with the glans, leaving an oval shape. This is a key maneuver to prevent meatal stenosis.

We found no report concerning meatotomy outcomes for meatal stenosis after hypospadias repair.

Stenosis developing more than 6–12 months after repair can also indicate balanitis xerotica obliterans (BXO), typically diagnosed by the white discoloration of tissues around the meatus. Correction requires complete excision of all involved tissues and replacement with oral mucosa using two-stage urethroplasty.

Urethral stricture

Neourethral stricture is an uncommon complication after hypospadias repair regardless of technique used. Most strictures are diagnosed because of symptoms, including stranguria, retention, or UTI.

Prevention

Strictures result from focal devascularization, emphasizing the need for careful tissue handling, especially in pedicle flaps. Proximal circumferential anastomoses joining tubularized prepuccial flaps to the native urethra may also stricture, and so wide, oblique junctions should be made.

Dissection of the urethral plate and proximal native urethra off the corpora to straighten ventral curvature followed by TIP repair resulted in 17% strictures, versus none in proximal TIP without this native urethral mobilization (5/29 (17%) vs. 0/47, $P = 0.01$). All were symptomatic within ≤ 1.5 years after repair, and ranged from 1 mm to 1 cm in length. Accordingly, we no longer recommend this maneuver combined with TIP urethroplasty [37].

Correction

One study using a prospective treatment protocol reported direct vision internal urethrotomy (DVIU) for hypospadias strictures < 1 cm was successful in only 2/18 tubularized flaps and 0/32 tubularized grafts, versus 8/11 onlay flaps and 7/11 urethral plate tubularizations, $P < 0.05$. Dilation after DVIU was of no benefit, and secondary DVIUs all failed. Therefore, a single DVIU is an option for hypospadias strictures < 1 cm after onlay flap or plate tubularization, but failure should lead to open urethroplasty [38].

Dorsal inlay grafting using oral mucosa was done in 37 hypospadias strictures with mean length 5 ± 2 cm. Recurrent stricture developed in three (8%) during mean follow-up of 23 ± 10 months [39].

Strictures leaving only a narrow strip of epithelium ($\leq$ approximately 4 mm) when the urethra is opened are corrected with excision of the fibrotic neourethra and two-stage oral graft urethroplasty [29].

Diverticulum

Diverticula are diagnosed from ballooning of the neourethra during voiding, typically with postvoid dribbling. Distal obstruction is not needed, as the tissues used in skin flap urethroplasties can stretch without anatomic blockage. Two reports indicated diverticulum developed in 7% and 11% after onlay and tubularized prepuccial flaps [40,41]. WS encountered diverticula in 6/10 (60%) Byar's flaps repairs with none having distal obstruction, leading him to abandon that procedure (unpublished data).

Prevention

Assuming experienced surgeons do not create skin flap neourethras that are too large, and given that diverticulum can form without distal obstruction, prevention may require use of alternative repair techniques such as TIP or two-stage prepuccial grafts.

Correction

Diverticulum repair involves assessment for distal obstruction, and then excision of excessive neourethra with retubularization and coverage with a dartos or tunica vaginalis flap. Vallasciani et al. [40] reported no recurrences during follow-up after an average of 9 years, while Snyder et al. [41] reported 1/13 (8%) developed a new diverticulum within mean follow-up of 24 months.

Recurrent ventral curvature

Approximately 10% of patients with distal hypospadias have ventral curvature that is nearly always $<30^\circ$ [3]. Approximately 50% of proximal hypospadias have curvature $>30^\circ$ after shaft skin degloving and excision of ventral dartos [42].

No studies report outcomes of single midline plication for curvature $<30^\circ$.

Curvature $>30^\circ$ has been corrected using multiple dorsal plications versus urethral plate excision and ventral corporotomy. One retrospective review of these options in which decision making was by surgeon preference without a standard protocol found recurrent curvature more common after plication during a mean of 2 years follow-up. However, if only those patients with recurrent bending thought sufficient to warrant additional surgery are considered, there was no difference in straightening by plication versus ventral lengthening (11/68 (16%) vs. 1/32 (3%), $P = 0.09$) [43].

One study reported outcomes in 16 patients during puberty after prepubertal ventral corporotomy with grafting. Of these, 13 reported straight erection, whilst two had persistent ventral curvature described as “mild” and not requiring additional surgery. There were three who reported sexual activity, of which one had poor-quality erection needing intracorporeal vasoactive drug injection [44].

Published retrospective series suggest ventral corporotomy with grafting has similar short-term outcomes whether dermal grafts, small intestinal submucosa (SIS) or tunica vaginalis grafts are used [45–47].

One series used three transverse ventral corporotomies without grafting for straightening ventral curvature so that two-stage graft urethroplasty could be done without concern for placing the urethroplasty graft onto a corporotomy graft. There was no recurrent ventral curvature in seven with corporotomy and dermal grafting or 11 having three transverse corporotomies without grafting [42].

Prevention

As discussed, there are no long-term outcome data regarding correction of ventral curvature $<30^\circ$ by dorsal plication.

Best treatment for curvature $>30^\circ$ is not clear. Because we use two-stage graft urethroplasty when the urethral plate cannot be preserved, we straighten

persistent bending $>30^\circ$ by three ventral transverse corporotomies without grafts.

Correction

Recurrent ventral curvature can result from contracture of ventral skin, neourethra, or ventral corporotomies, or from failure of dorsal plication. Curvature $<30^\circ$ previously straightened by dorsal plication usually can be corrected by repeat plication with dorsal corporal incision to ensure suture placement into the tunica albuginea. We use polypropylene suture. Curvature $\geq 30^\circ$ most often corrects by release of ventral shaft skin and/or excision of the neourethra. Failure of prior ventral corporotomies with or without grafts is uncommon.

Duration of follow-up

We reported urethroplasty complications occurring in 125/887 (14%) consecutive primary and reoperative TIP repairs. Of these, 34% were fistulas, 47% glans dehiscences, 7% meatal stenoses or neourethral strictures, and 2% diverticulum. Sixty-four percent were diagnosed at the first postoperative visit, and 80% within the first year after repair. Median time for diagnosis was 6 months for fistulas, obstructions and diverticulum, versus 2 months for glans dehiscences. After one year, 35 patients would need indefinite follow-up to diagnose one more urethroplasty complication [48].

Given the greater risk for complications after proximal versus distal urethroplasty, we recommend follow-up to approximately a year following distal TIP but schedule annual assessments after proximal TIP and two-stage repairs.

Cosmetic results

Although hypospadias surgical outcomes are largely expressed in terms of the urethroplasty complications discussed above, penile appearance is also important but few objective assessments have been published.

One study used a non-validated questionnaire given to parents after distal and proximal TIP, with responses compared to those of parents whose sons had elective circumcision. Consecutive boys returning for follow-up at 6 weeks were enrolled. There were no differences in scores rating either overall appearance or the specific

appearance of the meatus and penile skin between parents of normal boys versus hypospadias patients [49].

Two studies compared TIP versus Mathieu outcomes using photographs reviewed by blinded observers. Both reported TIP had superior cosmetic appearance [50,51].

We are not aware of similar cosmetic evaluations for other contemporary repair techniques, except for one report using photographs to compare outcomes of standard versus a modified proximal onlay flap repair designed to create a more slit-like meatus. A slit meatus was judged achieved in 20/55 (36%); however, only patients who already had a deeply grooved plate reliably had the desired meatal outcome. There was no difference in meatal appearance in patients with a flat or

cleft urethral plate who had the standard versus modified onlay repair [52].

A systematic review of adults who underwent hypospadias repair in childhood using techniques not often employed today (Denis-Browne, Ombredanne,) reported that about one-third of hypospadias patients reported an abnormal appearance of their penis compared to peers their same age. So did about one-third of non-hypospadias controls, although characteristics of the control groups were not specified. Patients with hypospadias were more likely to report dissatisfaction with penile size, and those with “severe” hypospadias were more likely to report dissatisfaction with penile appearance in general compared to controls [53].

DOS AND DON'TS

Do

- Incise the urethral plate deeply to near the corpora during TIP repair to achieve a sufficient caliber neourethra
- Perform two-layer subepithelial urethroplasty
- Cover the neourethra with a tissue flap to reduce fistulas
- Perform individual quality review of hypospadias outcomes and institute technical changes as needed to reduce complications
- Realize higher complications following proximal hypospadias repair and reoperations support subspecialization for these cases

Don't

- Tubularize the urethral plate too far distally during TIP repair to avoid meatal stenosis

CLINICAL PROBLEM/CASE: DISCUSSION

Factors that determine whether to perform a single-stage or two-stage repair depend on the nature of the urethral plate, size of the glans, and presence of chordee. In the case demonstrated, the urethral plate is very dysplastic with scrotal encroachment on the penile shaft and the meatus at the penoscrotal junction. There was significant chordee.

The glans size is less than 10 mm. A decision was made to undertake radical degloving of the penis, division of the atretic urethral plate, and excision of all chordee tissue to straighten the penis. The glans was splayed and a wide 15 mm × 20 mm inner prepuce free graft was anchored in place. This was tubularized 6 months later.

The determinants of a successful outcome are glans size >14 mm, subepithelial two-layer urethroplasty, and the use of barrier flaps. The type of dressing, use of postoperative antibiotics, or urinary diversion did not influence the success of the repair.

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CHAPTER 27

Phalloplasty for the biological male

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KEY POINTS

- Congenital absence or loss of the penis is a devastating condition
- Gender reassignment is no longer an option for this condition
- Phalloplasty is the gold standard treatment for this condition
- Maximal conservation of any penile tissue and incorporation in the phallus must be considered
- Phalloplasty must be performed by experienced surgeons
- Complication rate of phalloplasty is high
- Erectile implants after phalloplasty are feasible

CLINICAL PROBLEM/CASE

A 12-year-old prepubertal boy presents after primary exstrophy closure which was complicated by partial loss of the glans penis and an insufficient penile length. The parents are very concerned about how his penis looks and the boy himself worries a lot about his penile length. They

are looking to you for help and ask for penile construction with phalloplasty as they have read about this on the internet. On clinical examination, the penis has a hemiglanis is seen with a stretched penile length of 4 cm. There is a lot of scar tissue and the penis looks unesthetic.

- 1 What will be your advice to the parents and the boy?
- 2 How will you follow this boy?

Introduction

The biological male without a penis or with an insufficient penis remains a major challenge. Failure of penile development, trauma, medically-indicated penile amputations, and failed reconstructions of congenital anomalies are the main reasons for penile insufficiency (Box 27.1). Severe penile insufficiency and absence of a penis are devastating conditions for men, with significant psychological and physical impact. Although uncommon, it is a challenging condition to treat.

Possible treatment options are gender reassignment, tailoring of the penile stump, penile reattachment,

Box 27.1 Conditions leading to severe penile insufficiency.

Congenital conditions (disorders of sexual development):

- Aphallia or penile agenesis
- Ideopathic micropenis
- 46 XY disorders of sex development
- Exstrophy
- Cloacal exstrophy

Genital trauma:

- Injuries
- Surgery
- Penile amputation

Box 27.2 Treatment options for severe penile inadequacy.

- Endocrinological treatment
- Penile reconstruction
- Penile replacement (phalloplasty)
- Gender reassignment
- “Penile transplantation”
- “Tissue engineering”

phallic reconstruction (phalloplasty), and most recently penile transplantation (Box 27.2). In the past, sex reassignment to the female gender had been offered based on the principles applied to newborns with disorders of sexual differentiation and ambiguous genitalia. There is no evidence to demonstrate that the outcome of this policy is satisfactory. Indeed, long-term evaluation of a few patients shows contradictory results, which have triggered great controversy of this therapy [1]. The issue of gender reassignment is beyond the scope of this chapter. However, some recent reports have alerted physicians to the high incidence of gender identity disorder in gender reassigned children. Especially in cloacal exstrophy, the results have been disappointing [2,3]. Today, sex reassignment is no longer considered treatment of choice. Parents of children presenting with these conditions should be informed that a phalloplasty can be performed later in life.

Tailoring of the penile stump by means of penile degloving, division of the suspensory ligament, and rotational skin flaps has been reported [4,5]. However, this can only be applied to moderate penile injuries where there is still a reasonable penile stump.

When penile skin is lacking for reconstruction a reversed mesh graft can be used to cover the penile shaft. Widely meshed split-thickness skin grafts should never be used on the penile shaft. However, with the so-called reversed mesh graft (i.e. nonexpanded or 1:1 meshed skin grafts) only puncture-type perforations are made with a V1 carrier (Humeca®, Enschede, the Netherlands; Figure 27.1). These allow for a better fluid evacuation and thus a better skin graft take whilst still avoiding poor scarring and an unesthetic mesh pattern in the longterm. These non-expanded meshed split-thickness grafts have also

become a mainstay of complicated, staged urethral reconstruction [6,7]. McAninch [8] has described the use of (not too widely) meshed split-thickness skin grafts for scrotal reconstruction. Meshed grafts in this location may produce good esthetic results, but they will never measure up to the functional capacity of sheet or 1:1 perforated grafts.

In some cases good results can be obtained by combining techniques (Figure 27.2).

Penile reattachment can be attempted in the acute phase following traumatic amputation of the penis. The survival of the reattached penis depends on the viability of the amputated segment and the condition of the graft bed or penile stump. Reattachment must take place within 24 hours. Current reattachment techniques rely on microsurgical approximation of the dorsal structures and cavernosal arteries with uniformly good results. In traumatic amputation, salvage of the amputated segment with reattachment is the primary treatment option [9]. The outcome of erectile function after reattachment is, however, not clear.

The ideal candidate for genital replantation is a patient with a clean, sharp cut in which the amputated part has been cooled (Figure 27.3).

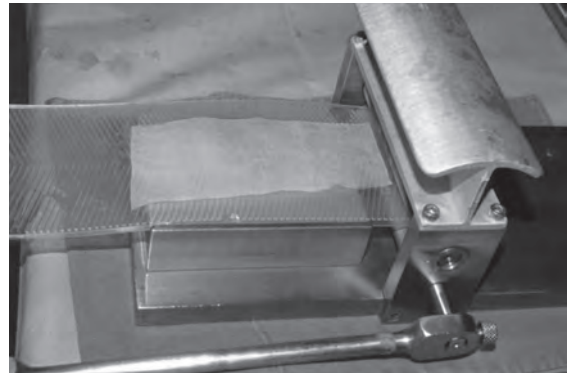
Recently, a single unsuccessful case report has been published on penile transplantation [10]. This technique is still experimental and is not a current treatment option. Future options, like tissue engineering, are as yet matters for science fiction rather than science.

Phallic reconstruction is another treatment option. The first phallic reconstruction was described by Bogoras in 1936 with a tubed abdominal flap [11]. Phalloplasty procedures have followed the evolution and advances made in plastic surgery. Originally, it was a complex, time-consuming, multistage procedure using tubed skin flaps or pedicled myocutaneous flaps with variable and suboptimal results [12–14].

In 1984, Chang completed the first successful microsurgical phalloplasty with a radial forearm free flap [15]. Since then, the radial forearm flap has been widely accepted as the best donor site for penile reconstruction and is nowadays the gold standard in penile replacement for female-to-male transsexuals [16–18]. This technique can also be applied to boys without an adequate penis. Defining penile insufficiency is difficult. To base the definition of inadequacy on length and appearance alone is impossible, especially in infants and young



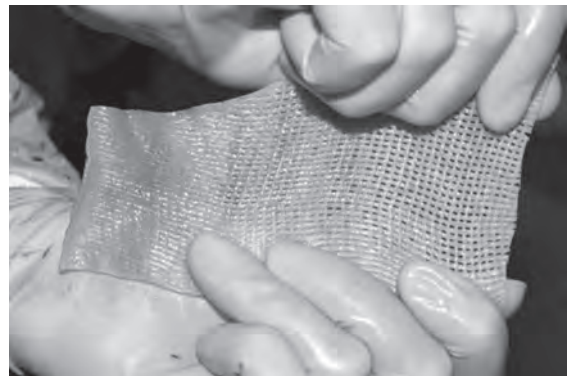
(a)



(b)



(c)



(d)

Figure 27.1 The so-called “reversed-meshed graft” or 1:1 perforated skin graft, obtained with a V1 mesh carrier (Humeca®, the Netherlands). (a) The 1:1 carrier which allows for perforation of the graft. (b, c) Carrier with a split-thickness skin graft in the skin graft mesher (Humeca). (d) The nonexpanded-only puncture-type perforated skin graft.

children. Penile inadequacy is an individual diagnosis that can only be made after puberty when sexual development is completed and the patient is sexually active. Puberty can change the final outcome of penile length and girth substantially. The number of children diagnosed with micropenis persisting into adulthood is limited [19].

As for penile reconstruction, many techniques have been described for penile augmentation. However, the results of most of these surgeries are very limited. Indeed, the reported outcome is often poor [20].

In this chapter, we will focus on the phalloplasty. It is considered the gold standard for penile absence or severe penile inadequacy, when endocrinological therapy is not beneficial.

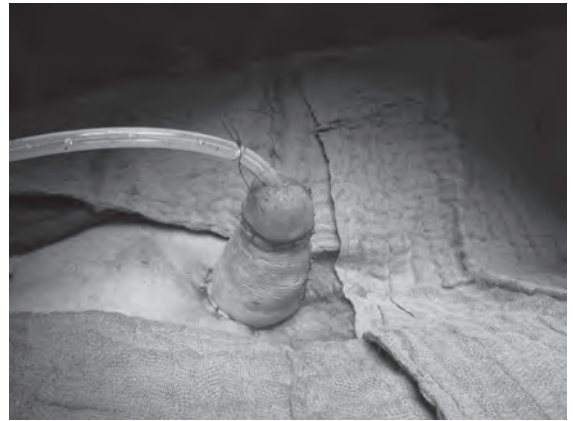
Phalloplasty

Surgical reconstruction of the penis (phalloplasty) is difficult because of the different cosmetic and functional requirements of the patients:

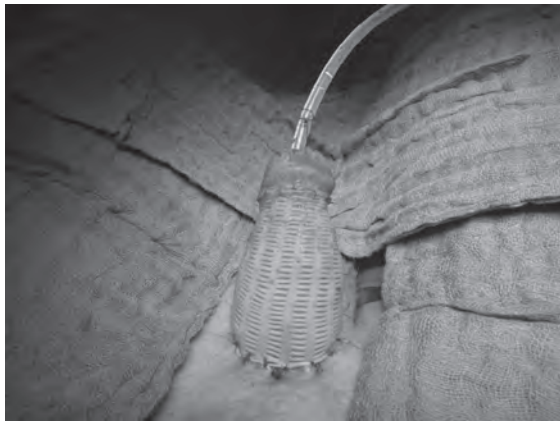
- 1 The reconstructed penis should be esthetically acceptable; it must be as normal as possible in appearance (minimal scar, glandular reconstruction, etc.).
- 2 The penile shaft must contain a urethra that extends to the distal tip and permits the patient to void in a standing position unless there is a concomitant condition that makes normal voiding impossible.
- 3 The penile shaft should allow the implantation of a penile stiffener in order to regain the possibility of



(a)



(b)



(c)

Figure 27.2 Example of a cripple penis where the stump was released and covered with reversed mesh graft. (a) Cripple exstrophy penis after multiple surgeries. (b) Lateral view after release and coverage with reversed mesh graft. (c) Dorsal view after release and coverage with reversed mesh graft.

sexual intercourse. Therefore, protective and erogenous sensation is needed.

- 4 The donor area should cause minimal morbidity with an acceptable scar that is easy to conceal.

Many of these objectives could not be obtained with the older methods in which phallic reconstruction required a complex multistaged procedure. Nowadays, microvascular free flap techniques come closer to achieving these objectives. Despite the multitude of free flaps that have been published (frequently as a case report), the radial forearm free flap is universally considered the gold standard in penile reconstruction [16–18,21].

Surgical technique

At Ghent University Hospital, more than 500 consecutive patients have undergone phallic reconstruction

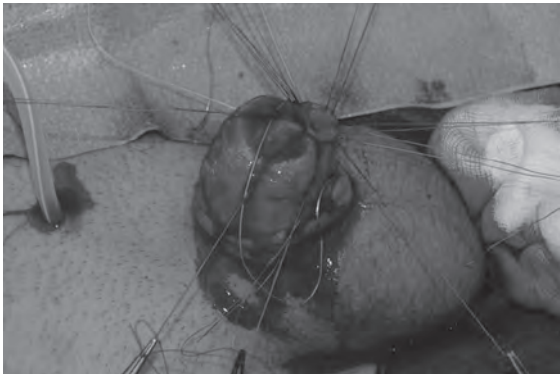
using a radial forearm flap. This experience is mainly in female-to-male transsexuals. We describe our current technique as well as the different changes and refinements made in the reconstruction for the biological male in this radial forearm phalloplasty procedure.

Depending on the underlying condition, we try to preserve any useful penile and cavernosal tissue. The urethral stump, if available, is prepared for connection with the phallic urethra and, also if available, a dorsal penile nerve is identified. A free vascularized flap of the forearm and the creation of a phallus with a tube-in-a-tube technique is performed with the flap still attached to the forearm by its vascular pedicle. A small skin flap and skin graft are used to create a corona and imitate the glans of the penis (Figure 27.4).

The free flap is then transferred to the pubic area where first the urethral anastomosis is performed.



(a)



(b)



(c)



(d)

Figure 27.3 (a) Self-inflicted penile amputation. (b) Preparation of microsurgical anastomoses. (c) Immediate postoperative results. (d) Late postoperative results.



Figure 27.4 End result with glandular reconstruction and tattooing of the glans.

The radial artery is then microsurgically connected to the common femoral artery in an end-to-side fashion, usually with an interpositional vein graft taken at the ankle. The venous anastomosis is performed between the cephalic vein and the greater saphenous vein. One forearm nerve is connected to the ilioinguinal nerve for protective sensation, and the other nerve is anastomosed to one of the dorsal penile nerves for erogenous sensation. All patients receive a suprapubic urinary diversion postoperatively.

In the first 50 patients the defect on the forearm was covered with full-thickness skin grafts taken from the groin area; later, patients were given split-thickness skin grafts harvested from the medial and anterior thigh.

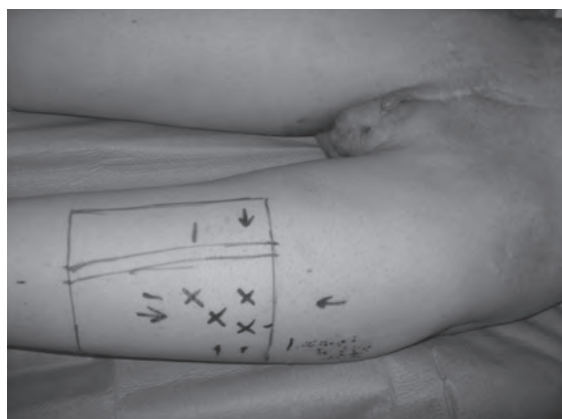
The patients remain in bed for one week after which the transurethral catheter is removed. Three to five days later the suprapubic catheter is clamped and voiding is started. It sometimes takes several days before good voiding is observed. The average admission period for the phalloplasty procedure is approximately 2.5 weeks.

Tattooing of the glans can be performed after a 2–3 month period, before sensation returns to the penis (Figure 27.4).

The development of perforator flaps has given rise to some new reconstructive options in patients with severe (congenital) penile insufficiency. Perforator flaps have the advantage of reducing the donor site morbidity, increasing the range of motion and combining different tissue flaps on one single pedicle. Although – overall – the free vascularized radial forearm flap described above is still considered as the “standard technique” in penile reconstruction, the pedicled antero-lateral thigh (ALT) flap has been shown to provide a valuable phalloplasty alternative specifically in patients with congenital penile insufficiency. This flap is a skin flap based on a perforator from the descending branch of the lateral circumflex femoral artery, which is a branch from the femoral artery. There are several reasons why in boys without a penis a pedicled ALT flap can be preferred above the standard radial forearm flap (Figure 27.5):

- A pedicled flap reconstruction (the flap has a sufficiently long pedicle) avoids the technically more complex microscopic procedure and might also shorten the operation time.
- A visible donor site scar on the forearm, often considered as the signature of female-to-male transsexualism, is avoided and the donor site on the leg can more easily be concealed.
- Previous reconstructive surgeries at the pelvis, groin area and lower abdomen (e.g. in case of bladder exstrophy) might have altered the local anatomy and vasculature making a microsurgical anastomosis more difficult.
- The subcutaneous fat layer is much thinner than in a (biologically female) trans-man, facilitating the (urethral) tube-within-a-(penile)tube reconstruction of the penis; moreover, many exstrophy patients empty their bladder by catheterization through a continent diversion (e.g. appendico-vesicostomy) and don’t require a urethral reconstruction in their phalloplasty. An ejaculatory opening can be left at the ventral aspect just above the scrotum.

It is very important to preserve and incorporate any useful glandular, penile and cavernosal tissue at the basis of the newly-reconstructed phallus in order to facilitate sexual stimulation and pleasure (Figure 27.5d). If available, a dorsal penile nerve is identified and connected with a cutaneous nerve of the flap; if not available, the lateral femoral cutaneous nerve is connected to the ilioinguinal nerve.



(a)



(b)



(c)



(d)

Figure 27.5 The reconstruction of a penis with a pedicled ALT flap used as a tube-within-a-tube technique (only possible in thin “boys” without a penis and after defatting). No real urethra was reconstructed here since the patient had a urostomy. (a) Preoperative view. (b) After flap dissection. (c) The flap is tunneled underneath the rectus femoris muscle. (d) Suturing and nerve connection (ilioinguinal nerve to lateral femoral cutaneous nerve).

Usually, it is recommended to perform a 3D angio CT scan preoperatively to provide detailed information on the perforator vessel(s) and the subcutaneous tissue layer.

Sexual function

Sexual function and pleasure is one of the goals in phallic reconstruction. For this purpose, in biological males any sensitive penile tissue left must be incorporated. Any glans tissue present can be incorporated in the base of the neophallus. This is important for sex-

ual stimulation and pleasure. Further erogenous and tactile sensations of the neophallus are obtained by microscopic anastomosis of, respectively, one dorsal phallic nerve and one ilioinguinal nerve to the cutaneous nerves of the flap [22].

Obtaining sufficient rigidity of the penis to allow penetration is extremely difficult because there is no good substitute for the unique erectile tissue of the penis. The radial forearm flap is too soft and can even demonstrate some atrophy of the subcutaneous fat with a loss of more than 20% of circumference. The use of bone or cartilage grafts has often resulted in complications and failure because of resorption,

curving, or fracture [23,24]. For sexual penetration, a penile stiffener is needed and, fortunately, the radial forearm flap has sufficient subcutaneous bulk to permit incorporation of a penile prosthesis. Incorporation of a penile stiffener can only be done after the phallus is endowed with sufficient protective sensation, which usually takes at least 12 months. Good protective sensation is critical in preventing breakdown and erosion of an internal stiffener [25,26]. Next to sensitivity, urethral function also has to be considered. Implantation of a penile prosthesis must be withheld until the urethra is stable, and the patient is free of voiding symptoms and urinary tract infection [25].

Unfortunately, high erosion rates (20–50%) are reported [16,25]. One of the reasons could be the less vascularized skin and subcutaneous tissue of the neophallus (in comparison with a native impotent penis), which can lead to chronic ischemia after implantation of a stiffener and subsequently diminished resistance against infection and perforation.

Despite the complications and difficulties, the satisfaction rate after phalloplasty is high and the results cosmetically pleasing (Figure 27.6). None of our patients regret the surgery. An important boost concerning the self-esteem level is observed in each

patient, which is a very important outcome factor postoperatively.

Complications

Despite the good outcomes described, phalloplasty is associated with a high complication rate. Most complications are related to the urethral reconstruction [16,27,28]. The main complications are fistulae and stenoses whenever the urinary tract is attached to the native urethra. Despite the high prevalence of these complications, the literature on treatment of these complications is sparse. Consequently, always consider whether the urethral reconstruction is necessary, as some boys will not need urethral reconstruction as the bladder is augmented and diverted. In addition, it is important to consider the ejaculation of the patient.

Is he actually ejaculating? Where? If so, do we want to reconstruct the urethra for ejaculation alone, or do we want to keep the ejaculation where it is? Preference should be given to keep the ejaculation where it is as reconstructing a urethra just for ejaculation can result in higher complications and possible anejaculation due to the length of the urethra and the weakness of the ejaculation due to the underlying condition.

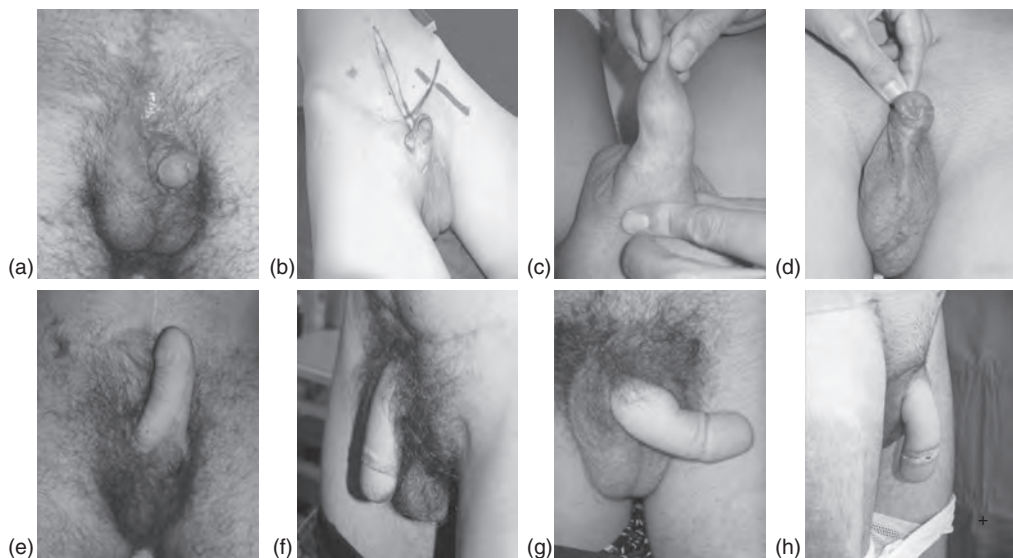


Figure 27.6 Results of phalloplasties in biological males. (a, b) Crinkle exstrophy. (c) Epitheloid sarcoma of penis. (d) Crinkle hypospadias with absence of corpora. (a–d) Before surgery; (e–h) after surgery.

Urethral fistulas

Remove all scar tissue and try to bring well-vascularized tissue to the area that needs reconstruction. In radial forearm flap phalloplasty, we have to consider that the tissue of the urethra is skin and not mucosa. Larger stitches with cutting needles should be used. The duration of bladder drainage is unknown, but in our practice we prefer to drain for 12 days. When using local skin flaps, consider possible future hair growth in the urethra. A good alternative is the use of buccal mucosa [29].

Urethral stenosis

In our experience we first attempt an endoscopic incision of the stenosis, if the stricture is relatively short. We leave a catheter for 12 days, which is much longer than after urethrotomy for urethral stenosis in normal urethras. We have to remember that part of the urethra is composed of skin, and healing of skin lesions is much slower than mucosal healing.

If endoscopic incision fails, we perform a formal urethroplasty. End-to-end anastomosis or Heineke–Mikulicz type reconstructions with longitudinal incision over the stenotic area closed transversally. For longer strictures or complex and repetitive stricture, two-stage urethroplasty (Johansson) must be considered. The stenotic area is longitudinally incised and the borders of the stricture are sutured to the surrounding healthy skin. The urethra remains opened until the skin and urethra are well healed, which usually takes a minimum of three months. During the second stage of the urethroplasty, the urethra is closed again and covered with skin.

Experience with phalloplasty in biological males

There are only a few series published on this topic. Perovic reported phalloplasty in 24 patients without a functional penis using the extended pedicle island groin flap. He suggests this technique as an available alternative to the microsurgical free tissue phalloplasty [30].

Sengezer et al. suggested total penile reconstruction with sensate osteocutaneous free fibula flap. With this technique, promising results were obtained in

18 patients without a functional penis for different reasons [31]. Gilbert et al. were the first to describe the application of a radial free forearm flap for phallic reconstruction in 11 boys without a functional penis. Satisfactory results were obtained [32].

We performed phalloplasty in 19 males with the use of the radial forearm flap: eight boys with inadequate penis after exstrophy repair; one boy with penile loss after multiple hypospadias repair; six boys with disorders of sexual development (DSD) and micropenis; one boy with a penile epithelioid sarcoma; one boy with idiopathic micropenis; and two men who traumatically lost their penis. There were no complications concerning the flap. Two complications were reported in the early postoperative period: one pulmonary embolism and one severe hematuria. Four patients developed urinary complications (stricture and/or fistula) for which a secondary procedure was necessary.

In a further five boys – two with cripple exstrophy and three after cloacalexstrophy – phalloplasty was done with an ALT flap.

Patient satisfaction after surgery was high in 22 cases and moderate in two cases. Psychological evaluation confirms this, especially on the self-esteem level. Ten patients underwent erectile implant surgery. In two patients, the erectile implant had to be removed because of infection (unpublished data).

In our series, one adolescent patient presented with epithelioid sarcoma of the penis and the phalloplasty was performed in a one-stage procedure with the penectomy [33]. This could be an option for some patients undergoing penile amputation if oncologically acceptable.

The reported success of phalloplasty in boys without a functional penis has convinced us that penile reconstruction is the optimal treatment for this condition. It has extremely good results and improves their self-esteem and their physical and psychological wellbeing. But the complication rate of the erectile implants is high.

Phalloplasty opens new horizons for the treatment of penile agenesis, micropenis, crippled penis, shrivelled penis, some DSD conditions, traumatic amputations in which the amputated segment is lost for replantation, iatrogenic amputations, and cloacal exstrophy.

DOS AND DON'TS**Do**

- Send boys with genital inadequacy to an experienced team
- Work together with an experienced plastic surgeon when using free flaps
- Give your patients a realistic perspective on what phalloplasty is. Wait until after puberty before offering phalloplasty to your patients

Don't

- Reconstruct complex congenital urogenital anomalies when you do not have the experience. When confronted with a cripple penis do not attempt surgery if you have no knowledge of all available reconstructive techniques

CLINICAL PROBLEM/CASE: DISCUSSION

Prepubertal phalloplasty is controversial and should be discouraged. There are issues with growth at puberty. The penis will grow with puberty and phalloplasty can only be offered to boys with a really insufficient penis. Supposing the penis develops with puberty to a length of 9cm, then phalloplasty is no longer an option but esthetic reconstruction should be the first choice treatment.

Furthermore, waiting until after puberty allows the boy to explore for sensitive sexual tissue. Supposing that he would be a good candidate for phalloplasty after puberty, it is likely to preserve any erotic sensitive tissue and incorporate it in the neophallus. Another issue is ejaculation. Before performing a phalloplasty the information on if, how and where the patient ejaculates is important as it is an issue in the reconstruction.

Finally, performing the operation later leaves the decision with the patient and he can give his own informed consent on the procedure.

Until the final decision, a good follow-up with an experienced child psychologist is necessary.

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CHAPTER 28

Complications of female genital surgery and disorders of sex development I

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KEY POINTS

- The success of genital reconstructive surgery in sexual function and reproduction may not be tested until many years after the initial surgical procedure
- Long-term complications of genital reconstructive surgery are common and the most frequent complication is vaginal stenosis
- Assessment of the long-term complications of genital surgery must include sexual function evaluation
- The vagina has no role in the prepubertal girl and vaginal surgery can safely be deferred until puberty if a uterus is present and later still if a uterus is not present
- Revision surgery for vaginal stenosis is necessary in the majority of those undergoing vaginoplasty in early childhood
- Clitoral surgery damages sexual sensation and has a negative impact on sexual function

CLINICAL PROBLEM/CASE

Case 28.1

A 15-year-old with congenital adrenal hyperplasia (CAH) presents to the gynecologist because she is unable to use tampons during her periods. The girl had a feminizing genitoplasty when she was 18 months, during which a flap vaginoplasty and a clitoral reduction were performed. Subsequent follow-up was satisfactory.

- 1 What is the likely reason for her difficulty?
- 2 How can this complication be avoided?

Case 28.2

A 17-year-old patient with a diagnosis of Mayer–Rokitansky–Kuster–Hauser (MRKH) syndrome is presenting 2 years following an intestinal vaginoplasty with a persistent vaginal discharge. The discharge is copious, malodorous and slightly blood tinged.

- 1 What is the likely cause of the patient's symptoms?
- 2 Are there any other risks of intestinal vaginoplasties?
- 3 Were there any alternatives to the initial management of this patient?

Introduction

Disorders of sex development (DSD) include a proportion of patients that are born with ambiguous genitalia or an absent vagina and will need some form of genital reconstruction to align the appearance of their genitalia

with their sex of rearing and enable future sexual function. Until recently, there was an uncontested practice to perform genital operations in infancy. However, a series of publications from the late 1990s onwards helped to identify the long-term complications of such procedures, which were mostly obvious in adolescence and adulthood.

Types of surgery

Reconstructive genital surgery is most commonly performed in the following two situations:

- To create a neovagina in conditions where the genitalia are female and the uterus and vagina are absent, i.e. Rokitansky syndrome or complete androgen insensitivity syndrome (CAIS).
- To feminize the genitalia when a child with ambiguous genitalia is assigned to a female sex of rearing. A uterus may be present such as in congenital adrenal hyperplasia (CAH), or may be absent as in 46XY DSD.

Timing of vaginoplasty

The vagina has no function in the prepubertal girl. If a uterus is present, then a passageway for menstruation is required at puberty. If the uterus is absent, then the vagina is not required until sexual activity is planned. If presentation is at adolescence with primary amenorrhea, then the timing of any vaginal reconstruction is uncontroversial. The procedure can be discussed with the patient and planned to fit in with need for intercourse and academic and/or work commitments. Nonsurgical alternative treatments such as vaginal dilation can be offered and may make surgery unnecessary.

However, if the diagnosis of vaginal agenesis is made at birth or during childhood, the situation is more controversial. In the past, creation of a neovagina was often recommended during early childhood. Progressive passive vaginal dilation is not appropriate for children, which left surgery as the only option. The need for repeat reconstructive surgery at adolescence has also been demonstrated whatever method of vaginal construction is used [1].

In the neonate with associated complex urinary and gastrointestinal anomalies, vaginal reconstruction is usually performed at the primary reconstructive procedure. The rationale for this is that there may be only one opportunity to access the pelvis, as surgery will be highly complex and re-entry to the abdomen at a later stage hazardous. Although this sounds logical, the few studies on this group of patients have shown that, despite early vaginoplasty, obstructed menstruation is common and repeat vaginoplasty often required [2]. Consideration should be given to following the same

principles in this group of girls and deferring surgery until puberty in the presence of a uterus or until sexual activity if the uterus is absent.

Assessment of outcome

The long-term assessment of vaginoplasty outcomes can be difficult. Patients' satisfaction will be influenced not only by the procedure but also by the clinical and psychological implications of their underlying condition as well as other treatments such as hormonal or steroid therapy. Other factors such as infertility will in most cases cause additional distress.

When assessing outcomes of pediatric vaginal reconstruction it is important to remember that the major aim of surgery is to allow the adult patient to have comfortable and pleasurable sexual intercourse. Anatomical success as quantified by a surgeon does not guarantee success in sexual function. Motivation for reconstruction is often based on aspirations for normality not just in sexual anatomy but behavior and experiences [3]. Female sexual function and dysfunction is based not only on biological but on psychosocial components as well. Long-term assessment of vaginal reconstruction must include such information; otherwise the whole point of surgery is missed and the data meaningless. Other outcome measures include a passageway for menstruation and tampon use. Although these outcomes should be easier to assess, there is still little data. Some authors have graded the outcome as "excellent" if the vagina was thought to be suitable for intercourse and "satisfactory" if the vagina permitted menstrual flow but did not allow intercourse [4]. This may appear logical but women may not perceive being unable to have penetrative intercourse as a satisfactory outcome of vaginoplasty.

Risks and complications of vaginoplasty

Intestinal vaginoplasty

Intestinal vaginoplasty lines the neovagina with a segment of bowel keeping the vascular pedicle intact. The procedure is usually performed via a laparotomy although a laparoscopic and robotic approach have been reported [5,6]. The main advantage of this procedure

is the low risk of vaginal stenosis and the avoidance of postoperative vaginal dilatation. Dilation is occasionally required at the perineal anastomosis but the vaginal canal itself should retain its original size. Other advantages include adequate vaginal length and natural lubrication.

Satisfactory sexual function outcomes have been reported in women with intestinal vaginoplasty [7]. However, these results should be balanced against the risks of major surgery with high associated morbidity and persistent symptoms, such as vaginal discharge, diversion colitis, the risk of neovaginal prolapse. In many cases – especially in the absence of prior genital surgery – women will achieve satisfactory sexual function with less invasive techniques such as dilation. Intestinal vaginoplasty should be reserved for those women who have failed previous vaginal reconstruction or have other associated complex bowel and urinary anomalies, where dilation and less invasive procedures are not possible. It should not be used as a first line treatment for vaginal agenesis.

When an intestinal vaginoplasty is the only option, surgery should where possible be deferred until adulthood. This means that potential problems of vaginal discharge and bleeding do not trouble the patient during childhood. In addition, the risks of malignancy are deferred and adult women are more able to comply with the extra vaginal examinations required for surveillance of an intestinal vagina [8]

McIndoe–Reed procedure

The McIndoe (Abbe–McIndoe–Reed) technique is still commonly performed throughout the world, as it does not require abdominal surgery and is of a relatively low initial morbidity. A potential neovaginal space is created between the rectum and the bladder. A split-thickness skin graft is then taken from the thigh, buttock, or abdomen and is mounted on a mold and left in the neovaginal space for 7 days. The graft will then epithelize, lining the neovaginal space.

Immediate morbidity is low although complications such as urethral and rectal fistulae have been reported, especially with older, firmer vaginal molds. One troublesome complication of the McIndoe technique is the formation of visible scars at the origin of the skin graft site and this may be unacceptable to some young women.

Overall, the main long-term complication of this procedure is vaginal stenosis, which has been reported in

up to 50% of women [9]. Stenosis can lead to painful intercourse or no intercourse at all, and it is imperative that the patient maintains her neovagina postoperatively by regular sexual intercourse or dilator use. Repeat surgery after a failed McIndoe–Reed procedure is difficult as the use of an intestinal segment can be hampered by excess scarring. Satisfactory long-term results have been reported for the McIndoe operation with up to 100% sexually active after the procedure [10]. There is, however, scanty information in the literature on sexual function or sexual pleasure in this group. As with intestinal vaginoplasty, there have been several case reports of carcinoma developing in the neovagina – in this situation a squamous cell carcinoma [11]. Prolonged postoperative follow-up is necessary with regular vaginal examination and prompt attention to any unusual onset of bleeding or discharge.

The risks of stenosis, need for dilation, and potential malignancy risk make this procedure also unsuitable for children, and surgery should be deferred until late adolescent or adulthood. With the increasing use of dilators and the advent of laparoscopic procedures such as the Vecchietti and Davydov procedures, it is probably that the McIndoe–Reed procedure will become less popular.

Laparoscopic Vecchietti and Davydov procedures

The Vecchietti procedure allows creation of a neovagina by passive traction rather than dilation [12]. An acrylic “olive” with attached tension threads is placed at the vaginal dimple. The threads are passed under laparoscopic control from the vaginal dimple through the abdominal cavity and then to a traction device on the abdominal wall. The tension threads are tightened daily to stretch the vagina. This procedure requires elasticity of the vaginal skin and is not suitable for women with vaginal scarring from previous genital reconstruction. The Davydov procedure is also performed laparoscopically [13]. A perineal incision is made first to create a neovaginal space. Then peritoneum from the pelvic sidewalls and the Pouch of Douglas is freed and directed down toward the vaginal incision to line the sidewalls of the vagina. The top of the vagina is created by suturing a vaginal “roof” of large bowel and peritoneum. This procedure is more suitable for those women with previous vaginal scarring, as the vaginal skin is not required to stretch.

The short-term morbidity of these procedures is low although ureteric, bladder and ureteric damage can

occur [13,14]. Good short-term anatomical and functional results have been reported [12–14]. More long-term results are awaited with regards to sexual function and complications, in particular urinary symptomatology which may be increased in patients undergoing the laparoscopic Vecchietti procedure due to the displacement of the urethral plate [15].

Complications of feminizing genital surgery for ambiguous genitalia

As for vaginal reconstruction discussed above, immediate complications of feminizing genitoplasty need prompt and appropriate attention. However, it is the long-term complications that will impact upon the success of the procedure. Long-term complications such as a poor cosmetic appearance or vaginal stenosis may be easy to identify. They may also be more difficult to assess such as sexual dysfunction and poor psychosexual and psychosocial wellbeing.

Cosmetic appearance

The cosmetic appearance after genital surgery is variable, but up to 40% of women are reported to have an unsatisfactory genital appearance [16]. While immediate cosmetic outcomes may be good, the postpubertal appearance may be very different. Significant scarring and pubertal change may lead to irregular and lop-sided external genitalia. Poor steroid control in CAH may contribute to clitoral regrowth and hypertrophy.

Urinary complications

The anatomical changes present in those born with ambiguous genitalia may lead to incomplete bladder emptying and pooling of urine in the common urogenital sinus. This may lead to reflux and subsequent urinary tract infections as well as postmicturition dribbling. Pediatric studies have reported persistent urinary symptoms including incontinence and reduced bladder capacity [17]. Studies looking at the long-term outcomes of adult CAH patients have shown incontinence as well as high levels of lower urinary tract symptoms with 70% of women complaining of troublesome lower urinary tract symptoms [18]. These studies were all observational questionnaire studies and more definitive urodynamic evaluation would be helpful to establish what the role of surgery is in causing or preventing urinary dysfunction.

Vaginal stenosis

Vaginal stenosis is the commonest complication of feminizing genital surgery [16,19]. In many cases, multiple repeat operations are performed during childhood and adolescence in an attempt to treat recurrent stenosis. However, these repeated attempts at surgical correction limit subsequent successful reconstruction by resulting in excessive scar tissue. Repeat surgery is of course associated with higher levels of complications. Frequent vaginal stenosis leading to repeat surgical correction is associated with an increased level of anxiety regarding intercourse and up to one-third of women experiencing specific difficulties with orgasm [20]. Other complications of multiple repeat procedures include recurrent urinary tract infections and persistent malodorous vaginal discharge. Menstruation may be obstructed and lead to hematocolpos requiring formal drainage and yet more vaginal surgery [21]. Deferral of reconstructive vaginal surgery until adolescence would avoid these complications during childhood.

Sexual function

Sexual function is likely to be affected by both vaginal size and clitoral sensation. Vaginal stenosis leading to pain or the inability to have penetrative intercourse will of course lead to poor sexual satisfaction. The clitoris has only one role – to contribute to sexual pleasure – and there is increasing evidence of the detrimental effect of clitoral surgery on sexual sensation and satisfaction.

Until recently, sexual function outcome data following childhood feminizing genitoplasty procedures have been sparse, with details about the assessment process often limited. Increasing concern from adult women who have undergone feminizing surgery has led to a recent focus of research on sexual function. Questionnaire studies assessing women after feminizing surgery demonstrate high levels of sexual dysfunction in all women after surgery when compared to normal controls [22]. When compared to women with ambiguous genitalia who had not undergone clitoral surgery, 39% of participants demonstrated specific difficulties with sensation and orgasm [23]. Subsequent objective sensation testing for women with CAH who had undergone genitoplasty procedures in childhood has demonstrated significantly impaired sensation to the clitoris in all women following genital surgery, when compared with normal controls, and this correlates to poorer sexual satisfaction [24].

Psychological outcomes

Genital surgery is likely to have an impact on psychosexual development and functioning although there is scanty information on either positive or negative outcomes. An accepted aim of surgery is to improve psychological wellbeing and failure to do so could be considered a complication. There is some evidence that in those who have undergone genital surgery, social and sexual milestones are reached later than age-matched controls and that these women are less sexually experienced and have expressed a lower level of sexual interest than control groups [25]. However, the specific contribution of genital surgery to these problems is difficult to separate from other factors associated with the various conditions assessed.

Conclusion

The short- and long-term complications of childhood reconstructive surgery on the clitoral and vagina are well recognized and documented in the medical literature.

Surgeons operating on the genitals of children have constantly refined their techniques over the years in an attempt to find procedures with lower complication rates and better outcomes. However, the nature of pediatric surgery means that any improvements will not be put to the test until many years later. There is at present no evidence that more modern techniques are less morbid or have a more positive contribution to make long-term outcomes. In addition, the constant focus on surgical techniques and interventions can mean that the psychosexual and psychosocial aspects become neglected.

The majority of women with vaginal agenesis and/or genital ambiguity will wish to have genital reconstruction prior to sexual activity. There is, however, little evidence to support any benefits of such surgery during childhood. Deferring reconstructive surgery until later in life means that unavoidable surgical complications – which can be significant – do not happen in childhood. Informed consent can be taken and complications may be better managed in an adolescent or adult woman who can weigh these up risks and balance them against her requirements for a functional vagina.

DOS AND DON'TS

Do

- Manage all patients with a DSD under a multidisciplinary team
- Avoid vaginal surgery until adolescence
- Avoid clitoral surgery in mild-to-moderate clitoromegaly
- Start vaginal dilation soon after vaginoplasty

Don't

- Ever consider intestinal vaginoplasty as a first line treatment for CAIS and MRKH syndrome. Vaginal dilation is the first line treatment for patients with CAIS and MRKH.

CLINICAL PROBLEM/CASE: DISCUSSION

Case 28.1 discussion

In most children with ambiguous genitalia assigned female, feminizing clitoral and vaginal surgery is carried out as a "one-stage" procedure at 6–8 months of age. The aims of surgery are to achieve a pleasing feminine appearance, to allow menstruation, to preserve sensation and permit normal sexual function, to promote normal psychosocial and psychosexual development and to prevent urological sequelae. Whether or not these aims have been achieved is usually not apparent until the individual has reached adulthood.

- 1 The patient is presenting with vaginal stenosis, which is the commonest long-term complication in feminizing genitoplasty occurring in over 90% of cases [16,19,20].
- 2 Vaginal dilation therapy using specially devised dilators is usually recommended soon after a vaginoplasty to maintain vaginal patency. Vaginal dilation should not be recommended to prepubertal girls. Education and support whilst performing vaginal dilation and deferral of a surgical vaginoplasty until adolescence should increase compliance and improve success rates.

Case 28.2 discussion

- 1 Persistent mucus discharge is a common side effect of an intestinal vaginoplasty. This can be foul smelling and lead to problems with self-esteem and confidence. In some cases it will respond to treatment by vaginal irrigation using short-chain fatty acids and steroid enemas. A significant number of women will need to douche regularly and always wear a pad [7]. Symptomatic diversion colitis has been reported postoperatively and can lead to heavy vaginal discharge with bleeding [26]. If the condition presents, however, symptoms can be alleviated with steroid vaginal douching. In some cases, removal of the intestinal vagina is the only solution.
- 2 There are several case reports in the literature of adenocarcinoma affecting the intestinal vagina, with a reported time to development of carcinoma anywhere from 7 to 50 years after the initial procedure [27,28]. Although these complications are uncommon, treatment is difficult and can lead to removal of the entire neovagina.
- 3 First line treatment for women with MRKH is vaginal dilation therapy [29], which entails the insertion of vaginal molds of increasing width and length and applying pressure on the vaginal dimple. Vaginal dilation poses no surgical or anesthetic risks and, as such, compares favorably against surgical treatment modalities. However, it can be time consuming and demoralising [30] and should be deferred until late adolescence or adulthood, when the patient is old enough to understand and comply with the suggested treatment regime. Success rates of 80–90% have been reported in the literature and are improved when undertaken within the supervision of a multidisciplinary team [31–33].

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CHAPTER 29

Complications of female genital surgery and disorders of sex development II

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KEY POINTS

- Feminizing genitoplasty can be divided into clitoroplasty, labioplasty and vaginoplasty
- Minor complications after clitoroplasty and labioplasty include hematoma and local infection and usually do not harbor any long-term implications
- Major immediate complication after clitoroplasty is ischemia and necrosis due to failure to identify and preserve the dorsal neurovascular bundle
- Long-term problems after clitoroplasty may include impaired clitoral sensitivity and sexual satisfaction
- Major complication after vaginoplasty is vaginal stricture, which may limit the ability to engage in sexual intercourse and also negatively impact sexual satisfaction
- Newer vaginoplasty techniques have the potential to prevent development of strictures and lead to better functional results
- Preoperative stimulation with topical estrogens may be helpful to avoid vaginal stricture
- There is no consensus around the ideal timing for feminizing surgery

Introduction

Female genital surgery for patients with disorders of sex development (DSD) can be fundamentally divided into three procedures: clitoroplasty, labioplasty and vaginoplasty. Ultimately, the main long-term complication of feminizing genitoplasty is a negative impact on sexual function, as a consequence of decreased clitoral/genital skin sensitivity, vaginal strictures and/or a poor cosmetic outcome. As each step of feminizing genitoplasty is associated with a distinct complication profile, we will address them individually.

Clitoroplasty

The goal of the clitoroplasty in the context of feminizing surgery is to reduce the size of the clitoris, thus

rendering extremely masculinized external genitalia (i.e. Prader 4 or 5) a feminine appearance whilst preserving its sensitive function. The first report of clitoroplasty (as opposed to clitoral amputation) for patients with enlarged clitoris dates back to the 1960s [1]. Modern clitoroplasty techniques gravitate around the concept of preserving the dorsal neurovascular bundle as summarized by Mollard [2] and Kogan [3] in the early 1980s. That principle was further substantiated by cadaveric studies in human fetuses demonstrating an extensive network of nerve fibers predominantly on the dorsal aspect of the glans [4].

Therefore any surgical manipulation of the clitoris can potentially damage the nerves, leading to decreased sensitivity of variable degrees.

Complications arising from clitoroplasty can be further categorized either as a function of time elapsed

since the procedure (short-term vs. long-term) or type of complication (i.e. functional vs. cosmetic). Key stages of the operation are summarized below, highlighting specific steps that should be undertaken to avoid complications:

- Clitoral degloving that mirrors what is done for hypospadias repairs. The correct plane of dissection lies between the dartos layer and Buck's fascia, which should not be violated in order to preserve the nerves. The mucosal strip should be divided, aiming to leave a distance of 1.5–2 cm between the glans clitoris and the urethral meatus.
- Dorsal foreskin is split longitudinally and the flaps obtained are used for labia minora reconstruction (labioplasty).
- Buck's fascia is incised longitudinally at 5 and 7 o'clock positions and dissection is carried out circumferentially, freeing it from the tunica albuginea overlying the corporeal bodies. Most authors agree that this dissection should be started as ventral as possible in order to avoid injury to the dorsal neurovascular bundle [5–7]. At the end of this dissection, the neurovascular bundle should be separated from the corporeal bodies for almost the entire length of the shaft of the clitoris. *Failure to adequately preserve the neurovascular bundle can lead to ischemia and even necrosis of the glans clitoris post-operatively, and injury to nerve branches can lead to decreased clitoral sensitivity.* Figure 29.1 exemplifies an overall poor long-term outcome from feminizing genitoplasty.
- The corporal bodies are dissected proximally to the level of the crura and at this point the surgeon decides either for removal or sparing of the corpora. In cases where a corporal sparing technique is chosen, both hemi-corpora are carefully separated in the midline septum to be subsequently placed in labial pouches [8].
- The glans clitoris is receded with an absorbable stitch to the periosteum of the pubic bone, at a distance of 1.5–2 cm from the urethral meatus (Figure 29.2).
- In instances where the glans is too prominent, glans reduction can be done either by wedge resection or by de-epithelization [9], although there are no data to support the impact of either technique on long-term functional and cosmetic outcomes. Figure 29.2 depicts an example of a poorly positioned and prominent glans after feminizing genitoplasty.



Figure 29.1 Long-term follow-up of feminizing genitoplasty performed in infancy depicting a small clitoris and a single perineal opening (persistent urogenital sinus).



Figure 29.2 Enlarged and abnormally positioned glans after feminizing genitoplasty.

Short-term complications

Minor short-term complications such as hematoma and edema are common and usually do not lead to long-term consequences. Hematoma formation can be avoided by placement of a pressure dressing for the first 24–48 hours postoperatively. Keeping a Foley catheter to decompress the bladder during that period for comfort is a reasonable approach. The most ominous complication of clitoroplasty is ischemia with partial or total necrosis of the glans. This is a rare complication, but case series on feminizing genitoplasty report anecdotal cases with an approximate incidence of 2–3% (1 case in series of 40–50 patients) [10,11]. Glans necrosis is an unacceptable technical complication that arises from the faulty dissection of the dorsal neurovascular bundle. When an adequate dissection is performed, contemporary series have reported no cases of clitoral loss [12,13]. Yang et al. performed a capillary perfusion test in 49 patients after nerve-sparing ventral clitoroplasty and demonstrated normal perfusion in all of them [13]. The main differential diagnosis of glans necrosis is venous congestion, which depicts a purple discoloration that tends to improve spontaneously without any intervention and may in some cases be mistaken for ischemia.

Long-term complications

Sexual function

The impact of surgery on clitoral sensitivity is the main point of contention around clitoroplasty. The fact that the clitoris does not have function until sexual maturity is reached has been a reason for some to advocate postponing clitoroplasty until later in life, providing the opportunity for the patient to consent for such intervention herself. However, leaving extremely virilized children unoperated can pose a stigmatizing burden on children and their families, especially for those without adequate psychosocial supervision. The ideal timing for feminizing genitoplasty is still an unsettled matter, lies beyond the scope of this chapter and has been addressed by other publications [14].

In a study of six adult patients with congenital adrenal hyperplasia (CAH) who underwent feminizing genitoplasty in infancy, clitoral sensitivity was assessed for temperature, vibration and light-touch and found to be highly abnormal in all of them [15]. The same group also documented decreased clitoral sensitivity in 24 women who had clitoral surgery compared to controls and to four CAH patients who never had surgery [16]. Sexual

satisfaction may also be negatively affected by clitoroplasty [16–18], although most patients reported in these series had surgery more than two or even three decades ago, and it is possible that the principles outlined above were not duly followed. Fagerholm et al. reported on 24 patients who underwent feminizing genitoplasty in infancy after 1980 and, despite a later sexual debut, overall sexual function and satisfaction were similar compared to controls [19]. Lesma et al. have described similar sexual satisfaction despite reduced clitoral sensitivity in CAH patients that underwent one-stage Passerini-Glazel feminizing genitoplasty compared to healthy controls [20].

Recently, it has been demonstrated that adult CAH patients bearing the null genotype (i.e. the most severe degree of 21-OH deficiency) depicted poorer sexual outcomes regardless of having had surgery or not [18,21]. This underscores the importance of offering long-term counseling, follow-up and psychosocial support for all patients with disorders of sex development, irrespective of age or type of surgical intervention performed, if any. Sexual function is also impacted by outcomes of the vaginoplasty performed during feminizing genitoplasty, and these aspects will be discussed in a separate section of this chapter.

Cosmesis

Although some authors have tried to develop references to guide surgeons on the “normal” appearance and distance between structures of the female external genitalia [22], others have demonstrated that there is a wide variation in the genital dimensions of women without external genitalia abnormalities [23]. Hence, it is challenging to define an acceptable cosmetic result for clitoroplasty. Creighton et al. studied 44 patients that underwent genitoplasty between 1979 and 1995 and found that 41% had a poor cosmetic outcome by their definition, which was requirement for further major surgery [24]. Stikkelbroeck reported on six operated CAH patients that reported similar satisfaction with the appearance of the external genitalia compared to controls using visual analog scales [25]. Large glans clitoris can be bothersome for some patients. The senior author has performed over 10 clitoroplasties in adult women concerned with an enlarged clitoris or glans and obtained self-reported positive results after surgery with regards to both cosmesis and function. Nonetheless, surgical procedures performed in this age group were perceived as being technically more challenging by the surgeon [26].

Clearly, the field of genital surgery for patients with DSD requires devoted and experienced professionals who are willing to follow their patients long-term after surgery in the context of multidisciplinary care. Another modifiable factor that plays a role in the cosmetic result of feminizing genitoplasty is adequate medical control, since poor compliance of patients with CAH can lead to recurrent clitoromegaly.

Labioplasty

The complications of labioplasty are largely cosmetic, although the functional role of the labia should not be underestimated, since the labia minora skin has been shown to be highly innervated in prepubertal girls [27], and the skin of the genital area overlying the clitoris seems to play a significant role in sexual arousal and pleasure [28].

Short-term complications of labioplasty are usually minor and include edema and hematoma, which are preventable by a pressure dressing and bladder drainage with a Foley catheter in the first few days postoperatively; in rare cases, infection and dehiscence can be observed. Venous congestion and ischemia as described for the clitoroplasty are also possible, albeit rare when careful surgical technique is exerted. Labia minora are usually created from flaps of the foreskin covering the phallus and labia majora from the rugated scrotal-type skin by means of a Y-V plasty. Occasionally, dehiscence of sutures after labial reduction is seen; however, healing usually takes place by secondary intention without undesirable effects on cosmesis (Figure 29.3).

Long-term complications are related to the appearance of the external genitalia and to a perception by the patient of “not enough” vs. “too much” labial tissue (i.e. absence vs. redundancy). Again, it should be stressed that there is a wide variability in the anatomical aspects of the genitalia among normal females who never had surgery [23] and that *de novo* virilization can ensue in cases of poor hormonal control.

Vaginoplasty

Vaginoplasty is a key step of feminizing genitoplasty and its main complication (vaginal stricture) can significantly hinder sexual activity and satisfaction;

therefore, it plays a central role in the functional outcomes of the feminizing procedure. In the last two decades, a number of new techniques as well as alternative access routes for old procedures have been described, thereby reflecting the limitations of traditional flap techniques used in the 1980s to create an adequately functional vagina.

The first step for a successful vaginoplasty is to perform a thorough radiological (genitogram) and endoscopic evaluation of the urogenital sinus (UGS) delineating vaginal size as well as its anatomical relationship with the UGS and the urethra. The following measurements are important:

- Length of the UGS, i.e. distance from the vaginal entry on the UGS and the perineal skin.
- Distance between the urethro-vaginal confluence and the bladder neck.
- Length of the vagina itself, measured as the distance between the point where the vagina enters the UGS to its dome when distended with fluid.

There is no consensus on what constitutes a high vagina or a short UGS, although some reports have suggested that a common channel ≤ 3 cm in infants would be the cut-off between a high and a low vaginal opening [29]. Others (including the authors) have implied that the length of the urethra is in fact more important than the length of the common channel [30,31]. Moreover, in our opinion a rudimentary, hypoplastic vagina (< 2.5 cm vaginal length) is very challenging to bring down to the perineum without jeopardizing its blood supply. Preoperative use of topical estrogen can make the vagina more robust, allowing for better traction and dissection during pull-through vaginoplasty. Even so, in the presence of very short and hypoplastic vaginas it may be worthwhile to postpone vaginoplasty until puberty, hoping that menstrual fluid accumulation will cause distension and increase in its size. Figure 29.4 depicts an example of a high UGS with a hypoplastic vagina.

Timing of vaginoplasty remains controversial [14], since some authors have demonstrated a high incidence of vaginal strictures requiring reintervention after puberty in patients who had surgery in their infant years [18,24,25]. The need for postpubertal reintervention should be analyzed under a critical lens; while some women may require a full redo reconstructive procedure, in the authors' experience most revisions are actually minor in the form of “introitoplasties” followed by



(a)



(b)



(c)

Figure 29.3 Dehiscence labial sutures: (a) and (b) demonstrate initial appearance and (c) depicts the long-term outcome with minimal residual scarring.

dilatations that lead to a reasonable caliber of the vagina to allow menstruation and coitus to occur. Minor strictures after vaginoplasty performed in infancy should

be weighed against the difficulty of obtaining tissue for reconstruction of the distal vagina when vaginoplasty and clitoroplasty are not performed simultaneously.

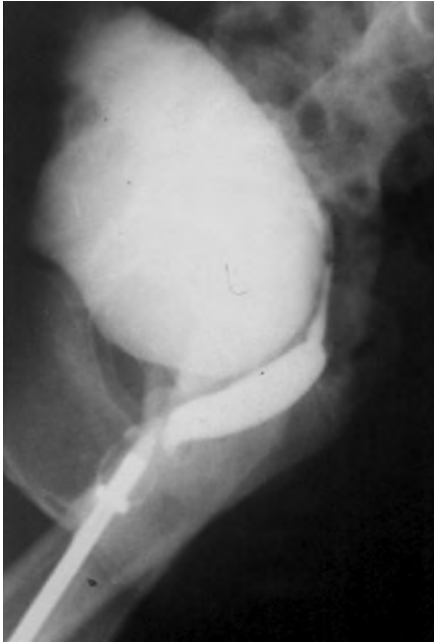


Figure 29.4 High urogenital sinus and hypoplastic vagina.

The main complication after vaginoplasty is the development of strictures, which in turn may lead to difficulties with menstruation and sexual intercourse. Other complications are more specific to the technique being used, hence we will categorize them as:

- Flap vaginoplasties – Fortunoff and Passerini-Glazel.
- Mobilization of the urogenital sinus as a unit – Total (TUM) or Partial (PUM).
- Vaginal pull-through – anterior perineal or transrectal approach (ASTRA).

Flap techniques

For a long time, flap techniques were the only ones available when performing feminizing genitoplasty and most long-term outcomes studies refer to patients who underwent such techniques. With the flap techniques, a perineal skin flap is used to bridge the gap between the vagina and the perineum. The Fortunoff flap was described in 1964 and consists of an inverted “U”-shaped incision in the perineum that is elevated as a flap and sutured to the posterior wall of the vagina [32]. Other than strictures, it can also lead to hair growth in the vagina after puberty.

In the Passerini-Glazel technique, the vagina is disconnected from the UGS and the so-called mucocutaneous plate from the UGS is tubularized and anastomosed to the vagina [12].

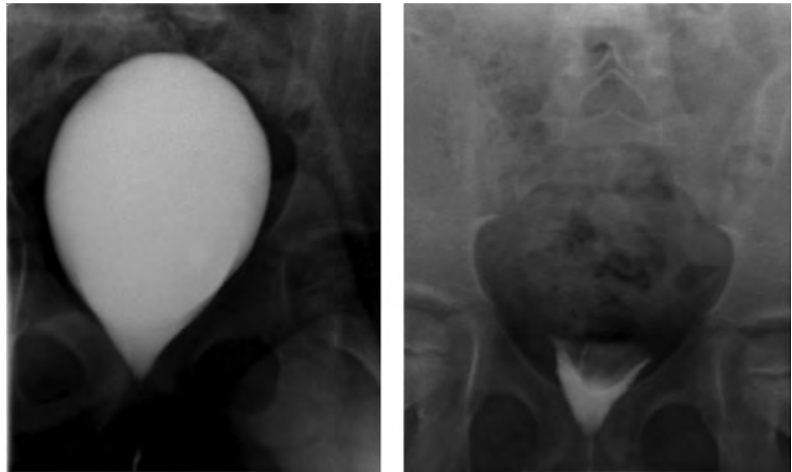
When flap techniques are utilized, only the posterior vaginal wall is mobilized; since the actual anterior vaginal wall is never mobilized, the urethra is invariably left in a hypospadiac position.

Mobilization of the urogenital sinus as a unit

In the last two decades, total and partial urogenital sinus mobilization (TUM and PUM, respectively) have been introduced to the surgical armamentarium of vaginoplasties. These are technical variations of the surgical approach to cloacal repairs described by Peña, aimed at avoiding the difficult separation of the vagina from the urethra and its potential complications (urethro-vaginal fistula development) [29,33]. UGS mobilization is usually performed in conjunction with a Fortunoff flap for patients with low vaginas (UGS common channel <3 cm) and although the follow-up is still limited (5–7 years at best), total urogenital mobilization (TUM) and partial urogenital mobilization (PUM) seem to produce better results than flaps alone [34–36], with most patients depicting a normal vaginal opening on inspection. Long-term functional studies are definitely warranted. PUM was devised as a conservative variation of TUM where the anterior dissection stops at the level of the pubo-urethral ligaments and, unlike TUM, these ligaments are not incised [35,37]. The theoretical advantage of that would be to limit disruption of the urinary continence mechanism due to unwarranted traction on the bladder neck. There has been a report of urge-incontinence following PUM in a child [34] and a recent comparison between the two techniques in terms of urinary continence showed no difference after 4 years of follow-up [38]. However, long-term follow-up is definitely warranted, especially after situations such as pregnancy and delivery, to ensure no urinary functional issues will arise. The authors have documented a low lying bladder neck in a girl who underwent TUM in infancy; although the patient was asymptomatic from a lower urinary tract perspective, this is certainly a finding that warrants further follow-up and observation (Figure 29.5).

In terms of other short-term complications, dissection in the posterior vaginal wall can lead to rectal injury, and urogenital mobilization procedures in postpubertal

Figure 29.5 VCUG in 10-year-old girl who underwent total urogenital sinus mobilization in infancy showing a low-lying bladder neck.



girls can be very tedious due to a significant amount of bleeding upon inadvertent (and very often unavoidable) entry into the spongiosum tissue of the UGS.

In addition, as mentioned above, attempting to perform mobilizations alone for high UGS often results in a hypospadiac urethra.

Vaginal pull-through

The most challenging aspect of feminizing genitoplasty for high UGS is the separation of the vagina from the urethra. Although flap vaginoplasties have been widely used even for high UGS, in our view the best approach for such cases is a vaginal pull-through, where the vagina is disconnected from the UGS and mobilized to the perineum. In this scenario, the UGS is used as urethra with part of its tissue used for the introitoplasty. This technique was initially described using an anterior perineal approach [39], which proved to be technically demanding due to poor visualization and possible injury to adjacent structures leading to fistulas (Figure 29.6). The use of a posterior sagittal approach improves visualization, facilitating separation of the vagina as well as allowing its dissection laterally to gain length for the pull-through, much like what is done for the surgical treatment of anorectal malformations [40].

In recent years, the anterior sagittal transrectal approach (also known as ASTRA) was introduced [41]; in this technique, with the patient in prone position, only the anterior wall of the rectum is opened longitudinally allowing the high vagina to be separated from the

urethra and mobilized to the perineum with improved exposure in a more ergonomic way for the operating surgeon without the need for a colostomy or extensive bowel preparation. Others have suggested that the vaginal pull-through can be performed without opening the rectum, as long as the patient is placed in a prone position [30].

From a surgical standpoint, a vaginal pull-through using the ASTRA approach is theoretically much more involved than a flap vaginoplasty and potentially associated with more complications, such as perineal sepsis, rectal dehiscence/stenosis, urethro-vaginal fistula,



Figure 29.6 Vesico-vaginal fistula after vaginal pull-through using a perineal approach.

fecal/urinary incontinence; nonetheless, complications so far have been exceedingly rare. In a series of 23 children with a mean age of 2.3 years who underwent a vaginal pull-through using the ASTRA approach, only one developed perineal infection and was treated with a diverting colostomy with no impact on the repair. After a mean follow-up of 3.4 years, no fecal or urinary continence issues have arisen and no cases of urethrovaginal fistulas have been recorded. All 15 patients who underwent subsequent examination under anesthesia portrayed visible separate urethral and vaginal openings; however, long-term follow-up will unveil whether strictures were prevented or not [31].

Despite the improved exposure offered by the ASTRA approach and the prone positioning, dissection and mobilization of the vagina in the infant can be challenging given its inherent friability. The use of topical estrogens on the genitalia 4 weeks before surgery in this age group, with the goal of increasing the robustness of the genital structures for surgical manipulation, has produced encouraging preliminary results and, when given perioperatively, could play a role in avoiding future development of strictures (authors' personal experience – unpublished data) (Figure 29.7).

Extensive vaginal strictures are one of the most difficult complications to treat in this population and the surgical approach will be contingent upon the length and/or location of the narrowed segment. Short segments can be treated with dilations if the stenotic tissue is found to be supple. In cases of recurrence, or when there is significant fibrosis likely unresponsive to dilation, we prefer to incise it at 3 and 9 o'clock position, covering the incised areas with full thickness buccal mucosa grafts. Once the graft is taken we wait for 3 weeks and start with progressive dilations. Because the dilation process will depend on the cooperation and compliance of the patient we usually wait to perform these procedures until the patient is cooperative and understands the reasoning for the treatment. There are controversies for the treatment of long-segment strictures. In cases of significant vaginal hypoplasia or little residual vaginal tissue and complete obliteration following procedures in infancy, early reconstruction of the vagina in adolescence may be indicated in order to allow menstrual fluid to be eliminated. Another option would be to block menstruation with hormonal treatment and postpone the procedure until the patient is psychologically ready.

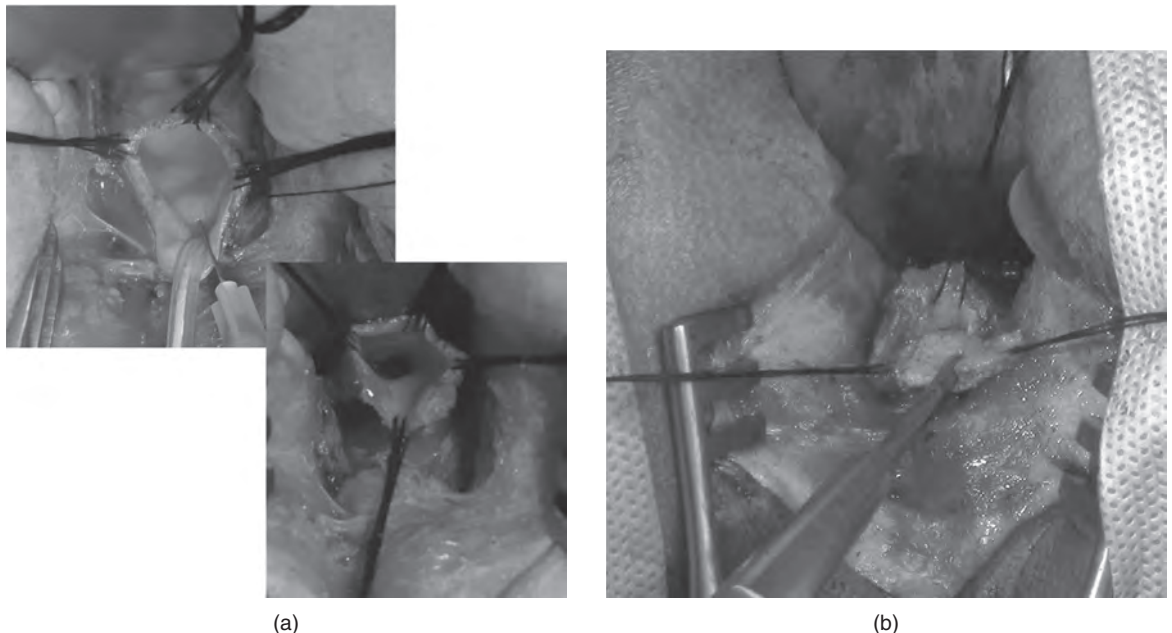


Figure 29.7 Prepubertal vaginoplasty in congenital adrenal hyperplasia (a) without and (b) with preoperative estrogen stimulation.

In cases where postoperative dilations are not feasible, we have preferred to perform a neovagina constructed with either sigmoid colon or ileum. Such neovaginas have the disadvantages of producing feculent intestinal mucus as well as developing colitis, especially when an excessively long segment of bowel is used in the reconstruction. In recent years, the use of buccal mucosa grafts for vaginal reconstruction has

been described [42–44]. The grafts are usually taken from both cheeks and meshed in order to cover the area of vaginoplasty. Preliminary results are encouraging but long-term follow-up is warranted to confirm the optimistic initial impression. Dilations are necessary post-buccal vaginoplasties; therefore, the surgeon must be sure the patient will comply to such recommendations.

DOS AND DON'TS

Do

- Discuss surgical decisions openly with patients and/or families in a multidisciplinary setting with strong psychosocial counseling
- Follow patients long-term in a multidisciplinary setting with strong psychosocial counseling
- Emphasize compliance with medical treatment.
- When performing clitoroplasty, develop a subtunical plane of dissection to spare the dorsal neurovascular bundle
- When performing vaginoplasty, perform contrast studies and endoscopy to assess the anatomy of the urogenital sinus and pick the best possible technique to avoid strictures and need for redo surgery

Don't

- Offer surgery for patients with mild degrees of virilization
- Attempt feminizing genitoplasty unless very familiar with the procedure

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CHAPTER 30

Persistent cloaca

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KEY POINTS

- Renal impairment occurs in up to 50% of patients
- Normal voiding incontinence is uncommon
- Social urinary continence can be created in up to 95% of patients
- Gynecological problems frequently occur at puberty and should be anticipated
- Patients with a long common channel have a poorer prognosis

CLINICAL PROBLEM/CASE

- Prenatal diagnosis at 20 weeks: pelvic cystic structure, bilateral hydronephrosis ? fetal bladder visible
- At birth: imperforate anus with single perineal opening; TOF with oesophageal atresia; VSD on echo; small bright hydronephrotic kidneys; butterfly vertebrae and short sacrum; absent left radius
- Early surgery: day 2 – thoracotomy and repair of TOF; colostomy formation; suprapubic catheter and vaginal drain

Infant treatment

- Early creatinine >200 but renal function stabilized with free drainage of SPC and vaginal drain
- Bladder assessment shows good bladder emptying but persistent hydrocolpos so started clean intermittent catheterization (CIC) via common channel
- Definitive surgery: posterior sagittal anorectal vaginourethroplasty (PSARUVP) and total urogenital mobilization (TUM) aged 10 months

- Colostomy closure aged 12 months
- GFR at 1 year 35 but stable

Preschool

- Aged 2.5–3.5 years: recurrent UTI; increase hydronephrosis and poor bladder emptying; worsening renal function
- Urodynamics: small capacity, poorly compliant bladder with detrusor overactivity. No leaks with high pressure contraction. Voids mainly by straining
- Cystoscopy: urethral stenosis and SPC
- By 4 years: end-stage renal failure and on transplant list
- Bladder augmentation (ileocystoplasty with Mitrofanoff) and ACE aged 4.5 years
- Aged 5: live related renal transplant (normal graft function)
- Started school clean and dry on Mitrofanoff CIC

Introduction

Persistent cloaca is the most severe type of anorectal malformation encountered in children. It is seen exclusively in girls and exists where a separate rectum,

urethra and vagina all fail to develop normally and drain via a single common channel onto the perineum [1]. This malformation is rare [1,2] with an incidence of 1 in 50,000 [1,2] and persistent cloaca has variable presentation and a wide spectrum of severity [3,4]

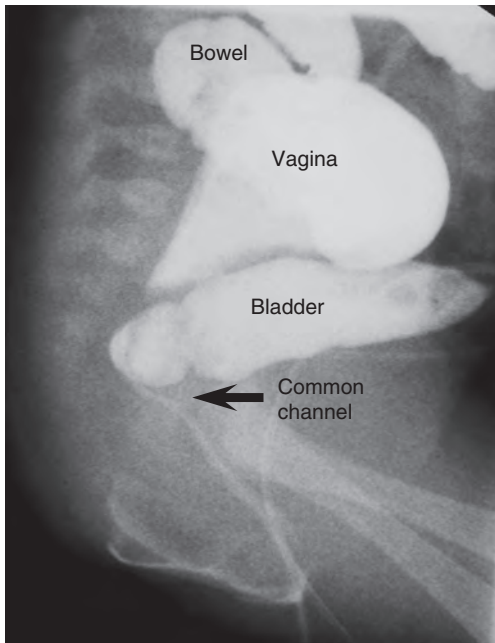


Figure 30.1 Radiological illustration of an infant with a long common channel cloaca.

(Figure 30.1). Cloacal anomaly should not be confused with cloacal exstrophy which is an abdominal wall defect with the bladder and hindgut exposed [5].

Children with persistent cloaca have a high incidence of associated anomalies and the defect may occur as part of a syndrome such as VACTERL or caudal regression syndrome [6,7]. The most commonly associated anomalies include urinary tract, genitalia, vertebral anomalies including sacrum, as well as cardiovascular, respiratory, gastrointestinal and central nervous system anomalies [8,9]. Persistent cloaca defect is often classified by the length of the common channel when measured endoscopically, with a short common channel measuring less than 3 cm and a long common channel more than 3 cm. The latter remains a difficult reconstructive challenge but with advances in surgical technique and perioperative medical care it is now possible to anatomically correct the defect in the majority of patients [3,5,10].

Historically, for many years cloacal malformations were called imperforate anus with urogenital sinus [11]. Patients therefore usually had surgical treatment for imperforate anus and the urogenital sinus portion would be dealt with at a later date. More recently, surgical treatment for cloaca has evolved and this coincided

with a new understanding of anorectal malformations after the scientific study of these defects by Stephens [12] and the development of new surgical techniques [13] such as the posterior sagittal anorectoplasty described and popularized by Peña [13,14]. Most surgeons now advocate simultaneous repair of urinary, vaginal and rectal components at the primary procedure, either by abdominoperineal or abdominal and posterior sagittal approach [15,16]. The ideal goals of the primary surgical reconstruction for patients with persistent cloaca is the achievement of bowel and urinary control for the child and also normal sexual function in adult life.

Diagnosis and early management

The diagnosis of persistent cloaca is suspected in a female infant with imperforate anus who on examination has a single perineal orifice [5]. The external genitalia often appear small with absence of the labia minora. A hypertrophied clitoris has been reported in some patients, which can lead to confusion with disorders of sex differentiation (DSD) (previously known as intersex disorders) [5]. Hydrocolpos is present in 50% of patients [5,10], which may be palpable on abdominal examination.

The goal of early management is to detect associated anomalies, and achieve satisfactory diversion of the gastrointestinal tract and appropriate drainage of the urinary tract if indicated [1,5]. In patients with renal impairment it is also important to have joint nephrology care [1]. A colostomy is performed to decompress the bowel and definitive surgery can then be planned when the patient is older, allowing the infant to feed normally and thrive [5,10]. At the time of colostomy formation, endoscopy should be performed to assess the length of the common channel and the internal vaginal and urethral anatomy. Further investigations to document the structural anatomy including distal colostogram and micturating cystogram should be performed before definitive surgical repair. Cross-sectional imaging such as MRI or CT with 3D reconstructions can display the internal anatomy beautifully [5].

Surgery

Using the posterior sagittal anorectal vaginourethroplasty (PSARVUP), the cloaca can be repaired by complete

separation of the three structures [15]. The rectum is identified by posterior sagittal approach and dissected from the vagina. The anterior vaginal wall is then meticulously dissected from the urinary tract and both rectum and vagina are mobilized from below (and above where necessary) before being placed in their normal positions. The previous common channel is used to create a neourethra [15].

In cases of a long common channel the vagina may be too short to anastomose to the perineum so additional skin flaps or intestinal replacement are used to create a vagina [17].

Peña later described the technique of total urogenital mobilization (TUM) for repair of persistent cloaca. This surgical maneuver has reduced operative time considerably and simplified the procedure. In this modification, the rectum is separated from the vagina via the posterior sagittal approach and then the entire urogenital sinus is dissected and mobilized en bloc [18]. The dissection proceeds laterally and anteriorly to the urethra and vagina in a circumferential manner. Avascular ligaments attaching the urethra and vagina to the pelvic brim are divided and the dissection continues around the vagina, anterior wall of the bladder and urethra until enough length has been gained to connect the vaginal edges and urethra to the perineum [18]. In cases of a long common cloacal channel, a combined abdominal, posterior sagittal approach with total urogenital approach may be necessary.

Outcomes

In infants with anorectal and cloacal malformations, for the first few years of life, surgical and medical treatment focuses primarily on the construction of a new anus and to restoring continuity to the gastrointestinal tract. The outcome literature for anorectal and cloacal malformations concentrates mainly on the results for fecal continence in children and ways of improving this by various surgical techniques [19]. Bladder dysfunction can easily be overlooked since surgery is performed at an age where urinary incontinence is considered normal. Recurrent urinary tract infection may be overlooked or attributed to coexisting vesicoureteric reflux or renal anomalies, which are prevalent in a high proportion of these children [20,21]. However, detecting bladder dysfunction at an early age is essential in avoiding deterioration in renal function [22,23].

Renal abnormalities and function

Structural abnormalities of the kidney are common in cloaca patients and are diagnosed in around 60% at presentation [24,25]. Renal dysplasia, duplex systems, pelviureteric junction obstruction and renal ectopia were the most frequently encountered anomalies [24–26].

A high incidence of renal failure is observed in cloaca patients. In one large retrospective review, half had chronic renal failure by 5.7 years, which was end stage requiring renal transplantation in 19% [24]. Renal impairment causes serious morbidity and there was an overall mortality rate of 6% from renal failure in this series [24]. Patients with structural abnormalities, particularly renal dysplasia, solitary kidney and vesicoureteric reflux, were statistically more likely to develop chronic renal failure [24].

Postnatally, it is important to relieve urinary tract obstruction early, correct upper tract abnormalities, prevent urinary tract infection and treat bladder dysfunction, thus dealing with the main preventable causes of renal deterioration [27]. Regular outpatient review and serial ultrasound scans of the kidneys throughout life is essential, particularly postsurgery to improve continence where extra vigilance is needed [24].

Continence

Around 60% of patients are continent of feces at long-term reviews [24,25,28]. However, only 28% are totally continent by spontaneous bowel movements and have satisfactory control [24,25] but around 30–40% need rigorous bowel management programs in the form of rectal washouts or antegrade enemas to achieve social continence [1,28]. However, psychological assessment of this group has shown that there is no association between the degree of incontinence and psychological adjustment [5].

Urinary continence

Urinary continence has been reported to be between 54–95% [1,5,16,28]. Hendren reports 64% of his patients void spontaneously and are dry by voiding, not mentioning the need for additional surgery [16]. However, in the Great Ormond Street series, only 22% void spontaneously and are dry. A further 12% of that group

Table 30.1 Major anatomical factors associated with urinary continence.

			Continent by			
			Void	CIC	Surgery	
Presenting anatomy		N	Continent			
Common channel	<3 cm	26	21 (81%)	8 (31%)	3 (12%)	10 (38%)
	>3 cm	24	19 (79%)	3 (12%)	3 (12%)	13 (53%)
Bladder neck	Good	29	27 (93%)	11 (38%)	4 (14%)	12 (48%)
	Poor	21	13 (62%)	0	2 (10%)	11 (52%)

CIC, clean intermittent catheterization.

have achieved continence by clean intermittent catheterization but 46% of their cloaca patients required major reconstructive surgery [1]. Peña's review also supports the above, as 48% of his patients required an intervention to achieve social continence of urine [28]. The surgical procedures performed were to augment the bladder to a sufficient capacity, reconstruct or close the bladder neck to prevent leakage of urine, and to provide a catheterizable channel for the child if the urethra was unsuitable for clean intermittent catheterization [1,26]. Multiple procedures were often necessary to achieve satisfactory urinary continence and independence for the child. Patients with short common channel and good bladder neck at presentation are much more likely to be continent by normal voiding [1] (Table 30.1).

The etiology of urinary incontinence in cloaca patients can be secondary to structural abnormalities of the bladder (including bladder atresia), bladder neck or urethra and sacral dysplasia. In addition, there is a high incidence of neurogenic bladder in this group, often associated with lumbosacral bony abnormalities or intraspinal lesions [5,32]. Cloaca patients are also potentially at risk of iatrogenic nerve damage during the surgical repair.

In a recent prospective study of anorectal patients, 90% of cloaca patients had bladder dysfunction on urodynamics at presentation [33]. The most common abnormal urodynamic finding overall was detrusor overactivity (hyper-reflexia) with bladder instability during the filling phase and high detrusor pressures during the voiding phase [33]. All patients then had a combination of PSARVUP and TUM performed.

After reconstructive surgery, there was a change in bladder function in 5 of 10 (50%) of the cloaca group and in 1 in 20 (5%) of ARM patients (Table 30.2). All

six of these patients subsequently required intervention by clean intermittent catheterization or urinary diversion and the observed change was statistically significant in the cloaca group.

In the prospective urodynamic study, four of the cloaca patients who had deterioration in bladder function postsurgery showed a change from detrusor overactivity pre-op to an inadequate bladder post-op on urodynamics [33]. All four had a long common channel (>3 cm). Other groups from Rome and Osaka report similar urodynamic findings of an atonic bladder in 65–80% of their patients post-reconstruction [34,35]. Posterior sagittalanorectoplasty alone does not appear to alter bladder function in anorectal patients [36] so deterioration in bladder function in cloaca patients may be associated with mobilization of the common channel to create a separate urethra and vagina. Peña reports urinary incontinence in 72% of those with a long common channel compared with 28% of those with a short common channel [2,25].

An atonic bladder after surgery is suggestive of damage to the nerve supply at a lower motor neurone level. The urinary tract and vagina share a common wall, therefore some dissection between these structures is necessary during repair of the cloacal anomaly [2,5].

Table 30.2 Status of urodynamics after surgical reconstruction.

	n	Bladder function		
		Stable	Improved	Worse
Cloaca	10	5	0	5
ARM	20	16	3	1

ARM, anorectal malformation.

Scott demonstrated that the peripheral nerve supplies were deficient in patients with sacral agenesis so even minimal trauma to these nerves in these patients can result in additional functional loss, which may not have been the case in children with normal nerve fibers [37]. As the majority of cloaca patients have pre-existing neurovesical dysfunction it may be that minimal disruption during surgical repair results in denervation and an atonic bladder [5].

It is therefore recommended that all cloaca patients should be screened for bladder dysfunction post-reconstruction.

Gynecological outcome

Cloaca patients have a high incidence of innate gynecological problems [38,39] but these may remain asymptomatic until menarche or early adult life. The mullerian and vaginal abnormalities found in patients with cloacal malformation show great variation depending on whether the confluence is high or low. Sixty percent [28,38,39] of cloaca patients have some degree of septation of the uterus and vagina, ranging from a partial septum in a large vagina with single cervix and uterus to a completely separated double vagina with double cervix and uteri.

In one long-term outcome study of 41 adult patients, two-thirds developed uterine function at puberty whilst 20% had primary amenorrhoea due to a vestigial uterus [38] (Table 30.3). Thirty-two percent were menstruating normally and 15 (36%) presented with hematometra/hematocolpos, typically presenting with cyclical abdominal pain at puberty (Figure 30.2). The most common cause of the obstructed uterus was stenosis of persistent urogenital sinus (no previous genital tract reconstruction) but a few developed stenosis of previous

vaginal reconstruction. All patients who developed an obstructed uterus required surgery [38].

Levitt and Peña also reported an obstructed uterus in 41% of their 22 cloaca patients [39]. In their series, the group of patients who presented as teenagers with cystic abdominal masses all had surgical resection of the cystic masses involving their fallopian tubes and uteri [39]. However, alternative treatments are favored by other institutions where patients are treated with an-ovulatory drugs (LHRH analogues) and delayed repair is performed involving vaginoplasty and/or division of vaginal septae, preserving uterine structures where possible [38,40].

The long-term outcome for the uteri left in situ is unknown and it has been suggested that endometriosis and infertility may be the result of an obstructed uterus [39,41]. There are case reports in similar patients who were able to conceive and carry a pregnancy to term [42,43], which has encouraged surgeons to create a passage for effective uterine drainage and to preserve the uterus and fallopian tubes where possible.

Adult gynecological follow-up has been reported in 21 adult patients [39]. It was encouraging to find that 86% had an adequate vagina with no menstrual problems



Figure 30.2 MRI scan of adolescent cloaca patient who presented with a 6-month history of cyclical abdominal pain.

Table 30.3 Outcome at menarche for 41 cloaca patients.

Outcome	n	%
Normal menstruation at puberty	13	32
Hematometra	15	36
Amenorrhea (vestigial uterus)	8	20
Early puberty (normal uterus and vagina)	3	7
Amenorrhea under investigation	2	5

and 12 (57%) are or have been sexually active. Half of these 18 women have progressed normally from their initial reconstructive procedure through menarche to adulthood without the need for further vaginal surgery [39]. An additional five (28%) had delayed primary vaginal reconstruction: three at puberty and two as adults, which was adequate to allow sexual intercourse later in life [39]. These results are significantly better than those seen in adult intersex patients where 98% of the 44 women who had feminizing genitoplasty in childhood needed further surgery in adolescence [44]. However, 19% of adult cloaca patients required additional vaginal surgery to facilitate intercourse. Even if the girl has no problems at menarche, her vagina may not be adequate to allow sexual intercourse later in life [39].

In the past 30 years, as surgical outcomes have improved, patient expectations have also changed. As the life expectancy, continence rates [1] and quality of life has improved, these young women now have the same expectations as their peer group, so sexual activity and fertility become an issue. For many women, it is important to preserve the uterus and therefore the potential of fertility, however small that may be.

Hendren reports the delivery of healthy babies to six of his cloaca patients [16]. If pregnancy does occur, these women will require considerable support, particularly those with impaired renal function, bladder

augmentation and catheterizable conduits [45]. Delivery by Caesarean section is usually recommended in patients where vaginoplasty has been performed [44].

Conclusion

Although the majority of cloaca patients can achieve social fecal and urinary continence with the surgical reconstructive procedures performed today, a large number will require additional, and sometimes multiple, urological procedures not only to achieve continence but also to treat bladder dysfunction and to protect the upper tracts [1]. Half develop renal failure so most patients, particularly those with severe malformations, will need regular review and lifelong surveillance [24]. Due to the high incidence of associated gynecological problems, all these girls should be reassessed at early puberty. Additional surgery may then be necessary to create a vagina for menstruation and for sexual intercourse, which is possible in the majority [39]. As more of these patients reach adult life better data will become available on long-term outcomes [25].

Persistent cloaca still remains one of the most challenging conditions to treat in pediatric surgery and urology and these patients should be cared for by a dedicated team with specialist experience in this area.

DOS AND DON'TS

Do

- Refer a patient with persistent cloaca to a dedicated team with specialist experience in this area
- Screen early for associated anomalies, particularly urinary tract and spine
- Check bladder for bladder dysfunction, especially post-reconstructive surgery
- Reassess gynecology close to menarche as additional surgery may be necessary to create an adequate vagina for menstruation and for sexual intercourse

Don't

- Confuse with cloacal exstrophy, which is an abdominal wall defect with the bladder and hindgut exposed
- Forget to monitor renal function throughout life and prevent causes of renal deterioration

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PART VII

Renal Impairment Surgery

CHAPTER 31

Hemodialysis and peritoneal dialysis

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KEY POINTS

- The incidence and prevalence of chronic kidney disease in childhood is increasing
- Transplantation is the gold standard for the management of end-stage kidney disease. Dialysis is sometimes necessary
- Choice of mode of hemodialysis or peritoneal dialysis depends on family preference, and clinical and environmental circumstances. Peritoneal dialysis is more “child-centred”
- For hemodialysis, “no needle” dialysis via a central venous catheter is better tolerated, especially by younger children, and central vessels should be reused as far as possible in cases of repeated access. In older children, vascular access in the form of arteriovenous fistula may be more durable
- For placement of peritoneal tubes, laparoscopy has the advantage of placement under direct vision, with an extraperitoneal tunnel to fix the catheter in the pelvis

Introduction

Renal replacement therapy in childhood has evolved in tandem with the adult experience. Whilst the ultimate aim is a kidney transplant, preferably without recourse to dialysis, hemodialysis and peritoneal dialysis remain crucially important therapies for acute and end-stage chronic kidney disease.

The incidence and prevalence of the population of children receiving renal replacement therapy seems to be increasing [1], with most marked increases being in the 12–16-year-olds. At transfer to adult services according to [1], around 85% of patients had a functioning transplant but a third had one or more reported co-morbidities.

The ethos of providing dialysis in childhood has been to promote and support development as normally as possible in an environment familiar to the child. In practice, home peritoneal dialysis is preferable in this sense. Pediatric dialysis units in the UK are regionalized, and twice- or three-times weekly trips to hospital for hemodialysis can be disruptive and expensive. In addition, the cardiovascular response to hemodialysis (with

episodes of hypotension during treatment) may make this a less attractive option.

Children on dialysis are short compared to their peers, their blood pressure is higher and control of metabolic bone disease and anemia is challenging (although improving over time) [2]. There is also evidence that hemodialysis increases the risk of subsequent allograft failure [3].

Techniques of vascular and peritoneal access will be considered separately.

Hemodialysis access

Technique

Broadly speaking, vascular access for dialysis can be by means of an indwelling central venous catheter (CVC) for “no needle” dialysis or by establishing a high flow conduit which can be punctured for access (e.g. an arteriovenous fistula (AVF), or using a prosthetic graft).

In small children, needled conduits can be technically difficult to establish and maintain, and access requires repeated needling which can be traumatic for the child.

In the older child, an arteriovenous fistula may be far more acceptable, and there is good evidence for the durability and lower infection rates of AVFs compared with CVCs [4] bearing in mind patient selection on the basis of tolerability of repeated needling. The following section considers CVCs.

If dialysis is anticipated to last for more than a few weeks, a tunneled cuffed line is preferable, as they are more durable, more comfortable and less obtrusive, and have fewer complications such as displacement in comparison to temporary lines. In the acute setting, a percutaneously-placed (by the Seldinger technique) line is reliable in the short term [5,6], which can then be removed or, if longer-term access is required, revised with a tunneled line.

The upper body central circulation is preferable, in particular the right internal jugular vein. Access to the catheter is easy, the route and distance to the heart is shorter, and the lower body vessels are preserved, for either future access or for transplantation. The vein is accessed either by needle and guidewire or by open approach through a small transverse neck crease incision. Current standards would recommend the use of ultrasound localization of the vein for puncture [7]. The line is tunneled from a convenient exit site on the anterolateral chest wall. It is important to ensure a gentle curve from venotomy to exit site as an acute angle may cause a kink. The line must be sized approximately prior to this maneuver to ensure the cuff is placed within the subcutaneous tissue. Reasonable surface landmarks for the right atrium include the midpoint between sternal notch and xiphoid, and the right nipple to indicate line length. When the line has been placed via the peel-away introducer, or directly through a venotomy, the tip position must be confirmed by on-table fluoroscopy.

Many hemodialysis lines have proximal and distal lumens (Figure 31.1) although dialysis may be performed

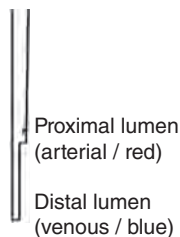


Figure 31.1 Schematic showing the separation of the lumens of a dual lumen hemodialysis catheter.



Figure 31.2 Single lumen hemodialysis catheter.

through a single lumen as illustrated by the catheter in Figure 32.2, so adequate flow must be ensured through both lumens. The largest line passable with ease is preferable, in accordance with Poiseuille's law determining flow. The tip of the line frequently needs to be placed within the right atrium, especially in smaller children, when the catheter can become occluded by its apposition to a vein wall if it is not intra-atrial. A reasonable practical approach to estimating adequate flow is to withdraw blood manually using a 20ml syringe at the time of insertion: flow should be smooth and constant over a few seconds through both lumens. Some manipulation of the line is frequently required to maximize the flow; constant reconfirmation of tip position by fluoroscopy is therefore imperative. On occasion, two smaller caliber lines can be used to provide two lumens. In very small infants, customized strategies for achieving adequate flow may be required for hemodialysis access [8].

Outcome

One of the striking features of reports of dialysis outcome are the myriad ways in which events and survival are reported, in terms of frequency of events per unit time of catheter use, or events within a group of patients. Nonetheless, all of the reports give a flavor of events and catheter survival and some are summarized below.

The European Pediatric Dialysis Working Group reviewed and published its contributing centers' experience of vascular access [9]. Sixty percent of patients

were dialyzed via CVC, 38% via a AVF and 2% via a graft. Infections were the commonest complications of CVCs, with bacteremia occurring with a frequency of 1.9 per 1000 catheter-days and exit site infection 1.8 per 1000 days. Infection requiring CVC removal occurred with a frequency of 0.9 per 1000 days. Line malfunction requiring catheter change occurred 2.1 times per 1000 catheter-days.

Favorable outcomes of more permanent means of dialysis access compared with CVCs have been reported [4] but it needs to be emphasized that patient selection for fistulae and grafts is crucial. In the author's unit, AVFs are usually reserved for teenagers, and frequently in those with failed previous transplants where issues like antigen sensitization have made the prospect of long-term dialysis much more likely.

Dialysis in small children can be formidable, and the complication rates have been found to be correspondingly higher than in older children. In a group of children under 10 kg, one group reported an average of 0.32 catheter changes per year, a mean rate of line infection of 0.14/patient-year with the average lifespan of a CVC for hemodialysis of 13 months [10]. In the under 2s from the author's center [11], around 70% of both groups on hemodialysis and peritoneal dialysis required catheter removal for reasons other than end-of-use, emphasizing the challenges of maintaining CVCs in babies and the importance of counseling families when embarking on renal replacement treatment, especially in small children.

Complications

Bleeding at the time of operation can usually be controlled by local measures, although it is reasonable to have a saved blood group. With ultrasound localization, the risk of hemo- or pneumothorax should be low, but a plain chest radiograph is recommended after Seldinger technique venous cannulation. Occasionally, passage of the line towards the heart can be difficult. This is particularly so for left-sided access or with redo lines. For redos, it is reasonable to confirm patency of the central veins by Doppler ultrasound; a formal venogram is rarely required, although may be indicated by clinical features of central vein occlusion (such as prominent chest wall veins, or plethora, or chronic limb swelling) with equivocal Doppler studies. Magnetic resonance venography may be helpful. Flow may be poor in the line which may be due to tip position (in the SVC), occasionally because of an acute angulation in the subcu-

taneous tunnel, or may be due to thrombus. Infection associated with the line may supervene at any time. The latter two are the commonest complications of central venous catheters [12].

Preventing and managing complications

As mentioned, fluoroscopy is mandatory for line insertion, and it can be useful to manipulate the line under screening if it proves difficult. The line may bend or kink at the confluence of internal jugular and subclavian, or may take a route into the contralateral neck or arm drainage. Usually, perseverance with manipulation under screening is adequate to place the line. Placing small amounts of torque on the catheter during manipulation may be helpful, as may be the suspension of ventilation. Elevation or depression of a shoulder may alter the configuration of a jugular/subclavian confluence sufficiently to allow a line to pass. We have found a hydrophilic guidewire (Terumo UK Ltd, Egham, Surrey) to be useful in manipulation into the right heart. The introducer sheath and/or catheter can then be passed over the guidewire.

We prefer the jugular veins for access. Subclavian puncture is possible, but may lead to a higher risk of arm venous thrombosis, which in turn might affect establishment of an arteriovenous fistula [12]. We prefer to try and preserve the arm veins if possible for the sake of creating an arteriovenous fistula later in life. If the inferior vena cava needs to be used, the same principles of access apply, although the subcutaneous tunnel is often very awkward as it either crosses the hip joint, putting the line at risk of kinking, or requires the line exit site to be on the thigh which can be obtrusive and uncomfortable. Our preference is the left saphenofemoral junction or femoral vein, as the preferred site for the transplant kidney is on the right.

Where central veins become very narrow (internal jugular vein stenosis is seen after 10% of insertions, subclavian vein stenosis in over 40% [13]) or obliterated, with so-called "end stage" access, there may be a role for specialist interventional vascular radiology for access [14].

Line sepsis should be managed in the first instance with appropriate antibiotics, and most infections can be cleared in this way [15], although clinical deterioration or failure to dialyze will necessitate removal of the line.

If dialysis is difficult because of poor flow (usually with high venous pressures), it is useful to confirm the tip position by means of plain radiography. A relatively "fresh" line (within a few days of its insertion) can be withdrawn

safely with appropriate analgesia or sedation, if improved flow can be obtained in this way. A contrast study may also be helpful, in demonstrating line thrombosis, and venous run-off. Thrombolytic line locks (e.g. urokinase, or tissue plasminogen activator) may be useful, although more extensive thrombosis sometimes requires thrombolytic infusion (e.g. tissue plasminogen activator) with repeat contrast imaging to document progression.

If a line requires revision within a few days of insertion the neck may be reopened and the line manipulated aseptically. We have found this generally unrewarding, however. Revising a line within 4 weeks or so of its insertion requires dissection of the vein for control above and below the venotomy. In an older line, when there is an established tract, it is reasonable to control the tract, and replace the line directly, or over a guidewire according to the surgeon's preference.

Symptoms or signs indicating a limb venous thrombosis require removal of the catheter; venous hypertension may warrant consideration of removal.

Other modes of hemodialysis access

As mentioned above, other modes of hemodialysis require needle puncture access, although they may be suitable for older children for whom hemodialysis will be required for a prolonged period (e.g. if sensitized after a failed transplant), or in transition-to-adult units where these modalities are more commonly encountered. The preferred technique is the arm arteriovenous fistula (AVF). If reasonable vessels exist at the wrist, it is advisable to fashion the anastomosis there, as this preserves the elbow brachiocephalic site for a later date. The fistula may fail to mature or thrombose later. Pseudoaneurysms may also form which require the access site to be abandoned. Generally, AVFs are fashioned in the non-dominant arm, proceeding distal to proximal. If no native vessels are suitable for arteriovenous anastomosis, then a prosthetic loop graft may be used. These are most commonly used in the thigh. Graft thrombosis, aneurysm formation, bleeding, and stenosis at the anastomosis of graft and native vessel may occur.

Peritoneal dialysis access

Technique

As with hemodialysis access, catheters for peritoneal dialysis (PD) may be intended for short- or longer-term

use. The latter are usually cuffed and tunneled to an abdominal wall exit site. There are three broad techniques for insertion:

- Closed (percutaneous).
- Open (minilaparotomy).
- Laparoscopic/laparoscopic assisted.

The principles of the open and laparoscopic techniques are broadly similar but differ in terms of catheter fixation. This will be discussed later. In the setting of acute renal failure in a sick child, a catheter can be placed on the ward under sedation using local anesthetic. A needle puncture is made into the peritoneum (aspirating to ensure no visceral injury has occurred) in a similar way to the passage of a Verres needle for laparoscopic surgery, and a guidewire passed. After dilatation of the tract, the catheter is passed into the abdomen and flushed to ensure adequate influx and efflux of dialysate. Peritoneal dialysis (PD) may be commenced immediately. It is possible to tunnel the line if desired, but the usual indication for the percutaneous technique is for acute, short-term PD.

The open and laparoscopic techniques require general anesthesia. In the open approach, a small incision is made above the umbilicus, and the catheter placed into the pelvis. Many surgeons choose to perform an omentectomy to lower the potential risk of catheter entrapment and failure of dialysis [16]. The abdomen is closed and the catheter tunneled in a gentle curve to a suitable site on the abdominal wall. Again, as the preferred site for placing a transplant is in the right iliac fossa, it is usual to tunnel the PD catheter to the left iliac fossa.

The laparoscopic approach uses one or more ports to place the PD catheter, again with or without an omentectomy. At the author's unit, we prefer the laparoscopic-assisted approach described by Najmaldin [17]. The essence of the operation is a single supraumbilical incision, through which an omentectomy can be performed (in children the greater omentum tends to be flimsy and easy to manipulate through a very small hole), and a laparoscope inserted. A needle and guidewire is then introduced to create a long extraperitoneal tunnel, through which a peel-away sheath is passed. The PD catheter is then seen to pass under direct vision into the true pelvis, fixed by a long extraperitoneal tunnel to avoid flipping (Figure 31.3). The catheter can then be tunneled in the usual way, and the laparoscopy port closed. PD catheters are available in a variety of configuration. We have used coiled double-cuffed

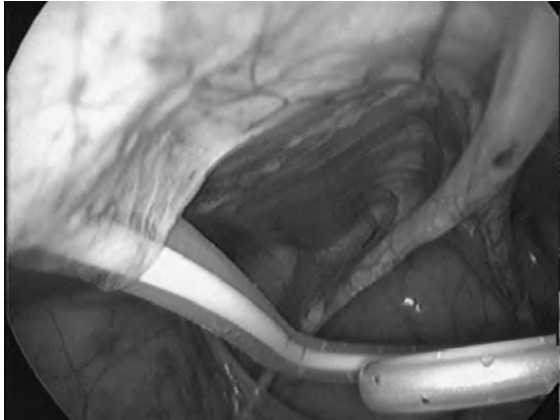


Figure 31.3 Intraoperative view of a peritoneal dialysis catheter, showing the position of the coil, and fixation by an extra-peritoneal tunnel.

tubes; a variety of sizes are available, including very small tubes suitable for neonatal use (Figure 31.4). Laparoscopy allows a more versatile approach if concomitant operations are required. We have used laparoscopic assistance for insertion of feeding gastrostomies at the same time as placement of a PD tube [18].

Outcome

Dialysis can generally be commenced on the same day as operation if desired. This is useful for PD in acute renal failure, or the rapid establishment of PD in a child who becomes dialysis-dependent quicker than

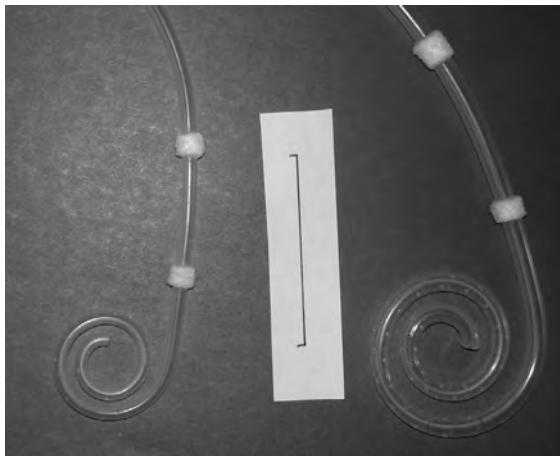


Figure 31.4 Coiled PD catheters suitable for neonatal use. The marker shows 5 cm.

anticipated. Catheters used early (within days) tend to have more mechanical problems, of which leak is the most important [19,20,21]. On average, the frequency of problems with PD seems to be of the order of one episode per 6 months of dialysis [22]. Peritonitis, exit-site and tunnel infections, and catheter occlusion make up the majority of the problems. Catheter survival is of the order of 80% at 12 months, 60% at 24 months and 35% at 48 months [22,23]. Younger children (less than 2 years old) have increased risk of catheter removal for problems [23,11].

A team approach is crucial in all aspects of chronic kidney disease management, and dialysis is an exemplar. Infection-related outcomes were seen to improve markedly with the introduction of specialist nursing care for the PD population [24].

Complications

The presence of adhesions complicating previous surgery may make PD ineffective or impossible. Nonetheless, previous surgery is not necessarily a contraindication to PD [25]. Bleeding or infection may occur early. Leak of dialysate may be seen early, and may necessitate suspension of PD. One innovative solution to dialysate leak has been the use of fibrin glue [26], although this is not widespread. Exit site infection or wound infection likewise may be seen early. PD peritonitis, characterized by cloudy (or fibrinous) PD effluent, pain and fever may occur at any stage and, according to culture, may or may not be resolvable with antimicrobials. Presentation with an acute abdomen can complicate PD, and sometimes the differentials (including acute appendicitis) can be difficult to exclude.

Dialysis (clearance) may become ineffective, reflecting peritoneal failure, or the child may have symptoms or fail to drain in or out. Fibrin sheath formation can occur causing a flap-valve effect. The tip of the PD catheter can migrate, or become entangled with intestine, omentum (if not excised), or ovarian fimbriae [22,27]. Occasionally, the subcutaneous cuff can erode through the skin.

Preventing and managing complications

Bleeding should be manageable by local control. One exception to this is the rare occurrence of visceral or blood vessel injury arising from closed technique puncture of the peritoneal cavity. Surgical exploration is mandatory if there is a suspected intra-abdominal

injury. It is important to ensure that the field is relatively bloodless: clots may occlude the PD catheter and interfere with dialysis. Care needs to be taken when tunneling the catheter, to ensure that the tube remains in the fat plane and does not transgress muscle. Laparoscopy has the advantage of allowing a thorough peritoneal inspection after the catheter has been inserted. Bleeding of sufficient magnitude to need re-exploration should be rare.

At our institution, we have always undertaken omentectomy. There is evidence regarding its efficacy [28,16], but in children, the greater omentum's mobility and flimsiness makes its removal a sensible step to obviate the risk of tube entanglement.

Dialysate leak is eminently manageable by lowering exchange volume, or suspending PD temporarily. A small volume leak may be inconsequential, especially if PD is crucial because of uremia or hyperkalemia. Our early experience mirrors that of others [29]: PD can be commenced satisfactorily within hours of insertion, rather than allowing the catheter to "rest" for days.

Exit site, wound infection or PD peritonitis should be managed conservatively in the first instance, according to culture. Intraperitoneal heparin is sometimes useful if the effluent is fibrinous. Rarely, fibrinolytics (e.g. urokinase) may be used. One important culture is fungal. Almost without exception, a PD catheter must be removed in the presence of fungal infection: even prolonged antifungal treatment is very unlikely to clear the organism. Likewise, recurrent bacterial infection may indicate revision of the catheter. Ideally, if a PD catheter is removed as a consequence of infection, it is prudent to wait a number of weeks prior to reinsertion.

Clarifying tube position, and fixation is one real advantage of the laparoscopic-assisted approach. It allows a thorough inspection of the peritoneum, and adhesiolysis if needs be, and the long extraperitoneal tunnel makes tip migration unlikely. It allows very accurate position, under direct vision, of the tip of the catheter in the true pelvis.

If PD becomes ineffective, or inflow and outflow are poor or symptomatic, a plain radiograph reveals the orientation and tip position. A contrast study may be helpful in demonstrating free flow (if a fibrin sheath or loculation is suspected). If the catheter has migrated or flipped out of the pelvis, under fluoroscopy the PD catheter can be manipulated by means of a guidewire.

An unwell child, with abdominal symptoms and signs particularly those of intestinal obstruction, might indicate the presence of encapsulating sclerosing peritonitis. This can occur even after the removal of a PD catheter, can be associated with significant morbidity or mortality, but the latter in children is mercifully rare [30,31].

Laparoscopic exploration of a malfunctioning catheter can be helpful [32], certainly if there is no associated infection. The tube can be released if it has become entangled or encased with fibrin. If excessively mobile, it can be looped within a suture placed at the dome of the bladder to further fix the tube (we have not seen this in laparoscopically-placed tubes on account of the fixation afforded by the extraperitoneal tunnel). If the catheter requires revision with a new catheter, it is straightforward to use the peel-away Seldinger technique to create a new extraperitoneal tunnel.

Extrusion of the cuff is uncommon, but may be expected if the cuff is at the exit site, or if the tract is just under the skin rather than in the fat plane (or if the fat plane is attenuated, as in a neonate). Occasionally, chronic infection is seen in association with the cuff, rarely overgranulation or a pyogenic granuloma. A conservative approach to preserve the tube is to shave the cuff down to the level of tube (done simply with a regular blade) although there is a risk of tube puncture. If the symptoms associated with the cuff are refractory, or the tube is breached, a revision is required.

Conclusion

Whilst the gold standard of renal replacement therapy is a successful kidney transplant, dialysis is an important mode of therapy. In very small infants in whom a decision has been made to treat but renal replacement is required in the workup to transplantation, dialysis is crucial but may present formidable challenges in this unique patient group. Many families opt for PD, and preservation of vascular access is one advantage en passant since these patients will almost inevitably require more intervention later in life. Nonetheless, dialysis is limited by peritoneal failure and by loss of central veins. The concept of "end-stage access" is very real and a cause of significant morbidity, or death. This is a major driving force towards early transplantation.

DOS AND DON'TS**Do**

- Involve a multidisciplinary team in planning and counseling families for renal replacement therapy. Specialist nurses are key. Growth and nutrition are important
- Plan dialysis with transplantation in mind. Try to avoid lower body vessels for hemodialysis. Try to avoid the right side of the abdomen for peritoneal catheter exit sites
- Involve interventional radiology services for difficult or revision vascular access

Don't

- Use the subclavian veins preferentially for hemodialysis access
- Forget about arteriovenous fistulae and grafts for hemodialysis, especially in older children and those about to transfer care to adult units

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CHAPTER 32

Kidney transplantation

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KEY POINTS

- Transplantation is the gold standard renal replacement treatment
- Living donor kidneys are preferable
- Pre-emptive transplantation is ideal. Avoiding dialysis preserves access sites and may prolong graft life
- Uropathies are disproportionately represented in the etiology of pediatric end-stage kidney disease. Pretransplant urological workup is therefore mandatory
- An abnormal urinary tract demands vigilance, but can be a safe means of drainage
- Nonadherence with medication is commonplace and contributes to graft failure, especially in teenagers. Transition to adult care needs careful attention

Introduction

A successful kidney transplant is the gold standard renal replacement therapy. Although over time there has been a trend toward longer graft survival [1], a child who receives a transplant will almost inevitably require further renal replacement in due course. Pre-emptive transplantation is the counsel of perfection: avoiding dialysis may confer a survival benefit to the graft [2–4], preserves access sites for dialysis and, if native function can be preserved to some extent, can avoid metabolic, biochemical, and fluid balance problems, and psychosocial issues associated with chronic hospitalization. At present, between 20% and 30% on average children receive a pre-emptive kidney transplant in the United Kingdom and North America.

There is inevitably a period of medical and surgical workup before transplantation, and these issues (including matching, virological and immunization protocols) are reviewed extensively elsewhere [5–10].

A major rate-limiting step in kidney transplantation is the availability of an organ from a deceased or living donor. Live donor programs introduce an additional

element to workup in that stringent donor workup is necessary to ensure fitness for donation and assess in detail vascular anatomy. Organ allocation systems prioritize children [1,11]. The current median wait on the deceased donor list for a kidney in the United Kingdom (pediatric) is 354 days [11], which has doubled over the past 10 years. In the USA in 2006, stratified by age cohort, the waiting time was 360, 430, and 569 days (ages 1–5, 6–10, and 11–17 years, respectively).

Urological workup of the recipient

Structural or functional urinary tract problems account for about 15% of the pediatric transplant population [1]. An abnormal urinary tract's effect on native kidneys is well documented [12]. A urethral leak point pressure of greater than 40 cmH₂O represents an “unsafe” lower tract. There is a suggestion that, in children, a “safe” pressure may be 30 cmH₂O or less [13].

Pretransplant urodynamics are mandatory in children with a uropathy. In the author's unit, we extend the indications for urodynamics to include those children

with lower urinary tract symptoms, known vesicoureteric reflux and recurrent urinary infections.

The commonest single uropathy in the transplant population is posterior urethral valves (PUV). A paradoxical situation in PUV is that urodynamics change, probably evolve, over time [14]. Thus, it is more appropriate to undertake urodynamic studies in the immediate time before listing for transplantation rather than earlier.

With the urodynamic findings in mind, appropriate measures can be implemented to lower pressures, correct reflux if necessary and provide capacity and drainage.

VUR is a risk factor for urinary tract infection (UTI). UTI is the single commonest infectious complication to follow a kidney transplant, and is commoner still in children with an underlying uropathy [15]. If recurrent UTI is a problem, it might be reasonable to consider an antireflux procedure. It is crucial to consider the contribution to overall capacity that might be made by dilated ureters. Of course, dilated ureters can be useful as a means of augmentation cystoplasty.

In boys with recurrent UTI, circumcision alone [16] may lower the subsequent risk, or may be combined with another procedure.

Pretransplant native nephrectomy is indicated for a number of reasons, including malignancy, refractory hypertension, and refractory protein loss. Recurrent infection, stones, and high-grade VUR have also been included amongst the indications, but a recent review of the author's center's cumulative practice has identified a trend towards more conservative management of the native kidneys [17]. The arguments for preservation of the native upper tracts include more straightforward fluid management of patients with ERF, preservation of native biochemical and endocrine function.

Technique

The kidney is prepared on the backbench before induction of the recipient's anesthesia, to ensure that the organ is usable. The vessels are dissected toward (but not into) the renal hilum, taking care to preserve perinephric tissue associated with ureter (shown schematically in Figure 32.1). Multiple vessels need to be given particular attention, as there may be an increased risk of thrombosis, although one multivariate analysis implicates donor atheroma as a risk factor, rather than

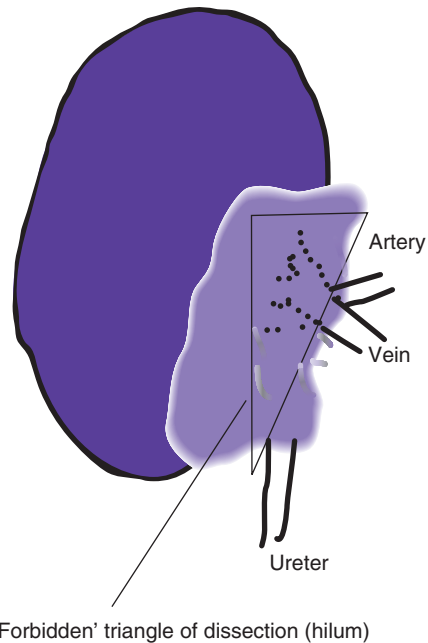


Figure 32.1 Forbidden dissection.

multiple vessels per se, and that reconstruction does not necessarily disadvantage the graft [18,19].

Lower pole arteries are important to preserve predicting that they supply the ureter. Small upper polar vessels can be tied safely. Where multiple vessels have been retrieved on a common vessel patch (Carrell patch), this can be used for the donor–recipient anastomosis, or the donor vessels anastomosed back-to-back or end-to-side (Figure 32.2) in a common anastomosis

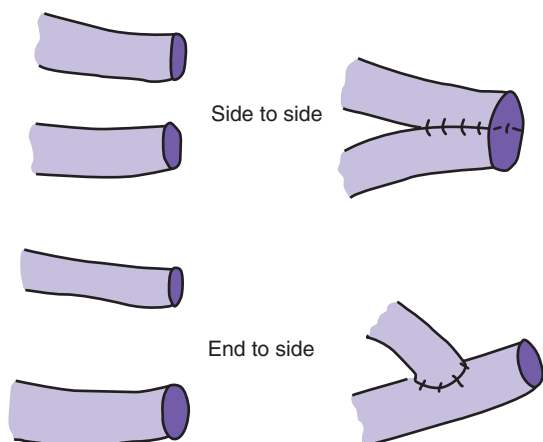


Figure 32.2 Side-to-side and end-to-side vessel reconstruction.

with the recipient vessel. It may be more convenient to do separate anastomoses of multiple vessels although this reintroduces the risk of thrombosis. For the venous drainage, a single large donor renal vein is sufficient, and accessory veins may be tied safely. However, vascular reconstruction of the renal veins is reasonable if circumstances dictate.

It is common practice to take a pre-implantation biopsy. The recipient undergoes general anesthesia and a central venous line and urinary catheter are placed. Induction antibiotics, immunosuppressive agents may be given at this point.

The choice of incision and approach for kidney transplantation is largely a matter of choice and experience. It has been commonplace to use a curved iliac fossa incision (modified Rutherford Morrison), approaching the iliac vessels extraperitoneally. This is especially useful if the recipient is on peritoneal dialysis, since in the event of delayed graft function (DGF), dialysis can be continued. According to the habitus of the child, the incision can be elevated into a “hockey stick.” For infants, some surgeons prefer a transperitoneal approach, via a midline incision. This affords space, but has the potential disadvantage of transgressing peritoneum and can make subsequent access to the graft (e.g. for biopsy) difficult. In an extraperitoneal approach, the abdominal wall muscles may be cut, or mobilized in the pararectal plane, which we have found gives very good access to the retroperitoneum. The inferior epigastric pedicle is identified, and usually tied and divided (unless required for complex vascular reconstruction). The spermatic cord in boys must be identified, preserved, and retracted. The round ligament in girls can be divided. In the transperitoneal approach, the right colon and terminal ileum are mobilized to give access to the inferior vena cava (IVC) and aorta.

In infants and small children (up to approximately 20 kg), the aorta and inferior vena cava are the preferred recipient vessels, and provide high-flow conduits. For ease, the arterial and venous anastomoses are usually separated by a centimeter or two.

In recipients with vascular anomalies, or thrombosed venous drainage, the surgeon may need to be creative. A good “road map” of potential anastomotic sites is useful, and in the author’s unit we have mandated that all recipients with previous cannulation of the lower-body circulation or intra-abdominal surgery have detailed vascular imaging. Doppler ultrasound is straightforward and

relatively noninvasive. Reconstructed computed tomography (CT) is useful but has a radiation dose, and can require contrast which might affect native renal function. Magnetic resonance angiography is an alternative. We have found, anecdotally, that where venous drainage is adequate (whether through native “anatomical” vena cava or via collaterals), the transplant renal vein drains adequately. Small published series have articulated these issues [20,21]. Prothrombotic states or anatomical factors might make postoperative anticoagulation advisable.

Vascular anastomosis is achieved using a continuous 5/0 or 6/0 nonabsorbable monofilament, taking care not to take the back wall of the recipient vessel inadvertently. It is useful to place curved bulldog-type clamps to the donor renal vessels to test for and repair anastomotic leaks before reperfusion. The warm ischemic time for the kidney is noted when the kidney is reperfused. Bleeding not detected at the time of anastomotic test can be investigated and dealt with at this point.

The commonest mode of urinary drainage is transplant ureter to native bladder (ureteroneocystostomy, UNC). Running an irrigant through the bladder catheter distends the bladder to make it more easily identified, and additional reassurance can be sought by the use of dilute methylene blue. Particular care is needed in patients on peritoneal dialysis. The peritoneum is often very thick, there is often clear fluid within, and distinguishing this from a urinary bladder needs care. There are many ways to perform UNC: extra- or intravesical, stented, or unstented [22–24]. The use of a stent is debatable, but wound drainage advisable.

If the native ipsilateral ureter is easy to identify, or the bladder is hard to distend or mobilize, a ureteroureterostomy may be used. There is no need to divide the native ureter.

After hemostasis has been assured, a drain is left, and the wound closed according to the surgeon’s preference. Occasionally, especially in very small infants, the abdominal wall can be difficult to close primarily. In these children, it is reasonable to return after a few days for secondary closure of the abdominal wall (having closed the skin at the first operation). Some surgeons advocate primary prosthetic patches [25].

Adequate maintenance of the recipient’s central venous pressure and blood pressure makes high dependency or intensive care mandatory. Hypovolemia and hypotension are risk factors for thrombosis and DGF. They have also been shown to be independent risk

factors for “slow” graft function, represented by a slower than expected fall in creatinine post-transplant [26].

Subsequent recipient management is very individualized to units, and probably best managed on a protocol basis according to local preferences.

Outcome

Ninety-five percent of grafts function at one-year post-transplant. Living donor kidneys have marginally better survival [1]. Graft survival is poorer in the under-5s, plateaus into the early teens, then fall until the early 20s [27]. Overall, there has been a trend to increasing graft survival over time (Table 32.1).

In the 2010 NAPRTCS review [1], 2.6% of grafts had primary nonfunction. DGE, which specifies the need for dialysis within the first week post-transplant (and probably impacts on graft longevity) occurred in 5.2% living donor kidneys and 15.6% deceased donor kidneys.

About 50% of graft failure is accounted for by rejection. Acute rejection was reported to account for 13% graft failure in the NAPRTCS review [1]. There has been an improvement in acute rejection rates since the 1990s (Table 32.2). Although immediate immunological events are uncommon, acute rejection is common, with a fine balance to be made between the risk of this and the risk of overimmunosuppression. The diagnostic gold standard is transplant biopsy, but frequently in pediatric practice, an empirical course of high-dose steroids is often given for a deterioration in graft function. There is a diverse collection of conditions which can cause more prolonged and chronic deterioration of transplant function.

Table 32.1 Graft survival (%) at 1, 3 and 5 years post-transplant according to year of transplant and organ source.

	LD			DD		
	1 year	3 years	5 years	1 year	3 years	5 years
1987–90	89	81	75	75	63	55
1991–94	92	85	80	85	76	70
1995–98	94	91	85	91	82	74
1999–02	96	92	87	93	84	79
2003–10	97	92	84	95	84	78

Adapted from NAPRTCS Annual Report 2010 [1].
DD, deceased donor; LD, live donor.

Table 32.2 Twelve-month probability (%) of first rejection by transplant year.

Year	LD	DD
1987–1990	54	69
1991–1994	45	60
1995–1998	33	41
1999–2002	22	27
2003–2006	13	17
2007–2010	9	17

Adapted from NAPRTCS Annual Report 2010 [1].
DD, deceased donor; LD, live donor.

Kidney recipients with underlying uropathies seem to fare as well as those without. A survey of units in the UK [28] returned a cohort of 74 children (a total of 78 transplanted kidneys) with a spectrum of underlying urological abnormalities (the largest group being 39 boys with PUV) with a median follow-up of 36 months. Of 57 grafts that could be followed, 25/57 had recurrent UTI (44%). Eighteen grafts had deteriorating function and 15 were in boys with PUV. The cause of graft deterioration in PUV children was multifactorial, although the survey highlighted the importance of pretransplant urodynamics in this group. It is unlikely that any single factor can be amended to avoid graft failure in boys with PUV, although one study suggests that more conservative initial management of the boys with PUV as opposed to aggressive surgical intervention might be beneficial in terms of graft function [29].

Published series have reiterated that transplantation into abnormal unoperated and reconstructed urinary tracts is safe [30–36]. Reconstructive surgery is more commonly done before transplantation [37] and this obviates the anxiety about reconstructive urinary tract surgery in the face of immunosuppression. Some authors point out that the so-called “dry augment” can be problematic [38], and care should be exercised in reconstructing the urinary tract of an anuric patient. As always, the decision to undertake major reconstruction needs to depend on the support for teaching and learning intermittent catheterization and bladder irrigation, and carer vigilance for complications such as stones.

A peculiarity of pediatric transplantation (in common with pediatric practice in most patients with chronic

disease) is transition into adult medical care. Nonadherence to treatment (immunosuppression as well as other treatment regimens including catheterization) is one of the major problems facing the adolescent recipient. Graft longevity has been observed to drop during adolescence [27], and there is common consensus that transition is, at best, a difficult time [39].

Complications

Bleeding is usually an immediate or early event that is obvious clinically. Venous thrombosis occurs in up to 7% and arterial in up to 7.5% of recipients [40]. Although thrombosis can be immediate, it may manifest after several days, with loss of function or urine output, graft tenderness, pain, and fever. Occasionally, a venous thrombosis can present with graft rupture and torrential hemorrhage. Urgent assessment of graft perfusion (with Doppler and/or isotope renography) is mandatory.

Later on in the postoperative course, renal artery stenosis can occur causing graft deterioration, and usually hypertension. Immediate immunological complications (e.g. accelerated rejection) are very rare with detailed immunological pretransplant workup, modern donor–recipient matching, and evolving immunosuppression regimens.

Ureteric complications occur in up to 10% of transplants [41–43]. Urinary leak may present early, with a wound leak or swelling around the graft. It can be sufficient to cause obstruction (urinary and vascular: to the graft and ipsilateral leg). Lymphocele can also present thus, and ultrasound can be useful. Ureteric stenosis may manifest as deteriorating graft function secondary to obstruction, and a hydronephrosis may arise. In one series of modified Lich–Gregoir UNC [42], 1% had anastomotic obstruction, responding well to stenting for several weeks. In one series of uretero–ureteric anastomoses [43], 8.4% had a ureteric complication (14/166). Ultrasound, excretion renography, and antegrade pyelogram may be useful in the diagnosis of ureteric stricture. Reflux into the graft may manifest as dilatation, or progressive scarring and deteriorating function if associated with UTI. Stones may occur in the graft. If a ureter has been stented, particular note needs to be taken to ensure its timely removal: retained stents may become encrusted.

Infection is common in a transplant recipient, and UTI is particularly common. In uropaths, urinary prophylaxis is a reasonable step because of this. Other

infections as a consequence of immunosuppression demand vigilance.

Late deterioration of graft is very common, but multifactorial, encompassing the diagnostic potpourri that is interstitial fibrosis and tubular atrophy (IF/TA). Calcineurin inhibitors are well-recognized culprits, and careful thought needs to be given to ongoing immunosuppression. Recurrent UTI and lower urinary tract dysfunction are also important to consider. One of the biggest challenges in pediatric transplantation is the issue of nonadherence with medication.

Preventing and managing complications

As with all vascular anastomoses, technical problems are common causes of failure. Positioning the kidney to allow comfortable anastomoses and to allow the donor vessels to lie without kinks is important. Intact vessel ends are important to avoid inclusion of adventitia within an anastomosis, and to avoid an intimal flap. Postoperative anticoagulation may be considered on the basis of a pre-existing prothrombotic state, if multiple vessels are present or if there has been perioperative hypotension. One report suggests that thrombotic risk might be lowered by the administration of interleukin-2 antagonists [44].

Ongoing bleeding after implantation requires re-exploration. Missed hilar vessels during bench preparation can be tied once identified. Thrombosis may go undetected for hours. There are donor and recipient factors for thrombosis. Traditionally, kidneys from very small donors have been considered to be at higher risk of thrombosis, although a recent paper describing a single center's experience of en bloc transplants from mean 11.4 kg donors may challenge this [45]. Donor age above 60 years may also increase the risk of thrombosis, although this may reflect atherosclerosis, itself a risk factor for thrombosis.

Recipient risk factors are difficult to tease out but may include small age (or size) per se, a previous thrombotic tendency, technical problems with the anastomosis, and perioperative hemodynamic instability. The latter are crucially important for very small recipients.

Thrombectomy can be attempted but seldom seems to salvage the situation. Later deterioration in graft function (usually associated with hypertension) can be due to renal

artery stenosis, which is usually just distal to the anastomosis, and is amenable to catheter balloon angioplasty.

Lymphocele can manifest by prolonged wound drainage, perigraft or leg swelling, or deterioration in graft function. Small (or asymptomatic) lymphoceles can be managed expectantly. Symptomatic lymphoceles require drainage, either percutaneously, or by fenestration into the peritoneum. Meticulous vascular dissection during preparation of the recipient vessels is to be commended to lower the risk of lymphocele.

Ureteric complications are usually a consequence of ischemia of the transplant ureter. Attention to the “golden triangle” of the ureter’s blood supply during bench preparation has been described earlier, and at the time of UNC it is useful to demonstrate an active blood supply to the cut end of the transplant ureter. Diathermy should be used sparingly and with caution.

An early ureteric leak may manifest as wound drainage. The drain effluent should be analyzed to determine whether it is urine or serum. Adequate bladder drainage must be ensured, especially in small infants, and some early leaks can reasonably be watched for a day or two, as they may settle. The use of stents is debatable, and studies demonstrate advantages to both stenting and not stenting [22,23]. One factor in pediatric transplantation that may be important as a decision-maker is that a child almost certainly requires a general anesthetic for removal of a stent.

A ureteric leak may be managed by early re-exploration or by temporizing transplant nephrostomy and later planned exploration. The latter demands dissection through scarred tissue which in itself can be hazardous to the blood supply of the kidney. If there is sufficient viable donor ureter, a redo UNC can be fashioned. If there is insufficient viable ureter, the recipient native ureter can be used in the form of a ureteropyelostomy or ureteroureterostomy. With a capacious recipient bladder, a lateral bladder flap can be tubularized (Boari), or elevated to anastomose onto the transplant pelvis or ureter (similar in principle to the “bladder elongation psoas hitch” procedure [46]). Rarely, an enteric interposition is required, or drainage into a cutaneous stoma.

A ureteric stenosis may present late. Stenting is a good option, although balloon dilatation has been reported with success [47]. Obstruction or stenosis refractory to stenting or dilatation requires revision as described earlier, or long-term stenting with intermittent stent changes. Late operation for a ureteric complication is

often difficult and hazardous, as the allograft becomes encased in scar tissue.

Urinary infection is an important early problem with pediatric transplant recipients, particularly those with underlying uropathies, and strategies for the management of VUR in the native urinary tract have been outlined earlier in the chapter. There has been interest in graft VUR and its effect. One report, using baseline DMSA imaging, establishes a strong link between VUR, infection, and the acquisition of graft scars, and recommends initial UTI prophylaxis as well as vigilance for UTI [48].

Another suggestion of this study is antireflux urinary drainage. Many surgeons perform a modified Lich–Gregoir UNC as stated earlier. A traditional antireflux approach is the Leadbetter–Politano ureteric implantation, although this has been reported to have a higher incidence of ureteric complications [41], as well as the potential effects of an open bladder procedure. Uretero-ureterostomy is a potential consideration if the native ureters do not reflux. A potential “minimally invasive” approach mirroring that of native VUR is subureteric injection (commonly now with dextranomer/hyaluronic acid copolymer). Anecdotally this has been a valuable approach although obstruction and graft dysfunction have been reported, mandating caution [49].

Post-transplant care is as much multidisciplinary as that of pretransplant. Longer-term sequelae of immunosuppression such as infection and malignancy, CAN, disease recurrence in an allograft, blood pressure control, lipid and glucose control, etc. require close collaboration between nephrologists, pediatricians, pathologists, radiologists, and other allied professionals. Fundamental to pediatric practice is nutrition, growth, and development, which are paramount to pre- and post-transplant management. Some of these long-term issues, and others including quality of life, have been reviewed elsewhere [10].

Conclusion

Although a successful kidney transplant is the pinnacle of end-stage renal disease management, it remains merely a treatment rather than a cure. Graft survival is poorer at the extremes of childhood as well as the extremes of life, and the causes are multifactorial. Although the uropathies are represented disproportionately in the etiology of pediatric renal failure, graft outcome overall is as good

in this group, although particular attention is required in their pretransplant workup. Attention to detail in the operative procedure is a *sine qua non*. Transition into the adult services can be a trying time for patients and clinicians, and mandates particular vigilance of graft function. The transplanted child, with our current techniques of renal replacement therapy, becomes an adult who will

require further renal replacement. Pre-emptive transplantation preserves venous access sites and probably allows for improved graft survival. Living donor kidneys currently have better outcomes than those from deceased donor kidneys. Therefore, the gold standard for renal replacement in childhood remains a pre-emptive live donor kidney transplant.

DOS AND DON'TS

Do

- Plan treatment of chronic kidney disease with a transplant in mind. Optimize growth, nutrition, anemia and bone biochemistry. Plan dialysis access with care. Aim for pre-emptive transplantation
- Bear in mind that a pediatric transplant recipient will most likely require further transplant(s) and plan surgery (approach, vessels, dialysis access) accordingly
- Investigate the lower urinary tract thoroughly

Don't

- Assume that an abnormal urinary tract invariably needs reconstruction. Careful assessment and vigilance are the keys
- Overlook the importance of support and careful, clear transition to the adult services. This is a precarious time for a transplant recipient

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PART VIII

Urogenital Tumors

CHAPTER 33

Wilms tumor and other renal tumors

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KEY POINTS

- Preoperative imaging and recognition of vena caval or intracardiac extension of tumor allows for safe surgical planning
- Large tumors may distort vascular anatomy. Adequate exposure of the renal vessels is crucial in order to avoid injury to the aorta and its major vessels
- The most common postoperative complication following surgery for renal tumors is small bowel obstruction

CLINICAL PROBLEM/CASE

A 4-year-old girl presents with an abdominal mass found on routine exam. CT scan demonstrates an 8 cm left renal mass and a 6 cm right renal mass. Both are solid lesions located centrally in the kidney extending into the renal hilum. There is no evidence of extrarenal extension and there are no chest metastases.

- 1 Would you advise surgical exploration?
- 2 If you choose to proceed with preoperative chemotherapy, what would be the duration of treatment?
- 3 What are the risks/benefits of renal-sparing surgery in a child with bilateral renal tumors?
- 4 What are the late sequelae of treatment in a child with bilateral renal tumors?

Introduction

With an annual incidence rate of 8 cases per million individuals and about 500 new cases in the United States per year, Wilms tumor is the most common renal malignancy in children and the fourth most common childhood cancer [1–3]. The prognosis of children with renal tumors is excellent, primarily due to advances in modern chemotherapy. This is most notable for those patients with favorable histology Wilms tumor. Other renal tumors are not as responsive to adjuvant therapies and complete removal of the tumor remains the best chance for success. Even with the remarkable advances in survival from adjuvant therapy, surgery remains an integral part of the multimodality treatment of Wilms tumor. The surgeon has an important role in assessing the extent of the primary tumor, and how the

surgery is performed has an impact on the ultimate local tumor stage. This chapter reviews the most common complications associated with surgery for renal tumors of childhood and discusses the management of these complications. The majority of the literature available regarding complications of renal tumor surgery in children is from patients treated for Wilms tumor; however, the principles discussed below are also applicable to other renal tumors.

Surgical technique: general principles

Primary nephrectomy remains the procedure of choice for unilateral tumors. However, in the setting of a solitary kidney, bilateral renal tumors, or a history of predisposition syndromes (e.g. Beckwith–Wiedemann, WAGR,

Denys–Drash or Frasier syndrome, etc.), nephron-sparing surgery has taken on a more prominent role. Partial nephrectomy or tumor enucleation may also be considered in select cases of unilateral Wilms tumor, although this has only been evaluated in small series of patients [4–6]. Large prospective studies will be needed to confirm that there is no difference in local recurrence rates, progression free survival, or overall survival in the treatment of low stage, clinically localized unilateral Wilms tumor compared to radical nephrectomy [7].

All patients should undergo imaging prior to surgery with either computed tomography (CT) or magnetic resonance imaging (MRI). These studies may provide important information regarding local extent of tumor (e.g. enlarged lymph nodes, extension of tumor into adjacent organs) but prospective correlation with imaging and surgical findings has not been reported. Modern-day imaging obviates the need for routine contralateral renal exploration prior to nephrectomy [8]. Ultrasound is often the first imaging study obtained in a newly-diagnosed abdominal mass. It can help differentiate between cystic and solid nature of the tumor, and can also exclude caval thrombus that may be present in 4% of patients [9]. If the ultrasound is not conclusive or if there is concern for suprahepatic or intracardiac extension of tumor thrombus, then an echocardiogram or MRI should be performed [10,11]. Chest CT is recommended to identify pulmonary metastases and can identify some lesions not visible on plain chest radiographs [12].

There are a variety of surgical approaches for performing nephrectomy. Extraperitoneal flank incision should be avoided as it does not allow for proper staging. A transabdominal transperitoneal approach allows for gross inspection and exploration of the abdominal cavity and a generous transverse upper abdominal incision is most often employed.

The type of surgical incision has not been shown to make a difference in terms of operative spillage or incomplete removal of tumor [13]. An increased complication rate has been noted with thoracoabdominal incisions, but this may reflect the selection of this approach for extremely large tumors that may have an inherent increased risk of removal. For patients with intracardiac extension of tumor, a midline abdominal incision with median sternotomy may be the best choice. Cardiopulmonary bypass is often needed in such cases.

The role of laparoscopic removal of renal tumors in children has been explored. Duarte et al. reported eight

patients with unilateral nonmetastatic Wilms tumor who had received preoperative chemotherapy prior to laparoscopic nephrectomy [15]. In this small series, there were no conversions to open surgery, no tumor rupture, and no postoperative complications. There have also been case reports on laparoscopic nephrectomy in untreated Wilms tumors [16,17]. Its use in this setting will be limited due to the large size of these tumors, risk of tumor spill during removal, and the need for accurate surgical staging that requires removal of the tumor intact. It may find a role in other renal tumors, but differentiation of Wilms tumor from other childhood renal tumors by imaging alone is difficult. Recently, the role of laparoscopic partial nephrectomy for Wilms tumor has been reported as well [18,19]. This approach remains controversial due to theoretical concerns regarding increased risk of tumor spillage and peritoneal or port site recurrences [20].

Several aspects of surgical technique should be emphasized. Gentle manipulation of tissue is crucial to avoid tumor rupture with spillage of tumor because of an increased risk of local tumor recurrence when this occurs in children with Wilms tumor [21]. Adequate exposure facilitates staging and allows for inspection of the abdominal contents. Lymph node sampling is essential for correct tumor staging, but radical node dissection is not warranted [22]. Early ligation of the renal vessels before manipulation is recommended and the renal artery should be ligated prior to the renal vein. This will avoid distension of the vein, which can dislodge the ligature. However, in many patients early ligation is not feasible due to the large size of the tumor that obscures the hilar area and the great vessels. It is imperative that the surgeon clearly identify that the vessel being ligated goes to the kidney to avoid inadvertent ligation of the superior mesenteric artery (SMA), contralateral renal vessels or even the great vessels.

Extension of Wilms tumor into the inferior vena cava (IVC) or right atrium, which occurs in 4% and 0.7% respectively, poses a challenge to the surgeon [9,23]. Obstruction of the hepatic veins may lead to ascites, hepatomegaly, or hepatic dysfunction. Atrial thrombus may present with hypertension or congestive heart failure. Despite adequate preoperative imaging, cases of intracaval or atrial tumor thrombus are still diagnosed intraoperatively. The difficulty of the operation is increased when the diagnosis is missed preoperatively.

In patients with IVC or atrial tumor extension, nephrectomy can be carried out prior to cardiopulmonary

bypass and removal of tumor thrombus. Luck et al. described their technique of performing median sternotomy and preparation for cardiopulmonary bypass followed by laparotomy and mobilization of the kidney and tumor, leaving the renal vein intact [11]. Atriotomy and removal of all gross tumor through the atriotomy and the renal vein is performed simultaneously. Removal of caval thrombus may be achieved by venacavotomy either with manual extraction or with a Fogarty or Foley balloon catheter. In rare cases of tumor, the thrombus invades the vena cava wall precluding thrombectomy. In this situation, cavectomy is a reasonable surgical alternative [9,24].

Complications

In this section we describe the recognition, management, and prevention of intraoperative and postoperative complications of nephrectomy for renal tumors of childhood. Management of complications should be individualized for each child based on the clinical findings.

The overall surgical complication rate of nephrectomy for Wilms tumor appears to have declined over time, but remains substantial. In a comparison of the complication rates from the National Wilms Tumor Study Group 3 (NWTS-3) (1979–1987) to NWTS-4 (1986 and 1994), NWTS investigators found the complication rate decreased from 19.8% to 12.7% ($P < 0.001$) [14]. Small bowel obstruction (SBO) and hemorrhage were the most common surgical complications (5.1% and 1.9% respectively). Factors associated with increased risk of surgical complications included higher local tumor stage, tumor diameter ≥ 10 cm, and intravascular extension into the IVC or atrium.

Preoperative chemotherapy may influence surgical complication rates by reducing tumor size. A report from the International Society of Pediatric Oncology (SIOP), where nephrectomy was performed after 4 or 8 weeks of chemotherapy, was associated with an overall surgical complication rate of 5% [25]. The most common complication in the SIOP group was SBO. A prospective comparison of complications in patients enrolled in the NWTS-5 and the SIOP-93-01 trials demonstrated the overall complication rate for the SIOP patients was 6.4% compared to 9.8% in NWTS patients ($P = 0.12$) [26] (Table 33.1). Of interest, one report from a single center comparing complication rates between

Table 33.1 Comparison of complication rates from the SIOP and NWTS-5 trials.

	SIOP-93-01	NWTS-5	P-value
Number of patients	360	326	
Complication rate	6.4%	9.8%	$P = 0.12$
Intraoperative tumor spill	2.2%	15.3%	$P < 0.001$
Small bowel obstruction	1.1%	4.3%	$P = 0.002$
Stage III tumors	14.2%	30.4%	$P < 0.001$
Resection of other organs	6.9%	15.0%	$P < 0.001$

patients undergoing primary nephrectomy compared to those having preoperative chemotherapy did not demonstrate a difference in surgical complications [27].

Intraoperative complications

Hemorrhage

In NWTS-4, 1.9% of children had blood loss exceeding 50ml/kg of body weight [15]. This was substantially lower than the 6.0% rate noted in NWTS-3 ($P = 0.0003$). Review of the SIOP-9 data showed a 0.33% hemorrhage rate (defined as blood loss over 50ml/kg) during post-chemotherapy nephrectomy [25]. In a prospective analysis comparing complication rates among patients enrolled in SIOP-93 and NWTS-5 trials, there was a significant reduction in rates of intraoperative hemorrhage in the group that received preoperative chemotherapy compared to the group who did not [26]. In addition to a reduction in tumor size following chemotherapy, there is decreased tumor vascularity and thus a decreased risk of intraoperative bleeding.

It is important to recognize that up to 8% of patients with newly-diagnosed Wilms tumor may have acquired von Willebrand disease and preoperative coagulation screening is needed [28,29].

Vascular injury

Iatrogenic injury to the aorta and its major vessels can occur during nephrectomy. Large tumors can distort the anatomy and the great vessels can easily be mistaken for the renal vessels. Vascular injury can be avoided by proper surgical exposure and identification of the aorta, IVC, SMA, celiac axis, and contralateral renal vessels. Inadvertent ligation of any of these structures could potentially lead to a devastating outcome. Mesenteric

vessels may be adherent to the renal hilum. The renal artery should be carefully traced and seen entering the kidney prior to ligation. The surgeon may choose to forgo early ligation of the renal vessels until after mobilization of the tumor if it allows precise identification of the renal vessels.

There are several reports of injury to the SMA; however, the actual incidence may be underreported [30]. All patients had left-sided tumor where the aorta and its branches lie in close proximity to the tumor, and it occurred in children under 5 years old. In each reported case of iatrogenic SMA ligation, the injury was identified intraoperatively and repaired. None of these children experienced adverse events related to the bowel or vascular anastomosis. The operative management of SMA injury depends on the type of injury. Options for repair include primary end-to-end anastomosis, anastomosis of the cut end of the SMA to the aorta in an end-to-side fashion, or possibly interposition graft if there is inadequate length for direct anastomosis.

Unexplained intraoperative hypotension or cardiac arrest should raise concern for unrecognized vena caval or intracardiac extension of the tumor [23]. The tumor thrombus can break off and embolize during the course of nephrectomy. The renal vein and IVC should be palpated prior to manipulation of the tumor to evaluate for the presence of intravascular tumor.

Postoperative complications

Small bowel obstruction

The most common postoperative complication after nephrectomy for Wilms tumor is SBO, occurring in 3–7% of patients [14,25,31]. The majority of cases of SBO occur within the first 100 days after surgery [31]. The most common causes of SBO are bowel adhesions and intussusception. Factors associated with increased risk of SBO include intravascular tumor extension, resection of other organs, preoperative tumor rupture, residual disease, stage III, and possibly tumor spill. Interestingly, patients who underwent small bowel resection did not have an increased risk of SBO compared to those who underwent resection of other visceral organs. There was no statistically significant difference in the incidence of SBO in patients who received radiation therapy compared to those who did not [31]. Some data suggest that SBO is more common among patients with

Wilms tumor compared to other intra-abdominal solid-organ malignancies [32].

As noted above, the incidence of postoperative SBO is lower when nephrectomy is performed after preoperative chemotherapy. The rate of intestinal intussusceptions was similar between the two groups, but there was a higher rate of obstruction secondary to adhesions in children undergoing primary nephrectomy [25]. One explanation may be that nephrectomy after preoperative chemotherapy requires less extensive dissection.

Surgical techniques thought to reduce the incidence of SBO include gentle handling of the bowel, maintaining a moist serosal surface of the bowel, and avoiding foreign materials on the bowel [31]. Despite adherence to these surgical guidelines, adhesions are thought to form from both inflammatory and ischemic processes.

Chylous ascites

Disruption or obstruction of lymphatic drainage can lead to chylous ascites. The actual incidence of chylous ascites is unknown [33]. Extended lymph node dissection is not recommended for Wilms tumor, although lymph node sampling is mandatory [21,22]. Avoiding extended lymphadenectomy may help prevent the formation of chylous ascites. Intraoperatively, care should be taken to ligate any disrupted lymphatics.

Increased abdominal girth and poor feeding should lead to the suspicion of chylous ascites [34]. Diagnosis is confirmed by evaluation of the white milky fluid obtained during paracentesis or exploratory laparotomy which will reveal a high triglyceride content 2–8 times as great as plasma, specific gravity greater than serum, and protein content >3 g/dl [35]. Radiographic imaging also plays a role in diagnosis of chylous ascites, although it may be difficult to differentiate chyle from hemorrhage on CT scan [34]. In the supine position, a fluid–fluid level may develop with the nondependent layer consistent with fat density.

Several treatment algorithms for management of postoperative chylous ascites exist in the adult literature, but may not necessarily apply to the pediatric population [35,36]. The initial management of chylous ascites should be conservative, including total parenteral nutrition (TPN) or a medium chain triglyceride (MCT) diet. Small lymphatic leaks usually resolve with conservative management. Occasionally, chylous ascites may require surgical intervention, such as direct ligation of leaking lymphatic channels or placement of peritoneovenous

shunt. Weiser et al. reported on nine children with chylous ascites following surgical treatment for Wilms tumor [33]. Seven patients were treated conservatively and completely resolved in 6–68 days (mean 26). The remaining two patients underwent exploratory laparotomy, one after failed conservative treatment and the other after presenting with increased abdominal girth and signs of an acute abdomen.

Outcomes

Surgeon experience

Surgeon experience may influence complication rates. Results from NWTs-4 suggested a lower incidence of surgical complications among pediatric surgeons and pediatric urologists than among nonspecialized general surgeons [14]. The prospective study comparing the NWTs and SIOP found a trend toward a lower incidence of complications in more experienced surgeons who had performed >6 nephrectomies for tumors in the previous two years [26].

Bilateral Wilms tumor

Synchronous bilateral Wilms tumor occurs in 4–6% of all patients with Wilms tumor [29]. Surgery for bilateral Wilms tumor has its own unique set of complications.

Horwitz et al. demonstrated a 15.3% complication rate in their series of 98 children undergoing renal sparing surgery [37]. In a review of 188 patients entered on NWTs-4, surgical complications were identified in 28 patients (14.9%) [38]. The most common complication was bowel obstruction in 14 children. They reported extensive hemorrhage in one patient, vascular injury in one patient, and visceral injury in two patients. Only three children had complications related to the renal collecting system. There were two patients with a urine leak and one child with urinary obstruction. Others have reported urine leaks in up to 30% of patients undergoing bilateral partial nephrectomy for Wilms tumor [39]. Urine leak can often be successfully managed with cystoscopic placement of a JJ stent.

Conclusion

Surgery remains an integral part of the multimodal treatment of Wilms tumor and other non-Wilms tumors of childhood. How the surgery is conducted has a great impact on tumor stage and therefore patient survival. Increased awareness of surgical morbidity has resulted in a decreased overall incidence of complications. Prevention of surgical complications starts with adequate preoperative imaging and sound surgical technique.

DOS AND DON'TS

Do

- Start with a generous transverse upper abdominal incision for the open surgical approach
- Ensure that all patients undergo imaging prior to surgery with either CT or MRI
- Consider nephron-sparing surgical approaches in patients at increased risk for renal failure (patients with a solitary kidney, bilateral renal tumors, or a history of predisposition syndromes)
- Remember that gentle manipulation of tissue is crucial to avoid tumor rupture with spillage of tumor
- Be conservative in the initial management of chylous ascites, including total parenteral nutrition (TPN) or a medium chain triglyceride (MCT) diet

Don't

- Make an extraperitoneal flank incision as it does not allow for proper staging
- Carry out contralateral renal exploration if the kidney is normal on CT or MRI
- Avoid extended lymphadenectomy as this may help prevent the formation of chylous ascites

CLINICAL PROBLEM/CASE: DISCUSSION

- 1 The child who presents with an apparent bilateral Wilms tumor does not need to undergo biopsy prior to initiating treatment. Biopsy is an option on the current Children's Oncology Group renal-sparing protocol. Biopsy is needed if the diagnosis is uncertain, but if imaging is typical for Wilms tumor then one can proceed with chemotherapy. Primary resection of the renal tumors at diagnosis is not recommended. If surgery is performed, only biopsy is recommended. Complete excision of the tumor is not advised at diagnosis unless there are very small tumors which are not present in this child.
- 2 The current Children's Oncology Group renal-sparing protocol recommends that chemotherapy be initiated with three drugs: dactinomycin, vincristine, and doxorubicin. The patient is treated with 6 weeks of chemotherapy at which time repeat imaging is performed to assess the feasibility of surgical resection. If there is less than a 50% response to chemotherapy, it is recommended that a biopsy be performed to determine the histology of the tumor. This will dictate if any change in chemotherapy is needed. If there is a good response to therapy (greater than 50% reduction in tumor burden), then one can deliver an additional 6 weeks of chemotherapy without performing biopsy. If partial nephrectomy or renal-sparing surgery is feasible at 6 weeks, one can proceed with surgery if it is felt that the majority of the kidney can be preserved. Surgery is indicated in all patients after 12 weeks of chemotherapy. Studies have shown that further chemotherapy is not likely to reduce tumor burden, nor facilitate renal-sparing surgery. Delaying treatment any longer might also increase the risk for development of anaplasia which has been shown to occur at an increased rate in patients with bilateral Wilms tumor.
- 3 Children that have bilateral renal tumors have an increased incidence of renal failure. Prior studies from the National Tumor Study Group have shown that patients with bilateral Wilms tumor have an incidence of end-stage renal disease approaching 15% at 20-year follow-up. For this reason, it has long been recommended that a renal-sparing approach be adopted for these patients. The potential benefit of preserving renal tissue and decreasing the rate of renal failure must be balanced against the increased risk of renal-sparing surgery in a cancer patient. These risks include having positive margins and leaving residual tumor, as well as intraoperative tumor spill. All of these can lead to an increased risk for tumor recurrence and increased mortality. Local recurrence in Wilms tumor is associated with a significant mortality rate.
- 4 As noted in the prior discussion there is an increased risk of end-stage renal disease in patients with bilateral Wilms tumor. In unilateral Wilms tumor, most patients that develop end-stage renal disease have known syndromes or diagnoses that are associated with a high rate of renal failure. These include patients with aniridia and Denys-Drash syndrome, but also patients with genitourinary anomalies. All of these patients have a WT-1 abnormality that predisposes them to both renal tumors and renal dysfunction. Bilateral Wilms tumor patients, even those not associated with syndromes, are at an increased rate of renal failure. The most common cause of renal failure in bilateral Wilms tumor is progressive tumor necessitating removal of both kidneys. That is why an aggressive protocol to preserve renal tissue and avoid nephrectomy is mandatory. That said, careful attention to surgical approach to decrease the risk of recurrence is also needed. Other late sequelae of treatment for patients with bilateral Wilms tumor are not different than those with unilateral tumors and these include second primary malignancies, cardiac dysfunction related to both chemotherapy and radiation, and fertility issues, in particular for those who receive radiation therapy.

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CHAPTER 34

Rhabdomyosarcoma and related pelvic tumors of the genitourinary tract

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KEY POINTS

- Fifteen to 20% of rhabdomyosarcoma (RMS) arises from the genitourinary tract
- Children's Oncology Group (COG) treatment protocol includes primary chemotherapy after initial biopsy, followed by surgery and radiotherapy for local control
- RMS of the vulva, vagina, and uterus are generally chemosensitive, and radical primary surgery is rarely indicated
- Whilst emphasis is placed on bladder preservation, a significant percentage of children may suffer from bladder dysfunction after treatment
- Adverse effects of treatment include surgical complications and chemotherapy/radiation toxicities

CLINICAL PROBLEM/CASE

A 13-year-old male presents with gross hematuria and urinary retention. Sonography reveals a bladder mass causing bladder outlet obstruction, and cystoscopic biopsy is consistent with rhabdomyosarcoma (RMS). There is no evidence of enlarged lymph nodes or distant metastasis on imaging.

- 1 What is the most likely histologic subtype?
- 2 What is an important initial goal of early management in this patient?
- 3 What is the primary mode of therapy for this patient?
- 4 This patient undergoes a post-treatment biopsy that demonstrates mature rhabdomyoblasts only. What is the recommended management approach?

Introduction

Rhabdomyosarcoma (RMS) represents the most common soft tissue sarcoma in infants and children, with approximately 350 new cases diagnosed in the United States each year [1]. Of these, 15–20% arise from the genitourinary tract, the most common sites being the prostate, bladder, and paratesticular regions, followed by the vagina and uterus. A bimodal age distribution exists, with a peak incidence during the first 2 years of life and then again during adolescence. Three pathologic subtypes have been recognized. Embryonal RMS represents approximately 80% of cases and has an

intermediate prognosis. Sarcoma botryoides is a variant of embryonal RMS that arises in hollow organs such as the bladder or vagina and has a more favorable prognosis. Alveolar RMS (ARMS) comprises 15–20% of cases, with the remainder of cases classified as undifferentiated tumors. Both alveolar and undifferentiated RMS carry an unfavorable prognosis. Recent molecular studies have demonstrated an association of ARMS with translocations $t(2;13)$ and $t(1;13)$. Translocation fusion-positive tumors comprise 80% of ARMS and are more aggressive. Fusion-negative ARMS may have a clinical course similar to embryonal RMS.

Presentation and evaluation

Bladder/prostate (B/P) primary lesions may present with urinary retention, urgency, or gross hematuria. Vaginal RMS can present as an introital mass or with vaginal bleeding, and paratesticular RMS typically presents as a painless scrotal mass [2]. Initial evaluation usually begins with ultrasonography; however, formal staging requires chest CT, CT or MRI of the primary site and regional lymph nodes, bone scintigraphy, and bone marrow aspiration. Ultimately, biopsy of the primary site is necessary to confirm diagnosis and provide histologic staging.

Staging and treatment principles

Pre-treatment staging is based on the tumor/nodes/metastasis (TNM) staging system and includes primary tumor site, tumor size (≤ 5 cm or > 5 cm), regional lymph node status, and metastatic disease. In combination with stage and histology, the Children's Oncology Group (COG) also assigns treatment based on residual disease following surgery [3]. Current COG protocols use staging and clinical grouping to stratify patients into low-risk, intermediate-risk, and high-risk categories. Treatment for RMS includes systemic chemotherapy for all patients and local control involving radiation and surgical resection for most patients. Vincristine, dactinomycin, and cyclophosphamide (VAC) currently remains the "gold standard" chemotherapy regimen for RMS. Outcomes for high-risk patients have historically been very poor, with $< 50\%$ of patients surviving 3 years [4]. Therefore, the COG protocol for high-risk patients includes the use of standard VAC in combination with additional multi-agent intensive chemotherapy.

Initial surgical management

Previous management of B/P RMS involved primary resection and/or exenteration combined with chemotherapy and radiotherapy. Development of multimodal treatments comprised of chemotherapy, radiotherapy, and bladder-preserving surgical approaches expanded and progressed with the aid of multi-center trials led by the Intergroup RMS Study (IRS). This approach shifted the treatment paradigm to primary chemotherapy and radiotherapy after initial biopsy, followed by surgery.

One of the initial goals of early management is preservation of renal function. Bladder outlet obstruction from B/P RMS or vaginal RMS is best managed with urethral catheterization. The use of suprapubic drainage has been associated with the potential for tract seeding. If ureteral obstruction is present, the preferred management is with internal stents. However, in the setting of tumor involvement of the trigone, percutaneous nephrostomy tubes may be required; these may be subsequently internalized.

One of the principal goals of therapy for bladder/prostate primaries is organ preservation. Initial management is with biopsy, delaying definitive surgery until after chemotherapy and/or radiation therapy has caused shrinkage of the tumor. However, if complete resection of the mass is feasible at the time of biopsy with preservation of bladder function, the tumor should be resected in its entirety. Sarcoma botryoides typically grows on a pedicle or stalk and may be amenable to complete resection with bladder preservation. Anatomically-favorable lesions located at the dome of the bladder are also potential candidates for up-front partial cystectomy [5]. The response of prostatic tumors to chemotherapy may allow prostatectomy to be performed with preservation of the urethra and bladder [6].

Endoscopic biopsy of the primary lesion is frequently attempted using a pediatric resectoscope or cold-cup biopsy forceps. Since the loop size of the pediatric resectoscope is small, multiple samples may be necessary to establish an accurate diagnosis. Cautery artifact can mimic spindle cell appearance to the inexperienced examiner or destroy the sample entirely. Use of low cutting current when taking a loop biopsy can limit cautery artifact. Alternatively, the loop can be used to cut out a wedge of tissue that can subsequently be retrieved with grasping forceps [7]. Biopsy should be performed with an experienced on-site pathologist who can evaluate frozen section specimens and guide the surgeon.

If no mucosal abnormality is apparent on endoscopy or if endoscopic biopsy is inconclusive, needle or open biopsy may be required. If laparotomy is performed for biopsy, preliminary evaluation of the pelvic and retroperitoneal nodes at or below the level of the renal arteries should be performed [6].

Paratesticular RMS should be excised via radical inguinal orchiectomy and resection of the entire spermatic cord. Resection of the hemiscrotal skin and contents is required in the setting of tumor fixation or

invasion. If the tumor has been inappropriately biopsied or excised via a transscrotal approach, the risk of local recurrence and non-regional lymph node spread is increased. A subsequent inguinal exploration with removal of the remaining spermatic cord and a partial hemiscrotoectomy, including the prior scrotal incision, must be performed [8]. Since CT scanning has a 14% false negative rate in the detection of retroperitoneal lymph node involvement, the current COG protocols recommend that children 10 years of age and older should undergo ipsilateral retroperitoneal lymph node dissection (RPLND) as part of their routine staging prior to chemotherapy. Wiener et al. [9] reported the outcomes of patients treated during IRS-III (1984–1991), in which all patients with paratesticular RMS underwent ipsilateral RPLND, compared with patients treated during IRS-IV (1991–1997), in which only patients with lymph node involvement identified on CT scan underwent RPLND. Overall, 3-year failure-free survival was 92% for patients treated on IRS-III and 86% for those treated on IRS-IV. Adolescents were at higher risk of retroperitoneal relapse than were children less than 10 years of age, and their failure-free survival and overall survival rates were worse.

Vaginal/uterine RMS is typically chemosensitive; therefore, these patients should be treated with initial biopsies followed by chemoradiation and subsequent resection as necessary.

Secondary surgical principles

Pretreatment re-excision

Initial biopsy is often done before establishing a malignant diagnosis. This frequently leaves gross residual tumor, microscopically positive margins, or unclear margin status. In these situations, pretreatment re-excision (PRE) is advised. PRE involves wide re-excision of the previous operative site including adequate margins of normal tissue. This procedure is done prior to administration of chemotherapy or radiation therapy. PRE has been found to improve failure-free and overall survival [10].

Second-look operations

Second-look operations are performed to confirm clinical response, evaluate pathological response to therapy, and remove residual tumor in patients who achieve clinical complete or partial response after chemotherapy or radiation therapy. In patients with a complete

radiographic response, confirmatory biopsy should be performed at the site of the original mass [6]. In patients with B/P and vaginal RMS, anterior exenteration with preservation of the rectum should be considered if residual tumor persists after therapy or if there is early failure or progression of disease after therapy.

Rhabdomyoblasts only on post-treatment biopsy

Maturation of rhabdomyoblasts after chemotherapy has been observed by various investigators, and their clinical significance has been called into question [11]. Atra et al. reported a group of patients with residual “rhabdomyoblast” that did not go on to relapse during observation [12]. Analysis of post-cystectomy specimens has also demonstrated rhabdomyoblasts along with a reduction in cellularity in patients treated with chemotherapy, suggesting that this pattern may be indicative of response to therapy. Ortega et al. followed six patients with post-treatment biopsy showing mature rhabdomyoblasts [13]. All patients remained free of disease after a follow-up period of 37–237 months. The authors emphasized the importance of correctly identifying mature cells as those with a large, smooth solitary nucleus, no significant pleomorphism, no mitotic activity, and the absence of cell clusters suggestive of growth from a common precursor [13]. In 2006, the COG published a report supporting observation for rhabdomyoblasts only on post-treatment biopsy. Failure after apparent tumor cell maturation on biopsy has been reported; therefore, careful observation of these patients is required [14].

Reconstructive surgery

Despite advances in therapy, certain patients ultimately require radical cystectomy or anterior exenteration as a component of treatment. Many reconstructive options exist, including various continent catheterizable pouches and orthotopic diversion. When necessary, vaginal reconstruction may be performed with skin grafts, rotational flaps, or colovaginoplasty [5].

The timing of definitive reconstruction is a critical issue in these patients. Intraoperative frozen section at the time of definitive resection can be difficult to interpret. This becomes a significant issue if up-front continent reconstruction is performed at the time of extirpative surgery. Lander et al. described the use of Le Bag

continent reconstruction in three children, one of whom was 26 months. Unfortunately, despite initial negative frozen section analysis, permanent sections revealed residual viable tumor, requiring local radiation and chemotherapy [15]. Similarly, Merguerian et al. performed reconstruction at the time of cystectomy in their patients, but they simultaneously cautioned readers that frozen section is an unreliable predictor of residual disease, as several of their patients had residual disease requiring adjuvant therapy or re-operation [16]. In addition, early reconstruction of irradiated tissues may lead to impaired healing and an increase in postoperative complications. Furthermore, the ultimate effect of radiation on the bowel may be difficult to assess at the time of the initial surgery. The authors' preference is to delay definitive continent reconstruction until a disease-free period of at least 2 years has been achieved and the patient is motivated and mature enough to perform self-catheterization [17].

In a patient with a history of pelvic radiation, choice of bowel segment for reconstruction should be carefully considered. Transverse or sigmoid colon may be utilized to avoid reconstruction with irradiated bowel segments. At the time of extirpative surgery, it should be assumed that any urinary diversion, whether continent or not, may be the first stage of ultimate functional reconstruction. Therefore, certain principles such as avoidance of incidental appendectomy and preservation of maximal ureteral length should be adhered to during the initial operation [18].

Surgical complications in general

Surgical management of RMS is associated with early and late complications. As expected, complication rates are higher amongst patients previously treated with radiation and chemotherapy [18]. Primary tumor resection is associated with potential bleeding, abscess formation, urinary tract infection (UTI), and ureteral and rectal injury [19]. Seitz et al. reported a 28.8% complication rate amongst patients undergoing secondary tumor resection after neo-adjuvant chemotherapy or radiation, including bleeding, bowel injury, fistula and abscess formation, ileus, peritonitis, UTI, ureteral stricture, and erectile dysfunction [19].

Management of postoperative complications may require secondary procedures, including lysis of adhesions, repair of fistulae, total cystectomy for bleeding or fibrosis, intra-abdominal abscess drainage, colostomy

for rectal stricture, augmentation cystoplasty for low-capacity bladder, and lysis of ureteral obstruction [20]. Minimizing postoperative bowel complications requires careful selection of healthy bowel segments for urinary diversion. Some authors suggest retroperitonealization of the conduit or omental interposition to minimize enteric complications [21].

Urinary diversion carries its own complications, including ureteral obstruction, stomal stenosis, pouch stones, cutaneous fistulas, and electrolyte disturbances. In particular, continent catheterizable channels are associated with many potential complications, though they can often be managed with minimally invasive outpatient procedures. Leslie et al. reported on 169 patients who underwent creation of continent catheterizable channels between 1992 and 2007 [22]. None of these patients had a history of malignancy. At least one subsequent surgical revision was performed in 67 patients (39%). Indications for surgical revision included stomal stenosis at the skin level in 17% of patients, stricture in 8%, incontinence in 10%, and prolapse in 4%. Sixty-nine percent of stenosis and 65% of incontinence were identified during the first 3 years after initial surgery. Welk et al. reported a 21% complication rate associated with continent catheterizable channels in 67 patients [23]. Two out of four patients with superficial stomal stenosis were successfully managed with dilation under anesthesia; the remaining two patients required open revision. Two out of four patients with channel stricture were managed with endoscopic resection of the channelbladder anastomosis, whilst the remaining two patients required open revision. Three patients with stomal prolapse required a single operative revision.

Pelvic exenteration

Major early complications of pelvic exenteration include wound infection (24%), abscess formation (12%), fistula (12%), and malnutrition (12%), whereas late complications include hydronephrosis (35%) and bowel obstruction (24%) [21,24]. Late effects of surgery include sexual dysfunction, infertility, and bladder dysfunction.

Retroperitoneal lymph node dissection

RPLND for paratesticular RMS can be associated with significant morbidity. Heyn et al. reported a 10% incidence of bowel obstruction, 8% incidence of ejaculatory dysfunction, and 5% incidence of lower extremity edema among patients undergoing RPLND for paratesticular RMS [1]. However, most of these patients

underwent bilateral node dissection. The current recommendation for staging is a modified unilateral node dissection, and nerve-sparing techniques may limit the morbidity associated with RPLND.

Bladder dysfunction

Literature from IRS I–III suggests that bladder preservation is possible in approximately 60% of patients [22]. However, results of IRS IV (1993–1997) demonstrated that of the 55 patients who retained their bladder, only 40% had normal function as determined by history [25]. The true incidence of bladder dysfunction in patients treated for RMS is unknown. There is a paucity of objective data such as validated questionnaires and urodynamic studies regarding bladder function in children treated for RMS.

Soler et al. reported on eight patients with an intact bladder after treatment of B/P RMS who were evaluated with urodynamics. Three patients were asymptomatic with normal urodynamic studies. One patient had disabling dysuria and underwent continent diversion with transverse colon. The remaining four patients had urodynamic findings of reduced bladder capacity, with overactivity in two cases, urgency in three, and suprapubic pain during filling in one patient [26]. Yeung et al. evaluated 11 children with pelvic RMS. At a mean follow-up of 6.6 years, only four of the 11 children had a normal voiding pattern. In addition, four children had evidence of upper tract dilatation, and two went on to require surgery. Only four children underwent urodynamic testing; all four studies demonstrated decreased functional bladder capacity, but only one child had decreased compliance [27].

Anticholinergic medications may be useful for patients with frequency, urgency, and incontinence. In patients with intractable symptoms, bladder augmentation with or without bladder neck reconstruction and catheterizable channel should be considered. Bladder neck competence must be evaluated, as treatment for RMS may affect sphincteric function [18].

Complications of chemotherapy/radiation

Multi-agent combination chemotherapy and radiation are integral components of therapy for RMS [28]. These treatment regimens have serious potential acute and long-term toxicities (Boxes 34.1 and 34.2).

Box 34.1 Chemotherapy-related complications [28,34,44,45].

Acute effects	Myelosuppression, bacteremia, renal toxicity, fever, neutropenia, and mucositis
Late effects	Secondary malignancy (i.e. myelodysplasia and leukemia), cardiotoxicity (doxorubicin), and endocrine dysfunction (i.e. gonadal failure, pubertal delay)

Bladder effects

Radiation therapy independently impacts bladder function. Early effects are caused by inflammation in the bladder wall and are typically reversible. Late consequences result from fibrosis of the bladder wall secondary to collagen deposition, causing urgency, frequency, hematuria, low capacity, and poor compliance [29]. Management may require clean intermittent catheterization or bladder augmentation.

Hemorrhagic cystitis is a known potential complication of chemotherapeutic agents such as cyclophosphamide and ifosfamide. Toxicity is due to the metabolic by-product acrolein. The incidence has decreased with the use of aggressive hydration, catheter drainage, and administration of mesna, a compound that binds acrolein [30,31]. Treatment options for refractory hematuria include the use of conjugated estrogens, intravesical instillations (aminocaproic acid, alum, silver nitrate, or formalin), and hyperbaric oxygen therapy. When conservative measures fail, surgical options include embolization, urinary diversion with bilateral percutaneous nephrostomy tube placement or ileal loop diversion, open packing of the bladder, or cystectomy [32,33].

Box 34.2 Radiation-related complications [44–46].

Acute effects	Urinary frequency/urgency, diarrhea, skin irritation, and fatigue
Late effects	Impairment of bone growth, delayed puberty, growth retardation, radiation cystitis, radiation enteritis, fibrosis, sphincter incompetence, rectal stricture, secondary tumors, distal ureteral strictures, and difficult biopsy interpretation

Fertility and sexual dysfunction

Radiation and, to a lesser extent, chemotherapy can compromise fertility due to endocrine dysfunction. Spunt et al. reviewed late effects of pelvic RMS treatment in females over approximately 20 years. All patients had received chemotherapy, and most underwent radiation therapy as well. Seventy-seven percent of these females had endocrine effects including ovarian failure. Vaginal stenosis requiring surgical reconstruction was also identified [34]. Pelvic radiation is known to cause severe narrowing of the introitus with resultant sexual dysfunction; in extreme cases, pubectomy may be required. In a review of late effects of treatment for B/P RMS, Raney et al. reported that two of 25 men were incapable of having an erection, while four of 16 suffered from anejaculation. In females, two of seven had abnormal menses, and only a single child was born among three women considered fertile [35].

Though still considered experimental, gonadal cryopreservation is an active area of research that could benefit survivors of childhood malignancy who suffer from infertility. In premenarchal girls, immature oocytes can be matured *in vitro* and cryopreserved for future fertilization. Once the patient is well enough, autotransplantation can be performed into the normal ovarian site or a heterotopic location. Michaeli et al. reported successful ovarian cryopreservation in 20 females under the age of 15 between 1997 and 2011 [36]. Successful transplantation of ovarian tissue has not yet been described in pediatric patients; however, 14 healthy babies have been born to adults after autotransplantation of cryopreserved ovarian tissue. Since prepubertal boys cannot benefit from sperm banking, cryopreservation of testicular tissue as well as hormonal or cytoprotective drug manipulation in order to enhance spontaneous recovery of spermatogenesis are active areas of research. Although not yet feasible in humans, animal data appear promising [37].

Secondary malignant neoplasms

Patients treated with radiation and chemotherapy have a higher risk of developing secondary malignancies, which may include bone or soft tissue sarcomas, breast cancer, colon cancer, leukemia, and cutaneous melanoma. Cohen et al. reported a risk of subsequent malignancy to be six times the expected rate [38]. In a review of 26 female survivors of pelvic

RMS, three cases of secondary malignancies were discovered, including osteosarcoma, colon adenocarcinoma, and cervical squamous cell *in situ* [34]. In 2011, the Italian Cooperative Study RMS 79 published long-term results of 163 patients treated from 1979 through 1987. Seven of 16 patients with late events went on to develop secondary malignancies. Three patients with a history of genitourinary RMS went on to develop rectal cancer, breast cancer, and cardiac angiosarcoma [39].

Special populations

Infants and young children less than 36 months of age with RMS pose a unique treatment challenge given their limited ability to cooperate, the increased radiosensitivity of underdeveloped organs, and the increased potential for secondary malignancy over a long lifetime. Puri et al. reported on 20 infants and toddlers who were treated for RMS with multimodal therapy including external beam radiation. Seven of these patients developed long-term toxicity including altered ambulation, recurrent infection, and secondary malignancy [40]. On the other hand, radiation therapy has been shown to be critical in preventing local recurrence. Results from IRS I-IV indicate that 46 of 83 patients with operative bed recurrence after resection had either limited adjuvant radiation or none at all. This deviation from radiation protocol was especially prevalent in younger children. Importantly, 75% of those with local recurrence did not survive [41]. The use of radiation for local therapy in very young patients remains controversial and is at the discretion of the local treating institute.

Recurrence

Most relapses occur within 3 years of initial diagnosis and portend a poor prognosis. Management of early recurrence includes local radiation for patients with residual disease and involvement of lymph nodes [42]. Exenteration has been attempted as salvage therapy after chemoradiation. The estimated 5-year survival rate after relapse is 64% for botryoid embryonal RMS, 26% for other embryonal tumors, and 5% for alveolar or undifferentiated pathology [43].

Conclusion

Despite improved overall survival, children with RMS continue to suffer from a wide range of

treatment-related side effects and complications. Meticulous follow-up including careful evaluation of bladder function is required to assess the sequelae of current therapies.

DOS AND DON'TS

Do

- Protect renal function by acting immediately to relieve obstruction
- Perform second-look operations when indicated. Second-look operations are useful to confirm clinical response, evaluate pathological response to therapy, and remove residual tumor in patients who achieve clinical complete or partial response after chemotherapy or radiation therapy
- Delay definitive continent reconstruction until a disease-free period of at least 2 years has been achieved and the patient is motivated and mature enough to perform self-catheterization

Don't

- Perform exenterative surgery up-front, as primary chemotherapy and radiation decrease tumor burden and facilitate organ preservation
- Perform exenterative surgery on the basis of mature rhabdomyoblasts on post-treatment biopsy
- Omit radiotherapy, as data support that this leads to higher local recurrence rates

CLINICAL PROBLEM/CASE: DISCUSSION

- 1 The most common histologic subtype is embryonal RMS. Embryonal RMS represents approximately 80% of cases and has an intermediate prognosis. Sarcoma botryoides is a variant of embryonal RMS that arises in hollow organs such as the bladder or vagina and has a more favorable prognosis. Alveolar RMS comprises 15–20% of cases, with the remainder of cases classified as undifferentiated tumors. Both alveolar and undifferentiated RMS carry a less favorable prognosis.
- 2 One of the initial goals of early management is preservation of renal function. Bladder outlet obstruction from B/P RMS or vaginal RMS is best managed with urethral catheterization. The use of suprapubic drainage has been associated with the potential for tract seeding.
- 3 The current management trend is toward organ preservation. Treatment protocol involves primary chemotherapy and radiotherapy after initial biopsy, followed by surgical resection.
- 4 Exenterative surgery should not be performed on the basis of mature rhabdomyoblasts on post-treatment biopsy. In 2006, the COG published a report supporting observation for rhabdomyoblasts only on post-treatment biopsy. Failure after apparent tumor cell maturation on biopsy has been reported; therefore, careful observation of these patients is required.

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CHAPTER 35

Testicular tumors

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KEY POINTS

- Inguinal orchiectomy is the standard approach to testicular tumors in adolescents and in children with an elevated age-adjusted alpha-fetoprotein (AFP) level
- Testis-sparing surgery should be considered in prepubertal patients with a normal AFP level
- Scrotal violations are probably not critical, but local tumor spillage may require adjuvant treatments
- The complication of ejaculatory dysfunction following retroperitoneal lymph node dissection (RPLND) can be avoided in nearly all patients with a nerve-sparing technique
- To minimize the risk of surgical complications, meticulous technique, protection of retracted viscera, and a working knowledge of vascular surgical techniques is important when undertaking an RPLND

CLINICAL PROBLEM/CASE

Case 35.1

A 14-year-old boy presents with painless scrotal swelling noted by his physician during his school physical. He denies recent trauma and has no associated symptoms. On exam, the patient is Tanner 4 and the right testis is firm and enlarged. A scrotal ultrasound (US) is obtained, and a representative image is shown here.

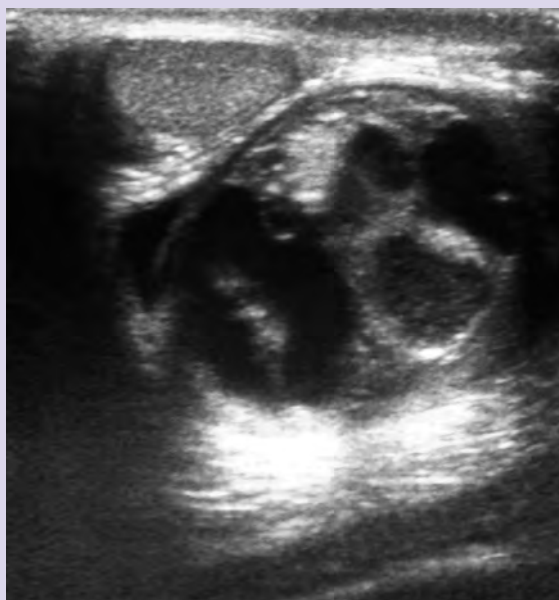
- 1 What is the differential diagnosis of this lesion? What is the next step in the evaluation?
- 2 What is the correct surgical approach to radical orchiectomy and why?
- 3 Pathology confirms a mixed germ cell tumor; tumor markers normalize after surgery, however CT abdomen/pelvis demonstrates limited paracaval lymphadenopathy. He undergoes bilateral nerve-sparing RPLND. What is the rationale for nerve-sparing?



Case 35.2

A 4-month-old infant presents with an enlarged firm right testicle. It is not painful, and the child seems otherwise healthy. Scrotal US reveals a complex cystic mass nearly replacing the testicle. The AFP level is 300 ng/dl.

- 1 What is the most likely diagnosis?
- 2 What will be your operative plan for the patient?
- 3 The frozen section reveals a mature teratoma. The testis is closed and returned to the scrotum. The final pathology report confirms the frozen section diagnosis. What oncological evaluation or treatment is indicated?

**Introduction**

Testis tumors are rare in children and fall into two categories in the pediatric population: prepubertal and postpubertal tumors. Tumors occurring in adolescents are the same as those seen in older adults, with non-seminomatous mixed germ cell tumors (NSMGCT) predominating. Benign tumors are uncommon in this population. The incidence of testis tumors in prepubertal children is 0.5 to 2.0 per 100,000 children, accounting for only 1–2% of all pediatric tumors [1]. Teratomas are benign in children and nearly all malignant tumors in children are yolk sac tumors. Unlike adolescents and adults, a large proportion, perhaps even a majority of prepubertal testis tumors are benign [2,3]. Therefore, the management of testis tumors in prepubertal and postpubertal boys differs significantly.

The initial radiographic evaluation of children with a suspected testis tumor is limited. Because many prepubertal testis tumors are benign, a metastatic evaluation is usually deferred until tissue confirmation of the tumor's histology is obtained. However, when a malignancy is suspected (for example in children with an elevated alpha-fetoprotein (AFP) level or in adolescents) a computerized tomography (CT) scan of the abdomen

may be obtained preoperatively. Imaging of the primary tumor is sometimes helpful. Ultrasonography is most often employed and is able to distinguish a testicular tumor from a benign extratesticular lesion or from a paratesticular rhabdomyosarcoma. The extent of testicular involvement can also be determined, which is helpful if testis-sparing surgery is being considered. The ultrasonographic appearance of specific testis tumors has been described. Unfortunately, ultrasound findings are too inconsistent to allow a definitive diagnosis.

Tumor markers play an important role in the evaluation and follow-up of childhood testis tumors. AFP is the most important tumor marker in prepubertal patients. AFP levels are elevated in 80–90% of children with a yolk sac tumor, and AFP has a biological half-life of approximately five days [4]. It should be kept in mind that AFP levels are normally quite high in infancy. An “elevated” level in a boy less than 1 year of age does not rule out the possibility of a benign tumor, such as teratoma [5]. In addition to AFP, the beta subunit of human chorionic gonadotropin (hCG) is an important marker in adolescent testis tumors, but this is rarely elevated in children because the histologic types that lead to elevated human chorionic gonadotropin levels are rarely encountered in prepubertal testis tumors.

Surgical approach

The standard approach to a malignant testis tumor or paratesticular rhabdomyosarcoma is an inguinal orchiectomy. Through an inguinal incision, the spermatic cord is isolated and controlled with a noncrushing clamp or Rommel tourniquet. The testis is then delivered into the inguinal incision with the tunica vaginalis intact. Once a testis tumor is confirmed by direct palpation, the cord is ligated and divided at the internal ring.

Increasing consideration has been given to performing testis-sparing surgery for benign testicular tumors [2]. This is particularly attractive in prepubertal patients because more than one-third of tumors are benign in this population. The preoperative evaluation plays a significant role in patient selection for testis-sparing surgery. An elevated AFP level in a child over 1 year of age virtually always reflects the presence of a yolk sac tumor and precludes a testis-sparing approach. However, in infants (who normally have high AFP levels), and older children with a normal AFP, the likelihood of a benign tumor is considerable. This is also true in boys presenting with virilization. For these patients, an inguinal exploration should be considered so that testis-sparing surgery may be performed if a benign histology is confirmed. The initial approach is the same as for an inguinal orchiectomy. Once the testis is delivered into the inguinal incision (the cord having been controlled), the field is draped off with towels and the tunica vaginalis is opened. The tumor is excised or enucleated and sent for frozen section. If a benign histology is confirmed, then the testicular defect is closed with absorbable suture and the testis is returned to the scrotum. Reports from small series suggest that testis-sparing is safe and effective in preserving testicular tissue with no reports of local recurrence of benign tumors managed in this fashion [6]. If a malignancy is detected, or the frozen section is nondiagnostic, then an orchiectomy is performed.

After orchiectomy, children with malignant testicular tumors or paratesticular rhabdomyosarcoma require additional evaluation and therapy. The type of adjunctive management selected will depend on the histology of the primary tumor and the results of radiographic and biochemical studies. The intensity of follow-up also depends on the malignant potential of the primary tumor. Prepubertal patients with stage 1 yolk sac tumor whose AFP normalizes postoperatively are managed

with observation. Patients with radiographic or biochemical evidence of metastatic disease, and those stage 1 patients who recur on observation, are treated with three or four cycles of platinum-based multi-agent chemotherapy. Retroperitoneal surgery is undertaken only in the rare event of equivocal lymph node involvement on CT scan with normal markers or in the event of a persistent retroperitoneal mass following chemotherapy. Retroperitoneal lymph node dissection is employed more frequently in adolescents with NSGCT. It may be undertaken in select patients as a staging and therapeutic approach to clinical stage 1 disease or in stage 2 patients with limited retroperitoneal disease. It is also undertaken for patients with a persistent mass following chemotherapy – a more common occurrence in adolescents than in prepubertal patients. RPLND also plays a role in the management of patients with paratesticular rhabdomyosarcoma. It is indicated as a staging procedure in adolescents with clinical stage 1 disease and is also indicated in all patients (pre- or postpubertal) with apparent lymphatic involvement on CT scan. From a surgical point of view, then, all patients with a testis tumor will undergo orchiectomy or testis-sparing tumor excision, and a select group of patients will undergo an RPLND as well.

Orchiectomy: outcomes and complications

Inguinal orchiectomy provides excellent local control for testicular cancer in prepubertal and postpubertal patients. Local recurrence is extremely rare and occurs almost exclusively in the context of local tumor spillage. Guidelines for the surgical management of testicular cancer have emphasized the importance of avoiding scrotal violation in removing these tumors. Despite this standard, a significant number of patients in single institution reports and in multi-center clinical trials have had scrotal violations occur. Studies in prepubertal and postpubertal patients suggest that these violations do not affect overall survival [7,8]. Scrotal violations occur in three settings. First, patients with testicular cancer who have previously undergone unrelated scrotal surgery, such as an orchidopexy for an undescended testicle, may be at risk for inguinal lymphatic spread. Whilst routine inguinal lymphadenectomy is not recommended, careful observation of the inguinal lymph

nodes during evaluation and follow-up is prudent. A second form of scrotal violation involves intact tumor extraction through a scrotal incision. Finally, patients may have a scrotal violation with potential tumor spillage such as might occur with a transcrotal biopsy or violation of the tumor capsule during a transcrotal excision. Local recurrence in the absence of tumor spillage or violation is exceedingly rare and can occur whether the approach is scrotal or inguinal. An inguinal approach is still recommended as a way to avoid tumor spillage or cutting across the tumor that may involve the spermatic cord. However, a scrotal violation in the absence of tumor spillage probably does not require any additional local therapy, although once the error is recognized a separate inguinal incision should be made to remove that portion of the spermatic cord. If tumor spillage does occur during a scrotal violation, then consideration should be given to hemiscrotectomy or other adjuvant therapy.

Complications following inguinal orchiectomy are rare. Bleeding occasionally occurs and is best prevented by close attention to hemostasis in the soft tissues of the scrotum and appropriate control of the spermatic cord at the internal ring. Bleeding from the spermatic cord can be difficult to control once the cord is divided and retracts into the retroperitoneum. This can be prevented by controlling the cord with a suture ligature and a more proximal free tie. The cord cannot retract from the suture ligature and the more proximal free tie will prevent tracking into the retroperitoneum of any bleeding inadvertently caused by the suture ligature. Following the orchiectomy, the scrotum should be inverted into the inguinal incision for fulguration of any bleeding tissue or vessels. When postoperative bleeding does occur, whether in the scrotum or retroperitoneum, it is best managed conservatively without exploration. In addition to the direct consequences of bleeding, a retroperitoneal hematoma can interfere with the interpretation of staging CT scans since a retroperitoneal hematoma may be confused for adenopathy [9]. When a pelvic mass without higher retroperitoneal involvement is seen on a post-orchiectomy CT scan, a retroperitoneal bleed is more likely than metastatic disease. Magnetic resonance imaging can sometimes make the distinction in difficult cases. However, to prevent this confusion it is best to obtain abdominal imaging prior to orchiectomy in cases suspicious for cancer.

RPLND: outcomes and complications

In 1977, Donahue described the classic suprahilar bilateral retroperitoneal lymph node dissection for testicular cancer [10]. The operation held significant morbidity, including loss of ejaculatory function due to disruption of the lumbar sympathetic chains and hypogastric plexus. However, in the era when chemotherapy and radiation were largely ineffective for metastatic disease, radical surgical clearance was of paramount importance, and the associated morbidity was acceptable. Over the years, both the effectiveness of chemotherapy and an increased understanding of retroperitoneal neuroanatomy have led to an evolution in the approach to RPLND [11]. Currently, the nerve-sparing technique is employed at most centers for patients with low-stage disease undergoing RPLND – whether unilateral or bilateral (Figure 35.1).

Since RPLND is rarely indicated in prepubertal patients, there are no data to support the feasibility of nerve-sparing for this group. For children with paratesticular rhabdomyosarcoma and limited positive retroperitoneal nodes, a modified unilateral template is appropriate. The same is true for children with yolk sac tumor and a persistent mass following chemotherapy. Whether children with more extensive lymphadenopathy should undergo a more extensive bilateral dissection is unclear since there are no data delineating the oncologic benefit in these rare clinical situations. The uncertain oncological benefit must be weighed against the increased morbidity of the more extensive operation. In prepubertal children with yolk sac tumor, normalization of AFP, and an equivocal node(s) on abdominal CT, an excisional node biopsy is adequate for staging purposes. The result of the biopsy will then determine whether adjuvant chemotherapy is indicated. Unless otherwise noted, the discussion below reflects the results for RPLND primarily in adults which should be similar to those for adolescents with NSGCT.

The oncological effectiveness of RPLND is reflected in the very low incidence of retroperitoneal recurrence [11–13]. Surgical complication rates of primary RPLND for low-stage disease range from 11–32% with lower rates in more recent series [13–15]. In a series of 478 patients treated at Indiana University, the most common complication was superficial wound infection accounting for nearly half of the patients with complications [14]. Most major complications were related to

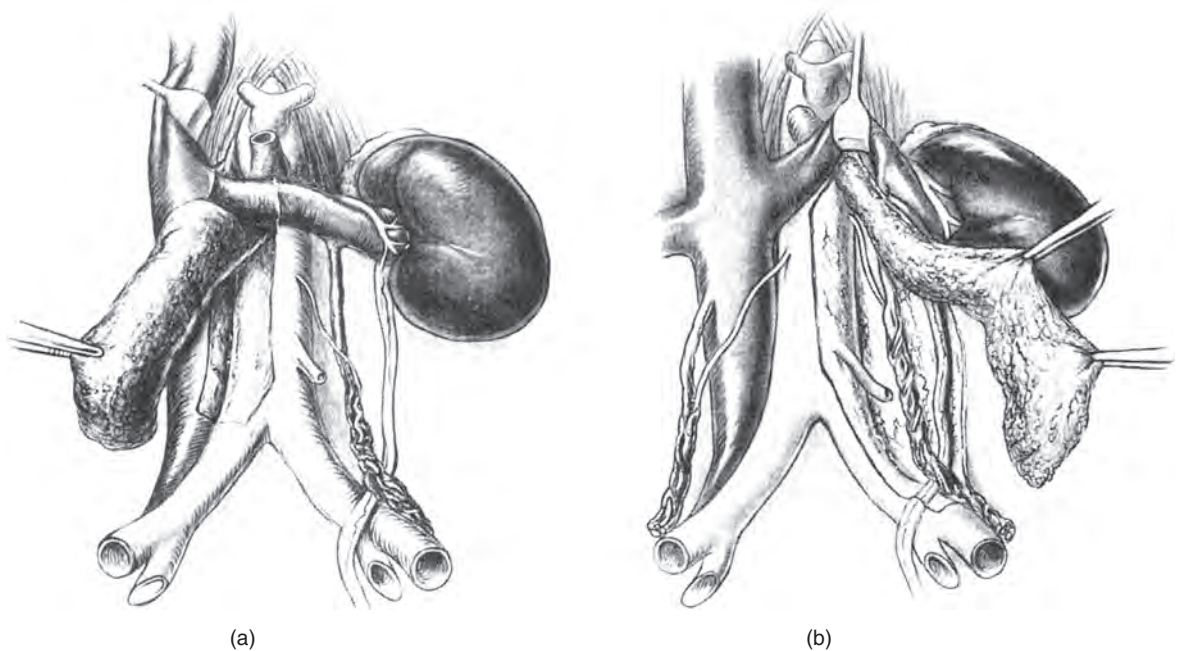


Figure 35.1 (a) The right-sided modified unilateral template in which the interaortocaval lymph nodes are included, while the left para-aortic lymphatic tissue remains undisturbed. (b) The left-sided modified unilateral template includes the upper interaortocaval group and left para-aortic lymphatic tissues. The lower right para-aortic region remains undisturbed. (From Donohue et al. 1998 [11]. Reproduced with permission of Elsevier.)

small bowel obstruction or atelectasis. Complications were twice as common in patients undergoing a bilateral dissection as those undergoing a unilateral approach. Ejaculation was preserved in 98% of those undergoing a nerve-sparing approach. The more common complications following RPLND in a recent study from Germany are shown in Table 35.1. Other complications occurring in under 1% of patients include late bleeding, pulmonary embolism, small bowel obstruction and deep vein

thrombosis. In this study of nerve-sparing RPLND, antegrade ejaculation was preserved in 93%. Laparoscopic approaches to RPLND have been reported with greater frequency in the last decade in both primary and post-chemotherapy settings. In a review of 100 patients undergoing post-chemotherapy laparoscopic RPLND for stage II disease, Steiner et al. reported minimal complications with a lymphocele in one patient and chylous ascites in another, and no evidence of recurrence after a mean follow-up of 74 months. Antegrade ejaculation was preserved in 95% of patients [16].

Table 35.1 Complications of RPLND.

Complication	Incidence (%)
Superficial wound infection	5.4
Ileus	2.1
Chylous ascites	2.1
Lymphocele	1.7
Hydronephrosis	1.3

From Heidenreich et al. 2003 [13]. Reproduced with permission of American Urological Association.

Loss of ejaculation/infertility

The key to preserving ejaculation is maintenance of the sympathetic chains which arise behind the vena cava on the right side and dorsolateral to the aorta on the left side (Figures 35.2 and 35.3). The T12–L4 nerve fibers travel antero-caudally to decussate on the anterior surface of the aorta and form the hypogastric plexus as they course over the aortic bifurcation. The nerve roots should be identified early in the dissection in order to minimize this complication. With nerve-sparing

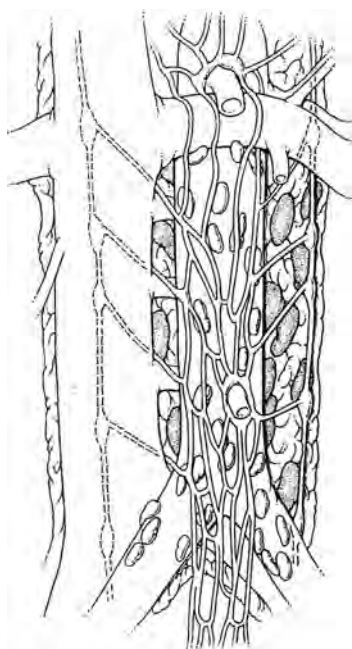


Figure 35.2 The lumbar sympathetic nervous system and its relation to the great vessel. Note the sympathetic ganglia L2–L4, the hypogastric plexus and terminal nerve trunks. (From Donohue et al. 1998 [11]. Reproduced with permission from Elsevier.)

techniques, antegrade ejaculation is preserved in nearly all patients (Table 35.2). However, in patients with more advanced disease, this may not be possible. When retrograde ejaculation occurs, it can be treated with a short course of imipramine [17]. Electroejaculation or testis biopsy can also be employed to achieve a pregnancy [18,19].

Whilst nerve-sparing modifications of the RPLND have preserved ejaculation, fertility rates among patients who have undergone an RPLND are affected by other aspects of their treatment and the underlying disease itself. Interestingly, fertility rates are higher amongst men with stage 1 disease who undergo nerve-sparing RPLND than among those managed on surveillance [20]. This is due to the fact that a higher percentage of men on surveillance ultimately require more intense systemic therapy for recurrence. In men undergoing nerve-sparing RPLND for low-stage disease, fertility rates of 75% and 84% have been reported [11,21]. Interpreting fertility results in testis tumor patients following RPLND is difficult since the causes are multifactorial. Following unilateral orchiectomy alone, sperm cell count is highly impaired 1–4 weeks after surgery in 60–70% of patients, but improves over 2–3 years. Recovery of spermatogenesis is delayed by a year for those receiving chemotherapy, and high-dose therapy

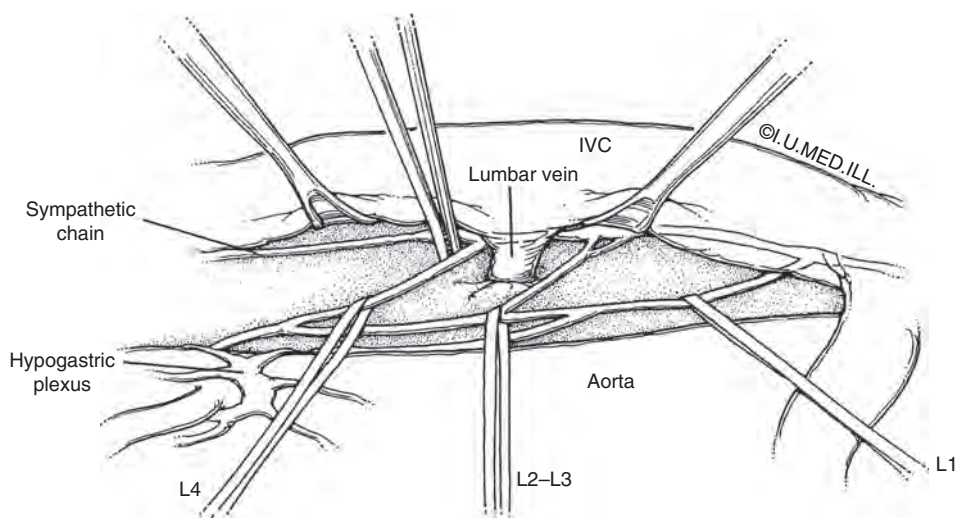


Figure 35.3 The right postganglionic fibers L1–L4 arise from the right sympathetic trunk dorsal to the cava, emerge into the interaortocaval and preaortic area, where they come in with splanchnic fibers in larger trunks. These are prospectively identified and isolated before lymphadenectomy. IVC, inferior vena cava. (From Donohue et al. 1998 [11]. Reproduced with permission from Elsevier.)

Table 35.2 Oncologic and ejaculatory results of RPLND.

Study	Type of dissection	Retroperitoneal recurrence (%)	Ejaculation preserved (%)
Weissbach et al. 1990 [12]	Bilateral	1.5	34
	Unilateral	2.4	74
Heidenreich et al. 2003 [13]	Nerve-sparing (88% unilateral)	1.2	93
Donohue et al. 1998 [11]	Unilateral nerve-sparing	0.6	100

reduces the chances of ultimate recovery [22]. Nearly all men undergoing chemotherapy for testis tumor become azoospermic, although most recover spermatogenesis within four years [19]. Semen cryopreservation should be offered to all Tanner 4 or 5 patients undergoing treatment for testis cancer. Patients as young as 12 years old can supply an adequate specimen [23].

Long-term data in children undergoing therapy for testis cancer are sparse. The Intergroup Rhabdomyosarcoma Study Committee reported the long-term health consequences of 86 children treated for paratesticular rhabdomyosarcoma from 1972–1984, most of whom had some form of RPLND. Elevated follicle-stimulating hormone or azoospermia occurred in more than half for whom data were available [24].

Bleeding

Major intraoperative bleeding is a rare complication but poses a significant problem when it occurs. Surgeons undertaking RPLND must be skilled at basic vascular surgery techniques, as a significant percentage of patients may require repair of an intraoperative vascular injury. Repair of inadvertent injuries to the vena cava or aorta can be easily accomplished with fine vascular suture. Inadvertent injuries to the renal arteries can be more problematic, occasionally resulting in nephrectomy [25]. Rarely, resection and replacement of portions of the vena cava or aorta may be necessary in patients with advanced disease. Immediate availability of an experienced vascular surgeon is essential when undertaking these more complicated cases.

Ileus/small bowel obstruction

Some degree of ileus is inevitable following a major transperitoneal operation. The risk of prolonged ileus and small bowel obstruction (SBO) may be minimized

by limiting manipulation of the bowel. Retractors such as self-retaining Deaver or sweetheart retractors that lift up and away from the field can be helpful. Minimizing the duration of bowel retraction is also important. Self-retaining retractors should be released on a regular basis (e.g. every 45 minutes) and the bowel examined for impending injury. Once the operation is complete the bowel should be returned to its normal anatomic position. Some surgeons favor tacking the bowel back in place with absorbable sutures, though the efficacy of this approach is unproven. Prolonged nasogastric tube (NG) drainage has not been shown to prevent prolonged ileus or SBO and the NG tube can be removed on the first postoperative day. Early ambulation may also stimulate bowel activity. SBO may be more common in children. Festen reported a 2.2% incidence of SBO following 1476 abdominal operations in infants and children [26]. Eighty percent of cases occurred within three months of surgery and 70% were due to a single adhesive band.

Patients who recover bowel function postoperatively and then re-present weeks or months later with complete bowel obstruction with no flatus or stool should undergo immediate laparotomy. When partial SBO occurs, it can usually be managed conservatively with NG suction, intravenous fluids, aggressive monitoring for and correction of electrolyte abnormalities, and serial radiographs [27]. Akgur et al. found that conservative management of SBO in children was successful in 74% of cases. However, these patients were more likely to suffer future recurrent SBO than those managed initially with lysis of adhesions (36% vs. 19%) [28]. An elevated white blood cell count with a left shift and persistent localized pain following nasogastric decompression suggest possible compromise of the bowel and early exploration should be considered

in those cases. In addition to SBO, patients presenting with abdominal pain, nausea and vomiting following RPLND may have pancreatitis. This is presumably secondary to elevation and retraction of the pancreas when obtaining exposure. Serum amylase and lipase should be obtained in patients with persistent symptoms postoperatively.

Deep vein thrombosis

Whilst very rare in children, pulmonary embolism is a potentially fatal complication of RPLND in adults. Most surgeons prefer to avoid the prophylactic use of anticoagulants because of concern for lymphatic leaks and lymphoceles. PAS stockings have been shown to be equally effective for preventing DVTs and should be considered for adolescent patients. For PAS stockings to be effective, it is crucial that the stockings be applied and activated before the induction of anesthesia. They should be maintained postoperatively and early ambulation is encouraged. Frequent physical examination and prompt radiographic evaluation of signs of DVT with early intervention are important for preventing pulmonary embolism.

Chylous ascites

Chylous ascites can be a debilitating complication of RPLND which usually presents with abdominal distension. The diagnosis can be made by paracentesis. The aspirated fluid will be high in triglyceride, total protein and cholesterol concentrations and have a cellular differential that is primarily lymphocytes [29]. The complication can be prevented by meticulous attention to hemostasis and control of lymphatic channels. All significant lymphatic vessels should be ligated or clipped. Ligatures should also be applied at the superior and inferior limits of the dissection, particularly where the lymphatic tissue exits the field at the renal hilum. Chylous ascites most commonly occurs in patients with prolonged courses of preoperative chemotherapy and in those with a large intra-operative blood loss, particularly during a post-chemotherapy RPLND [30]. Most patients can be managed without surgery. Conservative therapy includes a combination of paracenteses, medium chain triglycerides and total parenteral nutrition with a selective use of abdominal drains [30,31]. Whilst occasional patients will respond to observation or dietary changes, most require TPN. With conservative management, the ascites will

usually resolve within a few months. Use of somatostatins may also be helpful [32]. Those that fail conservative management will require placement of a peritoneovenous shunt. Occasionally, early re-exploration and ligation of the offending leak can be accomplished [33]. Feeding the patient a high-fat diet up to six hours before surgical exploration may facilitate intraoperative identification of the offending leak. There is little information regarding the management of chylous ascites in prepubertal patients, though the same approach as used in adults seems reasonable [34].

Surgical approach and morbidity

Whilst an anterior transperitoneal approach is still favored at most institutions, alternative approaches have been employed in an attempt to reduce morbidity. An extraperitoneal approach has been advocated for excision of residual masses following chemotherapy [35]. Reported advantages include the ability to excise coexisting thoracic masses when a thoracoabdominal extraperitoneal approach is used, improved access to nodes above and behind the renal vessels, and a more rapid postoperative recovery. Increasingly popular has been a laparoscopic approach – particularly for primary RPLND in patients with low-stage disease. An initial report of 20 patients in 1994 utilizing a modified unilateral template reported ejaculation in all patients and no retroperitoneal recurrences [36]. However, significant complications, most commonly bleeding, occurred in 30% of patients. In contrast, a 2001 report of 125 patients undergoing laparoscopic RPLND reported only two conversions for bleeding, no major complications and only 13 minor complications [37]. More recent reports describe the use of laparoscopic nerve-sparing bilateral RPLND, demonstrating good feasibility and minimal morbidity. The oncologic efficacy of this approach remains under evaluation [38]. Retroperitoneal recurrence is a serious problem and no compromise on the extent of dissection should be accepted to accommodate a laparoscopic approach.

More recent reports suggest that laparoscopic RPLND utilizing a modified unilateral template and, in some cases, nerve-sparing, can be performed with less morbidity and an improved quality of life compared to open techniques [16,38–40]. Laparoscopic RPLND has also been performed retroperitoneally [41–43].

Complications in post-chemotherapy RPLND

Not surprisingly, complications are more common in patients undergoing RPLND following chemotherapy for disseminated disease. A report of such patients from Indiana University reported a 30–50% intraoperative complication rate and a 7–14% postoperative complication rate including ascites, wound infection, prolonged ileus, pancreatitis, acute renal failure, and atelectasis [44]. Patients with higher stage disease had a higher rate of complications and more frequent need for additional intraoperative procedures including nephrectomy, IVC resection, bowel resection, hepatic resection/biopsy, arterial grafting, caval thrombectomy, adrenalectomy and cholecystectomy. Christmas et al. reported on 98 patients undergoing a post-chemotherapy RPLND. Some type of vascular procedure was required in nearly all patients, including 14 nephrectomies, 35 IMA ligations, two aortic grafts, and two IVC ligations [45]. Complete macroscopic resection of residual disease was possible in 97% of patients. In another series, 10 of 710 patients undergoing post-chemotherapy RPLND required intraoperative or postoperative aortic grafting (for aortic rupture or aortoenteric fistula) [46]. Pre-emptive intraoperative grafting should be considered when there is extensive sub-adventitial dissection, a duodenal enterotomy or extensive serosal bowel violation. A 7% rate of caval resection or thrombectomy has been reported in patients undergoing a post-chemotherapy RPLND [47]. Not only are complications more common in those undergoing a post-chemotherapy RPLND, but the occurrence of complications in these patients has been shown to adversely

affect their disease-free survival [48]. Ejaculatory dysfunction is also more common following post-chemotherapy RPLND due to the difficulty in preserving the sympathetic nerves in this group of patients. Among 472 patients undergoing post-chemotherapy RPLND, Coogan et al. reported that 20% were amenable to a nerve-sparing approach [49] and 76% of these patients reported normal ejaculation.

Conclusion

Orchiectomy and testis-sparing tumor excision are relatively straightforward procedures with low complication rates. However, meticulous hemostasis and adherence to oncological surgical principles are important. RPLND is the operation that carries the greatest risk of complications for patients with testicular cancer. Loss of ejaculation – an expected outcome decades ago – can now be avoided in nearly all patients with low-stage disease. Knowledge and implementation of sound vascular surgical techniques is crucial to anyone undertaking the operation. Chylous ascites, while rare, can be a major complication requiring prolonged management until resolution. Finally, efforts to avoid the complications of any major intra-abdominal operation – atelectasis, ileus, SBO, DVT, and PE – should be made. These efforts include attention to all aspects of preoperative, intraoperative and postoperative management. Fortunately, most patients undergoing operations for testicular tumors are young and otherwise healthy. But complications still occur, particularly in patients undergoing post-chemotherapy RPLND.

DOS AND DON'TS

Do

- Obtain AFP and hCG levels prior to orchiectomy
- Use an inguinal approach in the surgical management of testis tumors
- Consider testis-sparing surgery in a prepubertal patient with a normal AFP
- Consider nerve-sparing techniques for stage I and II disease when possible to preserve ejaculatory function
- Expect a higher complication rate in the setting of post-chemotherapy RPLND

Don't

- Compromise the margins of resection in an attempt to maintain ejaculatory function
- Compromise oncologic principles when considering a laparoscopic approach to RPLND

CLINICAL PROBLEM/CASE: DISCUSSION**Case 35.1 discussion**

- 1 The differential diagnosis includes germ cell tumors, such as embryonal, teratoma, seminoma, or a mixed tumor and stromal tumors. Measurement of baseline levels of tumor markers AFP, hCG and LDH are useful in predicting tumor type and ascertaining residual disease after orchiectomy.
- 2 Radical orchiectomy is performed using an inguinal approach. The spermatic cord is mobilized and controlled with a tourniquet, and the testicle is then delivered into the incision. A scrotal approach should not be undertaken in the setting of testis tumor because of the risk of altering the predicted pattern of lymphatic drainage and subsequently, the pattern of metastatic spread.
- 3 Antegrade ejaculation is mediated by sympathetic fibers that originate from the thoracolumbar spinal cord to form the hypogastric plexus. Prospective identification, meticulous dissection and preservation of the sympathetic chains, postganglionic sympathetic fibers and hypogastric plexus allow for the highest rates of preserved ejaculatory function after RPLND.

Case 35.2 discussion

- 1 The most likely diagnosis is a teratoma of the testis given the cystic appearance and an AFP level that is not particularly high for a 4-month-old boy.
- 2 This patient should undergo an inguinal exploration with control of the spermatic cord at the internal ring. The testis should be delivered inguinally within the tunica vaginalis and then draped off from the surgical field. The testis tumor should be enucleated and a frozen section obtained.



- 3 None, as mature teratomas are universally benign in prepubertal patients.

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PART IX

Trauma

CHAPTER 36

Genital trauma

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KEY POINTS

- Genital trauma is rare in the pediatric population
- Neonatal penile trauma is most often iatrogenic from circumcision injury
- Ultrasonography is a valuable adjunctive tool to physical exam in evaluating scrotal trauma
- Examination under anesthesia should be used to fully assess the extent of genital trauma
- Sexual abuse should always be considered in cases of genital injury, particularly when the history and physical exam findings are suspicious or discordant.

CLINICAL PROBLEM/CASE

An 8-year-old female is brought to the emergency room after sustaining a straddle injury walking on a railing. The appearance of the injury is shown in Figure 36.1 with a catheter in the urethra. She is stable and has no other injuries.

- 1 How should the injury be assessed?
- 2 What other associated injuries must be evaluated?
- 3 How should the injury be treated?
- 4 Are there any long-term sequelae that merit follow-up?



Figure 36.1 Anterior vaginal tear following straddle injury resulting in posterior displacement of urethra.

Introduction

Trauma is the leading cause of death in children. Genitourinary tract injuries are rare, accounting for <3% of trauma cases [1]. The majority of these injuries are due to blunt trauma such as motor vehicle collisions [2]. In developed countries, iatrogenic injuries are especially prominent in the neonatal period whilst school-age children are prone to sports-related injuries, kicks and straddle injuries [3]. Recent literature regarding pediatric genital trauma has focused on identifying and treating sexual abuse injuries as well as distinguishing these injuries from accidental trauma [4]. This chapter reviews common etiologies for traumatic genital injuries and evaluates current diagnostic and treatment options. Risk factors, presenting signs and the evaluation of sexual abuse will also be reviewed.

Penile injuries

Penile injuries in the neonate are most commonly iatrogenic [2]. The risk of circumcision-related injury due to clamp (e.g. Mogen, Gomco, Plastibell) circumcision is approximately 0.2% and may range in severity from mild penile skin loss to more significant glans or urethral injuries [5]. Self-inflicted or non-iatrogenic pediatric penile injuries are rare [6]. Traumatic injuries include degloving of the penis and penile amputations. Zipper injuries may result in contusion or pressure necrosis of the prepuce [7]. Another common etiology is the “hair tourniquet” leading to preputial edema and inflammation and, less commonly, more significant injury to the corpora or urethra [2]. As children begin toilet training, toilet seat injuries become common [4]. Similar contusions or lacerations can occur from rubber band-powered toys during bathtub playtime in this population. Penile lacerations and amputations have been rarely reported from dog or other animal bites [8]. Finally, blunt perineal trauma is the most common etiology for high-flow priapism secondary to a traumatic arteriovenous fistula in rare cases [9].

Treatment

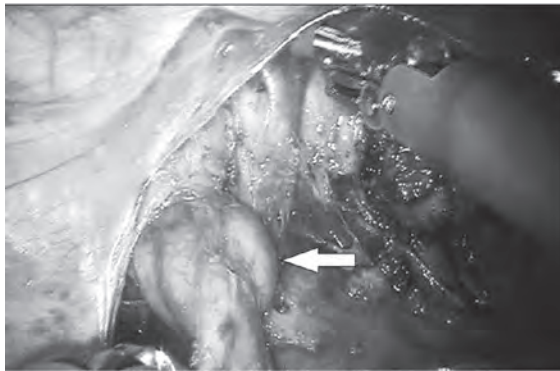
Superficial contusions and lacerations are generally managed with topical antibiotic ointments, but may require minor surgical debridement and skin reapproximation depending on the mechanism, patient anxiety

level, and severity of injury [4]. Empiric antibiotics may be prescribed to decrease the risk of cellulitis, although there are few data to support this practice. Where significant penile skin is lost due to neonatal circumcision, full thickness skin grafting of the excised prepuce or shaft skin may be performed in the immediate post-injury period [10]. In cases where prepuce is not available, healing by secondary intention may be considered, although it can involve frequent, often painful, dressing changes [1]. Late complications include penile trapping, distal urethral or meatal stenosis, and unsatisfactory cosmetic outcomes. However, post-circumcision healing in the neonatal period is generally excellent if the injury is recognized and treated promptly.

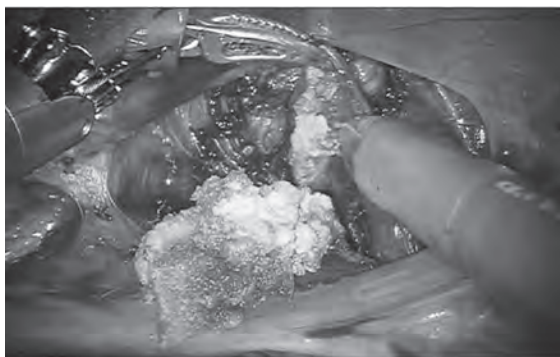
Zipper injuries to the penis more commonly occur in uncircumcised boys and may be treated with mineral oil to slip the trapped skin from the zipper [8] or by cutting of the median bar of the zipper with bone cutters [12]. In cases where conservative measures are inadequate, operative intervention may be necessary. Devitalized skin should be excised with primary reapproximation or healing by secondary intention in cases of delayed presentation or concern for contamination [13]. However, most procedures may be performed in the emergency room under local anesthesia. The use of a benzodiazepine such as midazolam may facilitate examination. Formal conscious sedation is not generally required [2].

Tourniquet injuries are generally superficial, but may extend to the corporal bodies or urethra. Often the initial diagnosis is mistaken for balanitis or paraphimosis due to significant associated preputial edema. Prompt removal of the constricting band may require an exam under anesthesia for adequate exposure. Identifying a hair tourniquet may be particularly difficult, especially in the case of blond hairs. Loupe magnification during an exam under anesthesia helps considerably in this situation. Local wound care may be used for isolated skin necrosis, while more severe cases of corporal injury may require primary repair of cavernosal defects to minimize the risk of fibrosis and preserve erectile function [2].

Amputation injuries may be managed with primary reanastomosis up to 8 hours after injury [14] (Figure 36.2). Buccal mucosa grafting has been suggested to reapproximate the coronal sulcus and provide an improved cosmetic result in cases of glans amputation [15]. Partial amputation injuries may be



(a)



(b)

Figure 36.2 (a) Amputation injury showing probe in proximal urethra and catheter through amputated glanular urethra. (b) Postoperative repair appearance.

treated nonoperatively with intermittent application of epinephrine-soaked sponges to control hemorrhage in the absence of associated urethral injury. If urethral involvement is present, formal hypospadias repair may be required [11]. As with any other body part, the amputated penis should be cooled as quickly as possible to reduce ischemic injury.

Genital injury due to animal bite should be assessed under anesthesia to fully assess the extent of trauma. Broad spectrum antibiotics and tetanus prophylaxis should be administered prior to treatment [9]. Debridement of devitalized tissue and aggressive irrigation of the wound have been shown to reduce the risk of wound infection from 59% to 12% [16]. Split thickness skin grafts should be applied to denuded areas to minimize wound contraction. Investigation for potential

abuse or neglect should be initiated in these cases to reduce the risk of future injury [9].

High-flow priapism may be managed nonoperatively with spontaneous resolution reported within days to weeks after injury [10]. If conservative measures fail, embolization with autologous clot is the preferred treatment, preserving erectile function in 80–100% of patients. However, up to 44% of patients may recur. In cases of recurrence, repeat embolization with nonabsorbable materials may be attempted [17]. Open surgical ligation is a measure of last resort due to the high risk of associated erectile dysfunction [10].

Scrotal/testicular injury

Injury to the testis is commonly associated with straddle injuries, for example bicycle crossbar injuries, where the testis is forced against the pubic ramus, causing tearing of the tunica albuginea. Injuries may also result from hits or kicks to the scrotum during sporting events or roughhousing. Due to its higher position in the scrotum, the right testis is more prone to injury than the left [18]. Injuries tend to be less common in infants due to the smaller size and increased mobility of the testes during infancy. Significant testicular injury associated with minor trauma in these patients should raise the possibility of intrinsic testicular pathology, including malignancy.

Trauma to the scrotum without underlying testicular injury tends to resolve within a short time. In patients with pain that initially resolves after a short period but recurs after several days, traumatic epididymitis should be considered [19]. However, pain persisting longer than two hours after trauma is suspicious for more significant testicular injuries, including testicular torsion or rupture [20].

Testicular torsion associated with scrotal injury, whilst uncommon, has been reported. Cases of delayed testicular torsion following blunt scrotal trauma, sports-related injuries, and bicycle riding have also been reported [20,21]. A history of trauma is the presenting symptom in 4–8% of all cases of testicular torsion [20,22]. Acute scrotal swelling may also be associated with intraperitoneal pathology, such as appendicitis, peritonitis, liver laceration, or splenic rupture [23–25]. In the child with a history of abdominal trauma, scrotal pathology in the absence of scrotal trauma should

prompt a thorough evaluation [25]. In the newborn period, adrenal hemorrhage should also be considered. Retroperitoneal imaging with ultrasonography aids in differentiating this diagnosis from other etiologies of scrotal swelling or ecchymosis.

Treatment

If physical examination and imaging, such as ultrasound, are indeterminate or suggest significant testicular injury, early scrotal exploration is recommended (Figure 36.3). Rates of salvage in cases of testicular rupture have been reported as high as 90% if performed within 72 hours [26]. Early exploration may also lead to decreased convalescent times and reduced risk of infection.

In cases of suspected torsion, early exploration is also recommended. Testicular salvage rates for torsion appear to increase with earlier exploration, with little benefit seen after 72 hours [18]. In contrast, nonoperative management has been associated with orchiectomy rates of approximately 45% [1]. Animal studies suggest that early application of ice to the scrotum may aid in the preservation of seminiferous tubules in testicular torsion [27].

Cases of isolated hematocele may be followed nonoperatively in the absence of impaired testicular flow. Isolated epididymitis may also be treated with supportive care including scrotal elevation and nonsteroidal analgesics. In the absence of ischemic changes, testicular fracture without disruption of the tunica albuginea may



Figure 36.3 Scrotal laceration after falling from a bunk bed without associated testicular injury.

be observed. If nonoperative management is selected, follow-up with both physical exam and ultrasonography should be used to monitor resolution of injury.

Vaginal injuries

Vaginal injuries are relatively rare in the pediatric population and are typically due to straddle injuries [28,29]. Straddle injuries may be associated with falls from bicycles, monkey bars, tree limbs, or ladders. Injuries may also be due to foreign body insertion or blunt trauma associated with motor vehicle collisions. Pelvic fractures may result in both penetrating injury due to bone spicules and traction injury due to shear forces [30]. Due to the proximity of the urethra and vagina and the susceptibility of the urethrovaginal septum to injury, traumatic urethral injuries in girls should prompt an evaluation for associated vaginal injuries [31].

Diagnosis

Prompt diagnosis is essential to avoid fistulae, stenosis, or other long-term complications of unrecognized vaginal injuries [32,33]. Physical exam may show evidence of labial bruising, bleeding at the introitus, vulvar edema, or hematuria. Patients with pelvic fractures should also be evaluated for urethral injury [28]. Examination in the prone position with the knees to the chest allows for adequate examination of the hymen, vagina, and anus without significant patient discomfort [34].

Examination in the emergency room can underestimate the extent of injuries. Lighting may be inadequate and patients are often unable to fully cooperate with a sensitive or uncomfortable exam, leading to an incomplete examination [35]. Although an examination may be attempted with oral or intravenous sedation in the emergency department, prior studies have suggested a significant increase in the extent of injuries identified on exam under anesthesia as compared to evaluation in the emergency department [35]. Based on these findings, exam under anesthesia should be performed when any doubt exists as to the extent of the injury. Relative indications for evaluation under anesthesia include a young or uncooperative patient, inability to see the full extent of the injury, vaginal hemorrhage, expanding hematoma, or concomitant injuries which require examination under anesthesia [36].

Treatment

Operative management is seldom required for genital trauma in young girls [29]. However, sexual abuse or assault may be identified in up to 25% of girls with genital injury [37]. Cystoscopy, vaginoscopy, and rectal exam should be performed to fully evaluate associated injuries [1]. Continuous flow of vaginoscopy aids in complete evaluation of the vagina, allowing for removal of foreign bodies and coagulation of isolated mucosal bleeding [38]. Gentle coaptation of the introitus during endoscopic examination with irrigation allows for better distention and visualization of the vaginal vault.

For more extensive vaginal lacerations, primary repair should be performed if possible, to reduce the rate of vaginal stenosis and urethrovaginal fistulae [31]. Vaginal lacerations may be repaired in layers with absorbable sutures (Figure 36.4). Perioperative use of antibiotics may help reduce the risk of secondary infection and wound dehiscence. Postoperative care includes the avoidance of extremely lower extremity abduction, sitz bathing, and the use of topical antibiotic ointments. For injuries that extend into the introitus, the use of permanent monofilament sutures in an interrupted fashion is recommended to reduce post-repair dehiscence risk. Long-term follow-up is essential to rule out postpubertal development of vaginal stenosis or hematocolpos, especially in patients with extensive urethrovaginal injuries [39].

Associated rectal injuries

Although rectal injuries are associated with increased morbidity in the adult population, they tend to be rare and less severe in children. Prolonged delay in diagnosis and presence of associated anorectal injury may increase postoperative complication rates in children with traumatic genital injuries. Complications in these patients include wound infection, dehiscence, and fistulae [40]. To minimize these risks, patients with a delayed diagnosis of genital injury with concomitant anorectal injury may require temporary diverting colostomy [41].

Sexual abuse

Sexual abuse has been defined as “the engaging of a child in sexual activities that the child cannot comprehend, for which the child is developmentally unprepared and cannot give informed consent, and/or that violate the social and legal taboos of society” [42]. It is the most common etiology for genital trauma in the pediatric population [43]. Approximately 10% of all reported child abuse is associated with sexual abuse. Up to 25% of girls and 10% of boys will undergo some form of sexual abuse by the age of 18 years [34]. However, studies suggest that less than 6% of sexual child abuse is reported [44]. Risk factors for abuse include children living without one or both of their biologic parents, those living with a stepfather, those with mothers who



(a)



(b)

Figure 36.4 (a) Perineal laceration without associated rectal or hymenal involvement after straddle injury. (b) Postoperative image after multilayered closure with deep vicryl and superficial chromic sutures.

are disabled or ill, and those whose parents have a significant amount of conflict in their relationship. Paternal violence may also be a risk factor for abuse. Socio-economic status, parental education level, and ethnicity are not significant risk factors for abuse [45].

A thorough history is essential to evaluate potential abuse. Separate accounts should be obtained from the victim as well as any witnesses and family members present. Inconsistencies between caregivers, the child, witnesses, and clinical findings may be subtle to discern but are important indicators of abuse [46]. Children are often reluctant to discuss their experience due to a sense of fear, shame, or guilt [47]. In interviewing the child, a supportive environment should be created, and a thorough history should be obtained using simple, open-ended, nonleading questions. Any spontaneous admission of abuse should be documented using the child's exact words, as these statements may be later accessed in legal proceedings [48].

Once abuse is suspected, child protection services should be contacted to assure protection of the child from potential future abuse. If more intense investigation is required, an individual with experience in this area should be designated to avoid repetitive questioning of the child [34]. Many medical centers have designated health care providers who are part of a child protection program. Their input and expertise can be invaluable in these situations given the medicolegal implications in this setting.

Physical exam should include a general physical exam as well as a thorough genital and anal exam. The reliability of the physical exam is dependent on the experience of the physician performing the exam [49]. The value of a second exam by a specialist adds significant value as approximately 23% of patients have their diagnosis changed or modified [50]. The exam should include inspection of the thighs, labia, clitoris, urethra, hymen, and posterior fourchette in girls. In boys, the thighs, scrotum, penis, and urethral meatus should be evaluated [34]. Cultures for sexually transmitted diseases should be obtained at the time of examination. Examination should be performed urgently as partial tears of the hymen, as well as abrasions and contusions, may heal quickly leaving little or no sign of previous injury [51,52]. Only 5% of children with a history of sexual abuse have an abnormal ano-genital examination, emphasizing the need for a high index of suspicion and highlighting the elusive nature of the diagnosis [53].

All findings should be carefully documented with photographs or drawings if necessary. Thorough documentation is critical and cannot be overemphasized. The use of colposcopy allows for magnification and improved lighting to detect subtle abnormalities [34,54]. It may also allow for simultaneous recording of the examination for documentation purposes. Although most children will tolerate examination in the emergency department, examination under anesthesia should be used if a speculum examination or more invasive evaluation of the urinary tract, vagina, or rectum is required [34,47]. The use of a Wood's lamp to aid in semen detection by UV fluorescence has been described, but is unreliable as a screening tool due to the variability of semen fluorescence with time and the inability to distinguish semen from urine, surgilube, and other commonly-used products [55,56].

Findings suspicious for abuse in girls include vaginal discharge and hymenal abnormalities, including attenuation, irregularity, scarring, mounding, or absence [49]. Also suspicious are vulvar and perihymenal erythema, friability or adhesions of the posterior fourchette, and vaginal synechiae. Hymenal bumps, rounding and notching may be normal variants and are not an indication of abuse in the absence of other findings [54]. In boys, suggestive findings include asymmetric rectal scars or tags, asymmetric rectal folds, venous engorgement, and perineal bruises or abrasions. Isolated genital burns are rare due to protection provided by the thighs and abdomen and should raise concern for abuse, with up to 46% of children with isolated burns having a suspected or confirmed etiology of abuse [57–59].

Isolated rectal injuries, particularly those occurring at the 12 or 6 o'clock positions, are significantly more common in victims of abuse compared to those with accidental injuries [58]. However, the majority of children under the age of 10 years who have suffered sexual abuse will have a normal genital examination [54].

Treatment should include management of any physical injuries identified on exam. In addition, social support services and psychiatric treatment should be made available to both the patient and family members as the morbidity of sexual abuse includes a significant psychological component [60]. Screening tools for prompt investigation in the emergency room have been reported but have yet to be validated [61].

Conclusion

Because pediatric genital trauma occurs less frequently than adult genital trauma, the involvement of experienced health care providers and subspecialists is needed to optimize care for these patients. The use of adjunct imaging studies such as ultrasonography has greatly improved our ability to accurately diagnose underlying conditions in this group of patients. However, the standard of care remains examination under anesthesia and surgical exploration when any doubt exists as to the

underlying diagnosis, and certainly when surgical intervention will reduce post-trauma morbidity, lead to organ preservation, and reduce long-term complications. Although trauma classification systems and algorithms have been developed for urinary tract injuries, few classification systems have been proposed for pediatric genital trauma [40], resulting in significant variability in the evaluation of genital trauma [62]. Multi-center studies and evaluation of trauma registries are needed to better elucidate the etiologies and best treatment practices for pediatric genital trauma.

DOS AND DON'TS

Do

- Manage superficial penile lacerations conservatively with topical antibiotic cream and clean, dry dressings
- Use ultrasonography as an adjunct investigation in cases of acute scrotum or scrotal trauma
- Perform early scrotal exploration if concerned for testicular torsion or testicular rupture
- Perform an exam under anesthesia in cases of vaginal injury to fully assess the extent of injury and rule out associated injuries

Don't

- Assume that a normal pelvic exam or minor genital trauma reliably excludes sexual abuse

CLINICAL PROBLEM/CASE: DISCUSSION

- 1 Examination in an emergency room setting is inadequate to fully assess the extent of the extensive anterior vaginal laceration shown in Figure 36.1. Studies have shown that blunt urogenital trauma consistently underestimates injury severity. Examination under anesthesia should be performed.
- 2 In addition to examination under anesthesia, cystoscopy, vaginoscopy and rectal exam should be performed to fully evaluate associated injuries such as urethral and rectal tears. A catheter should be left in place if there is evidence of urethral injury. Coaptation of the vaginal introitus allows for gentle distention of the vagina and optimal inspection of the vaginal vault and cervix.
- 3 Primary repair with layered, absorbable sutures should be performed to minimize the rate of vaginal stenosis. Perioperative antibiotics and copious irrigation should reduce the possibility of wound infection. Figure 36.5 shows how closure of the anterior dead space draws the displaced urethra back into a normal anatomic position and vastly improves the cosmetic appearance.
- 4 Long-term follow-up after a significant vaginal injury is essential to rule out postpubertal development of vaginal stenosis or hematocolpos.

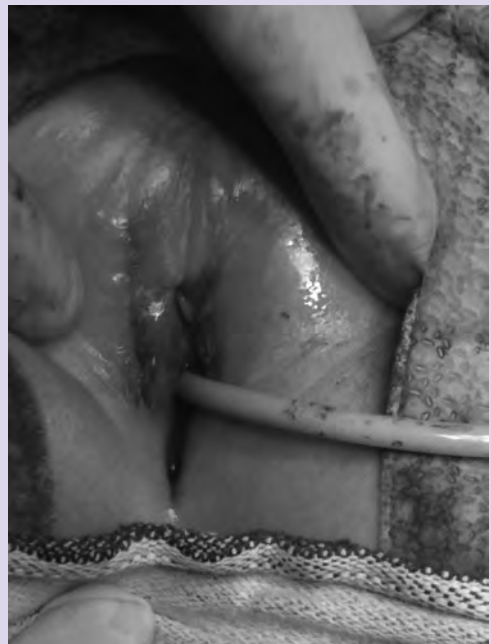


Figure 36.5 Postoperative appearance of injury after primary closure.

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CHAPTER 37

Urinary tract trauma

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KEY POINTS

- The majority of injuries to the kidney are blunt in nature
- Over the last 20 years, management of most pediatric renal trauma has shifted to a more conservative initial approach
- Accurate staging is necessary by means of a computed tomography (CT) scan for nonoperative management to be feasible
- Management goals are to preserve renal tissue and kidney function without significantly increasing morbidity and mortality risks to the child
- Early complications of renal injury include bleeding, urinoma, infection, devitalized tissue, and renovascular compromise
- Late complications include hypertension and loss of renal function

CLINICAL PROBLEM/CASE

An 8-year-old boy is brought to the emergency room 2 hours after being an unrestrained passenger in a high-speed motor vehicle crash. He is hemodynamically stable and there are no other apparent intra-abdominal visceral injuries.

- 1 What is the grade of the renal injury?
- 2 What are the treatment options for the injury?
- 3 What is the likelihood that a nephrectomy will be performed?
- 4 Are there any long-term sequelae that merit follow-up?

Introduction

Traumatic accidents are responsible for almost half of childhood deaths between the ages of 1 and 14 years. Motor vehicles are most commonly involved, with pedestrian accidents being a leading cause in the 5–9-year age group [1,2]. Injuries to the urinary tract and associated complications remain a source of significant morbidity following trauma in the pediatric age group. The kidney is also the most commonly injured abdominal organ in children.

Renal trauma

The majority (85–97%) of injuries to the kidney are blunt in nature. Penetrating trauma causes 3–15% of renal injury and is responsible for the majority of kidney trauma that requires surgery in children [3]. Some common causes of renal trauma in childhood are presented in Figure 37.1. Organized sports are an uncommon cause of serious renal injury in childhood.

When compared to adults, children are less frequently affected by penetrating injuries. However, the incidence

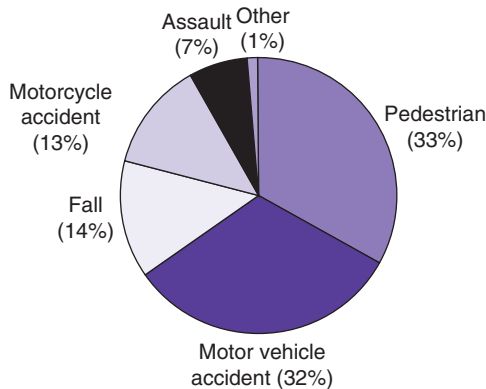


Figure 37.1 Etiology of childhood renal injury.

is on the rise, and reviews of firearm injuries in children show a significant rise in deaths due to firearms in recent decades [4]. Gunshot wounds are peculiar because they result in a “blast effect” with widespread damage away from the tract of the projectile. This may result in delayed tissue necrosis in areas that initially appear viable at the time of surgery. These patients can later present with bleeding, urinary extravasation, and abscess formation.

Imaging and staging

Radiographic imaging must be done efficiently and with accuracy so that important resuscitation measures are not interfered with in any way. Accurate staging is necessary, preferably by means of a spiral computed tomography (CT) scan in major renal injuries for nonoperative management to be feasible. A renal protocol CT scan consisting of noncontrast, contrast, and delayed phases makes it possible to evaluate the renal blood vessels, parenchyma, and the pelvicalyceal system, respectively. Box 37.1 shows the widely-accepted grading scale for traumatic renal injuries.

All patients who sustain a penetrating injury to the abdomen, flank, or torso must have further imaging to rule out significant renal injury. Previously, it was recommended that any child with a history of blunt trauma with any amount of hematuria should undergo further imaging. Recently, it has been noted that major injuries to the kidney are rare in children following blunt trauma in the absence of multisystem trauma, gross hematuria, or substantial microscopic hematuria (<50 RBC/hpf) at presentation [5–7].

Box 37.1 Grading of renal traumatic injuries.

- I** Renal contusion or subcapsular hematoma
- II** Non-expanding perirenal hematoma, <1 cm parenchymal laceration, no urinary extravasation, all renal fragments viable
- III** Non-expanding perirenal hematoma, >1 cm parenchymal laceration, no urinary extravasation, renal fragments may be viable or devitalized
- IV** Laceration extending into the collecting system with urinary extravasation, renal fragments may be viable or devitalized or injury to the main renal vasculature with contained hemorrhage
- V** Completely shattered kidney, multiple major lacerations of >1 cm associated with multiple devitalized fragments or injury to the main renal vasculature with uncontrolled hemorrhage, renal hilar avulsion

If the CT scan cannot be done because the child needs immediate exploration, then a single-shot intravenous pyelogram (IVP) is recommended. It serves to identify and document a normally functioning contralateral kidney if a situation arises where the involved kidney needs to be removed. A single-shot IVP can also provide important information about the anatomy and function of the involved kidney and limit the amount of exploration necessary. Sonography can also be valuable in the bedside evaluation of the severely injured child. This avoids the use of intravenous contrast which could potentially be nephrotoxic in a severely volume-depleted child.

Initial management and outcomes

Management goals are to preserve renal tissue and kidney function without significantly increasing morbidity and mortality risks to the child. The initial emphasis is on a prompt assessment of the condition concurrent with respiratory and hemodynamic stabilization of the patient.

Blunt injuries are mostly minor in nature and anatomically represented by contusions, minor perirenal fluid collections, and uncomplicated lacerations (grades I–III). They comprise 70–85% of pediatric renal injuries [3]. Management is usually conservative and involves bed rest and observation until gross hematuria clears and limited activity until microhematuria clears. The length of convalescence is from 2 to 6 weeks and is usually free of complications such as loss of renal function, hypertension, or hydronephrosis [8].

The management of major renal injuries (grades IV and V) is controversial and needs to be individualized. Over the last 20 years, management of most pediatric renal trauma has shifted to a more conservative initial approach. Early surgical intervention may lead to increased nephrectomy rates (up to 89%) and subsequent complications [9]. It has been noted that the complication rates are higher for conservative management of parenchymal lacerations of the kidney with significant (25%) devitalized renal tissue [10,11]. The challenge lies in the early detection and prompt evaluation of the severity of injury. Selective nonoperative management may be appropriate in the hemodynamically stable child, but only after accurate staging of the injury by a CT scan. Most centers have adopted an ICU protocol for children with a severe renal injury undergoing initial conservative management. Bed rest for the first 24–48 hours is mandatory, as well as continual monitoring of hematocrit, blood pressure, and clinical signs. The risk of bleeding is highest during this time period. Nonoperative management historically was thought to lead to surgery in a significant number of patients for complications of trauma such as bleeding, renal infarction, or segmental hydronephrosis [2].

The nephrectomy rate increases if surgery is necessary on other intra-abdominal organs, suggesting that the kidney may be removed as a damage control measure or due to insufficient experience with renal reconstructive techniques. Possibly, under these conditions, potentially salvageable kidneys are sacrificed to control hemorrhage. A recent review of the National Trauma Data Bank regarding grade IV and V renal injuries revealed that the risk of nephrectomy was consistently decreased in children managed conservatively regardless of injury characteristics [12].

Relative indications for exploration are significant urinary extravasation, nonviable renal tissue, arterial injury, and incomplete staging of the injury or significant extrarenal injuries. Partial nephrectomy is the operation of choice in this condition. Surgical management involves the preliminary control of the vessels of the kidney as well as the aorta. This is followed by debridement of devitalized tissue, hemostasis, closure of the collecting system, and repair or coverage of tears in the renal parenchyma. Initial trial of a JJ stent or a nephrostomy tube may be used in children with significant urinary extravasation. Initial conservative management of stable children with grade IV renal injuries

has been reported with close CT follow-up to rule out urinoma [3,8]. A low threshold for placing a ureteral stent should be maintained if there is evidence for ongoing urinary extravasation to avoid septic abdominal complications.

It is generally accepted that those with major renal injuries such as a vascular pedicle injury or a shattered kidney will require surgery. In most cases, these injuries are severe enough to warrant a nephrectomy, particularly considering the high incidence of life-threatening extrarenal injuries that these patients suffer. Results of emergency vascular repair for a pedicle injury or thrombosis in children are usually poor, particularly with the amount of warm ischemia that the kidney undergoes during the resuscitation and evaluation. An attempt at repair is generally limited to extreme situations such as a solitary kidney or bilateral injuries.

The outcome differs between blunt and penetrating injuries. Overall, patients in the penetrating injury group are more severely injured, have a higher 24-hour transfusion requirement, and have a higher nephrectomy rate. The rate of surgery is higher in penetrating injury, around 36% and, in contrast to blunt trauma, approximately half of these result in operative repair of the kidney [3,9,13]. Recent large series quote renal salvage rates of more than 98% in pediatric blunt kidney injuries with conservative management and selective surgical exploration when indicated. The expected exploration rate is under 10% [1,3,8]. It is almost universal in grade V injuries and the renal salvage rate in this group of patients is around 30% in the long term [14,15].

Early complications

Bleeding

The risk of bleeding is highest in the first 48 hours after the trauma and close observation is necessary during this phase. Strict bed rest and serial hematocrit monitoring is imperative. Observation may be continued if the child is clinically stable and the CT scan shows a stable hematoma (or stable amount of urine extravasation). Bleeding can recur even later and grade IV–V injuries generally need to have a follow-up imaging at around 3–6 months to document complete healing.

If at any time during the course of treatment the child shows signs of hemodynamic instability (hypovolemia with severe hypotension or is unresponsive to packed

red cells), then consideration should be given to either surgical exploration or to angiography and embolization. On exploration, expanding pulsatile or uncontained retroperitoneal hemorrhage indicates persistent bleeding. Primary angioembolization has been found to be useful in the treatment of isolated grade IV renal injuries with segmental artery bleeding or in the case of a grade V injury with a pseudoaneurysm [16]. It also has a role to play in delayed hemorrhage in grades II–IV renal injuries. It can be done only in a nonrenal failure patient with a definable segmental artery injury.

Urinary extravasation/urinoma

Extravasation of urine is a sign of a major renal injury resulting from a laceration of the renal pelvis, a parenchymal tear extending into the collecting system, a forniceal rupture, or an avulsion of the ureteropelvic junction (UPJ). Persistent urine extravasation can lead to urinoma formation, perinephric infection, and renal loss. UPJ disruption is very rare, being associated with forces of rapid deceleration involved in motor vehicle trauma and falls from heights. It is detected when there is major contrast leak in the medial and perirenal areas and the ipsilateral ureter is not visualized. Complete disruption needs immediate operative intervention. Pre-existing but heretofore unknown urinary tract anomalies such as a UPJ obstruction may predispose the kidney to injury.

It has been reported that almost 75% of children, even with a severe grade IV laceration, had spontaneous resolution of the urinoma, therefore a conservative approach has been suggested in these patients [15,17,18]. Possible long-term complications of a large urinoma include retroperitoneal fibrosis, pelviureteral and infundibular obstruction, infection, and hypertension. Initially, children are treated with parenteral antibiotics followed by the appropriate oral antibiotic. Monitoring by CT scan is essential. Symptomatic or worsening urinary extravasation can be managed either by percutaneous drainage of the collecting system or internal (JJ) stenting. It has been recommended that if resolution of extravasation does not occur within 2 weeks of conservative management, then ureteral stenting may be done. These measures have been found to be helpful in this scenario, more than percutaneous drainage of the urinoma itself. Stents may need to be maintained for as long as 10 weeks and bladder drainage for as long as 2–3 weeks. Patients who failed a conservative approach

with or without endoscopic drainage (less than 10%) require open operative intervention. Half of these operations result in nephrectomy [15,19].

Infection

Perinephric abscess can result from a hematoma, urinoma, or devitalized fragment of kidney. The incidence may be increased by concomitant pancreatic or bowel injuries. It may need intervention either by the percutaneous or open routes to achieve drainage. Sepsis commonly affects those with multi-organ trauma and judicious initial use of antibiotics is advised.

Nonviable tissue

Large devitalized fragments (involvement of 25–50% of affected kidney) are associated with significant complications when managed nonoperatively. Nonviable tissue can result from both blunt and penetrating trauma, and it can lead to short-term complications such as persistent urine leak and abscess (which might require surgical intervention) and, in the long term, hypertension [10,11,15,19,20]. Immediate surgery has been shown to reduce the morbidity in these patients. Therefore, it is suggested that renal injuries with large devitalized fragments and associated urine extravasation or a retroperitoneal hematoma to undergo surgical exploration [21]. The surgical procedure of choice is a partial or polar nephrectomy. Delayed necrosis with possible fistula formation is more likely to happen in penetrating trauma, especially if it is due to a gunshot. The blast effect may lead to delayed tissue necrosis resulting in bleeding, urine leak, or an abscess in areas which may be viable at time of initial presentation.

Renovascular complications

Renal vascular injury has been reported in 9–31% of children with renal trauma, the incidence being nearly twice that seen in adults [22]. Detection of pedicle injuries is generally delayed and repair beyond 14 hours reduces the chances of renal salvage. These injuries can involve either main or segmental renal vessels and are classified as avulsions, lacerations, or occlusions (secondary to thrombosis or dissection).

Traumatic occlusion of the main renal artery

Deceleration injuries cause the intima of the main renal artery to rupture, since it is low in elastic fiber content (the muscularis and the adventitia are more flexible).

Intimal disruption may create a subintimal false lumen, resulting in decreased renal blood flow, renal ischemia or infarction, hypertension, or arterial occlusion due to a thrombus. This can be diagnosed by prompt CT scan or arteriography. Surgical revascularization may be considered only in hemodynamically stable patients with a warm ischemia time of >5 hours [22]. These patients frequently have multiple injuries and the mortality rates are high. Therefore, doing a revascularization procedure is not often practical and a nephrectomy is usually done due to time constraints and to control hemorrhage.

Late complications

Renin-mediated hypertension

Hypertension (early or late) in children with renal injury is rare. It arises through activation of the renin angiotensin system from renal ischemia. Overall incidence of 1–2% has been reported in large series [22–26]. A restrictive fibrous capsule may develop around the injured kidney, reducing the renal blood flow by compression of the parenchyma (Page kidney). This is usually due to a prolonged urine leak or an inadequately treated urinoma. Other mechanisms are stenosis or occlusion of the main renal artery or one of its branches or the development of a post-traumatic arteriovenous fistula. Medical management is usually successful but surgery may be necessary. Surgical options include vascular or endovascular reconstructions, capsulotomy, or nephrectomy. Long-term monitoring of blood pressure is recommended. A prospective study is needed to assess the precise prevalence of hypertension after renal injuries and to define the length of follow-up needed to assume the patient is no longer at risk [27].

Loss of renal function

The goal of management of renal injury in children is to preserve functioning renal parenchyma. Renal nuclear scans (DMSA) may be done to document and track recovery. Preservation of renal tissue is less successful in children with renovascular trauma as well as severe concomitant injuries with shock and extensive blood loss. Conservative management of minor renal injuries (grades I–III) usually result in almost complete healing. Grades IV and V show some evidence of volume loss (22% and 50%, respectively) [25,26]. Surgical reconstruction after major blunt or penetrating trauma preserves more than one-third of the kidney in 81% of cases

[23]. The incidence of post-traumatic renal failure is low [26]. It has been reported in 6.4% of patients with renal injury who also have associated renovascular injury [25].

Morphological abnormalities

Conservative management of high-grade renal injuries in children can result in residual morphological changes such as single or multiple scars, cystic lesions with septae or segmental hydronephrosis. Figure 37.2 shows a follow-up ultrasound 5 years after a grade IV renal injury managed conservatively with urinary diversion by percutaneous nephrostomy.

Ureteral injuries

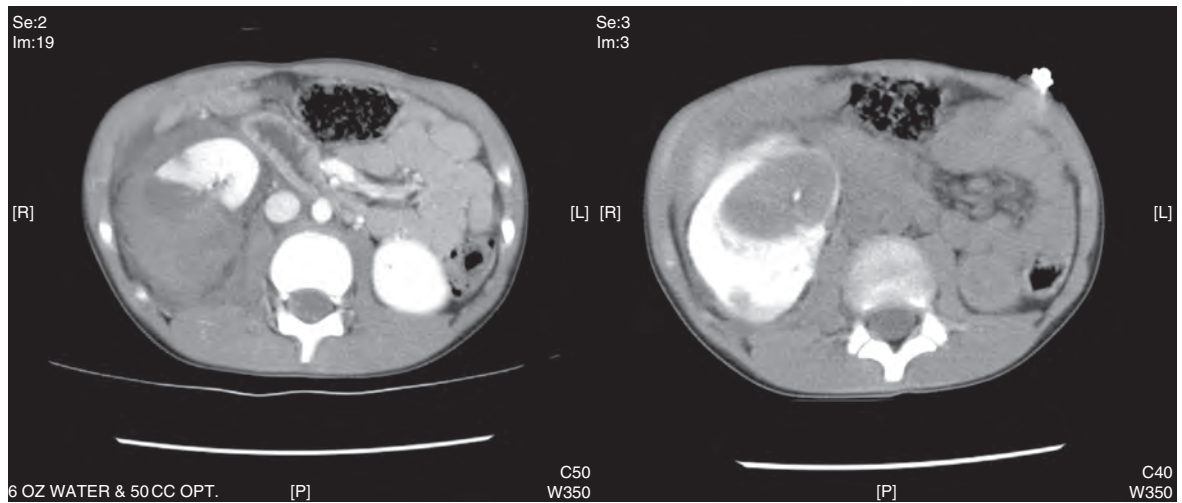
Isolated ureteral injuries are rare and constitute around 3% of genitourinary trauma [28]. Blunt traumatic disruption of the upper ureter and ureteropelvic junction, though very rare, is more common in children than in adults by a ratio of 3:1. This should always be suspected after severe blunt trauma or penetrating injury.

Penetrating injuries are extremely rare in children and are almost always accompanied by injuries to other organs. Gunshot wounds are also rare in children, but the mechanism of injury makes the ureter a target even if it is not in the path of the bullet. Iatrogenic injuries to the ureter have increased since the advent of laparoscopic and ureteroscopic interventions in childhood. They also happen during pelvic surgical procedures. Common injuries are mucosal injury during ureteroscopy with possible ureteral perforation, false passages, complete avulsion, or loss of a ureteral segment.

Diagnosis and management

Initially, these injuries may be unrecognized and a high index of suspicion is required in their evaluation. It is essential to evaluate for ureteral injury in any child with significant blunt abdominal trauma and multiple associated injuries. Though rare in the child, any penetrating injury should be suspected of injuring the ureter as well.

CT scan is the primary method of evaluation in these patients with multiple injuries. Extravasation of contrast may be confined to the medial perirenal space. If a rapid sequence spiral CT is done, then delayed films must be performed. On delayed images there is absence of contrast material in the distal ureter if there is complete ureteral transection. A complete high-dose intravenous urogram can be done in the resuscitation suite. The findings include contrast extravasation,



(a)



(b)

Figure 37.2 (a) Computed tomography scan of a right high-grade renal laceration with retroperitoneal hematoma in a 7-year-old boy (left). Delayed images of the same kidney are shown on the right. (b) Ultrasound showing cystic changes in the upper pole of the right kidney 5 years after conservative management of the injury.

delayed function, or mild ureteral dilation or deviation (Figure 37.3). Retrograde ureterography is probably the most accurate method of assessment of ureteral integrity, but is sometimes not practical in acute trauma. Intraoperative evaluation can be made by inspection of the ureter, with the aid of an injection of methylene blue into the collecting system or intravenous indigo-carmin can be used.

The timing and type of the intervention depends on proper injury staging, the patient's overall condition, and timing of diagnosis. When recognized early, the ureter should be repaired immediately. Repair should

be by means of a tension free, spatulated anastomosis. If possible, the repair should be wrapped with omentum or retroperitoneal fat. Stent or nephrostomy placement is advised in all cases. Surgical options are direct reanastomosis, ureteral reimplantation (with possible vesico-psoas hitch or Boari flap), transposition of ileum or appendix, transuretero-ureterostomy, and autotransplantation of the kidney. Nephrectomy may be done only in a life-threatening situation with a normal contralateral kidney.

If immediate definitive repair is not possible, then one should wait for several months before attempting

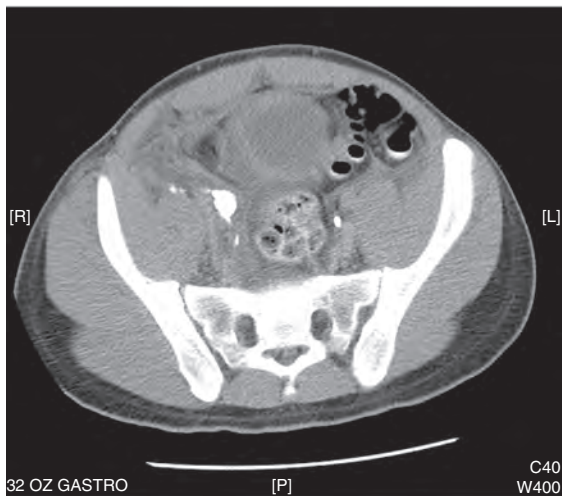


Figure 37.3 Computed tomography scan of a right ureteral injury from penetrating trauma in an 11-year-old boy.

reconstruction. In the case of an unstable patient or if there has been a delay in diagnosis, a temporary diversion may be done. Percutaneous nephrostomy with or without ligation of the ureter or a cutaneous ureteros-tomy may be done initially for damage control.

Most iatrogenic injuries are diagnosed intraopera-tively. Identification of the ureters by passing ureteric catheters prior to a difficult abdominal or pelvic dis-section is a valuable aid to prevent these injuries. Pass-ing a safety guidewire prior to ureteroscopy is helpful in maintaining access through the ureter during the procedure. Trying to retrieve large fragments of stones without breaking them up can lead to avulsion of the ureter. Most ureteroscopic iatrogenic injuries and their complications can be managed by JJ stenting for a short period of time.

Complications

Urinary extravasation can present as an enlarging flank mass in the absence of signs of bleeding. The initial management of a JJ stent or a percutaneous nephros-tomy is appropriate. Urinoma or abscess may be drained percutaneously. Most patients heal without stricturing.

Ureteral injuries, especially with a delayed diagnosis, may be complicated by ureteral strictures. Usually they can be managed by balloon dilatation, internal stent-ing, or endoureterotomy. Stricture length and duration determine whether conservative management is going

to be successful. These methods have a higher failure rate with longer strictures. Previous infection, urine extravasation, and poor blood supply may be factors causing longer strictures. Open or laparoscopic repair, if required, should be delayed for 1–3 months while infec-tion and inflammation subside.

Hydronephrosis is due to transient obstruction as a result of contusion or a stricture. Ureteral stenting is usually adequate treatment unless there is a long stric-ture. Ureterocutaneous and ureterogenital fistulae can occur later in the course of illness and may respond to a period of stenting during which the fistula undergoes spontaneous healing. If stenting with or without proxi-mal diversion is unsuccessful, open repair is needed. Renal failure and anuria may occur in children with bi-lateral injuries or when a ureter of a solitary kidney is affected. Temporary dialysis with the appropriate drain-age procedure may be done.

Bladder injuries

Pediatric bladder injuries are rare and are usually as-sociated with severe pelvic injuries with bony fractures. Blunt trauma is the most common cause of bladder in-jury in children. Inappropriately fastened lap belts in-crease the risk of this injury. Iatrogenic injury is also known, especially with the increasing use of laparo-scopic surgery. Augmented bladders and children with previous pelvic surgery are also at greater risk. An aug-mented bladder may perforate spontaneously or with trauma [29].

Bladder rupture may be intraperitoneal or extraper-itoneal or a combination of the two. Compared with adults, the bladder in a child occupies a more abdomi-nal position when full. It is therefore more susceptible to external injury and intraperitoneal rupture accounts for one-third of bladder injuries [30]. Extraperitoneal rupture almost always occurs due to pelvic fracture. It has been reported that bladder injury in children with a pelvic fracture is rare and occurs 1% of the time [31]. This lower incidence in children has been attributed to the elastic nature of a child's pelvis and its attachments.

Diagnosis and management

Diagnosis depends on precise studies including CT scan, cystography, and IVP. The bladder must be adequately filled, and oblique as well as post-drainage films should

be taken. Currently, CT cystography is a very sensitive and specific test. Children with intraperitoneal rupture develop hyponatremia, hypokalemia, elevated serum urea, and creatinine (urea nitrogen rises out of proportion when compared to creatinine), whereas those with extraperitoneal rupture do not do so. Hematuria is a cardinal sign and the patient may be unable to void.

The majority of injuries that present solely with hematuria are bladder contusions and require no specific treatment. Most extraperitoneal injuries may be treated by bladder drainage, either urethral or suprapubic, for 7–10 days. If bladder drainage is not efficient in the first 48 hours, then open repair should be done and a suprapubic tube placed. It has been recommended that these tears may be treated by catheter drainage if the urine clears of blood promptly, the catheter drains well, and the bladder neck is not involved in the laceration.

Major extraperitoneal injuries and intraperitoneal injuries are usually treated by primary operative repair. Prophylactic antibiotics need to be started on the day of the injury to prevent infection of the associated pelvic hematoma and continued until 3 days after catheter removal. Major extraperitoneal injuries include open pelvic fracture, rectal perforation (both of which have a high risk of infection when treated conservatively), and bone fragment projecting into the bladder, which is rare. When a laparotomy is done for other associated injuries, it is advised that the bladder be opened and repaired if a bladder injury is suspected. Massive injuries of the bladder and lower ureter are best drained by a temporary diversion. Occasionally, nonoperative treatment can be successful in cases of small intraperitoneal bladder tears in children [32]. The treatment consists of bladder drainage, percutaneous intraperitoneal tube

drain, and antibiotics in children who present early. Surgery is done in these children if bladder drainage is inadequate, there is prolonged drainage from the peritoneal drain, or if the clinical situation deteriorates. A follow-up cystogram must be obtained 10–14 days after the injury in the case of conservative management, or 7–10 days after bladder repair. Only then is the bladder catheter removed.

If there is involvement of the bladder neck, trigone, prostate, or vagina in the injury, immediate formal repair of all these structures is necessary. This type of injury is more common in children and early repair prevents complications. A small vaginal injury may be left open. A large laceration may need repair through the opening in the bladder. The bladder is then drained by both urethral and suprapubic tubes. Placing omentum between the bladder and vaginal repairs may reduce the risk of fistula formation.

Complications

Complications of bladder perforations include clot retention, ileus, pelvic abscess, and urinary fistula. Other complications are urge incontinence and areflexic bladder that might require intermittent catheterization. Bladder stones may occur from retained sutures. Pseudodiverticulum can occur due to a bony spike. An acute complication of intraperitoneal rupture is peritonitis, which can be fatal. Late complications of bladder repair are rare. Acute, self-limiting urinary frequency is common. Bladder neck injuries can result in bladder neck stricture and urinary incontinence. In females, they can also result in stress incontinence, sexual dysfunction, and vesicovaginal fistula. Impotence in males is related to wide separation of the pubic bones.

DOS AND DON'TS

Do

- Perform complete and accurate staging of renal injuries by computed tomography (CT) when considering nonoperative management.
- Document normal contralateral renal anatomy by single shot intravenous urogram (IVU) if emergent abdominal exploration is indicated.
- Monitor for late renin-mediated hypertension in cases of renal injury managed nonoperatively.

Don't

- Delay proceeding with placement of a ureteral stent or nephrostomy tube in grade IV renal injuries with ongoing urinary extravasation.

CLINICAL PROBLEM/CASE: DISCUSSION

- 1 The left kidney does not uptake intravenous iodinated contrast in this delayed image of a CT scan. This implies a vascular pedicle injury and, by definition, this would be a grade V renal injury. Deceleration injuries cause the intima of the main renal artery to rupture, since it is low in elastic fiber content (the muscularis and the adventitia are more flexible). Intimal disruption may create a subintimal false lumen, resulting in decreased renal blood flow, renal ischemia or infarction, hypertension, or arterial occlusion due to a thrombus. This can be diagnosed by prompt CT scan or arteriography.



- 2 In most cases, these injuries are severe enough to warrant a nephrectomy, particularly considering the high incidence of life-threatening extrarenal injuries that these patients suffer. Surgical revascularization may be considered only in hemodynamically stable patients with a warm ischemia time of less than five hours.
- 3 Results of emergency vascular repair for a pedicle injury or thrombosis in children are usually poor, particularly with the amount of warm ischemia that the kidney undergoes during resuscitation and evaluation. An attempt at repair is generally limited to extreme situations such as a solitary kidney or bilateral injuries.
- 4 If the kidney is successfully salvaged surgically, then long-term monitoring for hypertension is indicated. If the kidney is removed, then long-term assessment for hyperfiltration injury to the remaining solitary kidney should be considered.

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PART X

Surgery for Urinary Incontinence

CHAPTER 38

Augmentation cystoplasty and diversion

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KEY POINTS

- Preoperative counseling is important so that patients and family understand the lifelong potential side effects of inclusion of the bowel in the urinary system and potential short- and long-term complications
- Patient compliance with clean intermittent catheterization and irrigation are important points to emphasize in order to minimize problems associated with urinary tract infections, mucus, and stones
- A high index of suspicion is required in patients after augmentation cystoplasty to identify perforations early and limit morbidity and mortality. A CT cystogram is the gold standard for diagnosis
- Regular life-long follow-up is necessary for all augmentation cystoplasty patients in order to identify and manage the expected side effects associated with the use of bowel in the urinary tract and to prevent long-term complications

CLINICAL PROBLEM/CASE

You are contacted by an outside ICU physician to discuss a 14-year-old male myelomeningocele patient who was admitted to their ICU 2 days ago with urosepsis. The patient has not improved with IV antibiotics and has poor urine output. The physician says the patient has had some bladder surgery and does in and out catheters at home and they have continued with that while he is in the hospital.

- 1 What are important questions to ask the outside treating physician about the patient's history?
- 2 What should you tell this physician about optimal bladder drainage in this setting? Is continued in and out cathetering sufficient and appropriate?
- 3 What imaging would you want to see to help diagnosis the patient's underlying problem?
- 4 What would be your initial plan of management if he were found to have a bladder perforation?

Introduction

The objectives of this chapter are to familiarize you with the basics of an augmentation cystoplasty (AC), the common issues that can be expected postoperatively and the complications that can occur. An AC is intended to increase the capacity and improve the compliance of the bladder in patients with noncompliant dysfunctional bladders refractory to all conservative measures. Most commonly, this is necessary in children with a neurogenic bladder from spina bifida, sacral agenesis, tumors, or spinal cord injury. Emptying of the bladder

before and after AC is performed by clean intermittent catheterization (CIC) and can be done via the urethra or a concomitantly constructed catheterizable channel. Typically, AC is done using ileum. Alternatives to ileum include the use of other gastrointestinal (GI) segments like cecum, sigmoid, or stomach. Other techniques include the use of ureterocystoplasty and autoaugmentation. Use of biomaterials and tissue engineering techniques are still experimental. This chapter focuses on the use of ileum as it is the most commonly-used segment for AC. Table 38.1 is a comparison of different techniques.

Table 38.1 Comparison of four different techniques.

	Ileum/colon	Gastric	Ureter	Autoaugmentation
Outcome				
Capacity [12]	Significant increase	Significant increase	Improvement in carefully selected patients [27]	Up to 93% no improvement in urodynamic parameters
Compliance [12]	Significant improvement	Significant improvement	Improvement in carefully selected patients [27]	Poor on long-term follow-up
Continence [12]	Up to 96% improvement/resolution in one series	Up to 89% continence	Good medium-term results in up to 90%	Poor in long term
Upper tracts [12]	Stable	Stable in up to 91%	Stable	Deterioration in over 50%
Other [12]		Patch contraction, ureteral obstruction Hematuria dysuria syndrome in up to 25%. Long-term complications greater than ileum/colon	High reaugmentation rate of 82–91% where case selection in inappropriate, ureteral necrosis	Not shown to be useful with respect to continence and urodynamic parameters
Complications				
Mucus	Common	Decreased mucus production	None	None
UTI [14]	13–23%	8%	Occurs	Occurs
Stones [14]	18–30%	Decreased incidence		
Metabolic [14]	Hyperchloremic metabolic acidosis, not significant with normal renal function	Decreased chloride absorption	No	No
Perforation [21]	5–19% (highest with sigmoid colon, ileum 8%)	5%	Can occur	Can occur
Malignancy [24]	1.5% per decade	2.8% per decade		

Preoperative considerations

AC is major reconstructive surgery and alternatives to augmentation should be exhausted, including when safe and appropriate. Conservative management with CIC $\pm$ anticholinergic therapy, and/or intradetrusor injection of botulinum toxin are options. Proper education of the patient and family as to the pre-, peri-, and postoperative expectations is of the utmost importance. Many of our younger patients respond well to incorporation of a child-life or patient advocate to help explain the procedure to them using dolls that have catheters going into the belly to help these patients become familiar with their postoperative state. The patient and family should be familiar with the expectations for the need to perform CIC via the urethra or created abdominal channel, need for irrigation of mucus, and the importance of lifelong follow-up with the surgeon or a physician familiar with patients after AC.

All patients should have an upper tract evaluation with an RUS and a comprehensive lower urinary tract evaluation with urodynamics, in addition to a complete history and physical exam. This is important for the selection of GI segment or alternative techniques to use for the AC. Most commonly, this will be a standard AC with the use of ileum, but this chapter highlights the potential complications of using ileum and where alternatives may be beneficial. The necessary concomitant procedures should be planned based upon the preoperative evaluation. This should include the consideration of performing the following:

- Catheterizable stoma (to facilitate emptying the augmented bladder via CIC).
- Bladder outlet procedure (bladder neck or sling for incompetent bladder neck with incontinence).
- Bowel-emptying procedure (antegrade continent enema (ACE) may be beneficial in those patients with contaminant neurogenic bowel and chronic constipation).

- Ureteroneocystostomy (is vesicoureteral reflux (VUR) present; although the decreased storage pressure associated with a successful AC will be sufficient for resolution of VUR, in most cases of primary VUR).

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 pre-existing conditions and goals of surgery. Ileum is the most commonly-used segment in AC because of its ease of use, familiarity to the surgeon and because it has been shown to be the most compliant segment [1]. The contraindications to use of bowel for AC are intrinsic bowel disease (Crohn's disease, ulcerative colitis), short gut syndrome (i.e. some cloacal exstrophy patients), after radiation treatments in which the bowel segment of choice would have been in the field. Relative contraindications would include the use of a GI segment in which current metabolic derangements would be exacerbated (i.e. use of colon or ileum in renal failure patients with chronic metabolic acidosis) [2].

It is our preference that all patients are admitted the day prior to the procedure and follow our standardized pathway for a mechanical bowel preparation. This includes all patients who are to undergo an AC with or without a concomitant antegrade continent enema procedure. Preoperative labs are drawn at the time of initial IV placement. IV fluids are run at 125–150% of maintenance to compensate for fluid losses during the bowel prep. An NGT is placed in those patients too young to take the volume of Golytely orally. The volume of Golytely is continued until there are no solid particulates left in the resultant diarrhea. The NGT is removed as soon as bowel prep is completed. Patients are started on Reglan to facilitate gastric emptying. Neomycin is started at the initiation of the bowel prep and stopped after three doses. Preoperative antibiotic coverage is started with cefoxitin (nafcillin and gentamicin are used if the patient has a VP shunt, vancomycin and gentamicin if they are allergic to penicillin and have a VP shunt). Soap-suds enemas are given for those with significant distal impaction to facilitate emptying. The patient is allowed a clear-liquid diet until midnight.

Intraoperative considerations

Intraoperatively, care should be taken with the position of the patient and all bony prominences should be padded to prevent unintended injury during the lengthy procedure.

The ileal segment is selected with adequate quality and length of mesentery that is at least 15–20 cm from the ileocecal valve and is approximately 20–30 cm in length. The ileal segment is opened on its antimesenteric border and reconfigured in a U shape. The bladder is opened in the midline first and extended anteriorly towards the bladder neck and posteriorly in the classic clam shape but then also incised laterally from the midline on each side to show a star shape. These incisions to open the bladder widely will eliminate the possibility of an hourglass shape of the augmented bladder postoperatively. A suprapubic tube (SPT) is always placed out of the native portion of the bladder to assist in postoperative drainage.

It is our preference that an NGT tube is placed during the procedure and removed on the morning of postoperative day 1. Oral intake is initiated slowly once return of bowel function is seen clinically. 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 hours. After this has proven successful, the SPT is removed after 4 weeks.

Outcomes

With appropriate patient selection and intraoperative use of an adequate length of detubularized and reconfigured ileum the overall risk of reaugmentation secondary to persistently high pressures is low at about 5% [3]. Overall complications after major lower urinary tract reconstruction are relatively high (~40%) commensurate with the complexity of surgery and consistent with similar procedures [4]. Patients with an obese BMI are at increased risk [5]. This includes 34% that will ultimately need further surgery at the bladder level [3].

Complications

Early

Early complications that occur during the first 30 days are similar to those of major bowel and pelvic surgeries. Wound infections and dehiscence occur in 5–6.4% [6]

and, in a small study, are not proven to be increased by not bowel prepping of patients prior to AC [7]. Postoperative bleeding (0–3%), small bowel obstruction (3–5.7%) requiring reoperation, and mortality are rare [6].

Long-term

Long-term complications or side effects of the use of a GI segment in the urinary tract are commonplace and should be expected. Most importantly, the patient and family should be counseled extensively in the preoperative period about the management of mucus, stones, and metabolic derangements that are possible after AC. They also should be informed about the risk of perforation and malignancy prior to surgery.

Mucus production

Inherent to the use of the lower GI segments is the continued production of mucus. The ileum produces a mean of 30–40 g per day and the cecum may produce 50% more, and this does not appear to decrease over time [8]. Mucus mixed in the urine predisposes to UTIs, stones, and outlet obstruction, and is hypothesized to contribute to the risk of perforation [2]. Regular irrigation also has been shown to reduce complications associated with mucus production in the augmented bladder, such as UTIs and stones [9], and is a mainstay of postoperative management in our patients.

Urinary tract infections

Patients with a neurogenic bladder require CIC; this does not change after AC. One of the side effects of CIC is bacteruria. Bacteruria is common after AC (50–100%) [2] and most likely is related to CIC [4]. The urine of a patient after AC should not be regularly screened for bacteruria or treated, as asymptomatic bacteruria can be considered a colonization and not pathologic. In fact, prophylactic antibiotics and regular antibiotic irrigation are ineffective at maintaining sterile urine [10,11]. In our opinion, this contributes to bacterial resistance and makes those UTIs that are symptomatic, and clinically significant, more difficult to treat. Symptomatic cystitis is common and patients will often present with increased mucus or foul-smelling urine. In these cases, simply increasing fluid intake and bladder irrigation is often successful. However, urine cultures and treatment should be initiated with symptoms of suprapubic pain, hematuria, new or worse incontinence, or fevers. Febrile infections and pyelonephritis can occur in up to 13% of

patients after AC [12]. Rare deterioration of the upper tracts is seen if the AC maintains a low-pressure urinary system, but immunocompromised patients are at an increased risk [13]. Cultures that grow urease-splitting organisms should be treated and screened for stones.

Stone formation

Bladder stones are a common long-term problem seen in 18–30% of patients after AC [14]. The mucus production after AC is a nidus for the stone formation [15]. Those patients with an abdominal stoma are at an increased risk for stones compared to those that perform CIC via the native urethra secondary to having less-dependent drainage and therefore chronic stasis of urine [16]. A regular irrigation protocol can decrease the incidence of stones from 43% to 7%; this improvement was seen in AC patients regardless of CIC via abdominal stoma or urethra [9]. Most of the stones are struvite in these patients [17] and often associated with bacteruria associated with urease-splitting organisms. When diagnosed, these stones in an augmented bladder need surgical removal and can be approached in a number of ways. A transabdominal approach offers the ability to remove the stone intact but has the downsides of risk to the augmented segment, operative time because of potential for lysis of adhesions, and postoperative morbidity. An endoscopic route offers decreased morbidity in recovery but potential for damage to a catheterizable channel if the native urethra cannot accommodate the scope or is inaccessible. Endoscopy also limits the size of the stone that can be removed intact, therefore necessitating manual, laser, or ultrasonic fragmentation. This results in residual stones that cannot all be cleared and which serve as a further nidus for recurrent stone formation. In our hands, the most successful combination of removing the entirety of the stone but limiting the morbidity is to percutaneously access the bladder via the original SPT site with a laparoscopic trocar under direct vision through a cystoscope in a partially-filled bladder, then place all of the intact stones in a lap bag through the trocar to facilitate fragmentation without spillage of fragments. This results in decreased operative time and morbidity, and no residual fragments are left behind to serve as a nidus for recurrence [18]. We perform stone analysis and metabolic workups on our AC patients with recurrent stones and treat these urinary abnormalities in an effort to prevent recurrent stones along with a daily irrigation regimen.

Metabolic

The incorporation of gastrointestinal segments in the urinary tract can lead to systemic metabolic derangements related to the absorptive properties of the incorporated segment. Hyperchloremic metabolic acidosis from absorption of ammonium and chloride ions requiring oral bicarbonate is seen in 16% of patients and is low in those with normal renal function [6,19]. It appears that, when compiling data from the multiple conflicting reports, the potential chronic metabolic acidosis does not decrease linear growth or bone demineralization after AC [19]. However, in patients with underlying chronic metabolic acidosis from chronic renal insufficiency, the use of colon and ileum should be a relative contraindication and a gastric segment should be considered.

The risk of vitamin B12 deficiency and resultant megaloblastic anemia risks is mitigated by sparing the distal 15–20 cm of the terminal ileum, and is uncommon for the amount of ileum required for a routine AC. However, 7 years after AC, 21% of patients showed low B12 levels in one study [20], and vitamin B12 levels should be screened for in long-term patients after AC.

Chronic diarrhea is rare, but can occur if longer segments are used and the ileocecal valve is removed in the face of underlying neurogenic dysfunction (10–23%) [4].

Perforation

Perforation after AC is a potentially catastrophic complication. The incidence is 4.5–12.8% [21], without a specific segment shown to be consistently safer or more at-risk than another [4]. These patients can present at any time in the follow-up period, including more than a decade after AC. Often, they will have a history of decreased output with CIC, abdominal pain, abdominal distension, and fever. Neurogenic patients may not have typical symptoms and may present quite ill. The etiology is probably not always the same, but often is related to chronic over-distension from noncompliance or short-term gross negligence in cathing or emptying the bladder. It is unlikely to be related to single catheter trauma, although this theoretically could happen. Chronic microvascular ischemia, chronic infections and inflammation, and bladder hyper-reflexia also have all been hypothesized. Education regarding the possibility and presentation is important for families and patients. Often these patients are adolescents who have gained some amount of independence and become less

compliant. In some cases, it has been related to the patients becoming intoxicated and unable to cath despite consumption of a large volume of diuretics, resulting in over-distension and perforation.

A high index of suspicion is necessary, as patients are often misdiagnosed and mismanaged by physicians or staff unfamiliar with their surgical history and risks associated with AC. These patients can be very ill and deaths have occurred [22]. A CT cystogram has the greatest sensitivity and today is the standard for diagnosis. Initial conservative treatment often is safe and successful with maximal bladder drainage and IV antibiotics, possible interventional drainage of a large fluid collection, and possible temporary externalization of VP shunts [23]; however, some patients will require immediate or delayed repair of the perforation [4].

Malignancy

It has long been accepted that patients with an enteric augmentation are at an increased risk for bladder malignancy [4]. The etiology and exact risk are unknown and estimates are varied given the relatively rare incidence of malignant transformation in augmented bladders. Recently, this has been challenged in a well-thought-out review of a prospective registry in which patients with a previous ileal/colonic AC were compared to matched control patients who also performed CIC without AC. They concluded that there was not a statistically significantly increased risk in those patients who had undergone an AC (4.6% vs 2.6%) [24]. However, bladder cancer in both populations occurred at a younger age, was more advanced at diagnosis, and had a decreased survival rate.

Commonly, recommendations were to perform surveillance cystoscopy and cytology beginning 10 years post-augmentation, regardless of GI segment used. However, secondary to poor sensitivity and rare incidence with long lead-time prior to development of malignancy, this has been questioned and shown to be costly and ineffective for all patients [25]. Therefore, we agree with a targeted method of surveillance for those patients utilizing CIC with or without an AC who are potentially or known to be at higher risk, including those with a gastric AC, history of immunosuppression, history of systemic viral illness (EBV, CMV, BK), colon AC at the age of 50, those with >4 UTIs/year, gross hematuria, >50 RBC/HPF on urinalysis, chronic perineal or pelvic pain, or abnormal ultrasound findings [24,26].

Conclusion

In conclusion, a patient with refractory noncompliant dysfunctional bladder and AC is successful in improving the capacity and compliance of the bladder. An AC has low but real risks for early postoperative complications that are similar to those of other major bowel and pelvic reconstructions. A standard AC with ileum has the

expected side effect of mucus production with short- and long-term risks for UTIs, stone formation, metabolic derangements, perforation, and malignancy. Therefore these patients should have a complete workup prior to surgery with education for the patient and families regarding these risks. These patients need to be followed for life by their surgeon or transitioned to a surgeon familiar with AC and its inherent potential long-term complications.

DOS AND DON'TS

Do

- Open bladder widely to prevent hourglass formation and failed augmentation
- Irrigate regularly to evacuate mucus and prevent stones

Don't

- Treat asymptomatic bacteruria, unless urease-splitting organisms are present on culture and there is a history of stones
- Leave stone fragments when treating bladder stones, as these will be a nidus for stone recurrence

CLINICAL PROBLEM/CASE: DISCUSSION

- 1 A full surgical history is important regarding when and if the patient has had a bladder augmentation or other lower urinary tract reconstructive procedures. When and where did he have the surgeries done? Often if the physician is unfamiliar with the details, talking directly to the family is helpful. Also knowing from where to obtain records is helpful and sometimes necessary. This is important as a history of a bladder augmentation increases his risk for bladder perforation. You should have a high index of suspicion for perforation in a patient after augmentation.
- 2 If there is a concern for perforation an indwelling catheter should be placed via the native urethra and/or abdominal wall cath stoma.
- 3 A CT cystogram would be the gold standard to detect a bladder perforation.
- 4 Initial treatment with maximal bladder drainage, culture-specific antibiotics, IV fluid resuscitation, and consultation with interventional radiology for percutaneous drainage of a large extravescical and/or intraperitoneal fluid collection can be beneficial. If the patient has a VP shunt, consultation with a neurosurgeon is appropriate also. In the setting of bacterial peritonitis, the VP shunt may need to be externalized to prevent concomitant shunt infection. In a patient whose perforation is large and unlikely to heal with drainage or is severely unstable then an open exploration, abdominal washout and bladder augmentation repair will be necessary.

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CHAPTER 39

Appendicovesicostomy and ileovesicostomy

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KEY POINTS

- The success of a continent catheterizable channel depends on a flap-valve mechanism, a low-pressure storage of urine in the reservoir, adequate backing to the channel, and an intravesical length to tube diameter of 4:1 to 5:1
- The appendix is the most commonly-used channel, followed by the transverse tubularized ileal segment. Both techniques have similar continence outcomes and complication rates
- Difficulties in catheterizing the conduit due to kinking or stomal stenosis, stomal incontinence, and stones are the commonest complications needing surgical intervention

Introduction

The initial concept of a continent catheterizable channel and the subsequent utilization of this technique have dramatically improved the quality of life of many children with bladder dysfunction [1]. Following Mitrofanoff's initial experience, in which the appendix was implanted to create a flap-valve mechanism in the bladder, many other structures have been utilized as the efferent channel [2]. Advances in our knowledge of bladder function and structural characteristics of the channel have led to a better understanding of the complications that can result from this technique. Success of the procedure appears to be largely independent of underlying urologic disease, age of the patient, and specific configuration of the urinary storage reservoir.

Ideally, all patients should be able to achieve social continence using modern methods of continent urinary reconstruction. However, not all patients have the physical or cognitive abilities to perform clean intermittent catheterization (CIC). Additionally, patients who have undergone reconstruction may fail to demonstrate the adequate compliance with CIC required to maintain healthy intravesical pressures. In these patients, the incontinent ileovesicostomy (i.e. ileal chimney) can prove invaluable.

This chapter focuses on the most common problems that arise following creation of both continent catheterizable and incontinent channels and provides an approach to the management of these complications.

Surgical techniques

General

There are several factors that appear to be critical to the success of the continent catheterizable channel. From a mechanical standpoint, continence depends on a flap-valve mechanism in which there is maintenance of a positive pressure gradient between the lumen of the efferent limb and the reservoir [3]. To achieve this, the channel should consist of a supple tube which is tunneled submucosally and achieves an intravesical length to tube diameter of between 4:1 and 5:1 [4]. A urodynamic assessment of the efferent limb in 21 patients revealed that continence was generally achieved if the functional profile length (i.e. distance over which conduit pressure exceeds reservoir pressure) was greater than 2.0 cm [5]. The wall of the reservoir should be of adequate thickness to provide adequate support of the tube as it is compressed by intravesical

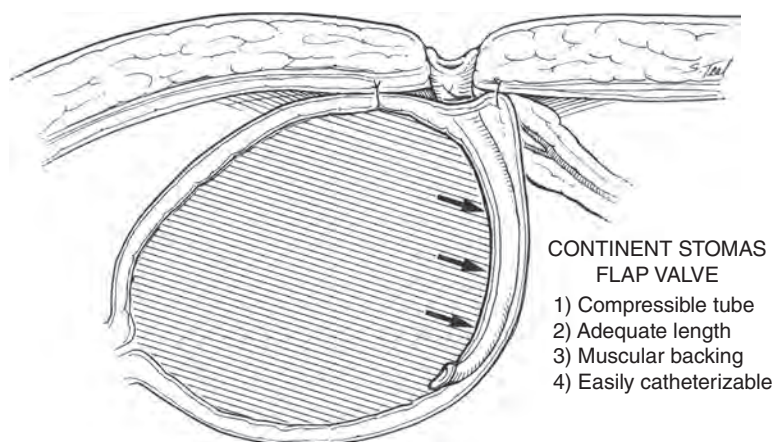


Figure 39.1 The flap-valve mechanism.
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forces (Figure 39.1). The ability to store an adequate volume of urine at low intravesical pressure is a necessity due to the fact that excessively high intravesical pressures can cause a technically adequate channel to leak (just as high intravesical pressures can cause an otherwise adequate ureterovesical junction to reflux) [6].

Anatomic characteristics of the tube are predictive of whether the channel will function adequately. The conduit should have a constant diameter throughout its length (at least 10 French) and have a predictable, healthy blood supply. It must be of sufficient size to achieve adequate tunnel length and traverse the abdominal wall.

The appendix and transversely tubularized bowel segments (TTBS; otherwise known as Yang–Monti channels) appear to be the best substrates due to the fact that they meet these criteria [2,7,8]. Other conduits, including ureter, vas deferens, Meckel's diverticulum, Fallopian tube, tubularized stomach, and foreskin, are either frequently unavailable (in the case of ureter) or have proven far less reliable for various reasons and therefore will not be discussed further in the context of this chapter [9–12]. The geometric relationships that exist between the conduit, reservoir, fascia, and skin also play an important role in determining the success of the channel and will be discussed further in the appropriate sections of the text.

Continent channels

The conduit

When a healthy appendix exists, care must be taken to preserve its mesenteric blood supply. A segment of 4cm length is all that is needed to create continence in most

cases. As a result, if the appendix is of adequate length, it can be divided with one segment used for the urinary conduit and the other utilized for the MACE procedure [13]. If the appendix is not deemed of adequate length, methods for cecal extension/tubularization can be utilized [14]. If a suitable appendix is not available or a decision has been made to use it for a concomitant MACE procedure then the TTBS technique can provide a uniformly suitable conduit for the efferent limb mechanism. For this procedure, a 2.5cm segment of ileum (or colon) is subtracted from the fecal stream [7]. The bowel segment is then opened along the antimesenteric border and subsequently tubularized in a transverse fashion over a catheter. This technique provides a tube with a centralized mesentery (Figure 39.2a). Anatomic considerations may dictate the need for a conduit with an offset mesentery. In these cases, the bowel segment is opened closer to one side of the mesentery than the other (Figure 39.2b). Two techniques have been described to create a longer channel for individuals with a thick abdominal wall. Monti described the use of a tandem tube in which two conduits were anastomosed to each other with interrupted absorbable sutures [7] (Figure 39.2c). Irregularities at the anastomotic site may result in difficulties with catheterization. The alternative technique of the Spiral Monti as described by Casale allows for the construction of a long tube with a more uniform, smooth lumen [15] (Figure 39.3).

Implantation

As previously noted, a firm reservoir wall is needed to provide adequate support of the channel so that

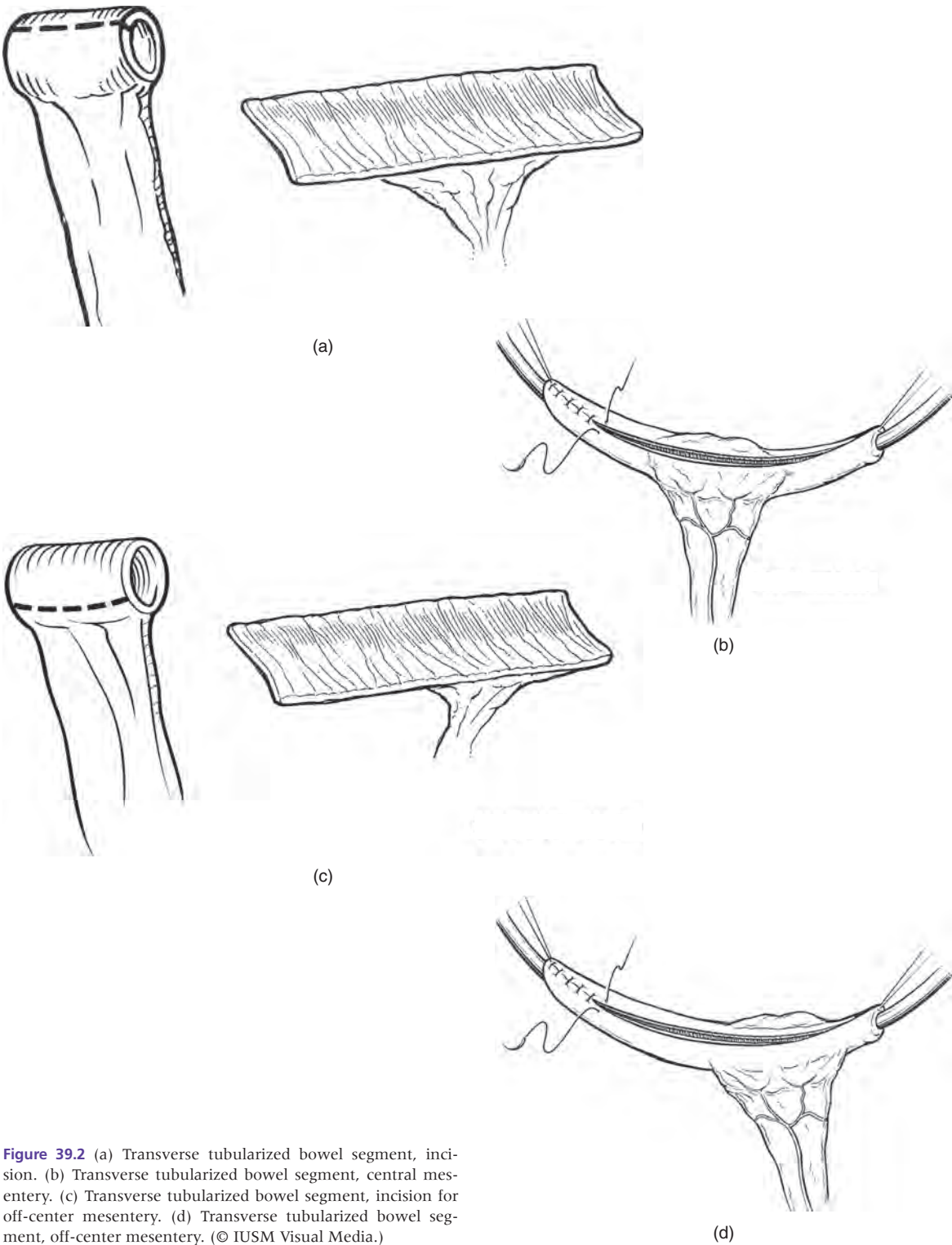


Figure 39.2 (a) Transverse tubularized bowel segment, incision. (b) Transverse tubularized bowel segment, central mesentery. (c) Transverse tubularized bowel segment, incision for off-center mesentery. (d) Transverse tubularized bowel segment, off-center mesentery. (© IUSM Visual Media.)

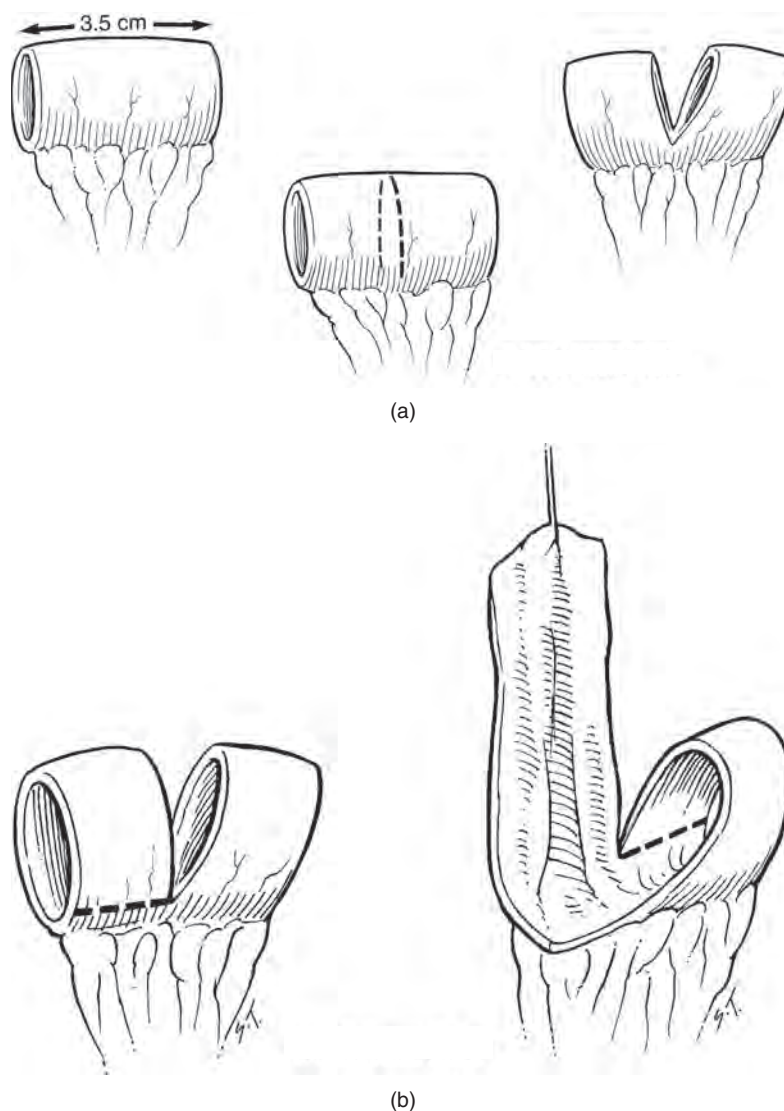


Figure 39.3 Casale spiral Monti: (a) initial incision; (b) subsequent incisions (*continued*).

intravesical pressure can compress the conduit [3]. The preferred site for implantation is therefore the native bladder wall. If native bladder wall is not available then tunneling into a tenia of the colon or the wall of a gastric augmentation is the next best option. Although an antireflux technique can be created between the conduit and the ileum, the thinness of the ileal muscle wall may result in suboptimal transmission of intravesical pressure to the tube. The conduit can be implanted into

the reservoir using either an intravesical or extravesical technique as long as the submucosal tunnel is of adequate length. The extravesical technique can be especially useful when intravesical surgery is not necessary (e.g. bladder augmentation, urethral lengthening procedure) (Figure 39.4). It is an effective and potentially time-saving method with stomal continence rates similar to intravesical techniques [16]. Robotic extravesical appendicovesicostomy is feasible. Mean operative time

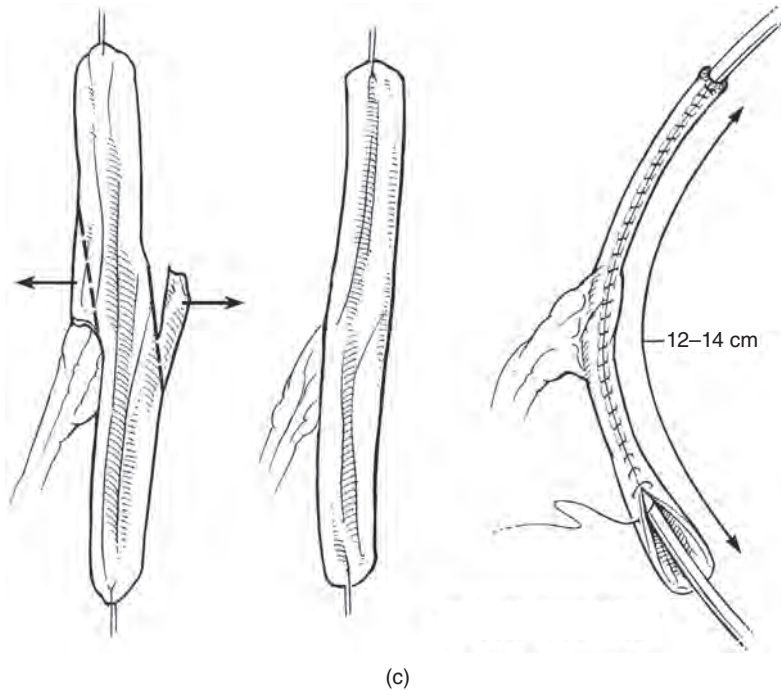


Figure 39.3 (Continued) Casale spiral Monti: (c) completed; (d) operative example. (© IUSM Visual Media.)

for isolated appendicovesicostomy has been reported to be approximately 6 hours [17]. It can be anticipated that operative times will decrease substantially as surgeons become more experienced with minimally invasive techniques.

Once the conduit is implanted into the reservoir, an appropriate location for its exit from the abdomen is determined. Many factors help determine the optimal location for stoma creation in a given individual. First,

a location must be chosen that will allow for a straight trajectory through the abdominal wall. Second, the reservoir must be fixed with permanent sutures at the point of exit of the conduit from the reservoir to the anterior abdominal wall (Figure 39.5). Failure to achieve these two goals will result in kinking of the tube at various degrees of bladder filling. Thickness of the abdominal wall relative to the length of the conduit may also play a role in deciding the stoma site. The umbilicus,

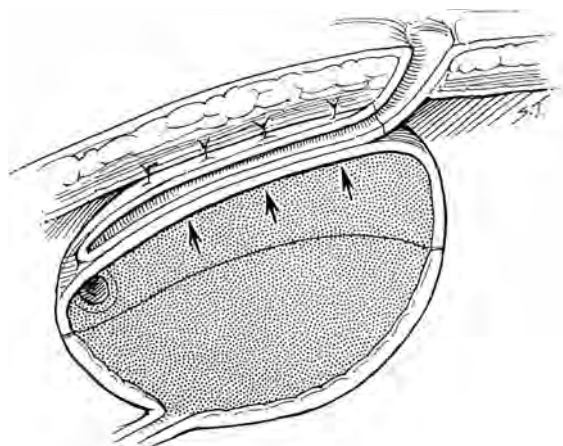


Figure 39.4 Extravesical anterolateral channel.

because it is the shortest course between the abdominal skin and peritoneum, is a unique location for catheterizable stoma placement. A stoma placed in this site is appealing, as it allows the cosmetic advantage of hiding the stoma more readily than in other abdominal locations. However, if the reservoir is not supple enough to be comfortably brought up to the fascial exit site of the channel then the unsupported segment of the channel can prove deleterious (see the section on subfascial conduit complications below). Finally, manual dexterity, patient preference, and gender must be considered when choosing the stoma location. Mitrofanoff himself raised a concern regarding the stretching/kinking of an umbilical-based conduit during pregnancy [18]. Anchoring of the appendix at the umbilicus also has the potential to tether the bladder, making it more difficult to safely retract away from the uterus during a C-section delivery.

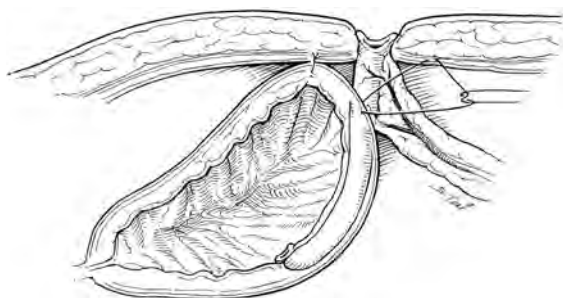


Figure 39.5 Proper anatomic relationships of the conduit to the bladder and abdominal wall. (© IUSM Visual Media.)

Stoma

The stoma itself can be created using a number of techniques. The anastomosis at the skin level should be performed in the absence of tension so as to avoid any compromise to the conduit's blood supply. Early descriptions of the technique proposed excising a circular skin segment, slightly wider than the conduit itself, and anastomosing it flush to the skin [1,2]. Various techniques have subsequently been championed which are believed to decrease subsequent stomal stenosis rates. Each method consists of developing a skin flap that maximizes the diameter of the junction between the skin and conduit. Included among these are the V- or U-shaped flap advancement into a spatulated conduit, VQZ-plasty and VQ-plasty [19–22]. Another potentially useful method for minimizing stomal stenosis with appendicovesicostomies is to harvest the appendix with a small cecal cuff [23].

Incontinent ileovesicostomy: the ileal chimney

The incontinent ileovesicostomy is an excellent surgical option that provides safe evacuation of urine from the bladder in patients who are not suitable candidates for or have failed continent urinary reconstruction [24–26]. This operation can dramatically simplify the care of patients with poor dexterity, impaired cognitive function and individuals who prove to be poorly compliant with catheterization schemes required to maintain low intravesical pressures. In contrast to ileal loop diversion, this technique does not require construction of uretero-ileal anastomoses and preserves the antirefluxing mechanism by leaving the ureters within the bladder. For this procedure, a segment of ileum with adequate mobility to reach the bladder and the abdominal wall is subtracted from the fecal stream. The proximal end is anastomosed widely to the dome of the bladder and the distal end brought out as a budded stoma [26] (Figure 39.6).

Outcomes

Continent catheterizable channels

Continence achieved with a Mitrofanoff tube is greater than 90% in most published series. Continence results appear to be independent of whether appendix or a segment of transverse tubularized bowel is utilized for the conduit. In Mitrofanoff's original series of patients treated between 1976 and 1984, 23 patients underwent

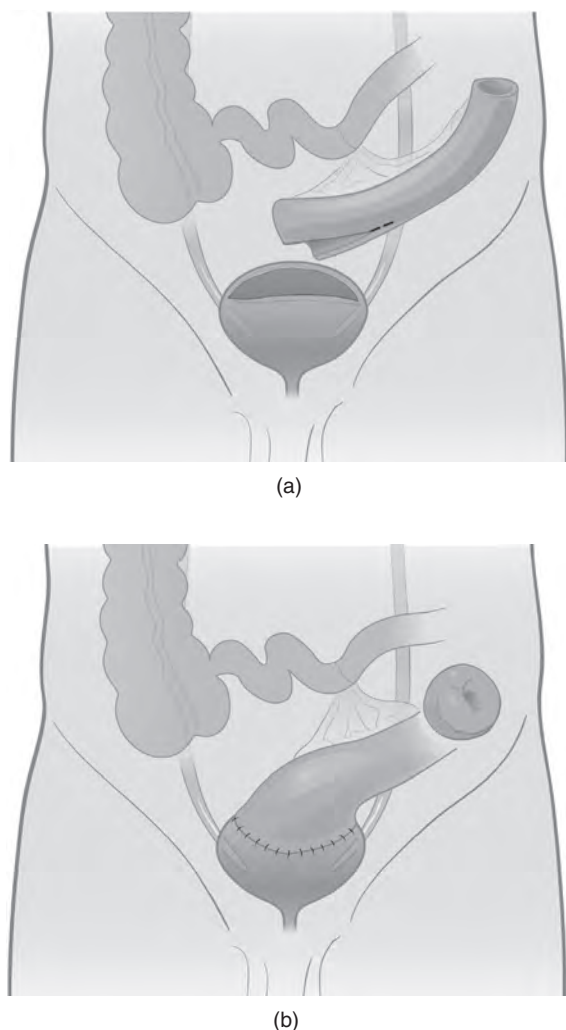


Figure 39.6 (a) Ileal segment harvested and spatulated in wide fish-mouth configuration. (b) Ileal segment placed anastomosed to bladder and brought out as rose bud stoma keeping the segment in an isoperistaltic configuration. (© IUSM Visual Media.)

creation of a continent catheterizable conduit (20 constructed from appendix). Mean patient age at surgery was 8 years and 4 months (range 3–16) and mean follow-up was 20 years (range 15–23). Bilateral upper tract deterioration was found in 10 cases secondary to elevated intravesical pressures. Bladder stones were found in five patients while complications directly related to the conduit included stomal stenosis or persistent leakage in 11 cases [18].

In Monti's series of 55 conduits (seven tandem channels: 48 single Yang–Monti channels) created using the TTBS technique, 91% continence was reported. After an average follow-up of 7 months, only one patient required a revision for stomal stenosis. Five patients experienced incontinence. One patient was rendered dry by adjustment of the catheterization routine, while the other four required two open revisions and two endoscopic procedures [4].

Castellan et al. reported three experiences with 45 Monti urinary channels (four tandem, 41 single), with mean follow-up of 38 months. Overall, stoma-related problems were noted in approximately 20% of patients. Three patients developed complete fibrosis of the channel whilst another three experienced stomal incontinence. Difficulties with catheterization were noted in four patients, with one undergoing stomal revision [27].

Narayanaswamy et al. reported their results with 94 continent catheterizable conduits, of which 25 were Monti channels (tandem/single 17:8). Mean follow-up was 2.1 years. Fifteen (60%) patients had problems with catheterization, with stenosis of the conduit, diverticular pouch formation, or both occurring in 13 of these patients. Six out of seven patients with pouch formation had a double Monti. The authors reported no difference in stomal stenosis rates between appendiceal channels and Monti channels [20].

Leslie et al., in comparing the Monti procedure to the spiral Monti procedure, revealed a 98% continence rate. Surgical revision of the conduit was required in 19% of patients (9% stomal revisions, 10% subfascial revisions). The only significant difference noted between the two procedures was a higher incidence of subfascial revisions for umbilical stomas in both groups. The need for subfascial revision was highest in the spiral Monti channels placed in the umbilicus [28].

The Toronto group, reporting on their 15-year experience of 159 continent catheterizable channels, reported a 39% surgical revision rate. No statistically significant differences in complication rates were noted when comparing use of appendix and reconfigured bowel or different stomal locations [29].

Incontinent ileovesicostomy

Leng et al. reviewed their experience of 25 men and 13 women with a mean age of 44.9 years who underwent incontinent ileovesicostomy. Mean follow-up was 52 months. Before ileovesicostomy the incidence of serious

complications associated with an indwelling catheter was significant, including poor bladder compliance in 50% of cases, urosepsis in 45%, hydronephrosis in 21%, renal struvite calculi in 18%, urethrocuteaneous fistula in 18%, autonomic dysreflexia in 13%, and bladder calculi in 2%. After conversion, 80% of this high-risk population maintained a normal upper urinary tract and normal bladder storage compliance. Other complications, including stomal stenosis, loop stricture, and bladder calculi, were noted in up to 5% of patients [25].

Recently, this technique has been described in a cohort of 18 children [26]. Average age at time of operation was 14.1 years. Average follow-up was 3 years. Eight children underwent the procedure due to a primary inability to perform CIC while 10 required the procedure secondary to poor compliance with CIC following continent urinary reconstruction. Renal function stabilized in all patients. No patient had developed intravesical calculi. Despite its apparent high success rate, others have reported that subsequent excessive weight gain can result in angulation of the channel resulting in reduced efficiency of drainage.

Complications

General

Complications can generally be minimized if proper catheterization techniques are utilized. Although the mucosa of the bowel makes lubrication theoretically unnecessary, generous utilization of lubricant is recommended. The author will have his patients fill a 5 ml syringe with lubricant and gently instill this directly into the stoma prior to catheterization to maximally lubricate the channel.

When difficulty with catheterization is encountered, families are told to contact their physician immediately so that a catheter can be placed across the channel. Failure to place a catheter across the site may allow the traumatized site to fibrose. The surgeon should have a low threshold for utilizing flexible endoscopy to evaluate the channel in these cases. Multiple unsuccessful attempts to place a catheter may simply extend the area of trauma. Our recommendation is to then leave the catheter secured in place for one week. This will allow most false passages or edema to resolve before catheterization resumes and hence minimizes the chance of exacerbating the injury. Complications can

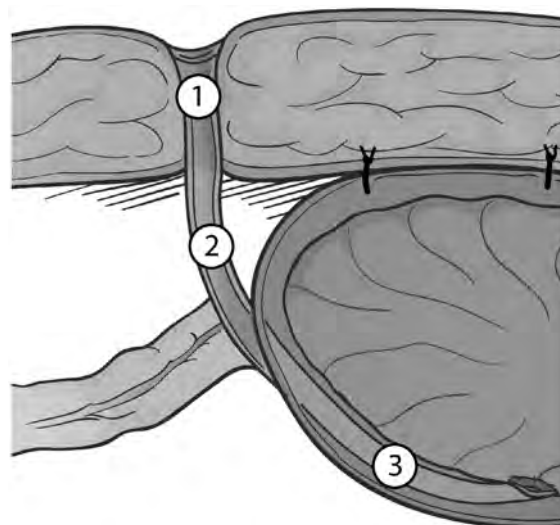


Figure 39.7 Channel anatomy divided up into three general segments: (1) transabdominal, (2) transperitoneal and (3) transvesical.

generally be grouped into three groups based on the segment involved (i.e. transabdominal, transperitoneal, transvesical) (Figure 39.7).

Stomal stenosis

Stomal stenosis is the most common complication of the Mitrofanoff procedure with reported rates ranging between 8% and 40% [9,23,27,30–33]. Stenosis generally appears to occur within the first 2 years following the initial surgery [23,30,34]. However, one report has demonstrated that this complication can occur as late as 15 years following the procedure, emphasizing the continued need for close follow-up of this patient population [18]. Initially it was felt that the well-constructed TTBS may have a theoretical advantage over the appendix in that the luminal diameter could be determined by the surgeon. This is in contrast to the appendix in which the luminal diameter is fixed and generally between 10 and 12 Fr. However, most studies have shown that the incidence of stomal stenosis does not differ significantly between TTBS and appendiceal conduits.

The use of a hydrophilic catheter can allow for continued use of the stoma that has experienced a degree of stenosis. Simple dilation can be enough but often recurrence will require surgical revision. Injections of triamcinolone around the stoma or topical steroid application have been used in an attempt to limit the local inflammatory

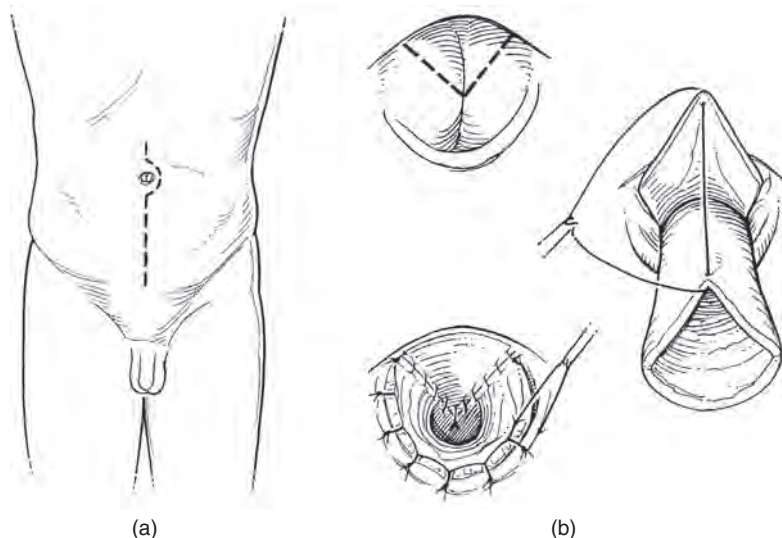


Figure 39.8 (a) Stomal stenosis. (b) Technique of repair of stomal stenosis. (© IUSM Visual Media.)

response that likely plays a role in stenosis [35]. Definitive treatment of stomal stenosis involves the creation of a new, laterally based V- or U-shaped flap, division of the stomal cicatrix along its most lateral edge, and creation of a widely spatulated anastomosis (Figure 39.8). A catheter is generally left in place for 3 weeks before catheterization is restarted.

Stomal prolapse

Stomal prolapse occurs far less frequently than stenosis (Figure 39.9a). The reported incidence of this complication ranges from 2–5% [36,37]. This can occur due to lateral placement of the stoma in a location that is not well supported by the rectus muscle (Figure 39.9b). Careful attention to placement of the channel through the rectus belly will provide better conduit support and should limit the chance of developing this complication. Additional contributing factors may include the creation of a larger-than-needed fascial opening and/or channel redundancy. Rapid reduction of the prolapse should be attempted to limit edema and subsequent vascular embarrassment (Figure 39.9c). Relocation of the stoma to a more medial location as well as improved anchoring of the intra-abdominal portion of the conduit are needed for definitive resolution of this complication (Figure 39.9d).

Subfascial conduit complications

If one is able to pass a catheter at the level of the stoma yet unable to advance the catheter into the reservoir,

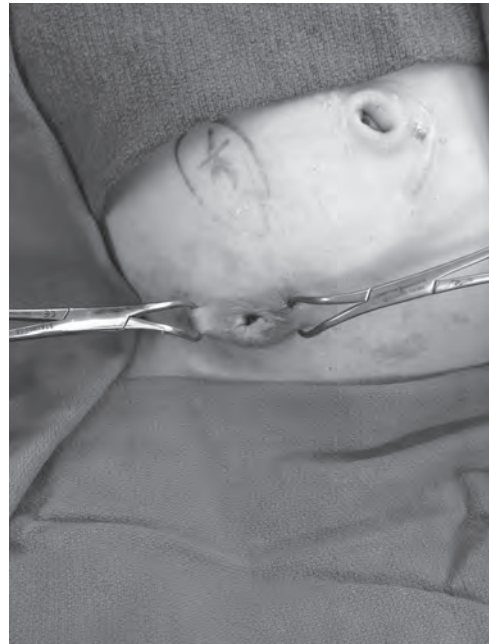
one of several conduit-based complications may exist either individually or in combination.

Kinking of the channel is one of the more common subfascial problems. This most commonly occurs when there has been poor fixation of the reservoir to the posterior rectus sheath. As a result, the bladder moves during filling, altering the angle at which the channel enters into the conduit (Figure 39.10). This problem can initially be overcome if one can decompress the bladder by placing a urethral catheter. In patients with an altered or obliterated bladder neck who experience acute urinary retention secondary to difficulty catheterizing the channel, an 18 gauge needle can be placed suprapubically to decompress the reservoir. After the bladder has been decompressed the angulation of the channel relative to the reservoir is often reduced and a catheter can be placed easily across the conduit. The long-term solution to the problem of conduit kinking may be to catheterize more often and not allow the reservoir to overfill. Persistence of the problem may require reoperation to more securely anchor the bladder to the fascia.

Conduit redundancy is another common etiology for catheterization difficulties. This generally occurs when the surgeon has left a portion of the catheterizable channel unsupported during the original procedure (Figure 39.11a). As a result of cumulative minor difficulties with catheterization, the channel becomes stretched and tortuous making future catheterizations progressively more challenging. For this reason, we have generally advocated bringing the conduit to a right lower



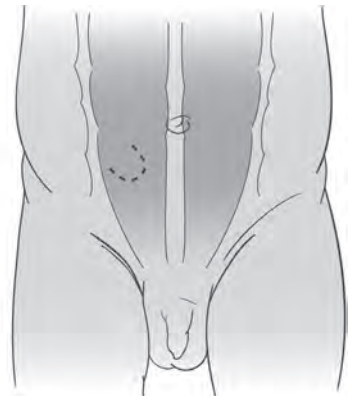
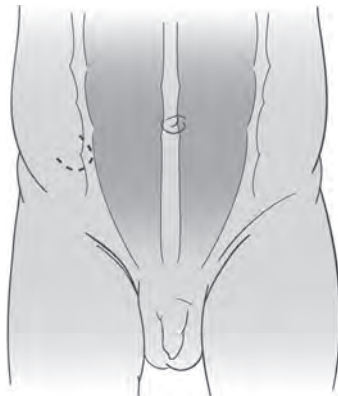
(a)



(c)



(b)



(d)

Figure 39.9 (a) Stomal prolapse. (b) Lateral location of stoma. (c) Stomal prolapse reduction. (d) Improper (left) and proper (right) stomal location with respect to the rectus muscle.

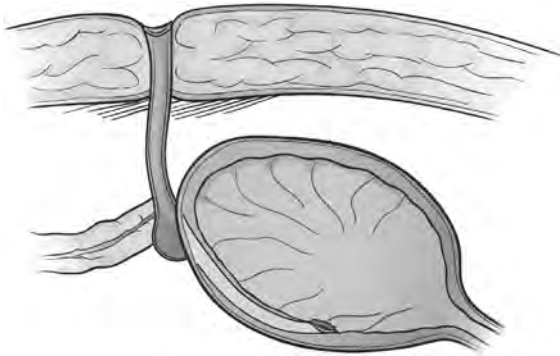


Figure 39.10 Channel course in an unanchored bladder leading to kinking of the redundant channel.

quadrant location unless the reservoir is of sufficient size to extend up to the umbilicus. Conduit redundancy can often be resolved by freeing-up the channel and putting it on additional stretch to straighten its course (Figure 39.11b). However, the continued presence of an unsupported free intraperitoneal segment of the conduit leaves the patient vulnerable to recurrence of difficulties with conduit redundancy.

False passages can occur in combination with kinking and/or conduit redundancy or in a well-supported conduit. In either case, the management is to place a catheter across the site and allow the tube ample time to heal before future attempts at catheterization are attempted. Even full thickness perforations can be successfully managed conservatively if no gross periconduit contamination has occurred. Special attention to the method of implantation when using the extravesical approach can prove particularly helpful in avoiding the creation of a false passage at the entrance into the bladder lumen. Figure 39.12 demonstrates the technique of conduit advancement that can prove invaluable in avoiding this later complication.

Stomal incontinence

Incontinence through the efferent conduit may be the result of an inadequate flap-valve mechanism, high intravesical pressures, conduit-vesical fistulas or a combination of these factors. Proper determination of cause is essential in determining the appropriate surgical treatment. Urodynamic evaluation with the catheter preferentially placed through the native bladder neck will establish bladder compliance and determine the conduit leak point pressure. A poorly compliant reservoir should be properly addressed with anticholinergic medication

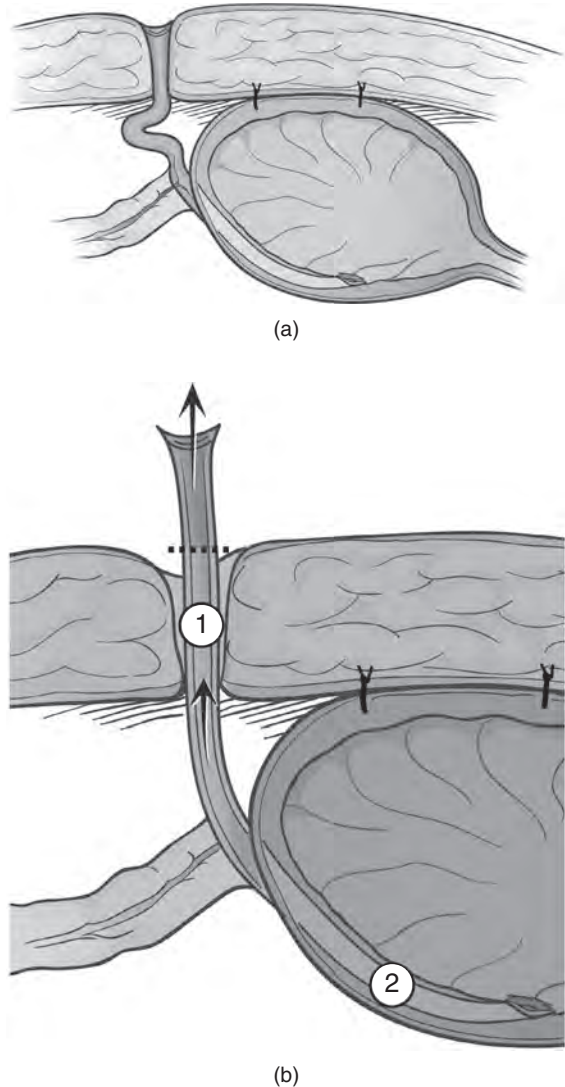


Figure 39.11 (a) Channel redundancy – the long extravesical course of the channel to the abdominal wall. (b) Technique for repair of channel redundancy. 1, site of resection of redundant channel after straightening; 2, intramural channel.

and/or bladder augmentation. An inadequate tunnel can be corrected by submucosal injection of biomaterial [38]. A similar technique may be utilized to manage a conduit-vesical fistula. The biomaterial can be injected transvesically in the same fashion as the STING procedure is carried out. Simultaneous evaluation of the channel via the conduit and through the bladder neck can prove invaluable in assessing the proper localization and assuring success of the injection [39]. If the

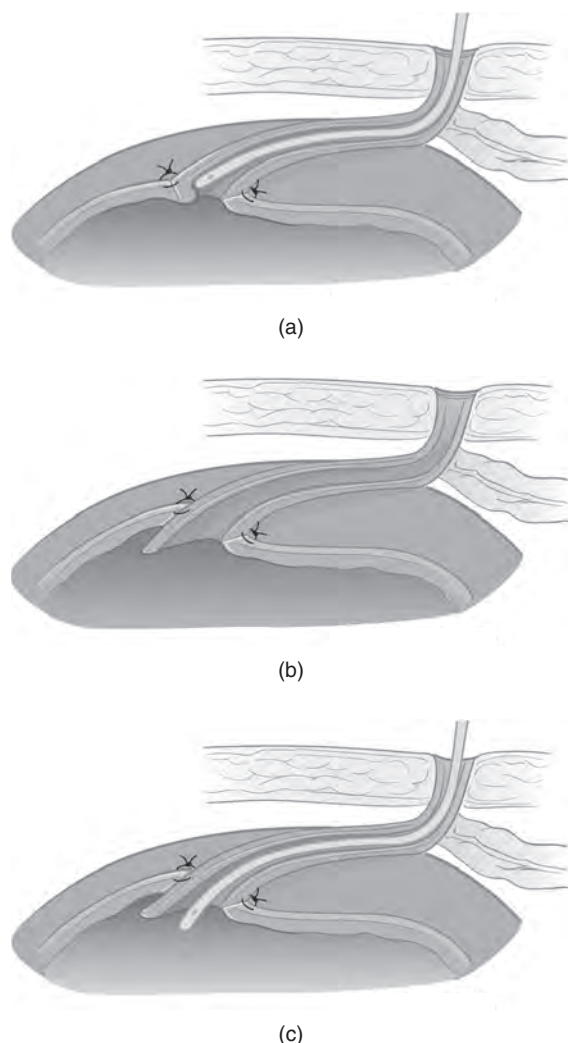


Figure 39.12 (a) Catheterization difficulty at the entrance into the bladder with a false passage created. (b) Technique of distal channel advancement to eliminate a false passage at the leading edge. (c) Trouble-free catheterization at the entrance into the bladder. (© IUSM Visual Media.)

urethra has been surgically modified, the material can be injected via the conduit with the bulking agent delivered submucosally at the 6 o'clock position. When this minimally invasive technique is utilized, the bladder should be drained by a route other than the channel so as to avoid the catheter molding the polymer. If the bulking agent is unsuccessful in resolving the incontinence then an open surgical procedure to re-establish a proper valve mechanism is indicated.

Stones

Urinary stasis is a well-known etiologic factor for stone formation throughout the urinary tract. Most patients with neuropathic bladder dysfunction and many patients with anatomic abnormalities of the bladder or bladder outlet (i.e. bladder exstrophy and posterior urethral valves, respectively) do not have the ability to spontaneously empty their bladder to completion. These patients are hence more prone to urinary stasis with subsequent precipitation of urinary solutes and creation of an environment amenable to bacterial overgrowth. Bladder stones have been reported with increased frequency in augments with coexistent bladder outlet resistant procedures and/or catheterizable abdominal wall stomas [40–42]. In one series, patients with an abdominal stoma had a 40% higher risk of developing reservoir calculi: the specific incidence depended on whether the patient emptied via an abdominal wall stoma versus the native urethra (66% vs. 15%). The incorporation of a gastric segment when an abdominal stoma is created may decrease the risk of calculus formation [35]. Leng et al. reported the development of stones in approximately 5% of patients following creation of an ileal chimney. In contrast, Kaefer et al. found no cases of stone formation in patients who irrigated their conduit daily using a catheter placed via the ileal segment into the bladder [26].

Methods for bladder stone removal include open cystolithotomy, percutaneous cystolithotomy and endoscopic cystolithotomy via the efferent conduit. Evacuation of stones from the bladder that is drained by the ileal chimney is straightforward. The large diameter of the conduit allows for easy passage of endoscopic equipment into the bladder and removal of stones intact from the bladder. In contrast, although it may be tempting to attempt stone removal via a continent efferent conduit, any manipulation of the conduit does carry with it the potential for injury and other options should be strongly considered if there is any difficulty passing endoscopic equipment or the stone is of significant size.

Perhaps the most important aspect of managing bladder calculi in patients following genitourinary reconstruction is the prevention of further stones. The patient and parents must clearly understand that there is a high probability of stone recurrence if measures are not taken to reduce risk factors. A number of series have demonstrated a clear reduction in bladder calculi

when a postaugmentation bladder irrigation protocol is instituted following bladder augmentation or creation of an ileal chimney [40,41,43]. Hensle et al. compared the incidence of stone formation in two distinct patient groups following bladder augmentation. Of 91 patients who did not perform postaugmentation irrigation, 39 (41%) developed bladder calculi with a mean time to presentation of 30 months. In contrast, only three of 42 (7%) patients who did perform postaugmentation irrigation developed reservoir calculi with a mean time to presentation of 26 months.

Therefore, lifelong daily bladder irrigation of the bladder is imperative to evacuate all mucus. Mitrofanoff emphasized the importance of creating a continent catheterizable channel of large caliber so as to allow more rapid drainage and minimize the chances of leaving residual urine within the bladder. In patients with continent catheterizable stomas, mucus may build up in the most dependent portion of the bladder. If the bladder neck is still accessible, it may be of benefit to periodically irrigate via the more gravity-dependent bladder neck to minimize buildup of mucus.

DOS AND DON'TS

Do

- Determine whether the patient has adequate cognitive ability, manual dexterity and compliance to be a suitable candidate for a continent urinary reconstruction with a catheterizable channel. If the patient is not deemed a suitable candidate consider vesicostomy, ileal loop or ileal chimney (incontinent ileovesicostomy) as an alternative means of providing suitable bladder drainage
- Address other urinary storage parameters including bladder capacity, bladder outlet resistance, and status of the vesicoureteral junction prior to surgical intervention
- Institute vigorous postsurgical bladder irrigation to avoid bladder calculus formation

Don't

- Leave an unsupported free intraperitoneal course for the conduit
- Place the conduit in a position that crosses the anterior midline of the bladder in conditions that may later require bladder augmentation

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CHAPTER 40

Surgery for fecal incontinence

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KEY POINTS

- The antegrade continence enema (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

CLINICAL PROBLEM/CASE

A 6-year-old female with a history of myelomeningocele status post-ACE creation with complaints of leakage back of the saline flush with small amount of stool material at the stoma site.

- 1 What is the most likely etiology of the leakage of the soilage at the stoma site?
- 2 What would be the first recommendation to help troubleshoot the problem above?

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 imbricating it over the cecum and bringing to the skin

as a catheterizable channel for flushing the colon. 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 soilage 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 fairly low with stoma-related problems being the most common. One of the more frustrating complications is stool leakage caused by an incompetent valve. Whilst sometime open revision is appropriate, newer minimally invasive techniques, such as Deflux injection can resolve the problem [7]. As in all reconstructive cases, patient selection with thorough preoperative patient education and good surgical technique are instrumental in the 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 pre-existing 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 coexisting 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 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 coexisting 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. 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 one 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 Golytely 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. Soapsuds 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 technique currently used. 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 90–95% [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. [3] as intracorporeal imbrication can prolong the procedure significantly.

At 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 45 degrees 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 French 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 Vicryl 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 intra-abdominal 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 have been 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 1 the patient is encouraged to mobilize and the diet advanced after flatus is passed. As soon as the patient is started on an oral diet, the ACE is flushed with 60ml of normal saline daily and slowly advanced to the maximum volume. Pain is initially managed with intravenous Toradol and Tylenol. 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. Whilst preoperative discussion is important, it is the postoperative close follow-up and care that prevents early misunderstandings and complications. The patient is evaluated three 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 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 minutes to 1 hour 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 40.1) [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 one month to obtain the volume necessary to

attain fecal continence and complete bowel evacuation. Thus, discussion of expectations cannot be over-emphasized.

Complications

Our patients usually go home on postoperative day 2 or 3 depending on how well they are tolerating oral intake. Problems relating to the stoma and catheterization in general make up the majority of the chronic and long-term complications. The best management pathway should involve the clinician's analysis of what the problem is and how best to help the involved parties – that is, the caretaker and the patient.

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. One risk factor that has been identified to predict an increased rate of stomal stenosis is 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.

Table 40.1 Surgical outcomes based on primary diagnosis.

Diagnosis	Full success	Partial success	Failure
Myelomeningocele	63	21	16
Anorectal malformation	72	17	11
Hirschsprung's disease	82	9	9
Constipation	52	10	38
Miscellaneous	44	25	31

From Curry et al. 1999 [8]. Reproduced with permission of Elsevier.

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. Mucus 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 unflushed 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 a 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 days 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.

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 a 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. 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. 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. Our institution supports a local sports organization for people with disabilities (GODSA: Greater Oklahoma Disabled Sports Association) to give affected patients the possibility to enroll in sports programs to increase their activity level. 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.

DOS AND DON'TS

Do

- Create a tunnel length at least five times the appendix circumference to ensure continent valve
- Create adequate fascial opening to allow passage of the channel without causing obstruction but tight enough to prevent a hernia

Don't

- Underestimate the need for a well-established team that delivers excellent instructions in the management of the stoma

CLINICAL PROBLEM/CASE: DISCUSSION

- 1 The likely etiology of the leakage during flushing of the stoma is impacted hard stool at the base of the channel.
- 2 By passing the catheter about halfway into the stoma, one can usually then flush the hard stool at the base of the channel rather than advancing the catheter to the base or slightly past it and having liquid return back to the skin surface.

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CHAPTER 41

Surgical problems in transitional care

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KEY POINTS

- Children who have undergone major urological reconstructive surgery require long-term follow-up through adolescence into adulthood
- Understanding complications that may occur in adulthood following such reconstructive surgery is important to facilitate transitional care
- The aim of transitional care into adulthood is to allow the individual to achieve their aspiration for quality of life, form relationships and have a family

CLINICAL PROBLEM/CASE

A 25-year-old man with ambulant spina bifida has been lost to follow-up for several years. He has returned because he has developed a suprapubic fistula with clear

fluid draining out from it. You discover that he has had a cystoplasty and a bladder neck reconstruction in childhood and has recently had an increase in the number of UTIs he suffers with. A CT KUB shows a large stone in his bladder and a smaller stone in his prostatic cavity.

Introduction

It is a tribute to the courageous advances in pediatric urological care that this chapter needs writing and its remit will expand with time. Much of this book will center on those treatments – this chapter will look less at surgical complications and more at long-term outcomes – although the two are not mutually exclusive. In the modern age, patients with major congenital urological anomalies have a realistic expectation of a good quality of life into adulthood – well beyond survival and continence. The exciting and challenging consequence of that is to match that achievement with aspirations for long-term quality of life, including the ability to work, form relationships and have a family. The impact of repeated major surgery and hospital visits over years coupled with an initial anomaly create many questions for both patient and carer about their

relationship with normality. By definition, no surgeon can create normality. In this context, our job is to try and improve that which needs it, restore function and ensure safety. In conveying our treatment plans to patients these limitations are vital to understand as we have to set clear and realistic expectations with patients and, probably, their parents.

This chapter looks at the aspects of urological function and the needs for follow-up and revision surgery. The relationship between urology and nephrology is important in ensuring the safety of upper tracts. The care of patients with disorders of sex development is advancing in its understanding. The long-term aspiration of all patients to achieve relationships with sexual function and fertility is an important part of long-term wellbeing. This chapter aims to provide an insight into aspects of this care and potential surgical issues for these patients.

Urological function

The bladder

The surgical problems that may arise with an adolescent bladder fall into three categories:

- 1 Problems with an existing reconstruction.
- 2 Bladder changes as a result of pre-existing disease.
- 3 New problems arising in adolescence.

The first of these is discussed below; the second two are beyond the remit of this chapter as they are not surgical complications. Suffice to say that patients with major anomalies such as posterior urethral valves or a neurogenic bladder need careful monitoring as they pass through puberty and into adult life; despite improved survival, long-term prognosis can be difficult to predict.

Problems with an existing reconstruction

Once bowel is introduced to the urinary tract patients require life-long follow-up. It is important to remember that bowel continues to behave as bowel despite contact with urine. The most obvious and apparent sequela to that is the continued production of mucus. This varies between individuals and may change with circumstance – puberty, dietary elements and pregnancy are all factors that our anecdotal experience suggests may vary mucus production. Patients appear to vary in their compliance with bladder washouts and the individual need has to be tailored. Some will never have washed out in childhood – this may not need to change if a patient is asymptomatic. If there are difficulties such as infections or catheter blockages, a regime of around twice per week may begin and this can go up or down as required. Any washout regime needs to be coupled with a healthy fluid intake.

Metabolic changes

Metabolic changes are common but the majority are subclinical and do not require treatment. It is our practice to routinely check vitamin B12. The cost of checking and replacing this is low. The consequence of a missed low vitamin B12 is an irreversible neurological deficit: subacute combined degeneration of the cord. To avoid ambiguity or error over the segment of bowel used we check vitamin B12 in all augmented/substituted patients. Bicarbonate and chloride levels also require monitoring, as a hyperchloremic metabolic acidosis is seen with a significant reduction in bicarbonate and a rising chloride and needs replacement with oral bicarbonate. Otherwise, malaise, bone demineralization and

confusion or (eventually) coma may result. Subclinical acidosis is identifiable in many patients and does not need treatment. Life-long monitoring is essential.

Stones

The incidence of stones in bladders varies with the type of drainage. The risk of stones increases along the following hierarchy:

Spontaneous urethral drainage < Urethral ISC
< Mitrofanoff drainage

The lowest incidence is cited at 2% in those with a cystoplasty draining urethrally but this increases fivefold with urethral catheterization and tenfold with Mitrofanoff drainage [1]. These may be removed urethrally where possible, percutaneously if a Mitrofanoff channel is the only route of entry to the bladder and with an open cystolitholopaxy with multiple or larger stones (Figure 41.1).

Infection

Asymptomatic bacteriuria is common with an incidence of significant urinary tract infections (UTIs) gauged at between 4% and 43%. Bacteriuria alone should not be treated for fear of generating resistant forms; however, symptomatic urinary tract infection needs early



Figure 41.1 Cystoplasty with bladder and prostatic cavity stone.

empirical antibiotic treatment with simultaneous urine culture to ensure the correct antibiotic has been selected. Otherwise, the risk of systematic symptoms and infections that are difficult to clear seem to increase.

Malignant change

Malignancy remains a concern in reconstructed bladders. The incidence is approximated at 1.2% and risk factors appear to be some forms of congenital disease (e.g. bladder exstrophy), smoking, immunosuppression and some forms of chronic inflammation, e.g. TB. The case for routine annual cystoscopy is not supported by evidence as cancers appear during screening intervals. The safest course appears to be thorough investigation of patients who develop symptoms such as pain, hematuria or recurrent UTIs. Gastrocystoplasty patients appear to be at higher risk of malignancy and therefore the case for annual cystoscopy of these individuals appears to be stronger [2,3].

Continence

Artificial urinary sphincter

The artificial urinary sphincter has earned its place as a very successful device for the treatment of prostatectomy incontinence. There are published data relating to its use in some congenital conditions. In those with a neuropathic bladder it has its greatest application. It shows an 86% continence rate at 10 years with a 50% need for revision surgery [4,5]. The risks are erosion and infection. A long-term patient is much more likely to need replacement of the device – with an average mechanical life expectancy of 10 years. Our institution has a number of patients who have sustained good function with a single sphincter for much longer than this – so this is by no means absolute. The sphincter is most effectively placed at the bladder neck, requiring major open abdominal surgery. The procedure can be performed simultaneously with a cystoplasty if required. The biggest risks for infection or erosion of the device are female patients and a need for intermittent self-catheterization. Either infection or erosion necessitates removal of the device and the need for the resulting inflammation and tissue damage to settle – usually a period of 6 months. The device has been used but there is less evidence about its outcomes in other conditions such as bladder exstrophy and incontinence following posterior urethral valves.

Bladder neck support

In some patients with incontinence there is a clear failure of bladder neck support – neuropaths are one example – with sphincter failure. In these cases, it may be possible to provide additional bladder neck support without placing an artificial sphincter. There is a significant chance that a patient will need to self-catheterize as a result of the increased outlet resistance but for many this will offer a vastly improved quality of life compared to the incontinence they were suffering. The preferable methods include a colposuspension in women or, if that fails, it is possible to harvest an autologous sling of fascia from the rectus fascial sling and place that around the bladder neck.

Bladder neck reconstruction

There are a variety of procedures that are really based around urethral lengthening and increasing bladder outlet resistance. In exstrophy/epispadias, where they are most widely applied, their success rates will rest at 48–57% [6,7]. This may be an acceptable starting point in childhood but unpublished data relating to newer techniques may offer better results. For an adolescent and young adult this may be a significant amount of leakage that and may necessitate additional procedures; one that may be of benefit is the rectus fascial sling. These situations become complex and require bespoke management. What is less clear is the outcome in women who later choose to begin a family and how their continence will be sustained both through pregnancy and flowing delivery.

Bladder neck closure

This is the option of last resort. It can be an excellent operation but is a huge commitment on the part of the patient to maintain regular drainage of their reservoir and on the surgeon to maintain a channel to provide drainage. In adult practice, transvaginal closure of the bladder neck may be possible but carries up to 30% failure rate. This may be improved by the harvest of a Martius (labial) fat pad. Transabdominal closure generally has a higher success rate but many of the patients undergoing the procedure will be complex and have had multiple previous procedures. Careful preoperative counseling is important with close follow-up and easy access for patients to contact the team that performed the surgery.

Urethra

We are really only just beginning to understand the importance of long-term follow-up in the treatment of

all pediatric surgery. This is not to say that many have not attempted or are not concerned about it but the logistics are very challenging. One of the most striking burdens will be that in hypospadias. With short-term follow-up and a failure to follow patients past puberty it has long been assumed that the complication rates for hypospadias surgery were very low. There are still those with a powerful voice that state there is no value to follow-up beyond the short term. This remains an unproven stance since there is no evidence to confirm or refute it – I need not point out the obvious ambiguity in this argument. We do know that analyses and studies that have looked at long-term follow-up have shown complication rates that were higher than expected. In an adult urethroplasty practice the overall burden represents about one-fifth of their work load – this percentage does not necessarily bear any relation to the number of hypospadias repairs performed [8].

Prospective long-term data are sparse but that which exists suggest puberty is an important factor and complication rates may be as high as 25%. Standardization of assessment, technique and follow-up would greatly assist this but remains a long way off. Those with insight continue to work towards meaningful data and allow more subtle selection of those who need functional correction and the implications for those parents who are choosing cosmetic correction. This will assist parent counseling. In literature reviews presented at the recent European Society of Paediatric Urology, about half of the papers reviewed did not specify a reoperation rate, with 30% not giving a length of follow-up [9,10].

Protecting the upper tracts

The principle role of the urologist in protecting renal function is the provision of a safe reservoir and the avoidance of obstruction or infection. The problem in many of these patients is that a urologist is trying to apply a normal range of established tests to an already abnormal urinary tract – in interpreting these it is important to be mindful of this. The ideal solution is regular follow-up and the ability to then follow a pattern to spot change or decline. There are three main problems.

1 Obstruction. Every urologist will understand this and we are all used to dealing with acute obstruction due to a stone. In interpreting annual imaging (USS usually) change is important. An increase in an

established hydronephrosis of whatever cause needs consideration. Bladder empty views are important and it is worth requesting a scan and checking that this has been done. If the hydronephrosis is maintained then renography (MAG3) is important. These can be difficult to interpret in some, and in difficult cases we have resorted to upper tract drainage and monitoring of creatinine or the use of a Whitaker test to help our diagnosis. Multidisciplinary discussions with a urologist, nephrologist and radiologist are helpful in providing an effective management plan.

2 Unsafe reservoir. Once again, change in appearance such as increasing hydronephrosis is important and in our practice would merit investigation with video urodynamics. These are relatively straightforward and high bladder pressures would need treatment to reduce them. Revising a cystoplasty to improve a reservoir can be a surgical challenge but it is perfectly possible to do. Important consideration needs to be given to groups where there may be a shortage of bowel for use in doing this, the most obvious being cloacal exstrophy. Under these circumstances, if further surgery is necessary discussion with a gastroenterologist is important and the minimum possible amount of bowel should be used to achieve the desired aim.

In our center we identified a small group of patients whose renal function continued to decline despite safe bladder pressures. These were investigated with simultaneous cystometrogram and dynamic renography. This study identified that the bladder was perfectly safe but that, above a critical bladder volume, the upper tracts did not drain. We taught patients about this and the need to drain their reservoir regularly to keep volumes below this critical level. By doing this, we were able to arrest the decline in kidney function, suggesting that, in some, volume rather than pressure-dependent deterioration can be identified and treated [11,12].

3 Intrinsic renal failure. Many of our patients are referred to us with a degree of renal impairment. Previous authors have stated that reaching puberty with a GFR of 30 ml/min when correct for body surface area is a strong predictor of end-stage renal failure in adulthood. It is vital that any urologist caring for these patients is alert to change and that they collaborate with a nephrologist. All patients arriving at our clinic are checked for blood pressure and proteinuria.

Proteinuria is investigated with a protein : creatinine ratio and a formal GFR where appropriate. Raised blood pressure can be difficult to interpret and, when noted, we ask the GP to perform a 24 blood pressure tape to understand whether this is “white coat” syndrome or more significant. Once renal deterioration has been detected and urological causes ruled out, on the advice of a nephrologist we would initiate renal protection with either an ACE inhibitor or angiotensin receptor blocker as a means of trying to slow decline. Multidisciplinary team working is very important for these patients [5].

Disorders of sex development

Early versus late surgery

There continues to be marked controversy about the timing of surgery in these patients, particularly in female XY disorders of sex development (DSD) and conditions that result in clitoromegaly. The proponents of early surgery will cite the relative ease of surgery and the advantages of minimal scarring. This is coupled with the argument that raising a female child with abnormal genitalia will cause increased psychosocial issues. It is possible to understand how a parent may view their daughter and express a desire for her to be normal. These seem like normal concerns and clinicians may view the solution as performing an operation to achieve these perceived advantages.

The argument against this are that many patients need revision surgery and therefore why perform an early operation when the later operation will be needed for many patients regardless of what is done.

Whilst scarring through childhood may be minimal – once a patient passes through puberty there is the risk of asymmetric growth and scarring (Figure 41.2) – this may well require surgical correction, with patients expressing genuine concerns about genital appearance.

The psychological perspective is challenging. There is little evidence to support the fact that surgery improves child–parent bonding or alleviates other psychosocial issues that are postulated. There is no doubt that both parents and child should be offered psychological support – the most obvious times will be with the parents at the time of diagnosis and with the child at the time of diagnosis disclosure. This care needs to be continuous and the team needs to be alert to other issues that may



Figure 41.2 Assessment at EUA of previously-operated genitaloplasty in infancy. Note intestinal vaginoplasty scarring and asymmetry.

arise. Some families and individuals will need more support than others and this is difficult to predict. These diagnoses are difficult to manage and challenging for patients, parents and the healthcare team. Surgery at any time needs to be considered carefully and the indications very clearly specified.

Gonadal preservation

In female XY there are two indications for gonadectomy. The first is the risk of androgenization of a female child or adolescent, and the second is in the context of increased malignant risk. Patients with complete androgen insensitivity are clearly at the lowest risk of androgenization. It is in this group that some young adult women are making a positive decision to retain their gonads. Previous data have suggested that these gonads are at low risk of malignancy; however, the default position for many of these patients was gonadectomy. There is little in the way of long-term data to confirm this belief. The wishes of these patients can be respected but they require careful counseling and regular follow-up. In our series, we used MRI and compared that with histology in those patients that decided to have subsequent gonadectomy. MRI was good at detecting paratesticular cysts and sertoli-only adenomas but did not show anything that would indicate intratesticular germ cell neoplasia (ITGCN) in the cases where that was detected. There were no invasive malignant tumors in our series so comment could not be made about that. The concern is that these testes are

intra-abdominal and cannot be examined either by the patient or clinician. Interval scanning seems reasonable but setting a safe interval to avoid the risk of late diagnosis is a concern [13,14].

Sexual and reproductive function

The penis

It is normal for adolescent men to suffer at least a period of anxiety about their penile function. This is no less true for men who have had surgery for penile anomalies. When examining studies comparing hypospadias patients with controls, about one-third of both groups felt their penile appearance was abnormal when compared to their peers, although those with hypospadias were less likely to be satisfied with their penile appearance [15]. In terms of sexual function, those with severe hypospadias were more likely to report ejaculatory problems such as weak ejaculation or the need for milking. Hypospadias patients showed lower satisfaction with intercourse when compared with controls and those with severe hypospadias were less likely to have formed relationships (although the mean age was lower than that for controls: 24.4 vs. 26.9 years, $P < 0.01$). There was no difference in sexarche between the two groups [9].

In conditions such as bladder exstrophy, scarring or erectile deformity may not be apparent until puberty. The authors' experience is that many patients have good erectile function although around 30% are affected by a severe dorsal chordee that prohibits penetrative intercourse. In some, this can be corrected by relatively straightforward ventral tucks; for others, complete penile disassembly may be required. This is possible because there is no cross corporeal circulation in the exstrophy penis, the dissection can be very difficult and care needs to be taken to identify the neurovascular bundles (Figure 41.3).

Clitoral and vaginal surgery

In our review of 52 female exstrophy patients, we confirmed sexual activity in 81% (4% had not had sexual intercourse and data were not available for 15%). This only represents one diagnostic group but suggests a good outcome for many patients. In groups requiring reconstructive genitoplasty or vaginoplasty, there is considerable discussion around the timing of surgery. We know that genital surgery is likely to affect sensation



Figure 41.3 Dissection of exstrophy penis showing the neurovascular bundles.

but whether this affects long-term sexual pleasure or orgasmic function continues to be questioned [16]. For those having early surgery, many will require additional intervention at puberty. For a few this will be a major revision and some may only need a minor revision, but prediction or selection of these is at this point impossible. Those surgeons and centers undertaking this kind of surgery need to be well-acquainted with the risks and benefits and be prepared to have a frank conversation with parents or patients about the options available. Decisions should be supported by a multidisciplinary team (MDT) as this provides important advocacy for the patient, ensuring that the decisions taken are in their best interests; psychology input and support is vital as part of this. The MDT also provides important backing for the medical team undertaking direct treatment (be that surgical or endocrinological). It is possible that such treatments may be questioned in time and continuous peer-review will be important in support of the contemporaneous decision making.

Fertility outcomes

Major congenital anomalies do not necessarily prevent fertility but they can place limitations upon it. In bladder exstrophy, we found that patients had undergone a mean of eight procedures. Sixty-eight percent of our female exstrophy patients were successful in becoming pregnant, although 79% had taken longer than one year to do so and 26% had required fertility treatment. Fifty-six percent achieved live

births but the reported stillbirth or neonatal mortality rate was 7/100 (10 times the expected UK data). These deaths were explained by congenital anomaly, extreme prematurity in twins and at least one delay in an emergency delivery. The need for caesarian section is an obstetric decision but it is important to ensure

appropriate support for any delivery. Appropriate urological support for an emergency caesarian section will be difficult to achieve in many centers. For this reason, we advocate elective caesarian section in major pelvic conditions (such as bladder exstrophy) and shared care throughout pregnancy [17].

DOS AND DON'TS

Do

- Always think about the long-term follow-up when carrying out major reconstructive surgery
- Work in a multidisciplinary team
- Consider the upper tracts and renal function
- Be clear about the indication for surgery

Don't

- Rule out surgery in childhood but do think about why and whether that is the best time
- Ignore new symptoms in cystoplasty

CLINICAL PROBLEM/CASE: DISCUSSION

The most common complications of a cystoplasty are mucus production and biochemical changes. When bowel is grafted into the urinary tract it still continues to behave as bowel – including its tendency to produce mucus. There has not been any treatment that appears to improve this. Some factors such as infection, diet or pregnancy may worsen production. A good fluid intake and bladder washouts are an important part of preventing a build-up. Catheter size is also important. A size 14 Ch catheter is ideal as it will predictably allow mucus to drain; a size 10 Ch is too small and will not. Many people will manage with a size 12 Ch. Metabolic changes are often subclinical but a net loss of bicarbonate in exchange for chloride and a tendency to absorb ammonium ions will create hypochloremic metabolic acidosis. If the levels are measured as normal or near normal, treatment is unlikely to be needed but abnormal levels or symptoms require oral replacement with bicarbonate. Other complications include bowel complications (change in bowel function, small bowel obstruction and fistulae) urinary tract infections (20–30%), stones, B12 deficiency (depending on segment used), perforation and the potential for malignant change.

The first and most obvious risk for bladder stones is having had a previous stone; any patient who has suffered stones in their cystoplasty should be given advice about how to reduce the risk of future occurrences. In a paper that examined the characteristics of those patients who did develop stones, the two most significant findings were the low 24-hour volume in stone formers and lower urinary citrate. An increase in fluid intake (≥ 2.5 liters per day), avoiding salty and high oxalate food (e.g. rhubarb, spinach, chocolate, peanut butter), along with a healthy balanced diet are important preventative steps. Ensuring regular bladder drainage is vital as are weekly bladder washouts to ensure there is no mucus build-up. Regular imaging follow-up is also essential in order to detect stones early and remove them before they increase in size.

The only option for removing large stones is with an open procedure. The patient has had multiple abdominal procedures and his Mitrofanoff channel is catheterized to allow filling of the bladder. An incision of between 5 and 10 cm is made (often using an old scar, size dependent on the size of the stone), the bladder is opened between stay sutures and the stone lifted out intact. The bladder is closed in two layers and left on free drainage for 10 days.

In our institution, once a patient is using their cystoplasty and has passed a trial of void/emptying, they are seen at 6 months with an ultrasound, MAG3 renogram and bloods (including renal function, bicarbonate, chloride, vitamin B12 and full blood count). The bloods and ultrasound are repeated annually and a GFR is performed every 5 years. Some would argue that a B12 assay is unnecessary if you know which bowel segment has been taken. We have many patients who have had previous surgery and for whom it is difficult to know which segment has been used. Therefore, since a B12 test is cheap and safe, we perform this routinely as part of our protocol as the consequences of missing a deficiency are irreversible neurological deficit.

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