Pediatric Minimal Access Surgery



Edited by
Jacob C. Langer
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Preface

The widespread use of minimal access techniques for performing surgical procedures has arguably been the greatest advance in surgery over the past 15 years. In addition to lessening the visible scar, the use of minimal access approaches may decrease postoperative pain, minimize postoperative ileus, shorten hospital stay, and decrease cost. In some cases this technology has been credited for other benefits such as decreased adhesion formation, better visualization of anatomy, and attenuation of the surgical stress response. At its inception, minimal access surgery was limited to the adult population. In the last decade, it has been widely adopted by pediatric surgeons, and applied in creative ways to the unique conditions and needs of the heterogeneous pediatric population.

There have been a number of books written on the topic of pediatric minimal access surgery. As the field is expanding rapidly, these books have grown from small monographs to larger, more comprehensive volumes. The focus of most texts on this topic has largely been technical and procedural as it is these advances that have allowed minimal access surgery to be applied in a seemingly limitless fashion in the pediatric population. As with many technological advances, however, there has been a tendency for evidence-based practice to lag behind the many other forces (e.g., economic incentives, academic interest, and consumer demand) that drive the creation and application of new and innovative techniques. In contrast to previous publications, this book focuses on the principles behind the use of minimal access approaches, and the evidence, to date, that has been accumulated to support their use. We recognize that for many conditions and operations, there is a dearth of significant evidence or that the evidence is extrapolated from the results of operations in the adult population, and thus may not be directly applicable to children. This first edition can be viewed as "embryonic" in its development. It is intended to stimulate readers into taking an evidence-based and principle-based approach when using minimal access technology. The "holes" in our outcomes knowledge need to be studied and filled in. We also hope that this book will stimulate an interest in contributing to the acquisition of evidence by having pediatric caregivers participate in proper trials and studies.

This book is dedicated to Ferne, Jessica, Benjamin and Alexander, Laura, Samantha, and Melanie, without whose support and understanding we would never have been able to complete this work. We thank all of the contributors for their time and patience. Most of all, we dedicate this book to all of the children, past and future, who have taught us and helped us to use our hands, minds and hearts in the pursuit of a better way.

Jacob C. Langer
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1

Introduction: An Evidence-Based Approach to Pediatric Minimal Access Surgery

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Progress in medicine is made in small steps. Many day-to-day decisions are made by trial and error, and the clinician routinely evaluates the results of an intervention and makes further decisions based on these results. Similar situations in subsequent patients are managed in a way which is based on the results of decisions made in previous patients. This process is routinely known as the acquisition of "clinical experience," which is accumulated over years and then taught to others both formally and informally.

Unfortunately, clinical practice which is developed in this way is not always in the best interests of patients, as the conclusions drawn from personal experience may be fraught with error from a wide number of sources. These include the normal variability of complex biological systems, such as human beings, the tendency for people to "see what they want to see" and to draw false conclusions based on the incorrect interpretation of clinical data, biases in patient populations, lack of physician equipoise, lack of adequate follow-up, and the tendency to generalize conclusions from one population of patients to others in which the conclusions may not be valid. In addition, the economic, political, and academic pressures on physicians in the modern world may result in the adoption of clinical practices which are not in the best interest of patients for a variety of reasons.

In recent years, there has been a move toward the adoption of "evidence-based" practice. The stimulus for this has come from several sources including the increasingly recognized need to improve patient safety (1), pressure from the managed care industry

to improve outcomes while decreasing health care costs, and the recognition by health care professionals that personal experience may lead to incorrect conclusions and the adoption of ineffective therapies. In addition, the need for multidisciplinary centers of excellence for rare and highly complex therapies (e.g., fetal and bariatric surgery programs) is being recognized and driven by outcomes-based practices. However, generating evidence is a time-consuming and expensive activity, which requires dedication and training. For this reason, there remains a paucity of evidence which can be used to guide practice in medical, and particularly in surgical conditions.

1. CREATION OF EVIDENCE

Evidence can be generated from a number of different types of studies, which result in a variety of levels of "quality." Table 1.1 shows one way of characterizing levels of evidence, although many other classification systems exist. The principles of generating quality evidence are the elimination of bias, the ability to rigorously evaluate the statistical significance of the findings, and the validity of applying or generalizing the results to a wide population.

Most evidence in the surgical literature comes from case reports or case series, in which a group of patients treated in a certain way are reported, and results of treatment are evaluated without any comparison to any other form of treatment. Increasingly, retrospective comparative studies have been done using "historical" controls, that is, patients with the same condition who were previously treated using another modality. Although more useful than a simple case series, the use of historical controls does not consider the possibility of changes in other aspects of treatment over time or changes in the natural history of the disease over time as well as bias in treatment assignment. Attempts to overcome these problems by using matched historical controls or case–control techniques improve the validity of the evidence to some extent.

Prospective studies are clearly superior to retrospective studies, as the data acquisition can be standardized and is less likely to be biased. The prospective, blinded, randomized, controlled clinical trial is the gold standard for evidence-based decisions. However, it is important to realize that randomized trials are associated with a number of logistical issues. They are extremely expensive and difficult to carry out, particularly when they require a multiinstitutional approach. The results are often highly specific for a super-selected patient population and may not be generalizable. Particularly in surgical trials, controlling the actual technique of a surgical procedure can be difficult and may introduce unanticipated biases, and the natural evolution of surgical technique in the hands of individual surgeons coupled with the normal learning curve for surgical procedures may also introduce bias. These trials often take 3–5 years to complete. Over that time period, the surgical technique may change or even become obsolete (2), and

Table 1.1 Levels of Evidence

I	Evidence from at least one properly designed RCT
II	Evidence from nonrandomized studies
	a. Well-designed controlled but nonrandomized trials
	b. Well-designed cohort or case–control studies
	c. Poorly controlled comparative studies
III	Opinions of respected authorities, based on clinical experience, descriptive studies, or reports of expert committees

there may be significant changes in referral patterns or institutional practices. One must achieve equipoise among all treating physicians. Patient accrual and the possibility of treatment outside of the trial are considerations for those trials of rare disorders. A difficult issue centers on the ethics of assigning/withholding innovative therapy to select patients. Before the trial begins, there must be a willingness on all stakeholders to abandon ineffective therapy if the results prove so. Negative trials, in which no difference between treatments is found, may not be statistically sound if the type II error is not calculated and reported (3).

2. APPLICATION OF EVIDENCE

The next step to applying an evidence-based approach to a clinical problem is to gather and evaluate the evidence and then to develop clinical practice guidelines which are based on the best evidence available. This can be a difficult task, considering the huge volume of scientific publications produced each month and the limited time most individual surgeons have. For specific questions, one can use electronic databases such as Medline, but this is time-consuming and the sheer volume of information may be overwhelming. For this reason, most clinicians rely on reviews to educate them on an ongoing basis.

Reviews of the literature may be classified as "narrative reviews" or "systematic reviews." Narrative reviews include literature reviews which are commonly published along with a new case report of a specific condition or technique, "collective reviews" assembled by a single author who may or may not be an expert in the field, editorials, and "review articles." All of these narrative reviews are subjective and tend to reflect the opinion of the author. In most cases, there is no attempt to evaluate the quality of the evidence presented, and the completeness of the review may also be questionable.

Systematic reviews are characterized by an attempt to minimize arbitrariness and to standardize and report the technique used for the review. A clear search strategy is developed, and some kind of grading system is used to report the quality of the evidence in each paper used. The technique for combining the evidence gleaned from individual studies is determined and reported. In essence, a systematic review uses research studies, rather than patients, as the study material.

The most organized and rigorous form of systematic review is known as "meta-analysis." This technique involves the collection and analysis of previous prospective randomized trials, using statistical methodology to combine the results of these trials and create a unified, statistically more powerful conclusion. Although this technique is widely used, there are many problems with it. The most important is the inability to standardize techniques and patient populations used in the trials and the inevitable compromises required to place differing trials into the same analysis.

There are a number of sites for the clinician to access systematic reviews. Perhaps the most well developed is the Cochrane database, initially founded by Archie Cochrane in the UK (http://info@updateusa.com/cochrane/abstract.htm). This database provides up-to-date systematic reviews on a wide variety of clinical problems, which are done at the highest possible level and which are updated regularly as the field develops. In addition, methodology reviews are developed and collected by the Cochrane Collaboration.

3. EVIDENCE-BASED PEDIATRIC MINIMAL ACCESS SURGERY

As with most new technologies, minimal access surgery began with case reports and case series describing techniques and preliminary results. Procedures such as cholecystectomy,

which are very common in the adult population, were the first to be described, followed by increasingly complex and uncommon operations. Next appeared comparative studies using historical controls, and only later were prospective controlled or randomized trials performed. However, in the adult population, a number of randomized trials have now been done for the more common operations such as cholecystectomy and appendectomy. In some cases, such as appendectomy, where the benefits of a minimal access approach are less clear, there have been enough trials done to warrant a number of meta-analyses, including one from the Cochrane collaboration (4).

Pediatric surgeons were slow to adopt minimal access surgery. Reasons for this included delay in the downsizing of instrumentation and optics and the fact that the more common operations in adults, such as cholecystectomy, are much less common in children. However, the pediatric minimal access surgery literature has followed the same pattern as the adult literature, with case reports followed by comparative studies with historical controls. At the time of this writing, however, there have been very few prospective randomized trials in the field of pediatric minimal access surgery (5,6).

Why has the pediatric surgery community not pursued what is clearly necessary to validate this technology and create good evidence for practice? One reason is that there has been a tendency to extrapolate the evidence from adult trials and apply them to children. Although this may be valid for some conditions and operations, there are many differences between children and adults with respect to underlying medical conditions, indications for surgery, pain tolerance, and postoperative recovery, which may make adult data irrelevant. Secondly, many of the pediatric conditions which are treated using minimal access techniques are relatively rare, and multicenter studies are necessary to do an adequate trial. Obtaining funding for such trials and overcoming the logistical issues are difficult. Thirdly, many surgeons have become convinced that the minimal access approach is superior and have lost equipoise, making it difficult for them to participate in what they would consider to be an unethical trial. Fourthly, the use of minimal access surgery has often been advertised by surgeons and hospitals as a business tool to attract patients, a factor which clearly interferes with the performance of a randomized trial. Until resources and greater collaboration allow for the broader application of high quality prospective clinical research, there will be a continued dependence on observational data in shaping the practice of minimal access surgery. However, this underscores the importance of maximizing the methodological quality of the research. Clearly the development of rigorous guidelines, similar to the CONSORT-mandated guidelines for randomized trials (7), is needed for nonrandomized data. With that said, there are a number of operations which lend themselves to a randomized trial because of the relative frequency, simplicity, and lack of any good evidence of superiority over other approaches. Examples include Ramstedt pyloromyotomy, thoracoscopic pleural debridement for empyema, and the Nuss repair of pectus excavatum. Each of these questions is associated with challenges which must be overcome, but each needs a properly controlled trial to provide appropriate evidence to guide practice in an evidence-based way.

This book will attempt to discuss the present state of knowledge about the use of minimal access surgery in children. We have attempted to provide the reader with current evidence, both from the children, where available, and from the adult literature, if appropriate. In many cases, the literature is still at the case series stage, and much more work needs to be done. The authors have also attempted to delineate principles which can guide the clinician in the use of these techniques and which can be used to design future studies for the acquisition of better evidence. This book is just a beginning. We hope that it will stimulate interest in an evidence-based approach and will encourage

pediatric surgeons to initiate and participate in studies and trials which will continue to advance the field.

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History of Pediatric Minimal Access Surgery

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1. EVOLUTION OF TECHNOLOGY

Substantial improvements in surgery have been made in the last 150 years. Since the introduction of antiseptic technique by Lister and the introduction of inhalation anesthetics at Massachusetts General Hospital in 1846, surgery has progressed at a rapid pace. Prior to this time, surgical procedures were avoided and, if performed, they were brief. The best surgeon was the fastest surgeon who caused less pain to his restrained and un-anesthetized patient (1).

Early on, the idea that "large problems required large incisions" dominated surgical thinking. Adequate exposure was the key to a safe and successful operation. Today, exposure is still essential for a safe and successful operation, except that it now can be achieved with minimal skin incisions and use of minimal access techniques.

Minimal access surgery (MAS) has its roots in the early 19th century. The first report was in 1805 by Bozzini (2) who attempted to view the bladder of a woman using the candle powered lichleiter scope, which he developed (Fig. 2.1). The medical community criticized him for his aggressiveness, and little was done to advance the technique until Desormeaux, in 1853, ignited a mixture of alcohol and turpentine to produce a light source (3) (Fig. 2.2). In 1868, Bruck introduced electrical illumination (4). He used a platinum loop heated by electric current. During the same year, Kussmaul performed esophagogastroscopy on a willing sword swallower (5). The incandescent bulb produced by Edison in 1880 tremendously improved visibility. In 1883, Newman used the miniature

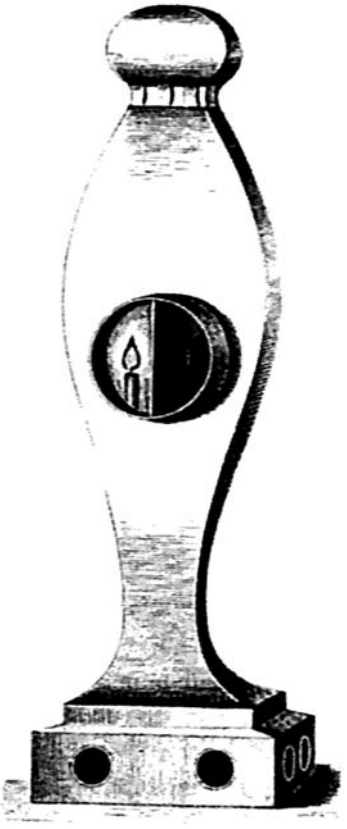


Figure 2.1 Bozzini's cystoscope using the lamp–mirror–candle system. (Courtesy of National Library of Medicine.)

version of the bulb mounted at the end of the cystoscope (6). The major problem with this device was that the light produced too much heat, making it potentially dangerous.

George Kelling (7), in 1901, reported the first celioscopic examination when he used a cystoscope to examine the abdominal cavity of a dog. In 1911, Jacobeus published his results of using laparoscopy and thoracoscopy for diagnostic purposes (8). He was the first to use the technique in humans and described pneumoperitoneum as the first step in performing laparoscopy. The first peritoneoscopy in the USA was also in 1911; Berheim (9) used a one half-inch proctoscope and an electrical headlamp to examine the abdominal cavity through the abdominal wall, and called it organoscopy.

Since then, multiple innovations have been made in instrumentation and technique. Fiberoptic transmission was patented in 1928, but it was not until 1952 that Fourestier et al. (10) described a method to transmit an intense light from outside the body cavity along a quartz rod to the tip of the endoscope. By 1957, this technology was used in flexible telescopes, and is now called the "cold light system." The next major advance was the development of the Hopkins rod lens in 1966 (11).

As the optics and illumination improved over the years, so did the techniques of pneumoperitoneum and entering the abdominal cavity. In 1918, Goetze invented a spring mechanism for abdominal puncture and gas insufflation (12). Ordnoff invented the trocar in 1920 (12). The trocar had a pyramidal tip and a valve to prevent the

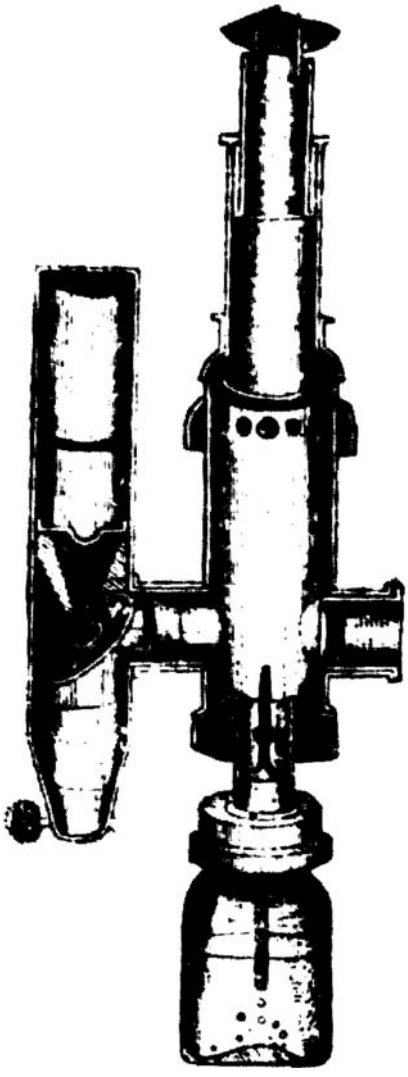


Figure 2.2 Desormeaux kerosene lamp. (Courtesy of National Library of Medicine.)

escape of pneumoperitoneum. In 1911, Fervers used oxygen and carbon dioxide but later turned to room air for pneumoperitoneum. Zollikofer preferred carbon dioxide to room air for insufflation (13). In 1938, Veress (14) modified Goetze's needle for the purpose of creating a pneumothorax for the treatment of tuberculosis. Since then, the Veress needle became the needle of choice to perform safe penetration of the abdominal wall. As the procedure became more widely accepted, increasing numbers of access related complications were observed. This situation prompted Hasson, in 1974, to introduce the open approach for trocar placement, which helped to decrease the incidence of bowel injury (12).

In 1929, Kalk (15) introduced many new instruments and ideas to apply a safe pneumoperitoneum (Fig. 2.3). He used a trocar with a spring-loaded stylet, introduced the 30° viewing scope, performed the procedure under sedation and local anesthesia, and used room air for pneumoperitoneum using the standard rubber bulb used with sphygmomanometers or rectoscopes. For several decades, manual air insufflation was



Figure 2.3 Hans Kalk, MD.

the method of choice. As laparoscopy became widely accepted, and as procedures become more therapeutic than diagnostic, requiring longer operating time and use of electrocautery, carbon dioxide became the gas of choice for the creation of pneumoperitoneum. The simple manual insufflators were no longer adequate to handle the longer operating times and flow requirements with multiple trocars and instrument exchanges, and this ushered in the introduction of modern insufflators.

Insufflation of the abdomen or chest cavities for MAS procedures has important physiologic effects. Much of this physiology has been studied in adults, but there has been very little work done on this subject in children. Pneumoperitoneum is required in the majority of cases for successful laparoscopy. There has been some debate in terms of which medium is best. Once the intra-abdominal volume exceeds the ability of the peritoneal cavity to expand without a significant increase in abdominal pressure, increase in pressure leads to detrimental physiologic effects. This is especially true when the cavity is small, as in children. Much work needs to be done on the physiologic effects of pneumoperitoneum in children. Coronary, hepatic, mesenteric, and renal flow may be impacted as well as cerebrospinal fluid pressure and pulmonary dynamics (16).

2. APPLICATION OF MAS TO SURGICAL PRACTICE

It took >100 years for MAS to embed itself into surgical thinking. During the 1960s and 1970s, gynecologists took the lead in the development of MAS while most of the surgical

community ignored the possibilities of this new technique (17). The surgeon was required to hold the scope up to his eye with one hand and operate with the other. The first laparoscopic appendectomy was performed in conjunction with a gynecologic procedure using this technique (18).

The application of MAS to general surgery began when Muhe (19) performed the first laparoscopic cholecystectomy in 1985. Mouret, Dubois, and Perissat, in 1987, helped popularize the laparoscopic cholecystectomy (20). The technical innovation that helped transition laparoscopic surgery into mainstream general surgery was the invention of video laparoscopy. This development allowed the camera to be attached to the telescope's eyepiece and the image viewed onto a television monitor (Fig. 2.4). Both hands of the surgeon were freed, and visualization of the operative field was available to the rest of the surgical team. With a team, it became possible to perform more technically demanding procedures.

Within several years, laparoscopic cholecystectomy became the standard of care. Since that time, MAS has been applied to numerous other procedures with good results. The sweeping success of this laparoscopic revolution has thoroughly changed the way surgery is performed.

Surgical procedures can be categorized on the basis of their complexity and can be divided into excisional, in which a structure is removed; ablative, in which tissue is destroyed; or reconstructive, in which structures are repaired, joined or connected. Excisional or ablative procedures are easier to perform than reconstructive procedures and are more easily adapted to endoscopic techniques. Operations can also be categorized as either high or low volume procedures. High volume procedures achieve success over a shorter period of time than low volume procedures because of the ability to learn the procedure more quickly and because of the market opportunity presented for technology development.

The success of laparoscopic cholecystectomy in adults was in large part due to the excisional nature of the procedure and the high volume of cases. Other excisional



Figure 2.4 Video laparoscopy.

procedures, such as cholecystectomy in children, have not been as quick to convert because of lower case volumes. Neither have other high volume procedures, such as coronary artery by-pass grafting, been as rapidly converted to a MAS approach because of the complexity and reconstructive nature of MAS (21).

3. APPLICATION TO PEDIATRIC SURGERY

The excitement that general surgeons first experienced with laparoscopic cholecystectomy during the 1990s was transmitted into a variety of other specialties. However, the widespread enthusiasm among general surgeons to perform minimal access procedures was muted initially among pediatric surgeons.

There had been a great resistance to MAS in the pediatric population for a number of reasons. It was traditionally felt that children did not experience pain. The costs of laparoscopy and thoracoscopy, with the use of disposable instruments and trocars, were felt to be too high. Equipment developed for adults was not small enough for infants and children. It was felt to be too hard to do, too hard to learn, and the cases were felt to take too long to set up and to perform. Many surgeons thought that laparoscopic and thoracoscopic cases really did not apply to children, and because pediatric surgeons already prided themselves on small incisions, they felt that MAS was unnecessary. Many felt MAS was not safe and its efficacy not proven.

In response to these criticisms, it has become clearer to many pediatric surgeons that shorter hospital stays, decreased postoperative pain, quicker return to normal activities, and parents' earlier return to work counterbalance the higher cost of MAS. Also, the current trend towards reusable instruments and trocars further lowers the cost of MAS. Continuing surgical education, training of the surgical team, and use of dedicated minimal access operating rooms, allow for faster turnover of patients in the operating room suite, as the surgeon and the staff become more comfortable with the procedure. Evidence continues to accrue which demonstrates that MAS is both safe and effective in infants and children.

Early attempts to make the telescope smaller resulted in unacceptable optics and poor vision. This was probably the greatest surgical obstacle to MAS in pediatric surgery. With the advent of improved fiber optic light sources, lens systems, and video cameras, a small telescope with superior optics and adequate light was possible. This process began in 1970, when Gans introduced the prototypic pediatric instruments to the USA (12).

Despite the previously mentioned impediments, pioneers in the field persisted, and now MAS is broadly applied to the surgery of infants and children. The techniques that were found to be useful in adults have now been applied in children. Pediatric surgeons developed innovative modifications of technique and instrumentation to account for the smaller working space in the pediatric patient. But even more importantly, the different spectrum of pathology has led to the development of many techniques, which are specific to the pediatric patient. The advances in pediatric MAS, particularly instrumentation, have subsequently been used in adults; adult surgeons often request the smaller telescopes and instruments developed for children. The modern pediatric MAS surgeon now uses elegant and delicate instruments with telescopes from 1 to 5 mm in size, has excellent optics that are steadily improving, and functions in a fully equipped operating suite devoted solely to MAS (Fig. 2.5).

Sir Willian Osler said, "Diseases that harm call for treatments that harm less". This quote represents the impetus for the development of MAS. Because of such influence,



Figure 2.5 MAS suite.

during the last quarter of the 20th century, especially during the last decade, there has been a paradigm shift in the technique used to perform surgery (22). Using MAS, surgeons have learned that they can greatly reduce the access trauma (the incision), the primary cause of pain and disability related to traditional surgery. With this approach, patients can now expect a less painful convalescence, a shorter hospital stay, a rapid return to full activity, and excellent cosmetic results (22). For parents, this means that the family unit returns to normalcy quicker, they can get back to work and their normal activities sooner.

4. FUTURE OUTLOOK

Advances in equipment and instrumentation have expanded the application of MAS to patients ranging in age from premature infants to teenagers. However, small infants present significant technical challenges related in part to the smaller working area. The performance of suturing and intracorporeal knot tying is very difficult and presents possibility for injury to surrounding organs. Robotic technology presents an attractive solution to these technical challenges. Robotic surgery holds the promise of minimizing the risk of injury to surrounding tissues and allowing controlled precise movements by filtering out the surgeon's tremor and scaling down instrument movement so that large movements in the console can be translated into much smaller repetitive motions at the instrument's tip. In the future, robotic technology will potentially play an important role in expanding the applications of minimally invasive pediatric surgery (23).

An additional advantage of the robotic technology is the ability to disseminate pediatric surgical expertise through telementoring and telepresence surgery (24). This will allow the robotic surgeon in one institution to guide the surgical care or complete a minimally invasive operation of a patient many miles away.

Integration of preoperative (3D CT imaging) or "biomaterial enhanced" operative imaging studies (e.g., use of fluorescence emitting dyes coupled to tissue specific compounds captured with infrared cameras) with real time MAS are likely to emerge in the

future as technology advances. This will enable the minimal access surgeon to "see" structures beneath the operative surface, for example, feeding vessels and tumor margins, further minimizing potential for errors and complications.

Charles Darwin in 1869 said, "It is not the strongest of a species that survives, but the one that is most adaptive to change". Present day surgeons must take this advice seriously. These are times of rapid change. As pediatric surgeons encounter newer and better technologies, they will integrate them into practice, always striving to improve the surgical care of children. The ultimate destination for the patient must be "surgical cure" (25). Application of MAS to pediatric surgical problems is an excellent example of this surgical dictum.

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3
Anesthesia for Pediatric Minimal Access Surgery

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1. INTRODUCTION

When minimal access surgery (MAS) was first introduced into the mainstream of adult surgery in the 1980s, anesthesiologists found themselves needing to adjust to a new set of variables in order to provide optimal intraoperative care for their patients. It became necessary to minimize the amount of air in the patient's gastrointestinal tract to improve surgical visualization, to continue neuromuscular blockade throughout surgery, to consider hemodynamic consequences of intra-abdominal insufflation and the sitting position, and to anticipate longer operative times. Thoracoscopy added the challenge of single-lung ventilation with double-lumen tubes or bronchial blockers in cases which otherwise did not require lung isolation. Intrathoracic insufflation of gas further shifts the mediastinum into the dependent, ventilated lung. Because of the additional burdens and duration of surgery, many patients were excluded from these novel approaches. Patients with significant cardiopulmonary disease were considered to be at

very high risk and, therefore, could not reap the benefits of reduced postoperative pain, improved cosmetic appearance, and in some cases, superior surgical outcome.

Pediatric patients had to wait until some of the kinks were worked out in adults, as well as for the development of appropriately sized instruments before they could undergo MAS. Children, however, represent the greatest beneficiaries of the potentially tremendous surgical advantages. They have the most to gain in terms of reducing adhesions and improving cosmetic results, and they are, in general, the most physiologically well-equipped to handle the additional stresses imposed by endoscopic surgery. They have large cardiac reserve and rarely have chronic pulmonary insufficiency; thus, they can tolerate intra-abdominal or intrathoracic insufflation of gas with minimal change in hemodynamic measurements. Furthermore, children rarely suffer any sequelae of the hemodynamic stresses caused by insufflation (e.g., tachycardia and hypo- or hypertension) because they do not have underlying coronary artery or vascular disease.

Since the 1990s, the production of small endoscopic surgical instruments has made the common application of adult surgical procedures possible for even tiny neonates. In many pediatric hospitals, MAS has accounted for a greater percentage of intra-abdominal and thoracic operations than open surgery. The complexity of the cases continues to increase, while the patient selection becomes ever more inclusive. Pediatric anesthesiologists have had to become adept at providing safe anesthesia for patients with a whole new range of problems. Optimal patient positioning for MAS often dictates that the baby is at the end of the bed, a long distance from the anesthesiologist and anesthesia machine. They are often in steep reverse Trendelenberg position or turned 90° on the bed. The need for maximal operative space has meant that nitrous oxide must usually be avoided. The use of cold gases for insufflation can make temperature maintenance more difficult. Decisions regarding postoperative pain control can be tricky. The decision to use neuraxial blocks, including caudals and epidurals, may need to be delayed until it is determined whether additional incisions need to be made (e.g., to remove large solid organs or masses or conversion to an open procedure). Discussion with the family regarding epidural placement should be done prior to surgery, so that if it is deemed appropriate, it can be placed at the conclusion of surgery.

To date, the large experience with pediatric MAS demonstrates improved cosmesis, reduced postoperative pain, earlier feeding, fewer intensive care unit (ICU) admissions, and shorter hospital stays (1–9).

2. GENERAL CONSIDERATIONS

2.1. Patient Selection

In the early experience with MAS, children without significant cardiopulmonary disease were the only ones thought to be amenable to the hemodynamic derangements imposed by gas insufflation into either the peritoneal or the thoracic space. Although these patients certainly represent the least challenging group, it has become clear that those who have significant cardiopulmonary disease are benefited the most. Postoperative pain is reduced and thereby may reduce splinting and atelectasis. Shorter hospital stays reduce the risk of acquiring nosocomial infections in these high-risk patients. Less manipulation of the bowel results in fewer adhesions and may therefore simplify subsequent surgery in ill children who are likely to need further surgery.

As with all anesthetics, safety begins with a careful history and physical examination. Derangements in cardiac and pulmonary performance should be sought in order to determine which patients may not readily tolerate the effects of gas insufflation. Mild

cardiopulmonary disease (e.g., asthma or a left-to-right intracardiac shunt) seldom represents a significant management problem. However, severe restrictive lung disease as with advanced neuromuscular disease or kyphoscoliosis may present a challenge. Likewise, children with severe cyanotic congenital heart disease may not present unique challenges for MAS. Patients with other severe, underlying medical disease need to be evaluated on a case-by-case basis. Derangements that were once thought to be contraindications to MAS now represents some of the ones that are best served by the MAS. Coagulopathies, for example, in theory represent a challenge because small amounts of blood in the surgical field diminish visibility by absorbing light. However, laparoscopic splenectomy for diseases such as idiopathic thrombocytopenia purpura and hereditary spherocytosis are now common. It is possible to do simultaneous splenectomy and cholecystectomy for patients with hemolytic disease avoiding one or two large, upper abdominal incisions.

Healthy children represent the ideal candidate for MAS because of their enormous cardiac and pulmonary reserves. They, in general, lack underlying atherosclerosis and therefore tolerate changes in heart rate, blood pressure, and cardiac output without sequelae such as myocardial infarction or stroke. Infants <6 month-old depend on increases in heart rate to compensate for alterations in pre- and afterload because they are unable to increase stroke volume until the contractile function of the heart matures. However, infants with significant congenital heart disease may not have the same reserve and alterations in pre- and afterload introduced by insufflation of CO_2 gas into the thoracic or peritoneal cavity may seriously compromise cardiac output. In particular, babies with single ventricle physiology, who rely on passive conduits for pulmonary blood flow, may not get sufficient preload to maintain oxygen saturation or blood pressure. Being vigilant with the maximum insufflation pressures allowed in both the thoracic and peritoneal cavities is vital in preserving optimal cardiopulmonary function in these delicate patients. Limiting thoracic insufflation pressures to 4–6 Torr and intraperitoneal pressures to ≤ 12 Torr has been used successfully even in sick neonates (10–12). Over the past decade, smaller and smaller infants have been successfully treated with MAS, recognizing that they may have the most to gain from these innovative techniques. Smaller incisions, often placed more remotely from the diaphragm than conventional open surgery, allow for better respiratory effort and function postoperatively. The need for reduced doses of respiratory depressant opiate analgesics may allow these babies to be extubated earlier as opposed to open surgery or avoid ICU admissions. The obvious benefit is the reduction in the risk of pneumonia. In a retrospective review of neurologically impaired children undergoing fundoplication, the incidence of postoperative pneumonia was shown to be 1.8% with MAS vs. a reported incidence between 14% and 40% following open fundoplication (13).

2.2. Patient Positioning

MAS necessitates the greatest possible degrees of freedom for the surgeon in order to accomplish a three-dimensional procedures with two-dimensional visualization. Conventional surgical positioning is vastly altered and frequently dictates that the patients be either at the far end of the operating table (e.g., for fundoplication) or turned 90° away from the anesthesiologist (e.g., for pyloromyotomy). Ensuring that the length of airway circuit and intravenous (IV) tubing is adequate are essential to avoid tension and dislodgement of endotracheal tubes (ET) and IVs. It is advantageous to have the ET taped to the side of the face toward the anesthesiologist so it is available for inspection and suction when necessary. The same is true for IVs so that malfunctioning can be expeditiously evaluated and corrected without interrupting surgery. Avoiding excessive abduction at

the shoulders can be a challenge for thoracoscopy in small children because the nondependent arm is often brought up over the head. Alternatively, the arm may be prepped into the field and therefore IV access must be avoided. Axillary rolls are used to avoid brachial plexus injury to the dependent arm and to improve respiratory excursion. Perineal surgery (e.g., pullthrough for Hirschsprung's disease) makes IV access in the lower extremity undesirable as the entire lower half of the body is usually within the sterile field. When alternative access is unavailable, IVs can be covered with sterile dressing and remain on the surgical field. They then, however, become unavailable for inspection should they cease to function during the operation. The reverse Trendelenberg position is frequently employed in order to have the bowel fall away from upper abdominal organs such as during a fundoplication or a cholecystectomy. Care must be taken to bolster patients from sliding towards the foot of the bed.

Recently, pediatric surgeons have been employing robot-assisted MAS (e.g., da Vinci Surgical System, Intuitive Surgical, Sunnyvale, CA) to provide an improved three-dimensional view. Robotic surgery means the primary surgeon is at a remote site in the operating room with the robotic controls, while an assistant is at the operating table to position the robot's arms. Surgery employing robotics necessitates that small children be elevated on foams and blankets so that the robotic arms are free to move without abutting the operating table. It is essential that ET placement and IVs are impeccably inspected and secured as access is severely limited to the patients. Moving the robot away from the patient entails disengaging the instruments prior to moving the robotic cart, which may be time consuming and cumbersome (14).

Open thoracic surgery is always performed in the straight lateral decubitus position. In contrast, thoracoscopy procedures require one of three positions depending on which area of the mediastinum is being dissected. Patients are nearly prone for posterior mediastinal surgery (e.g., esophageal atresia repair), nearly supine for anterior mediastinal surgery (e.g., thymectomy) and straight decubitus for middle mediastinal operations (e.g., lobectomy). For prone and semi-prone thoracoscopy, it is critical that care be taken to avoid kinking of the ET or inspissation of secretions in small ETs. Using warmed, humidified circuits is helpful in reducing the viscosity of airway secretions and should be considered for all prone cases, especially when small ETs are employed as these are the most difficult to effectively suction when they become occluded.

As MAS often requires steep positioning of small patients, whether it be reverse Trendelenberg position or 30° from prone, it is vital to ensure that the patients are secured on bolsters and either taped, seat-belted to the table, or supported in a molded "beanbag." Slipping of axillary rolls may lead to brachial plexus injuries if the dependent shoulder is allowed to abduct $>90^\circ$.

2.3. Anesthetic Considerations

The induction of anesthesia in infants and children is accomplished by either the inhaled or the IV route. Being mindful of the importance of the available workspace for the surgeon dictates certain elements of anesthesia practice. It is crucial to limit positive pressure ventilation by facemask as much as possible before laparoscopic surgery because intraluminal bowel air reduces the available workspace, making some procedures impossible due to poor visualization. Bowel obstructions from atresias or malrotation do not allow gas egress distally and may preclude a laparoscopic approach. Prompt suctioning of the stomach following endotracheal intubation may prevent air from passing the pyloru. Once in place, a sump-type suction catheter should remain at least for the duration of the surgery to keep the stomach and bowel as decompressed as possible. Nitrous oxide is best avoided for laparoscopy in order to reduce intraluminal bowel gas as much as

possible and for thoracoscopy to allow maximal oxygenation under adverse ventilation–perfusion conditions.

Endotracheal intubation is preferable for all MAS. However, short, pelvic procedures may be done with the use of a laryngeal mask airway. Brief, low-pressure insufflation has been reported to maintain barrier pressure at the lower esophageal sphincter and therefore should not pose a substantial risk of gastroesophageal reflux and aspiration in low-risk patients. However, surgeon preference may dictate intubating these patients in order to establish controlled ventilation with less abdominal muscle use and greater regularity of the respiratory pattern.

Endotracheal tube selection may be affected by MAS because of the need to ventilate against extrapulmonary pressure from peritoneal insufflation and the cephalad displacement of the diaphragm. Rounding up to the larger ET size for age or employing a cuffed rather than the more traditional uncuffed ET in small children may allow improved ventilation under adverse situations, for example, thoracoscopy in a child with underlying diffuse pulmonary disease. Uncuffed ETs that leak at < 15 cmH_2O may not allow adequate ventilation when the peritoneum is insufflated. The use of cuffed ETs allows air to be added or removed as needed to improve ventilation. It is imperative to use a heated, humidified circuit in small children undergoing thoracoscopy because it may prevent secretions and blood from becoming inspissated and impeding ventilation and helps to reduce heat loss from the respiratory route.

Monitoring is with routine American Society of Anesthesiologists (ASA) monitors including electrocardiogram, noninvasive blood pressure, pulse oximeter, capnograph, temperature, and precordial or esophageal stethoscope. Patients with underlying cyanotic heart disease may benefit from the use of arterial lines in order to determine acid–base balance and allow for prompt treatment of derangements. Urinary catheters should be used in all at least for the duration of laparoscopy, and particularly in the shortest cases because it aids visualization by decompressing the bladder, while providing information regarding volume status. Small patients in the lithotomy or prone position may fail to produce enough urine to be measured at the collecting urimeter because of pooling in the dome of the bladder. This must be considered in light of hemodynamic measurements before aggressive hydration is used to correct "inadequate" urine output. Gentle pressure on the lower abdomen by the surgeon or intermittent reverse Trendelenberg may augment the flow of urine into the collecting bag for a more accurate assessment. Central venous pressure (CVP) catheters are reserved for patients in whom volume status is critical and fluid shifts are likely to be great. Indwelling central venous lines are used for chemotherapy can easily be used to monitor CVP, when necessary.

The type of anesthetic administered varies by procedure and physiologic status of the patient. Commonly, an inhaled, potent volatile anesthetic (e.g., sevoflurane or isoflurane), combined with an opiate and a nondepolarizing muscle relaxant is used. The volatile agents contribute to muscle relaxation and thus offer the advantage of greater surgical exposure with less insufflation pressure and the potential for less CO_2 leak from around the trocars. Other than for brief, pelvic operations, muscle relaxation should be maintained throughout MAS to provide the best working conditions for the surgeon with the minimal insufflation. Upon exsufflation, muscle relaxation can be allowed to wear off and reversed during wound closure.

2.4. Pain Management

One of the greatest benefits of MAS is the reduction in postoperative pain. The contrast from open surgery is greatest when one considers the subcostal incisions used for conventional surgery like diaphragmatic hernia, fundoplication, splenectomy, and

cholecystectomy or the large thoracotomies used for lung resections. The four or five trocar sites may be pre-emptively infiltrated with local anesthesia (most often <1 cc/kg of 0.25% bupivicaine). There is good recent evidence that the pre-emptive infiltration of local anesthetics into trocar sites reduces postoperative pain when compared with infiltration at the time of wound closure suggesting that the inflammatory cascade may be thwarted (15–16). In the smallest patients it is impossible to adequately block a large cutaneous area without exceeding toxic doses of local anesthetics whereas the small trocar sites may be liberally infiltrated while remaining well within nontoxic doses of local anesthetic.

Traditional regional anesthesia employed to minimize postoperative pain are typically avoided with MAS thereby removing the attendant risk of performing these blocks in anesthetized children. The most recent rendition of MAS hernia repair employing a transcutaneous stitch technique probably does not even require a caudal block, the nearly routine block used by pediatric anesthesiologists for decades. The placement of a thoracic epidural catheter in anesthetized children has always been a contentious issue because of the possibility of injury to the spinal cord in a patient who is unable to report paresthesias prior to serious neural damage. MAS obviates the need for neuraxial blockade. Postoperative pain is easily controlled with local anesthesia and intermittent small doses of IV opiates or Patient Controlled Analgesia (PCA) for the first one or two postoperative days.

Recently, even operations which are not readily amenable to laparoscopy are being performed with laparoscopic assistance in an effort to reduce postoperative pain and its attendant negative effects on pulmonary toilet by making small, trocar incisions in the upper abdomen while allowing large masses or solid organs, for example, massive spleen or multicystic kidney, to be removed via low pelvic incisions. The impact on postoperative management of pain and pulmonary toilet is obvious.

While pain is significantly reduced following MAS, it is certainly not eliminated. There are several causes implicated in post-MAS pain. It is thought that pressure peaks from gas insufflation may have a noxious effect on the phrenic nerve, perhaps from stretch caused by displacement of the diaphragm. This, is turn, may cause endoneural ischemia and lead to the common postoperative referred shoulder pain following MAS. Subdiaphragmatic instillation of local anesthetics has been advocated by some. Additionally, dissolution of CO^2 may have an irritant effect on the phrenic nerve and the peritoneum by virtue of the acidic milieu it creates in addition to the distention. Complete exsufflation following MAS may help to prevent some of the discomfort. Physical characteristics of the gas insufflated my also have a role in postoperative pain. While cool gas is rapidly warmed by the body, it is speculated that dry gas may have a damaging effect on exposed membranes (17). Warming and, more importantly, humidifying insufflated gas may help to reduce postoperative pain and diminish heat loss from the patient, although in practice this does not appear to be a substantial problem.

Despite the small incisions, inflammatory mediators are induced by skin pain nociceptors. Pre-emptive infiltration with local anesthesia has been shown to reduce postoperative pain for open surgery perhaps by blocking this sensory input to pain receptors. Recent evidence shows that the use of selective nonsteroidal anti-inflammatory drugs (NSAIDs), that is, cyclooxygenase-2 (COX-2) inhibitors, prior to surgery has a significant impact on postoperative pain presumably by reducing the cascade of inflammatory mediators caused by surgery (18,19). Pre- and postoperative use may reduce the need for opiate medications with their attendant side effects of respiratory depression and delayed gastric motility. COX-2 inhibitors do not interfere with platelet function and, therefore, alleviate the concern for postoperative bleeding like older, less specific

NSAIDs caused in the past (e.g., ketorolac). Drains and chest tubes are a frequent source of postoperative pain. Liberal use of local anesthetics may help, particularly surrounding chest tubes where pain may impede respiratory effort.

2.5. Fluid Management

Conventional teaching for anesthesiologists is to replace deficit fluids, administer calculated maintenance fluids, and to replace ongoing losses, in particular blood loss and "third space" losses, mostly evaporative. Fluid deficits may need to be replaced earlier in the course of MAS than open surgery because the combined effects of induction of anesthesia, reverse Trendelenberg position and insufflation of CO_2 conspire to reduce preload leading to an exaggerated reduction in cardiac output and potentially blood pressure. Patients with cardiac shunts who are extremely sensitive to changes in intravascular volume in order to maintain pulmonary or systemic perfusion may need to be hydrated prior to insufflation.

In general, third space losses are inconsequential during thoracoscopy as opposed to the 4–8 cc/kg per h loss incurred during open thoracic surgery. Laparotomies, in which the bowel is exposed for prolonged periods of time, may result in losses of 10–15 cc/kg per h or more. Reducing these fluid losses to the ambient environment may lead to a substantially lower overall volume of crystalloid replacement and less postoperative edema. Laparoscopic replacement volumes are difficult to estimate but are significantly less than with a laparotomy and should be based on heart rate, blood pressure, and hourly urine or central venous pressure when available.

Blood loss may take longer to control with MAS than with open surgery because of the reduced freedom of movement and exposure for the surgeon. Small hemorrhages impede visualization because hemoglobin absorbs light and may make it difficult to identify the source of bleeding quickly. It may be necessary to make an additional incision to gain control. Quantifying the amount of blood loss represents a challenge because small pools of blood are difficult to estimate in two dimensions. The traditional weighing of sponges to estimate blood loss in small children is replaced by estimating the volume in large suction canisters. Blood may pool in dependent areas including the pelvis that are not obvious without deliberate inspection. For the anesthesiologist, however, having greater visualization into the field via television monitors allows for prompt assessment and intervention as needed.

3. CARDIORESPIRATORY EFFECTS OF MINIMAL ACCESS SURGERY

3.1. Laparoscopy

Surgical exposure for MAS depends on the continuous flow of gas in order to produce distention in the peritoneal cavity and lung collapse in the thoracic cavity. Carbon dioxide is currently used because of its physical characteristics. It is noncombustible, highly soluble, and does not cause serious cardiovascular compromise in the event of an IV gas embolus. Its absorption can be readily eliminated via an increase in minute ventilation in healthy patients.

Changes in cardiovascular function during laparoscopy are affected by insufflation pressure, intravascular volume status, patient position, and anesthetic agents. While the increase in end-tidal CO_2 is easily handled by increasing ventilation, the effects of the mechanical distention of the peritoneum are more hemodynamically significant. While

invasive hemodynamic studies are scant in infants and children, Sakka et al. (20) reported a decrease in cardiac index (CI) of 13% by transesophageal echocardiography (TEE) at an insufflation pressure of 12 mmHg and no reversal of this effect at 6 mmHg in small, healthy children Guegniaud et al. (21) looked at the hemodynamic effects of pneumoperitoneum during laparoscopy in 12 ASA Class 1 infants by using noninvasive continuous esophageal aortic blood flow echo-Doppler. Insufflation to 10 mmHg caused the cardiac output to decrease approximately 30%, but MAP was unchanged. They noted a significant decline in aortic blood flow and increase in systemic vascular resistance (SVR) as in the adult studies. Several studies using adult patients support the fact that peritoneal insufflation has significant hemodynamic consequences. Joris et al. (22) studied 15 non-obese healthy adults undergoing laparoscopic choplecystectomy under general anesthesia using invasive hemodynamic monitoring via flow-directed pulmonary artery catheters. The study showed that insufflation pressures of 15 mmHg caused an increase in mean arterial pressure (MAP) of 35%, decrease in CI of 20%, increase in SVR of 65%, and an increase in pulmonary vascular resistance (PVR) of 90%. When combined with general anesthesia and the reverse Trendelenberg position of 10°, CI decreased 50%. Dorsay et al. (23) also studied 14 healthy adult patients undergoing laparoscopic cholecystectomy by transesophageal echocardiography and found that with insufflation pressures of 15 mmHg CI decreased 3%, heart rate (HR) increased 7%, MAP increased 16%, and stroke volume index (SVI) decreased 10%. The addition of 20° head-up position decreased CI by 11%, SVI by 22%, and increased HR and MAP by 914 and 19%, respectively. A third similar study in adults by Mclaughlin et al. (24) using TEE and invasive CVP and arterial blood pressure monitoring found an increase in MAP of 15.9%, increase in systolic blood pressure (SBP) of 11.3%, increase in diastolic blood pressure (DBP) of 19.7%, increase in CVP of 30%, decrease in stroke volume (SV) and CI of 29.5% following positioning in the reverse Trendelenberg position. All hemodynamic derangements were reversible following exsufflation (24).

Healthy infants and children easily tolerate these hemodynamic stressors because of their huge cardiac reserve. Issues of tachycardia and hypertension leading to myocardial ischemia and infarction are virtually nonexistent. Children with underlying cardiac disease (congenital or acquired), may not benefit from this some luxury. In particular, children with severe cyanotic heart disease and single ventricle physiology represent a great challenge. They are the patients most likely to benefit from MAS with its reduced postoperative respiratory compromise from smaller incisions and reduced narcotic medication perhaps leading to decreased postoperative pulmonary dysfunction (25). These fragile babies often require gastrostomy tubes and fundoplications in order to grow and may suffer from other congenital anomalies that necessitate surgical correction.

Small babies with hypoplastic left heart syndrome (HLHS) often fail to thrive following the first stage Norwood procedure. This palliative procedure uses the anatomic right ventricle as the systemic "workhorse" ventricle while using a Blalock–Taussig shunt in order to create passive pulmonary blood flow from the subclavian artery. The ratio of pulmonary to systemic blood flow (Q_p/Q_s) depends on the balance between SVR and PVR. These patients are routinely placed on afterload reduction medication in order to unburden this morphologically weaker ventricle. Absorption of CO_2 from the peritoneum may lead to a rise in arterial CO_2 leading to increased PVR, decreasing Q_p, and diminished oxygen saturation. Ventilation must be carefully adjusted to maintain end-tidal CO_2 in the normal range so as to prevent a respiratory acidosis in addition to the potential metabolic acidosis which frequently occurs in these patients. It has been shown that end-tidal CO_2 may not be a reliable indicator of arterial CO_2 in infants and children with cyanotic heart disease undergoing laparoscopic procedures. Large gradients are seen between

end-tidal CO_2 and arterial CO_2 in these patients. Although the cause of this gradient is unknown, possible causes include the absorption of CO_2 across the peritoneum, dead space ventilation caused by decreased functional residual capacity, alterations in pulmonary blood flow, or reduction in cardiac output caused by insufflation (26). The increase in afterload posed by intraperitoneal insufflation could in theory cause this ventricle to fail. Using the lowest possible insufflation pressures, judicious hydration and careful monitoring of blood pressure have made MAS possible for these patients. Fluid administration is particularly critical because the passive pulmonary circulation relies on systemic blood pressure to maintain oxygen saturation. Sluggish flow through synthetic systemic-to-pulmonary shunts may precipitate thrombosis and death. Because of the lack of reliability of end-tidal CO_2 monitoring and delicate fluid balance, invasive arterial catheters may be necessary in all but the most basic procedures for this patient population in order to avoid severe acidosis from inadequate ventilation or poor peripheral perfusion. Low-dose inotropic support with dopamine or dobutamine may be necessary. When acidosis cannot be corrected with IV volume administration, reduction of insufflation pressure, and an increased minute ventilation, conversion to open surgery should be considered.

3.2. Thoracoscopy

A vast array of surgery is currently being performed with thoracoscopy in neonates, infants, and children including newborn anomalies. While decortication for empyema and lung biopsies have been standard thoracoscopic cases for years, more and more complex operations can now be done with minimal access techniques. Tracheoesophageal fistulae and esophageal atresias, in addition to lobe resections, patent ductus arteriosus occlusion and anterior spine fusion are among the more recent repertoire of pediatric operations amenable to minimally invasive repair. With these technical advances, pediatric anesthesiologists have had to address the issues of lung separation for optimal surgical exposure in patients for whom no double-lumen endotracheal tubes exist. Alterations in patient positioning and the cardiopulmonary effects of gas insufflation and single-lung ventilation now must be considered when planning an anesthetic for these children.

There is perhaps no greater benefit to patients from MAS than is seen with thoracoscopy. The reduction in postoperative pain and splinting is an obvious advantage, particularly in the sickest patients who would have required large incisions for relatively minor surgical procedures. Open lung biopsies in severely ill or immunocompromised patients may necessitate postoperative intubation and mechanical ventilation in an intensive care setting. Smaller incisions via thoracoscopy allows many of these patients to be extubated immediately following surgery because of the reduced pulmonary dysfunction from reduced pain and depressant medication. Patients with severe, diffuse pulmonary disease used to be considered poor candidates for MAS because of the potential for bronchopleural air leaks following surgery, but advances in stapling devices have made it possible for these procedures to be done safely and effectively.

Thoracoscopy is performed with gas insufflation at low flow (1 L/min) and pressure (4–6 Torr). Lung collapse and the working space are created by first producing a pneumothorax via a Veress needle. A valved trocar is then introduced. In small patients, where lung separation is the most difficult, gas flows of 1 L/min at a pressure of 4–6 Torr allow the lung to be mechanically pushed away allowing greater exposure. The hemodynamic consequences of gas insufflation at $\geq$5 mmHg with selective lung intubation in adult swine has shown a decrease in CI, MAP, and left ventricular stroke work index while pulmonary artery and CVP increased (27). This technique, however, is well tolerated even by small infants undergoing PDA ligation at flows of 1 L/min at a pressure

of 4 mmHg (10). The physiologic impact of one-lung ventilation can be profound, particularly in the smallest and sickest patients. While ventilation to perfusion ratios (V/Q) are well maintained in the supine and lateral position in larger, awake children, small children are prone to atelectasis. The hydrostatic gradient caused by gravity is reduced and the chest wall is more compliant. Therefore, functional residual capacity is closer to residual volume and closing volume. Atelectasis leads to compromised ventilation and V/Q mismatch. General anesthesia and mechanical ventilation cause further V/Q mismatch. Hypoxic pulmonary vasoconstriction, a mechanism whereby blood is diverted away from nonventilated lung, is blunted by many anesthetics, including inhaled, volatile anesthetics. All of these factors operate in concert to increase shunt fraction and reduce arterial oxygen saturation. Allowing some low-pressure ventilation to the operative, nondependent lung may help to improve oxygen saturation while low flow, low pressure CO_2 insufflation maintains the working space. Neonates <4 kg often do not tolerate single-lung isolation. Despite this, adequate lung collapse is achieved with a CO_2 pressure of 4 Torr with the ET positioned in the trachea. This revelation is what has allowed the repair of tracheoesophageal fistulae and esophageal atresias by thoracoscopy in newborns.

3.2.1. Techniques for Single-Lung Ventilation

Various parameters of the techniques used for single-lung ventilation are summarized in Table 3.1.

Double-Lumen Endotracheal Tubes. Double-lumen tubes have been the mainstay of single-lung ventilation in adults undergoing thoracoscopy. They utilize two coaxial, cuffed tubes, the proximal one in the trachea and the distal into either the right or left mainstem bronchus. This is ideal because it can provide complete lung separation while allowing either single- or double-lung ventilation whenever appropriate. Position can be confirmed by use of a fiberoptic bronchoscope (FOB) at any point during surgery. The nondependent operative side can be suctioned and continuous positive airway pressure can be administered when needed to improve declining oxygen saturation. The smallest available double-lumen tube, however, is a 26-French and is, therefore, appropriate for children 8–10 years old ($\sim$30–40 kg).

Table 3.1 Age, Airway Dimensions, and Device Sizes

Age (year)	Approximate trachea (mm)	Endotracheal tube ID (OD) (mm)	Fiberoptic bronchoscope OD (mm)	Balloon catheter French (mm)	Univent ID (mm)	Double-lumen tube French OD
<0.5	5	3.0–3.5 (4.3–4.9)	Up to 2.4			
0.5–1	5.5	3.5–4.0 (4.9–5.5)	Up to 3.1			
1–2	6	4.0–4.5 (5.5–6.2)	Up to 3.4	$\pm$5 (1.67)		
2–4	7.5	4.5–5.0 (6.2–6.8)	Up to 3.4	5 (5 Arndt)		
4–6	8.0	5.0–5.5 (6.8–7.5)	Up to 4.2	5		
6–8	9.0	5.5–6.0 (7.5–8.2)	Up to 4.2	5 or 6	3.5	
8–10	10.0	5.5–6.0 cuffed	Up to 5.2	5 or 6	3.5–4.5	26
10–12	10.5	6.0–6.5 cuffed	Same	6	4.5	26–28
12–14	11.5	6.0–7.0 cuffed	Same	6 (9 Arndt)	4.5, 6.0	32
14–16	13.0	6.5–7.0 cuffed	Same	Same	6.0–6.5	35
16–18	13.5	7.0–7.5 cuffed	Same	Same	7.0	35–37

Note: ID, internal diameter; OD, outer diameter.

Single-Lumen Endotracheal Tubes with Selective Mainstem Intubation. For smaller children and neonates, an assortment of techniques is available to attempt to separate ventilation. Nearly, all are fraught with significant failure rates meaning they cannot be effectively placed or they do not prevent spillover ventilation from occurring despite optimal positioning. The simplest technique is the use of a single-lumen tube advanced into the contralateral mainstem bronchus until breath sounds disappear on the ipsilateral side, taking care to avoid occluding the takeoff of the upper lobe bronchus. Left mainstem intubation presents a challenge, particularly in neonates. Turning the head to the right may help the ET advance into the left mainstem bronchus by directing the tip to the left. Rotating the ET 90° to the left or 180° (where the bevel faces right and points left) may be effective. Placing the child in the right lateral decubitus position may help to shift the mediastinum to the right and partially compress the right mainstem bronchus, preferentially allowing passage into the left. Use of a FOB as a guiding stylet is useful. However, in neonates, the small ET necessitates the use of a small, floppy 2.2 mm FOB which may not be stiff enough to help guide a tube into the left mainstem. Tube position may be confirmed with the use of a FOB either within the lumen of the ET or alongside it. This technique requires minimal excess equipment, but may fail to achieve an adequate seal of the mainstem bronchus, especially if size limitations preclude the use of a cuffed ET. Care must be taken to make sure the entire cuff is below the takeoff of the mainstem bronchus so that ventilation does not spill over to the operative lung.

Balloon Occlusion Bronchial Blockers. Balloon-tipped bronchial blockers including Fogarty embolectomy catheters and end-hole, balloon wedge catheters (Arrow International Corp., Redding, PA), and the Arndt Endobronchial Blocker (Cook Critical Care, Bloomington, IN) may be used to seal the bronchus on the operative side. The catheter may be placed into the trachea under direct vision with laryngoscopy. Following this the trachea is intubated with an appropriate sized ET. Using a FOB via a swivel adapter in the ET, the catheter is manipulated into the operative mainstem bronchus. Care is taken to assure that the ET remains above the carina. Conversely, the bronchus may be intubated first with an ET and then a guidewire is passed through and the ET withdrawn. The open-ended balloon-tipped catheter is then fed over the guidewire into the bronchus. The trachea is then intubated alongside the balloon catheter. Fiberoptic bronchoscopy is used to confirm placement of the balloon below the takeoff of the mainstem bronchus and position of the ET above the carina. The Arndt Endobronchial Blocker (Cook Critical Care) has a guide loop at the end of the blocker's balloon that can be placed under FOB guidance. The loop is secured to the FOB and introduced through an ET with the use of the Arndt Multiport Airway Adaptor, which permits uninterrupted ventilation during placement. The 5 French pediatric blocker can be used with the smallest FOB through a 4.5–5.0 internal diameter ET. A small lumen allows CPAP to be delivered, if needed and the blocker may be withdrawn without the need for reintubation should be patient require postoperative ventilation.

Regardless of the device used, placement is confirmed by inflation of the balloon and the loss of breath sounds on the operative side. The balloon should remain deflated until after insufflation of the chest so that the lung is able to collapse. The catheter is then firmly secured to the ET and face to prevent dislodgement.

Balloon-tipped catheters have the advantage of reliable occlusion of the operative side. They are, however, equipped with low-volume, high-pressure balloons and may cause trauma to the airway mucosa if overinflated. Inadvertent displacement of the balloon into the trachea upon insufflation will result in the inability to ventilate the patient. Also, the more readily available closed-tip, Fogarty-type, catheters do not allow suctioning of the lung or delivery of CPAP if oxygenation declines during surgery.

Univent Tubes. A Univent tube (Fuji Systems Corporations, Tokyo, Japan) is a conventional ET with a second lumen that contains a balloon-tipped, small tube that may be advanced into a mainstem bronchus via FOB guidance. These tubes are available as small as 3.5 mm internal diameter (ID) (outer diameter 7.5–8 mm, equivalent to approximately 5.5 ID conventional ET). Therefore, these tubes are useful for school-age children, but are not suitable for infants and toddlers.

Because the Univent tube employs an integral bronchial blocker, dislodgement of the blocker may be less likely. It also has a small lumen that can be used to provide an outlet for gas, delivery of oxygen, or the application of CPAP.

4. ANESTHETIC IMPLICATIONS OF INTRAOPERATIVE COMPLICATIONS

As with any modality, complications occur with MAS. Early in the evolution of MAS, operative times were long mostly because of the awkward nature of suturing with two-dimensional visualization. Technical difficulties often led to the conversion to open surgery. Prolonging surgery leads to increased fluid requirements, problems with temperature maintenance, atelectasis, and potential for delayed emergence from anesthesia. Experience now shows that over time, MAS has proved to be efficient and often superior to conventional open surgery. Remote reaches of the abdomen and thorax are accessed with improved visibility. Technical advances and positioning have made suturing and other two-handed procedures appear effortless. Still, complications unique to MAS remain and require vigilance to recognize and correct.

Because gas insufflation is continuous during MAS, it may dissect into tissue planes including across the diaphragm and into the mediastinum. Pneumomediastinum and pneumothorax can occur from breaches in the diaphragm caused by the heated tip of a cautery device and may go unrecognized until respiratory or cardiovascular compromise becomes apparent. Prompt discontinuation of insufflation and evacuation of gas from these spaces returns cardiopulmonary function to normal. Carbon dioxide may also cause subcutaneous emphysema when it tracks during malplacement of trocars or via the mediastinum. When it becomes severe in the neck, subcutaneous emphysema may impede respiration. It may be necessary to leave patients intubated to maintain airway patency until some of the emphysema resolves. Patients who require postoperative positive-pressure ventilation should have chest tubes left in place following thoracoscopy to avoid a pneumothorax.

Though the conversion rate to open surgery is declining over time, the complexity of cases dictates that occasionally hemorrhage or technical difficulties will arise. Converting to open surgery may necessitate changing the position of a patient while maintaining the sterile field. Vigilance regarding IVs and airway devices is of paramount importance. Confirming adequate ventilation and IV patency after repositioning is vital. Consideration should be given to epidural placement at the conclusion of surgery.

MAS has become the gold standard for many operations, once thought too complex for this innovative approach. It has become clear that over time the vast majority of pediatric surgery will be done with some form of MAS. Robotic surgery is the most recent rendition, allowing surgeons greater freedom of movement plus all the advantages of MAS. Pediatric anesthesiologists have had to adapt to provide safe operating conditions in an environment that fosters the success of novel techniques. Perhaps even more than for open surgery, communication between members of the team is critical. Difficulties arise during surgery (e.g., cephalad displacement of ETs) upon insufflation need to be addressed quickly. Often, small decrements in insufflation pressure or repositioning of ETs make

management of small patients vastly easier. Minor ventilatory changes may help to improve visualization by the surgeon by reducing inflation of the operative lung.

Monitors allow the anesthesiologist a view into the operative field that was unavailable before and provides valuable information regarding possible causes for cardiopulmonary changes. Prompt intervention can help prevent significant desaturation or hemodynamic decompensation. Communication is most important perhaps when significant bleeding occurs or the decision is made to convert to an open procedure.

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4
Minimal Access Neonatal Surgery

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1. INTRODUCTION

The emancipation of neonatal surgery is of relatively recent date. It began roughly after the First World War when William Ladd started his work at the Boston Children's Hospital. It only became established in most countries since Second World War. Rickham in 1969 attributed the rapid after Second World War development of neonatal surgery to four main factors:

1. Concentration of neonates with a surgical condition in centers draining large areas
2. Improvement in anesthesia and management of cardiorespiratory complications

3. Pre- and postoperative care
4. Improvement in surgical technique.

He regarded the last factor as the least important (1).

With increasing safety of anesthesia in neonates, coupled with the improvement in postoperative facilities, the length of the operation has become less important, allowing for complex operations to be performed. It appears that surgical technique is a major factor in the progress of neonatal surgery. Good exposure has always been and still is one of the fundamentals of surgery. In the era before muscle relaxation, this had to be achieved by large incisions and strong retractors. It seems likely that this has been one of the factors why Dennis Brown advocated transverse abdominal incisions in which both rectus muscles were severed and why he placed so much emphasis on retractors (2,3). Thanks to muscle relaxation, the access to body cavities can be much smaller without jeopardizing exposure.

Pediatric surgeons have always sought for less invasive ways of dealing with surgical conditions, for example, the transumbilical route for pyloromyotomy and the muscle sparing thoracotomy for esophageal atresia (4,5). Nevertheless, many incisions in the newborn today are still quite extensive, for example, the supraumbical transverse laparotomy, which gives a tremendous exposure yet at the cost of extensive skin, fascia, muscle, and nerve cutting. Moreover, such a laparotomy exposes most of the bowel and of peritoneal cavity to the surrounding air, promoting evaporation, and as consequence hypothermia and drying out of the tissues. Moreover, the contamination of the peritoneal cavity with air seems to be a potent stimulator of the stress response (6,7). Last but not least, the covering of the viscera and peritoneum after such "excellent" exposure with gauzes in order prevent dehydration and direct trauma may harm the covered structures as a result of tissue foreign body reaction and repetitive trauma due to friction. This is a chain of events believed to lead to adhesion formation and an ileus.

In adults, transient cellular and humoral immunosuppression after a different degree of operative stress has been well documented (8,9). Such immunological studies in the neonate are largely absent. In 1-week-old rats, however, immunosuppression up to 7 days depending on the degree of invasiveness of the procedure was demonstrated (10).

Incisions and the subsequent scars are permanent and do grow proportionally with the child. Moreover, as the newborn and especially the premature newborn lacks subcutaneous fat, the skin scar may become adherent to the fascia giving a poor cosmetic result.

It leaves little doubt that neonates with a surgical condition have had less than optimal care for many years as it was thought that neonates experience less pain. A great step forward in the care of neonates in general but of surgical neonates, in particular, is the increasing awareness that the newborn even the premature can mount a considerable endocrine and metabolic response to surgery and that neonates experience pain (11,12). The hormonal and metabolic responses of the neonate to surgery are directly proportional to the degree of surgical trauma (13). More and more evidence is accumulating that early, especially repetitive painful stimuli have a negative effect on behavioral development and decrease the pain threshold level (14).

Although pediatric surgeons were at the forefront of diagnostic thoracoscopy and laparoscopy, they were rather skeptical when endoscopic surgery boomed for adult patients at the end of the 1980s. Since then, however, minimal access surgery in children has quickly evolved. Newborn minimal access surgery has progressed, albeit more slowly, is not surprising in view of the small body cavities, the relatively large endoscopic instruments and the relative rarity of many conditions. There are many theoretical advantages of

using minimal access techniques in neonates, yet hard evidence that these procedures are superior when compared with procedures using a classic approach are largely lacking, except for the cosmetic benefits. The success of endoscopic surgical techniques is often expressed in terms of postoperative hospital stay or postoperative time to full tolerance of oral feeding. Such endpoints, however, can only be used when they have been exactly defined in advance. Few studies have looked at factors such as serum interleukins. Fujimoto et al. (15,16) found lower IL-6 responses in laparoscopically operated children. Iwanakawa et al. (17) did not found significant differences, but the group of patients, they studied, was very heterogeneous. Bozkurt et al. (18) studying older children undergoing emergency abdominal surgery also found no difference, but this may have been caused by the magnitude of the underlying pathology.

There are three body cavities in the neonate that are regularly approached endoscopically namely the ventricles of the brain, the chest, and the abdomen. Endoscopic techniques have revolutionized pediatric neurosurgery and especially the treatment of hydrocephalus (19,20). We will not elaborate further on this subject. The focus of this chapter is to assess the development and outcomes of neonatal laparoscopic and thoracoscopic procedures.

2. THORACOSCOPIC AND LAPAROSCOPIC INTERVENTIONS

2.1. Indications

2.1.1. Thoracoscopic Interventions

These can be subdivided into diagnostic and therapeutic procedures.

Potential indications are

- Great vessels
  - Interruption of a patent ductus arteriosus (21,22)
  - Division of vascular rings
  - Aortopexy
- Pericardium, for example, cyst excision
- Thoracic duct ligation
- Lungs
  - removal of a bronchogenic cyst
  - lobectomy for lobar emphysema and cystic adenomatoid malformation (23,24)
  - resection of a pulmonary sequestration (25)
- Esophageal pathology, for example, atresia, duplicate cyst (26,27)
- Diaphragmatic repair for eventration or hernia.

The most common thoracoscopic operation that has been performed in neonates, and even in prematures, is the interruption of a patent ductus arteriosus (21,28). Laborde, a pioneer in the field, published, already in 1995, a series of 300 pediatric patients. Schier and Waldschmidt (29) described their experience with thoracoscopy in children. Of the 22 children, three were neonates and two additional children were younger than 6 months. One neonate had a bronchogenic cyst. The remaining children had a diagnostic thoracoscopy. In 1997, Rothenberg and Chang (30) described their experience with endoscopic surgery in neonates and infants; six had PDA occlusion and five lung biopsy. At the Children's Hospital in Utrecht, 10 neonates have been thoracoscoped so far by the Department of Pediatric Surgery. Nine had esophageal atresia with distal fistula (31). Lung

and heart surgery at the Children's Hospital is done by pediatric cardio-pulmonary surgeons.

2.1.2. Laparoscopic Interventions

At the University Children's Hospital in Utrecht, the endoscopic surgical program started in 1992. Until August 2001, 1036 endoscopic surgical operations have been performed. Of these 112 were in neonates, which is 10.8% (Table 4.1). Roughly, half of the children had hypertrophic pyloric stenosis. The remaining children had a variety of pathology. If not only neonates, but also all children below the age of 6 months are counted then 303 children have had endoscopic surgery, which is 29.2% of the total population (Table 4.2). Again about half of these children had hypertrophic pyloric stenosis.

An increasing number of neonates have undergone minimal access surgery in the face of associated cardiac anomalies (32). In this series, 22 had nonduct-dependent lesions and underwent a variety of thoracoscopic and laparoscopic interventions using insufflation pressures of 6–8 Torr. There were no perioperative adverse events referable to the surgery or anesthetic technique.

The indications are shown in the following Tables 4.1 and 4.2.

In this series, there were only few laparoscopies for inguinal hernia. The indication for laparoscopy in these patients was when there was doubt about the diagnosis of a clinical hernia or in the case of a recurrence. Before the era of minimal access surgery, it has been common practice in North America to explore the other side in unilateral clinical

Table 4.1 Endoscopic Surgical Procedures Carried out in Neonates

Hypertrophic pyloric stenosis	46
Malrotation	10
Esophageal atresia	9
Ovarian cyst/torsion	8
Duodenal atresia	8
Anorectal anomaly	3
Diaphragmatic hernia	5
Hirschsprung's disease	3
Duplication cyst of the ileum	2
Anorectal malformation	2
Obstruction	3
Intersex	2
Sacrococcygeal teratoma	2
Gastrostomy	1
Jenunostomy	1
Meckel	1
NEC	1
Antireflux surgery	1
Jejunal atresia	1
Perforation NEC	1
Testicular teratoma	1
Jaundice	1
Inguinal hernia	1
Total	112

Table 4.2 Endoscopic Surgical Procedures Carried out in 1- to 6-month-old Children

Hypertrophic pyloric stenosis	107
Hirschsprung's disease	22
Jaundice	20
Antireflux surgery	9
Malrotation	8
Inguinal hernia	6
Diaphragmatic hernia	5
Ovarian cyst/torsion	3
Duplication	2
Intersex	2
Duodenal stenosis	1
Obstruction	1
Rectal prolaps	1
Intussusception	1
Recurrent pneumothorax	1
Anorectal stenosis	1
Cholelithiasis	1
CAPD catheter	1
Total	192

hernia. By laparoscopy of the contralateral internal inguinal ring, the incidence of negative explorations will undoubtedly drop.

3. UNIQUE TECHNICAL ASPECTS OF NEONATAL THORACOSCOPIC AND LAPAROSCOPIC SURGERY

3.1. Patient Positioning

Owing to the small size of neonates, they can be placed transversally or at the end of the operating table, allowing for a perfect in line position of the surgeon, operative field and monitor, which is ergonomically better.

3.2. Limited Working Space

3.2.1. Working Space in Thoracoscopic Surgery

A particular problem in thoracoscopic surgery in the neonate is the creation of an adequate working space. Single lung ventilation would be ideal but this is hard to achieve in small children. Main stem intubation of the contralateral lung is an alternative but is seldom selective enough. Bronchial blocking of the ipsilateral main bronchus with a balloon catheter is often mentioned as alternative but there are very few publications on its actual use. There are no publications on the effects of one lung ventilation in the newborn. In an experimental study in neonatal pigs, it was concluded that single lung ventilation was well tolerated (33). Three out of the eight animals, however, were hemodynamically very unstable and were excluded from analysis. In the remaining animals, the arterial blood pressure dropped in a statistically significant way. Moreover, the ventilation rate had to be increased by 25% in order to keep $PaCO_2$ within normal limits.

The single most common indication for thoracoscopic surgery in the neonate has been open ductus arteriosus. In most of these publications, no selective intubation was used. Instead, the lung was retracted with an additional instrument. An alternative to retraction is the use of CO_2 insufflation. Insufflation pressures of 4 up to 8 mmHg have been used. Flow should be low, for example, 100 mL min. Apparently, such a CO_2 pneumothorax is well tolerated and there are no reports on adverse effects. It is the author's experience that the ventilator settings have to be adjusted in order to create a new equilibrium. If an increased frequency is not enough, pressure should be increased. Sometimes, it is advantageous to manually assist the ventilation until a stable situation is reached. It will take $\sim$5 min before the lung on the ipsilateral side collapses, so the surgeon should be patient. Major CO_2 leaks should be prevented because of cooling down and drying out of the tissues. Alternatively, preheated and moistened CO_2 should be used. Apart from retraction or CO_2 insufflation, advantage should be taken of gravitational forces, for example, for posterior mediastinal structures to be approached a prone position is advantageous as the lung falls out of the way. It has been suggested that the prolonged use of a telescope in a small working like the chest in the neonate can produce hyperthermia, because of the energy released through the telescope by the light source (34).

3.2.2. Working Space in Laparoscopic Surgery

As in the chest, the limited working space in the abdomen in the neonate is a major problem. There are two ways to enlarge the working space: CO_2 insufflation and abdominal wall lifting.

As is well established, CO_2 insufflation has a number of disadvantages. There is the increased intraabdominal pressure and the use of CO_2 both having local and systemic consequences. Studies on the consequences of the CO_2 peritoneum in the neonate are largely lacking. In 14- to 19-day-old piglets, Graham et al. (35) showed that CO_2 insufflation at a pressure of 15 mmHg increased $PaCO_2$ by 31%, cardiac index by 10%, central venous pressure by 29%, and blood pressure by 17%. There was no increase in systemic vascular resistance or in inferior vena cava flow. If the increased $PaCO_2$ was controlled by increased ventilation, there was significant change in cardiac index, but blood pressure and systemic vascular resistance increased by 7%, whereas pressure in the inferior caval vein increased by 57%. In contrast, inferior vena cava flow decreased by 22%. Substitution of CO_2 with N_2O resulted in an unchanged cardiac index, but in an increase of blood pressure by 16%, of systemic vascular resistance by 22%, of central venous pressure of 35%, and in a decrease in inferior vena cava flow of 25%.

It has been shown that there is a direct transmission of the increased intraabdominal pressure onto the ventricle system of the brain (36). Recently, an increase in flow in the basilar artery in rabbits has been documented (37). It seems logical to assume that the same occurs in neonates but this remains to be proven. Whether such changes would be clinically relevant is another question to be answered.

The effects of the pneumoperitoneum on local hemodynamics in the abdomen in neonate have also not been properly studied. As blood pressure in neonates is proportionally lower when compared with older children, more profound effects on regional perfusion can be expected. During long lasting procedures, urine production is almost absent. Routine insertion of a urine catheter, therefore, makes no much sense. For the same reason, the administration of intravenous fluids should not be pushed for the sake of the diuresis alone as this will not lead to an increased diuresis but to overhydration, edema, and hemodilution.

Even when CO_2 is insufflated, the working space is limited. The abdominal cavity in a neonate can only contain about 300 mL of CO_2 at a pressure of 8 mmHg. Abdominal wall lifting using either an intraabdominal device or a subcutaneous wire has been described but has not gained wide acceptance (38,39). In abdominal wall lifting, ambient air has to enter the abdominal cavity. Contact of air with the peritoneum and viscera seems to be a potent stimulator of the immune system (6,7). Moreover, an abdominal wall lifting does not create a nice dome shaped space, often low pressure CO_2 insufflation is added.

3.3.3. Secondary Factors Influencing Working Space

1. *Optimal muscle relaxation*: Although there is no scientific evidence that muscle relaxation increases the working space, it is logical to assume that such a relationship exists.
2. *Cannula length inside the working space*: The less cannula length sticks inside the cavity, the more working space will be available. There is, however, a relationship between this length and the chance of the cannula being pulled out. Secure fixation of the cannula rather to the fascia than to the skin is, therefore, of paramount importance. The use of cannulae with beveled end should be avoided, as the beveled end has to be entirely in the body cavity thereby decreasing the working space.
3. *Length of the active tip of the instrument*: Manufacturers of endoscopic instruments have sized down the diameter of the instruments, mainly for use in adults. As a result, the length of the instruments and especially of their active ends has remained long. Such long active ends decrease the available working space. Moreover, as these long metal ends are not insulated, collateral damage when energy is applied can easily occur.
4. *Pathology-specific considerations*: For example, when the diaphragm has to be plicated for paralysis, a thoracoscopic approach will be difficult because of the high position of the affected hemidiaphragm. In congenital diaphragmatic hernia, the abdomen is smaller than normal. Moreover, withdrawal of the abdominal viscera from the chest into the abdomen will decrease the working space.

3.3. Cannula Position

It is common practice to insert the telescope and working cannulae relatively more remote from the target organ, the smaller the child is. Owing to the limited dimensions of the abdominal cavity and the spherical shape after insufflation, the more distally the telescope and instruments are inserted, the smaller the optical axis to target view, and the smaller the elevation angle will be. Moreover, such low positions result in small manipulation angles. It has been shown that optimal task performance is achieved when the optical axis to target view is 90°, when the elevation angle is $\sim 30^\circ$ and the manipulation angle 60° (40).

3.4. Cannula Fixation

The thickness of the wall of the thorax and abdomen in the neonate is so small that the wall does grip well enough onto the cannula. Moreover, when CO_2 insufflation is used, the cannulae have to be valved, which causes additional friction of the instruments inside the cannulae. As a result cannulae in the neonates are easily pushed in or pulled out.

Pushing in of the cannulae may not only harm the viscera but limits the small working space even further. Special fixation of the cannulae in the neonate is therefore essential. There are several ways of doing this. Georgeson (41) uses 1–2 cm long rubber sleeves cut from Red Robinson catheters. The sleeve is pulled over the cannula and fixed to the skin. This is a good way of fixing the cannulae. Moreover, it allows for adjustment of the inside length of the cannula by gliding the cannula inside the sleeve. However, an other material than rubber would be better in view of possible latex allergy.

Another way of fixing the cannulae is to suture the stopcock to the abdominal wall and to put a piece of tape, for example, a large SteriStrip® around the cannula and suture at skin level (42). In the neonate, it is advisable to put the suture not only through the skin but also through the underlying fascia in order to prevent that traction is applied on the loose skin only allowing the short inside part of the cannula to be pulled out of the working space.

There are expandable sleeve trocars on the market (Step®, US Surgical). The sleeve is introduced over a Veress needle through a stapwound into the cavity to be entered. The Veress is then removed and a cannula with blunt trocar introduced. The sleeve is thus radially dilated. The general experience with this kind of cannula in children has been good and only few complications have been reported (43). In a series of 2157 insertions, slippage occurred in 0.88%. Mean age of the children, however, was 7.2 years and mean weight 28.4 kg. The exact number of neonates in the series is not given but underrepresentation is likely.

A disadvantage of these cannulae is the poor relationship between outside diameter of the cannula with sleeve and internal diameter of the cannula, for example, a cannula with sleeve for 2–3 mm diameter instruments has an outside diameter of 7.2 mm. Moreover, the resistance inside the valve is rather high so that the cannula with sleeve is likely to dislodge in neonates unless additional fixation measures have been taken.

3.5. First Cannula Insertion

The safety of first cannula insertion is more a function of training and personal experience rather than the technique itself. An open technique is often advocated. On the other hand, there are data to support the use of the Veress technique, as it has been shown to be safe for many thousands of patients (43). Whether the umbilical region should not be used for the insertion of the first cannula because of the vicinity of the urachal remnant and the umbilical vessels, as stated by Waldschmidt and Schier (44) is debatable. The authors have had no problems with an open approach through the inferior infraumbilical fold. We pick up the inferior umbilical fold with a surgical forceps and the skin in the midline immediately below. With a curved pair of scissors, we make a small smile like incision in the picked up skin. Next, the fascia is freed and a very small transverse incision using electrocautery is made. As soon as the fascia is opened, a Mosquito type forceps is pouched into the abdomen with the beak of the forceps to the left of the urachal remnant and tangential to the abdominal wall. The pushing of the Mosquito should be done with a firm quick movement in order to avoid pushing off of the peritoneum. The point of the beak of the Mosquito should be freely movable inside the cavity. Only a small hole is made, so that the cannula will fit snugly. The cannula is inserted with a blunt trocar. Before starting insufflation, one should make sure that the cannula has pierced the peritoneum. One can argue that this is not a complete open insertion.

3.6. Insertion of Secondary Cannulae

The optimal place for the secondary cannulae is determined with the telescope and external landmarks (e.g., costal margin). The authors pierce the body wall with a pointed blade and insert then a cannula with a blunt trocar. Again the hole is kept to strict minimum place

so that the cannula fits snugly. Many pediatric surgeons experienced in the field, sometimes insert instruments directly through the wall into the cavity to be operated, which has the advantage that the hole in the body wall is smaller. This is only advantageous, however, when the particular instrument has not to be changed often.

3.7. Instruments

There are instruments with varying diameters and lengths. Instruments with a smaller diameter need smaller holes in the body wall and are less invasive from that point of view. On the other hand, smaller diameter instruments grasp less well and the application of more force may easily damage the tissues. Moreover, as such instruments are less blunt than larger diameter instruments, they may accidentally pierce organs, for example, the bowel or liver.

As far as length is concerned, it has been stated that the optimal ratio between the inside and outside parts of an instrument should be 2 to 1 (40). This ratio is hard to achieve in the neonate. The ratio will be at best inverse as otherwise the handles will clash. Long instruments have the disadvantage that they will have to be operated with the upper arms of the surgeon in abduction which is very tiring. The authors have started to use 20 cm long instruments in the neonate and feel more comfortable with them. A problem with the short and smaller diameter instruments is that the available variety of these instruments is smaller than the variety of the longer and thicker instruments, for example, clipping and stapling instruments, energy applying tools such as ultrasonic graspers. As said before, despite the miniaturization of instruments in terms of diameter and recently in terms of length, the metal end of most short and small diameter instruments has remained rather long. As a result, it is difficult to keep to whole metal end in view when working in a limited space, which predisposes for collateral damage when monopolar high frequency electrocautery is used.

4. CONCLUSIONS

For years, it was erroneously believed that surgery in neonates was associated with less pain compared with older children and that they were unable to mount a good response to stress. There is concern, now a day, about the long-term consequences of pain in the neonatal period on further brain development. Pain should not only be treated adequately but also be prevented as much as possible. Theoretically, minimal access techniques should be associated with less perioperative stress than open surgery. After a slow start of endoscopic surgery in children in general but in the neonate in particular, most operations that have been performed in an open way can now be performed endoscopically. The cosmetic results are undoubtedly superior but are the so-called minimally invasive procedures really minimal invasive? As long as this question is insufficiently answered through research the term minimally invasive should not be used. Pediatric endoscopic surgeons should be careful in thinking that what they do is really better. A continuous critical evaluation of endoscopic surgery in general, but in the neonate in particular, is the best guarantee that this type of surgery will progress along the least invasive ways.

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5
Clinical Outcomes in Minimal Access Fetal Surgery

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The rapid advances over the last 20 years in prenatal imaging and diagnosis, coupled with an increased understanding of the pathogenesis of neonatal disease, has led to the identification of the fetus as a patient and to the burgeoning field of fetal surgery. An increasing number of select fetal anomalies are currently amenable to prenatal intervention (Table 5.1). Life-threatening congenital anomalies have been historically treated by *open* fetal surgical techniques. Yet, a variety of significant complications including pre-term labor (PTL), premature rupture of membranes (PROM), pre-term delivery and maternal complications from the tocolytic therapy have lead surgeons to investigate innovative approaches to minimize these complications. In order to reduce maternal morbidity related to the hysterotomy and fetal morbidity due to exposure and manipulation, minimal

Table 5.1 Summary of Applications of Fetal Surgery

Defect	Effect on development			Open	Minimal access
LETHAL					
Placental vascular anomalies					
Twin–twin transfusion syndrome (TTTS)	Vascular steal through placenta	→	Fetal hydrops/demise Surviving twin with severe morbidity	Fetectomy	Photocoagulation of chorangiopagus
Twin reversed arterial perfusion syndrome (TRAP)	Normal cotwin hear pumps for both twins	→	High output cardiac failure, hydrops	Fetectomy	Selective reduction via umbilical cord ligation or radiofrequency needle
Obstructive uropathy	Hydronephrosis Lung Hypoplasia	→	Renal failure Pulmonary failure	Vesicostomy	Vesicoamniotic shunt Valve ablation
Congenital diaphragmatic hernia (CDH)	Lung hypoplasia	→	Pulmonary failure	Complete repair Temp tracheal occlusion	Temporary tracheal occlusion (PLUG)
Cystic adenomatoid malformation/ sequestration	Lung hypoplasia or hydrops	→	Respirator insufficiency Fetal hydrops/demise	Pulmonary lobectomy	Radiofrequency ablation
Sacrococcygeal teratoma	High-output heart failure	→	Fetal hydrops/demise	Debulk Complete resection	Laser vascular occlusion Radiofrequency ablation
Complete heart block	Low output failure	→	Fetal hydrops/demise	Pacemaker	Pacemaker
Pulmonary/aortic stenosis	Ventricular hypertrophy	→	Heart failure Single ventricle physiology	Valvuloploasty	Catheter valvuloplasty
Pericardial Teratoma	Heart failure	→	Fetal hydrops/demise	Resection	—
Ebstein's anomaly	Heart failure Pulmonary hypoplasia	→	Fetal hydrops/demise Pulmonary failure	Valve repair and atrial reduction	—

Congenital high airway obstruction syndrome	Overdistention by lung fluid	→	Fetal hydrops/demise	Tracheostomy EXIT strategy	Tracheostomy
Obstructive hydrocephalus	Hydrocephalus	→	Brain damage	Ventriculoamniotic shunt Ventriculoperitoneal shunt[a]	Ventriculoamniotic shunt
NONLETHAL					
Myleomeningocele	Chiari formation Exposed spinal cord Hydrocephalus	→	Paralysis Neurogenic bladder/ bowel Orthopedic anomalies	Repair	Repair
Tension Hydrothorax	Lung hypoplasia	→	Respiratory failure	—	Serial thoracenteses Thoracoamniotic shunt
Previable premature rupture of membranes (PROM)	PTL	→	Fetal demise Fetal/Maternal infection	—	Amniopatch Amniograft
Gastroschisis	Bowel exteriorization	→	Bowel preivisceritis Prolonged ileus	—	Amnioexchange
Amniotic bands	Limb/digit/umbilical cord constriction	→	Limb/digit deformity or amputation Fetal demise (cord occlusion)	—	Laser separation of bands
OTHER				—	
Stem cell/enzyme defects	Hemoglobinopathy Immuno-deficiency Storage diseases	→	Anemia Infection Neurological impairment	—	Stem cell transplants Gene therapy

Note: Refs. (228, 229).

access surgical techniques have been adapted to the fetal environs. Minimal access fetal surgery (MAFS) may allow for a broader applicability of fetal intervention, and extension of treatment to nonlethal and highly morbid fetal maladies.

Experimental animal research suggests that MAFS techniques may improve the outcome measures of uterine contractions and PROM. The data on PTL, however, remains equivocal. In 1995, Van der Wildt et al. (1) studied the uterine contractions of five mid-trimester Rhesus monkeys after fetoscopic access. Twenty-four hours after access, no uterine contractions could be measured. Of the three monkeys who did not die within 1 week of surgery, no premature uterine contractions were observed in the third trimester. On the other hand, in 1996, Luks et al. (2) studied 10 third trimester sheep and showed no difference in quality or quantity of uterine activity between control, endoscopic access, and hysterotomy. These differences between the two studies may, in part, be attributed to the behavioral differences between the primate uterus and the sheep uterus. The sheep uterus is thin and tolerant of injury, whereas the primate uterus is thicker and more unforgiving. The study of Luks et al. did show a decrease in uterine artery blood flow and uteroplacental oxygen delivery by 73% compared with control in sheep undergoing hysterotomy vs. *no* decrease after endoscopy. This can result in a decreased fetal pH, an increased serum lactate, and a redistribution of fetal blood flow (3,4). Thus, fetal homeostasis may be more stable with fetoscopy. Human clinical experience with fetoscopic intervention for congental diaphragmatic hernia has shown promise with decreased PTL, a decreased use of tocolytics and subsequent reduction in maternal complications and maternal hospital stay (5,6). A review of the evolution in technique for the fetal treatment of congenital diaphragmatic hernia (CDH) is illustrative of the overall rationale of MAFS toward ameliorating the pathophysiology of major congenital malformations.

1. CONGENITAL DIAPHRAGMATIC HERNIA

CDH is a condition that develops when there is an abnormal fusion of the four structures of the diaphragm: the septum transversum, the pleuroperitoneal membranes, dorsal mesentery of the esophagus, and the body wall (7). Abdominal viscera are able to herniate through the defect into the thoracic cavity. If the defect is large or occurs early, a large volume of viscera may herniate, and anatomic compression of the developing lung bud can occur. Compression of the lungs can stunt pulmonary development and possibly displace the heart and vessels. This leads to the recognized pathophysiologic sequelae of pulmonary hypoplasia, pulmonary hypertension, and postnatal respiratory failure.

CDH affects approximately 1 in 3500 live births (8). The clinical course for neonates ranges from exceedingly good with standard postnatal care to death despite all interventions. Historically, the reported mortality for neonates diagnosed with CDH at birth has been reported to be 30–50% (9), and the reported mortality for those diagnosed antenatally up to 88% (10,11) despite optimum postnatal care including extracorporeal membrane oxygenation (ECMO). This difference, termed the "hidden mortality" by Harrison et al. (11), reflects fetal death *in utero* or shortly after birth. Fetuses with the poorest outcomes can be risk stratified by prenatal ultrasound through the identification of liver herniation (liver-up) (12); and calculation of a lung-to-head ratio (LHR—length times width of right lung divided by the head circumference) (13,14). Fetuses with "liver-up" CDH have a 43% survival vs. 93% in those with "liver-down" CDH. Liver-down fetuses with LHRs less than 1 have 100% mortality, LHR between 1 and 1.4 have 60% mortality, and LHR greater than 1.4 have zero mortality. Some have argued that new nonsurgical

therapies have improved survival for CDH infants, but Stege et al. (15) contend that reported increases in survival for CDH over the 1990s have been due to selection bias and that newer therapies such as ECMO, high-frequency ventilation, and inhaled nitric oxide have had no effect on the mortality of ~62%. Most experts currently believe that a philosophical change toward permissive ventilatory care to include spontaneous respiration has had the greatest impact on survival (16,17).

The early dismal postnatal mortality rates (18) and the desire to combat the "hidden mortality" led to the first attempt to repair diaphragmatic hernia *in utero.* Experimental results in sheep and primates demonstated improved lung growth, pulmonary function, and neonatal survival (19–22). Although the open surgical repair (i.e., hysterotomy, partial fetal delivery, and repair of the diaphragmatic defect) was demonstrated to be feasible in humans (23–25), there were many factors limiting its usefulness including the global issues for open fetal surgery of PROM, PTL, and fetal morbidity.

In 1997, Harrison et al. (26) reported the results of a prospective, National Institutes of Health (NIH)-funded trial comparing open repair to standard postnatal care [which included ECMO support, when indicated]. Four fetuses with isolated left-sided CDH, significant lung volume displacement, and *no* liver herniation underwent prenatal repair, and seven were repaired after birth. There was no significant difference in survival (75% vs. 86%), and therefore the study concluded that fetuses with prenatally diagnosed CDH *without* liver herniation should be treated with standard postnatal care. Yet, the optimum treatment of fetuses with severe CDH as evidenced by liver herniation and a low LHR, remain unaddressed.

The UCSF group subsequently searched for ways to exploit the known observation that fetal lungs externally drained of fluid do not grow, whereas prevention of the efflux of fluid from fetal lungs via tracheal obstruction promotes lung growth (27–29). Several methods of reversible tracheal occlusion were devised and applied. The initial human experience at UCSF with tracheal occlusion (PLUG technique—plug the lung until it grows) (31) involved open surgery (Table 5.2) with placement of an intratracheal plug or external tracheal clips. These devices were removed at the time of birth using the *ex utero* intrapartum treatment (EXIT) strategy. Initially, eight fetuses underwent tracheal occlusion. The first occlusion device was an internal foam plug which produced the desired result on the lung but caused tracheomalacia. In the second case, a smaller plug was used to avoid tracheal injury but it failed to produce lung enlargement, likely due to leak around the plug. To overcome these problems, an external clip technique was developed using aneurysm clips (two cases) and subsequently hemoclips (four cases), which were easily removed.

Flake et al. (33) at the Children's Hospital of Pennsylvania (CHOP) reported their experience with open fetal tracheal occlusion with hemoclips (Table 5.3). From 1995 to 1999, 15 fetuses underwent open temporary tracheal occlusion. These fetuses had isolated, severe right- and left-sided CDH with low LHR. Five (33%) fetuses survived long-term, and of these, three had severe neurological deficits. Lung growth was variable but those occluded early (before 26 weeks' gestation) showed more consistent lung growth. This group observed that even in the fetuses with dramatic lung growth, lung function seemed impaired postnatally. They attributed this to prematurity, to the detrimental effect on the number and function of type II pneumocytes by tracheal occlusion (34–36), and possibly to altered lymphatic drainage impairing lung fluid clearance after birth.

The open tracheal occlusion procedures were performed by hysterotomy and therefore, were complicated by the ever present specter of PROM and PTL. To minimize uterine trauma and its sequelae, video-assisted fetal endoscopy (FETENDO) was

Table 5.2 Open Tracheal Occlusion (UCSF Experience) (31)

Case	Gest age at diagnosis	Gest age at operation	Gest age at delivery	Survival	Cause of death	Other
1	23	27	30	E		Foam plug
2	18	27	31	C	Plug not occlusive, pulmonary hypoplasia	Foam plug
3	18	25	29	B	Umbilical cord accident	Hemoclip
4	20	25	28	C	Intracranial hemorrhage, support withdrawn	Hemoclip
5	20	27	27	B	Tocolytic failure, IUFD	Aneurysm clip
6	20	27	34	D	Plug not occlusive, death at 4 months	Aneurysm clip
7	19	26	29	D	Bowel necrosis at 4 months	Hemoclip
8	18	27	32	D	CNS damage at 4 months, support withdrawn	Hemoclip
9	23	26	27	B	Hydrops from rapid, excessive lung growth	Hemoclip
10	21	29	30	C	Ipsilateral sequestration	Hemoclip
11	20	30	33	C	Tetrasomy 12 p, death at 4 h	Hemoclip
12	21	30	31	C	No biologic response to occlusion, death at 1 h	Hemoclip
13	26	30	33	C	No biologic response to occlusion, death at 30 h	Hemoclip

Note: A, Died intra-operatively; B, Died *in utero* but not intra-operatively; C, Died within 30 days of birth; D, Died after 30 days old; E, Long-term survival.

developed (6,37,38). The initial technique utilized a maternal laparotomy and three access ports. Under ultrasound guidance, the fetal neck was fixed in extension with a chin stay-suture and the tracheal midline is identified with the placement of a T-bar. A perfusion pump circulated warmed irrigation and suction fluid via an operative fetoscope. The trachea was dissected and occluded with two titanium clips.

A comparison of the FETENDO technique with open tracheal occlusion and standard postnatal care was reported in 1998 by UCSF in a retrospective study. From 1994 to 1997, the initial eight fetuses and an additional five fetuses (four of which were conversions from fetoscopy) underwent open tracheal occlusion. Thirteen underwent standard postnatal care and eight were treated with fetoscopic tracheal occlusion. The results were very promising for FETENDO with a 75% survival rate (does not include converted cases) compared with 38% standard care and 15% open surgery. Seven out of the eight FETENDO fetuses demonstrated a reliable physiologic response to the occlusion with consistent lung growth, whereas only five of the thirteen open fetuses had evidence of lung growth.

In July 2003, UCSF reported progress with the FETENDO technique and discussed 11 additional fetoscopic cases (Table 5.4) (39), making a total of 19 fetuses treated with FETENDO clips from 1996 to 1999. They reported an overall 68% survival 90 days after delivery, with an 86% survival for fetuses with LHR >1 and 63% for LHR <1. Only one fetus died *in utero* on postoperative day two. Obstetrical complications included 6 mothers

Table 5.3 Open Tracheal Occlusion (CHOP Experience) (33)

Enrollment and selection	
Total number of cases	15
Right-sided liver herniation	2
Left-sided liver herniation	13
Maternal morbidity	
Early preterm labor (POD 2, 5)	2
Complicated post-op course	3 • Readmission for tocolysis • Pulmonary edema and ventilation Vaginal bleeding and possible chorioamnionitis • Uterine irritability and cervical change Bedrest, PTL
Lung growth after fetal tracheal occlusion	
Late tracheal occlusion (27–28 weeks) with clear lung growth	3/7
Early tracheal occlusion (25–26 weeks) with clear lung growth	5/6
Survival	
Right sided/left sided	2/3 [total 5 (33%)]
ECMO required	4 (1 survived)
LHR (left-sided lesion only)	0.73
Average hospitalization for survivors	76 days
Average hospitalization for deaths	18 days
Causes of death	• Early PTL (2) • Atrial perforation with central line (1) • Inadequate lung growth, inability to be resuscitated (1) • Multisystem organ failure (6)
Long-term survival	
Tracheal stenosis	0
Severe neurologic deficits	3
Recurrent pneumonia	2

Note: POD, Postoperative day; ECMO, extracorporeal membrane oxygenation; LHR, lung-head ratio.

who developed pulmonary edema, 12 who had chorioamniotic membrane separation, and 12 who developed PROM. There was an additional fetal morbidity in the form of vocal cord paresis/paralysis, tracheomalacia, and tracheal stenosis in five patients. Although the early attempts at endoluminal plugging encountered problems with tracheomalacia and leak, this concept was revisited. The evolution of the endoluminal approach involved a gelfoam plug, an expanding umbrella, and finally a detachable balloon (30,40,41). In May 2001, UCSF reported two cases of CDH treated by fetoscopically placed detachable balloon (42). Maternal laparotomy was still performed, but only a single 5 mm trocar was used. Hydrodissection with a continuous perfusion fetoscope allowed for access to the fetal mouth and trachea, and a detachable balloon loaded on a catheter was placed in the trachea via the side port of the fetoscope. The balloon was inflated to optimize the seal but to avoid tracheal ischemia. Both fetuses survived and did well without any airway-related problems.

Table 5.4 FETENDO Clip Experience (5,39)

Case	Gest age at diagnosis	Gest age at operation	Gest age at delivery	Survival	Cause of death	Other
1	23	30	33	C	Multiple pterygium syndrome, support withdrawn	
2	16	30	31	E		
3	21	30	33	E		Vocal cord paresis, tracheostomy Died at 11 months, tracheostomy dislodgement
4	19	28	35	E		Vocal cord paresis, tracheostomy
5	20	29	35	C	No biologic response to occlusion	
6	19	27	31	E		Laceration, repair at EXIT
7	18	28	29	E		
8	19	27	32	E		
9	23	29	32	E		Died at 9 months, meningitis
10	24	28	31	C	No biologic response to occlusion	
11	20	26	30	E		Vocal cord paresis, tracheostomy Died at 15 months, tracheostomy dislodgement
12	16	25	26	B	Fetal demise	
13	19	26	27	C	Pneumonia, sepsis, pulmonary hemorrhage, ischemic bowel	
14	21	26	30	C	During CDH repair	
15	24	27	32	E		Laceration, repair at EXIT
16	21	26	33	E		
17	17	25	29	E		Vocal cord paresis, tracheostomy, malacia with multiple stents
18	17	26	31	E		Malacia with multiple stents Cotton procedure
19	17	26	32	E		

Note: A, Died intra-operatively; B, Died *in utero* but not intra-operatively; C, Died within 30 days of birth; D, Died after 30 days old; E, Long term survival; EXIT, *ex utero* intrapartum treatment.

Before this technique could be widely disseminated, the UCSF group embarked on an NIH-sponsored prospective, randomized trial to compare fetoscopic tracheal occlusion (balloon) with optimal postnatal care. In November 2003, the results of that trial were reported (43) (Table 5.5). Women with fetuses between 22 and 27 weeks' gestation and severe left-sided CDH (liver herniation and LHR <1.4) were randomized. After the enrollment of 24 women, an interim analysis demonstrated no difference in 90 day survival between groups (77% vs. 73% for postnatal care and tracheal occlusion, respectively). This was an unexpectantly high survival in the postnatal care group. It was determined that it would not be feasible to accrue enough patients to show a difference in mortality between groups and the study was terminated. There was a significant difference in gestational age at delivery between the two groups. The tracheal occlusion group delivered at an average of 31 weeks vs. 37 weeks gestation with standard care. This result combined with the survival statistics suggests that the benefits of lung development with tracheal occlusion may be offset by the costs to the fetus from prematurity. Another significant result was the direct correlation of LHR to mortality. The hazard ratio for death associated with an

Table 5.5 FETENDO Balloon vs. Postnatal Care Randomized Trial Data (43)

Parameter (%)	Standard care ($N = 13$)	Tracheal occlusion ($N = 11$)
Maternal age	28.5 ± 5.7	29.5 ± 5.6
Fetal sex (% male)	9 (69)	8 (73)
Gestational age at randomization	25.4 ± 1.3	24.5 ± 1.6
LHR	0.96 ± 0.20	0.97 ± 0.14
Maternal wound infection	0	1 (9)
PTL	4 (31)	8 (73)
PROM	3 (23)	11 (100)
Time from tracheal occlusion to PROM		24.8 ± 14.8
Time from PROM to delivery	<1	9.5 ± 8.5
Placental abruption	1 (8)	3 (27)
Mode of delivery EXIT/vaginal/Cesarean section	0/12 (92)/1 (8)	11 (100)/0/0
Gestational age at delivery	37 ± 1.5	30.8 ± 2.0
Birth weight	3.03 ± 0.48	1.49 ± 0.36
Survival LHR <0.79	0/0	0/1
Survival LHR 0.79–1.06	8/11 (73)	5/7 (71)
Survival LHR 1.07–1.39	2/2 (100)	3/3 (100)
Age at CDH repair	6.7 ± 2.2	5.7 ± 2.3
Prosthetic patch CDH repair	10/11 (91)	11/11 (100)
Age at successful extubation	35.3 ± 20.5	38.8 ± 15.5
Age at hospital discharge	62.1 ± 28.7	59.6 ± 17.9
Supplemental oxygen at discharge	4/9 (44)	4/8 (50)
Age at full enteral feeding	27.2 ± 10.5	31.9 ± 11.9
Fundoplication	8/11 (73)	7/11 (64)
Gastrostomy tube	3/10 (30)	1/9 (11)
Tube feeding at discharge	5/9 (55)	4/8 (50)
Antireflux meds at discharge	8/9 (89)	7/8 (88)
Weight gain at discharge	680 ± 490	570 ± 540

Note: LHR, Lung-head ratio; PROM, Premature rupture of membranes; EXIT, *Ex utero* intrapartum treatment; CDH, Congenital diaphragmatic hernia.

LHR >0.9 to an LHR ≤ 0.9 was 0.13. The results of this study have generated many questions that need further study. The optimal timing and duration of occlusion in humans still need to be determined. Also, developments in endoscopic instrumentation may improve the PTL rates with fetoscopy and therefore eliminate some of the morbidity of prematurity. Finally, fetuses with LHR <0.9 still have a very poor prognosis. Therefore, the Eurofetus group is developing plans for a randomized control trial targeting this particular subset of CDH fetuses (49). The unexpectedly high survival with standard care may be evidence of the advancement in perinatal care concurrent with advances in surgical treatment.

2. TWIN–TWIN TRANSFUSION SYNDROME

Twin–twin transfusion syndrome (TTTS) is a complication of monochorionic pregnancies. Of the total 20–30% of all twins are monochorionic, and ~10% of these suffer from varying degrees of TTTS (50). In diamniotic pregnancies, TTTS is defined by the presence of polyhydramnios [maximum vertical pocket (MVP) >8 cm] in the recipient twin's sac and oligohydramnios (MVP <2 cm) in the donor or "stuck" twin's sac (51). The syndrome results from an imbalance in net blood flow between the twins due to abnormal placental vascular communications. The donor twin typically suffers from growth retardation and progressive renal failure, whereas the recipient twin experiences overload cardiac failure, hydrops, and possibly *in utero* demise. Expectant management results in 80–100% mortality of both twins (44,46).

Quintero et al. (47) described a staging system to risk stratify twins (Table 5.6) based on retrospective data. The absence of urine in the donor twin bladder after 60 min of ultrasonographic observation coupled with critically abnormal Doppler studies in either twin (e.g., absent or reverse end-diastolic velocity in the umbilical artery, pulsatile umbilical venous flow, or reverse flow in the ductus venosus) were determined to be poor prognostic indicators. Hydrops in either twin, indicative of cardiac failure, was an extremely poor prognostic indicator, and finally death of either twin was usually followed by death of the other, or the delivery of an extremely compromised (usually neurologic impairment) twin. This group's 1999 study demonstrated a statistically significant difference in survival rates by stage; however, another study by Taylor et al. (48) reported that prognosis correlated with a change in stage rather than the stage on presentation.

Treatment strategies for TTTS have included expectant management, serial amniocenteses (amnioreduction), laser therapy, umbilical cord occlusion, and septostomy. Expectant management results in $<20\%$ survival (49). One may argue for a role of expectant management in stage I disease late in gestation.

Table 5.6 Twin–Twin Transfusion Syndrome Staging (47)

Stage I	+BDT
Stage II	−BDT
	−CAD
Stage III	−BDT
	+CAD
Stage IV	Hydrops
Stage V	*In utero* demise

Note: BDT, urine visible in bladder of donor twin by ultrasound; CAD, critically abnormal Doppler studies.

Serial amniocentesis of the polyhydramniotic sac to reduce the amniotic fluid volume has been shown to prolong pregnancy and improve survival by an unknown mechanism (49–52). This procedure has an overall success rate of 66% (at least one twin survival) with a risk of neurological impairment of 15% (45). A recent study by Johnsen et al. (53) examined 24 pregnancies with TTTS treated by serial amniocenteses between 1993 and 1999. A 79% of pregnancies had at least one fetus survive with 50% of both fetuses surviving. The mean gestational age was 34.6 weeks.

In 1990, De Lia et al. (54) demonstrated the feasibility of using Nd:YAG laser photocoagulation as a treatment modality for TTTS. In this technique, all placental vessels crossing the inter-twin membrane are photocoagulated using fetoscopy. By 1995 (55,56), this group demonstrated a 53% survival rate (28/53) with this technique. Ville et al. (57) used photocoagulation in a study with 132 pregnancies and demonstrated a 55% survival rate, a 73% survival rate of at least one twin, and a 4.2% rate of adverse neurologic sequelae after 1 year.

In a 1999 retrospective study by Hecher et al. (58), 73 of the patients treated with laser therapy were compared with 43 patients treated with amniocentesis. The two groups had similar survival rates (61% vs. 51%), but the laser group had a greater number of pregnancies with one or more survivors (79% vs. 60%), less spontaneous intra-uterine deaths (3% vs. 19%), lower incidence of brain abnormalities (6% vs. 18%), a longer interval between intervention and delivery (90 days vs. 72 days), and higher birth weights (1750 g vs. 1145 g). The next year, the same group (59) reported an overall 68% survival rate, with an 81% survival rate for one twin and a 42% survival rate of both twins in a large series of 200 pregnancies.

In 1998, Quintero et al. (60) introduced the concept of selective laser photocoagulation of communicating vessels (S-LPCV) vs. non-selective laser photocoagulation of communicating vessels (NS-LPCV). In NS-LPCV, all vessels crossing the inter-twin membrane are targeted. In the selective technique, only unpaired vessels are targeted. Arterio–venous communications are identified by noting that the terminal end of one artery does not have a corresponding returning vein to the same fetus but, rather, returns to the other fetus. Also, arterio–arterio and venous–venous communications are identified by following these vessels from one fetus to the other. In 2000, the group published data (61) by comparing these two approaches. There were 18 pregnancies in the NS-LPCV group and 74 in the S-LPCV group. Survival of at least one twin was higher in the S-LPCV group (83% vs. 61%) because there was a lower rate of intra-uterine demise of both fetuses (5.6% vs. 22%). There were more hydropic fetuses in the NS-LPCV group (27% vs. 5.4%). Feldstein et al. (62) described a similar but "super selective" technique denoted as the "SELECT" procedure (63) (sonographically evaluated, laser endoscopic coagulation for twins). In the SELECT procedure, a TTTS pregnancy, unresponsive to serial amniocenteses, was successfully treated by identifying the single offending arteriovenous anastomosis through spectral Doppler and fetoscopy. Only this putative anastomosis was laser coagulated.

Prospective, randomized clinical trials are currently underway in Europe and in the United States to compare the efficacy of amnioreduction and laser photocoagulation. The inclusion criteria for the multi-center US trial include monochorionic diamniotic pregnancies diagnosed with TTTS prior to 22 weeks' gestation, oligohydramnios in the donor twin and polyhydramnios in the recipient twin, decompressed bladder in the donor (stages II–V), and no structural abnormalities or known CNS abnormality by MRI. Patients will be randomized to either aggressive serial amnioreduction or selective fetoscopic laser photocoagulation before 24 weeks' gestation and stratified by pregnancies presenting prior to 20 weeks' and after 20 weeks' gestation. The pregnancies will be followed with

ultrasound, MRI, and echocardiography. The primary outcome measure is 30 day survival after delivery (with no treatment failure). Secondary outcome measures will include neonatal comorbidities and long-term neurodevelopmental assessment, short- and long-term maternal morbidities and postpartum correlation of imaging data, and placental examination. This trial differs from the European trial in the use of selective laser photocoagulation (vs. nonselective in Europe), and in the rigorous assessment of long-term developmental outcomes. Patient recruitment began in March 2002.

Umbilical cord occlusion is usually reserved for the severe case in which fetal death of one twin is likely. When one twin dies, the loss of blood pressure in the dying twin causes an acute hemorrhage from the healthy twin into the dying one. Acute hypotension usually causes death or neurologic damage in the remaining twin (64). The goal of umbilical cord occlusion is to eliminate the blood exchange between the two fetuses by ligating (65,66) or cauterizing (67,68) the umbilical cord of the terminal twin. This can limit the acute transfusion event, although some transfusion can occur through other anomalous connections. In a related study by Taylor et al. (68), bipolar cord coagulation was performed on 15 Stage III and IV TTTS pregnancies with a survival rate of the co-twin of 93%. This is marginally higher to the single survivor rates of laser coagulation of vessels of 85% for Stage III/IV (61). The overall survival for umbilical cord ligation is a little lower at 46% compared with 57% for photocoagulation (61,68). Quintero (51) quotes a survival rate of 76% with no incidence of cerebral palsy with umbilical cord ligation.

Septostomy was first described by Saade et al. (69) in 1995 as a method to equalize volumes in each fetal sac and minimize the number of invasive procedures. The technique used a needle to puncture the intertwin membrane, allowing fluid to accumulate around the oligohydramniotic fetus. The same group showed in 1998 that septostomy can provide a survival rate of 83% (20/24) which is comparable to more invasive methods. Johnson et al. (70) compared amnioreduction with septostomy and demonstrated a similar overall survival rate (78%). These groups believe that septostomy may provide similar benefits as amnioreduction with fewer numbers of procedures, more room for the "stuck fetus," and possibly a later gestation delivery. Some groups believe that the problems with septostomy make it a poor treatment option. Quintero (45) has demonstrated that there is no pressure differential between the two sacs (71,72); thus obviating the need for eliminating the membrane. He states that septostomy can result in a pseudomonoamniotic state fraught with the problems of cord entanglement and fetal demise. A randomized multi-centered trial is being considered to compare these options (73).

3. TWIN REVERSED ARTERIAL PERFUSION AND TWINS DISCORDANT FOR A LETHAL ANOMALY

Twin reversed arterial perfusion (TRAP) is a rare complication of monozygotic twin pregnancy and occurs in 1% of these pregnancies (74). The TRAP sequence is the most severe form of TTTS. A normal (pump) twin provides circulation for itself and an abnormally developing acardiac (perfused) twin. The acardiac twin is not connected to the placenta but, rather, directly to the umbilical cord of the pump twin. The acardiac twin is perfused by the normal twin pumping in a reversed direction into the acardiac twin. The pump twin is at risk for developing high output cardiac failure, hydrops, and death (75). The mortality for the acardiac twin is 100% and 35–55% for the pump twin (76–78). One prognostic indicator is the twin:weight ratio (weight of the acardiac twin expressed as a percentage of the weight of the other twin). Moore et al. (78) noted that in 49 TRAP pregnancies,

a 70% ratio predicted a 90% pre-term delivery rate, 40% hydramnios rate, and 30% congestive heart failure rate in the pump twin.

Some monochorionic twin pregnancies can be complicated by one twin that is discordant for a lethal anomaly, that is, one that has a likelihood of leading to *in utero* demise. Usually there is no placental anomaly. However, if the abnormal twin dies *in utero*, the normal co-twin may be impaired or die due to a hemodynamic "unloading" into the deceased co-twin. Interventions for TRAP that have been described include termination of pregnancy, expectant management with early delivery, treatment of polyhydramnios with indomethacin, (79) treatment of heart failure with digoxin (75,80), and early delivery of the abnormal twin by hysterotomy (termed sectio parvo) (81–83). All of these approaches carry significant risks to the mother and the normal fetus. Currently, the treatment goals for TRAP sequence, discordant anomalies and Stage V TTTS is selective reduction of the acardiac, anomalous, or hydropic co-twin. Depending on the placental anatomy, selective twin reduction can be done via umbilical cord embolization, ligation, ultrasonic transaction, laser coagulation, or thermal coagulation using monopolar, bipolar, and radiofrequency (RF) energy. Embolization, although widely studied (84–88), has fallen into disfavor because of high failure rates (23%) and pregnancy loss (32%) (75).

In 1994, Quintero et al. (89) described the first successful endoscopically guided umbilical cord suture ligation of an acardiac twin. The mother had an uncomplicated birth of a normal twin at 36 weeks. In 1998, DePrest et al. (90) reviewed 23 cases of cord ligation which had a survival rate of 73% but a high risk of PROM (40%).

Laser photocoagulaton of the cord is performed fetoscopically (91–93). The root of the anomalous twin's umbilical cord is targeted with an Nd:YAG laser. Lewi et al. (94) reported a consecutive series of 50 cases. Forty-six percent of the cases failed and were completed with bipolar coagulation. The Lewi study noted a 75% survival rate, and a persistent PPROM rate of 25%. Moldenhauer et al. (75) described two successful cases of laser coagulation through a 16 gage needle under ultrasound guidance.

Because of the high failure rate of photocoagluation, thermal coagulation has been attempted. A monopolar technique first performed in four cases was described by Rodeck et al. (95). In this technique, a sonographically guided wire is placed into or adjacent to the lumen of the aorta. Three of the cases in the Rodeck experience had good outcomes, in the remaining case, blood flow was only reduced not terminated. The acardiac twin stopped growing after 2 weeks, and the hydrops of the pump twin improved. The mother suffered from pre-eclampsia and the baby was delivered at 32 weeks' gestation with hyaline membrane disease and developmental delay. Rodeck contends that the sonographically guided needle and wire are safer and less expensive than other techniques and targeting the aorta vs. the umbilical cord is relatively easy. It can also be performed earlier in gestation (95–97).

Bipolar coagulation has been performed in 108 cases by a number of groups with excellent success (100%), but with a high rate of PROM (20%) (75). This procedure is performed under ultrasound guidance using a bipolar cautery device through an endoscopic trocar. The umbilical cord is grasped, thermal energy applied, and cessation of blood flow is confirmed by Doppler ultrasound. Bipolar energy via a single trocar has also been used to transect the umbilical cord (98–100).

Perhaps the quickest and simplest method for selective reduction of a compromised twin is the use of RF energy for cord ablation (100–102). Using sonographic guidance, a 14 gauge needle is percutaneously placed at the base of the umbilical cord. Energy is applied through a radiofrequency ablation (RFA) probe placed through the angiocath and blood flow ceases within 5 min. An additional advantage of this approach is an ability for the needle and probe to be placed through an anterior placenta without

complication. Tsao et al. (102) described 13 TRAP cases treated with RF ablation. Twelve of the 13 delivered a healthy twin, and one was prematurely delivered and died subsequently.

Tan and Sepulveda (103) reviewed treatment of acardiac twins through 2002. They identified 75 cases treated with minimally invasive techniques and divided these into two treatment modality groups—umbilical cord occlusion and intrafetal ablation. Cord occlusion included embolization, ligation, laser coagulation, and monopolar and bipolar thermocoagulation. Intrafetal ablation included alcohol injection, monopolar thermocoagulation, interstitial laser photocoagulation, and RFA. The overall twin survival rate was 76%. Comparing intrafetal ablation with umbilical cord occlusion, they found lower technical failure rates (13% vs. 35%), lower rate of premature delivery or rupture of membranes (23% vs. 58%), higher median gestational age at delivery (37 vs. 32 weeks), and a longer interval between treatment and delivery (16 vs. 19.5 weeks) with the intrafetal ablation techniques. This group, therefore, claims that the treatment of choice for TRAP should utilize intrafetal ablation. A summary of experience with vascular occlusion techniques is listed in Table 5.7.

4. OBSTRUCTIVE UROPATHY

Fetal lower urinary tract obstruction (LUTO) can lead to irreversible renal damage from renal dysplasia and to lung hypoplasia from oligo- or anhydramnios (104). Oligohydramnios can also lead to face and limb deformities, and bladder distention can lead to abdominal muscle deficiency. Obstructive congenital abnormalities occur in 1% of pregnancies, but only one out of 500 pregnancies have severe urologic manifestations (105). The most common cause of LUTO in males is posterior urethral valves and urethral atresia. Female fetuses with LUTO, typically have developmental abnormalities associated with syndromes (e.g., cloacal anomaly) that are not amenable to antenatal treatment (105). Neonates that manifest completely obstructing posterior urethral valves from early gestation have a 45% mortality rate (106) due mostly to pulmonary hypoplasia. Early oligohydramnios from LUTO (<22 weeks' gestation) has a mortality rate as high as 95% (107).

The clinical picture of fetal LUTO was reproduced in an elegant experimental sheep model by Harrison et al. (108) where ligation of the fetal lamb urachus and urethra *in utero*

Table 5.7 Outcomes of Vascular Occlusion Techniques for Fetuses With TTTS, TRAP, and Those Discordant for Lethal Anomaly (75,97,100–102)

Technique	Procedures	Gestational age at procedure	Success of occlusion	PROM	Total loss	Gestational age at delivery
Embolization	22	24 (18–27)	17 (77%)	2 (9%)	7 (32%)	34 (24–39)
Ligation	24	22 (17–26)	21 (88%)	7 (39%)	9 (35%)	30 (24–37)
Monopolar coagulation	13	20 (16–24)	11 (85%)[a]	1 (8%)	4 (31%)	36 (32–42)
Bipolar coagulation	108	21 (13–28)	108 (100%)	21 (21%)	19 (18%)	33 (24–41)
Radiofrequency ablation	16	20 (17–24)	15 (94%)	3 (19%)	2 (13%)	38 (24–40)

[a]The two failures required a second occlusion.
Note: PROM, premature rupture of membranes.

produced bilateral hydronephrosis and severe pulmonary hypoplasia with a resultant high perinatal mortality. The same group was able to ameliorate these effects by relieving the obstruction with a suprapubic cutaneous cystostomy (109). These experiments suggested that the fetus with early and severe obstructive uropathy may be salvageable.

In 1982, Harrison et al. (110,111) reported the first human clinical experience with antenatal surgical intervention for fetal LUTO. Of 26 pregnancies with hydronephrosis, 9 underwent antenatal interventions. Three who were diagnosed by percutaneous vesicoamniotic shunting to have poor renal function were aborted. Four had percutaneous vesicoamniotic shunts successfully placed. Only one underwent open fetal surgery with the creation of bilateral ureterostomies. Three of the five unaborted fetuses died postnatally and two survived. Of the three nonsurvivors, one had multiple anomalies and two had irreversible kidney damage. With five out of nine fetuses having renal failure, this experience illustrated the need for more accurate and earlier diagnosis to identify fetuses that may benefit from prenatal intervention.

In 1994, Johnson et al. (112) reported the evaluation of 24 cases of fetal obstructive uropathy and proposed an algorithm for identifying fetuses for antenatal treatment (Fig. 5.1). The steps include (1) a detailed ultrasound exam identifying the signs of LUTO and also screening for other structural abnormalities, (2) fetal karyotype analysis, and (3) three serial fetal bladder aspirations (over 3–5 days) with analysis of fetal urinary electrolytes and protein to assess kidney function (Table 5.8). The third aspiration is believed to most accurately reflect fetal renal function. Candidates for fetal surgery need to have a normal male karyotype, no other lethal anomalies, a favorable urinalysis, and favorable-appearing kidneys by ultrasound (i.e., no evidence of corticomedullary dysplasia or cystic changes).

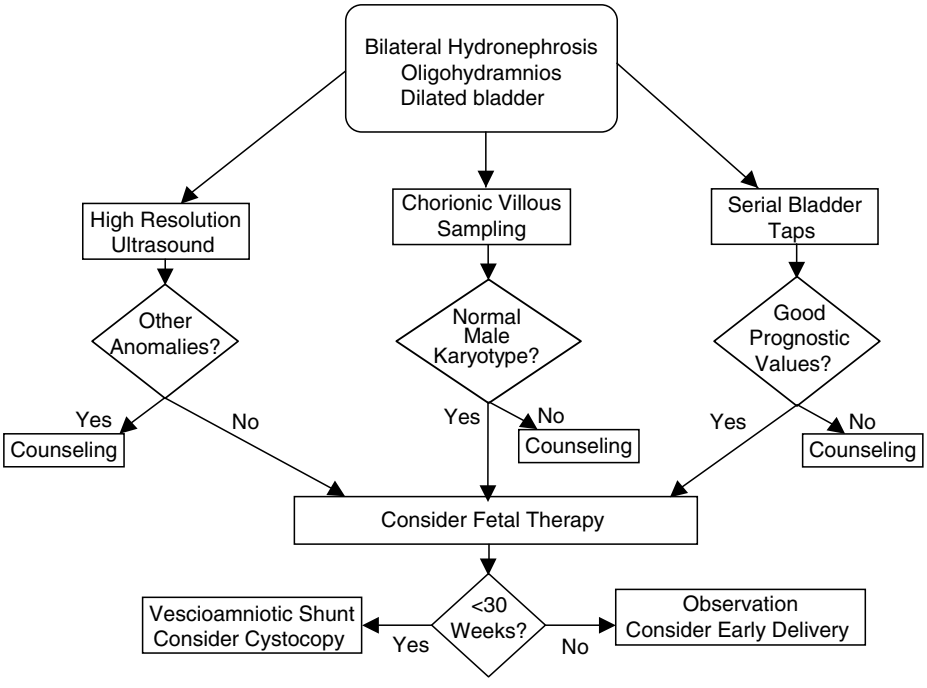


Figure 5.1 Management algorithm for fetal obstructive uropathy [Johnson et al. (112)].

Table 5.8 Favorable Levels of Fetal Urinary Electrolytes and Protein (112)

Electrolyte	Cutoff
Sodium	<100 mg/dL
Chloride	<90 mmol/L
Calcium	<8 mg/dL
Osmolality	<200 mOsm/L
B2-Microglobulin	<4 mg/L
Total protein	<40 mg/dL

Current treatment therapies for LUTO include percutaneous sonographically guided vesicoamniotic shunt placement and direct valve ablation via fetal cystoscopy. In early surgical experience, open fetal therapy was performed but was abandoned due to the advent of percutaneously placed shunts which resulted in less morbidity to mother and fetus than the open hysterotomy approach (113).

Subsequently, there have been a number of large series examining clinical experience in using vesicoamniotic shunts. Vesicoamniotic shunting is performed percutaneously with ultrasound guidance. Since the initial double-J catheter proved to be prone to obstruction and dislodgement, it was subsequently replaced by a double pigtail catheter by Rodeck et al. (114). The pigtail shunt is placed between the bladder and the amniotic cavity to temporarily divert urine, relieve the obstruction, and potentially treat oligohydramnios. The first large series was reported by Elder et al. (115) who reviewed all cases of antenatal intervention for fetal obstructive uropathy through the end of 1985. Of the 57 interventions (from bladder aspirations to vesicostomies), 21 were technically successful and 10 unsuccessful. The complications included shunt migration or poor drainage (11) and shunt extrusions into the peritoneal cavity (2). Of the nine patients with oligohydramnios treated by shunt, only two survived, three were terminated and four died. Forty-five percent of fetuses reviewed had complications (although not all of these had shunt placement).

In 1986, the report of the International Fetal Registry was published (116). The registry described 73 catheter shunt placements for fetal obstructive uropathy with a 41% survival rate (48% if elective terminations not included) and a 4.1% procedure-related mortality rate. The majority of deaths were perinatal due to pulmonary hypoplasia. Although this was encouraging and, again, demonstrated feasibility, efficacy could not be established due to the selection bias in this data set.

In 1990, Crombleholme et al. (117) presented 19 cases of vesicoamniotic shunt placement with encouraging results. Shunting restored amniotic fluid levels and prevented pulmonary hypoplasia in 9 of 17 (53%) fetuses with oligohydramnios. Overall survival for shunted fetuses was 58% (11/19). The report divided the cases into those with good prognosis and poor prognosis according to the prenatal diagnostic evaluation. In the poor prognosis group, 3/10 survived with shunting and 0/14 survived without. In the good prognosis group, 8/9 survived with shunting and 5/7 survived without.

In 1993, Lipitz et al. (118) examined a series of 25 fetuses with LUTO, 14 of who underwent vesicoamniotic shunting. Of these fetuses, two were electively terminated, one died *in utero*, five died in the newborn period, and six survived (43%). Five of the six shunted survivors had evidence of renal dysfunction. Three out of eleven (27%) survived without a shunt with two having evidence of renal damage. Although the numbers in this study were small and there was one procedure-related death, these data suggest shunting is worthwhile.

In 1996, Freedman et al. (119) reviewed 55 cases of fetal obstructive uropathy. Of these 28 were shunted with an overall survival of 61%. Of the poor prognosis fetuses, 50% of those shunted survived whereas none of the nonshunted survived. Of the good prognosis fetuses, 64% of those shunted survived, and 45% of the nonshunted survived. Of the 13 fetuses with posterior urethral valves, 6/9 (67%) shunted survived [vs. 0/4 (0%) nonshunted]. Of nine prune-belly fetuses, 5/5 (100%) of shunted survived [vs. 1/4 (25%) of nonshunted]. None of the 11 fetuses with urethral atresia survived.

Coplen (120) reviewed these five large series and noted that even with 169 successful shunt placements, fifteen others were unsuccessful or had serious complications including three severe maternal infections and seven procedure-related intra-uterine deaths. Other complications included catheter obstruction, migration, and fetal injury. Failure to restore amniotic fluid levels met with 100% mortality, and 40% of survivors still suffered from end-stage renal disease.

In 1999, Freedman et al. (121) looked at the long-term outcomes for children who underwent antenatal intervention (Table 5.9). Of 34 patients who underwent vesicoamniotic shunting from 1987 to 1996 at the Children's Hospital of Michigan, seventeen survived, but three were lost to follow-up. The 14 children evaluated had a range of diagnoses with most having prune-belly syndrome or posterior urethral valves. Of the long-term survivors, 43% had normal renal function, but 36% required renal transplantation. Some suffered multiple urinary tract infections postnatally that may have worsened any baseline renal dysfunction. The authors concluded that antenatal intervention for children with severe LUTO resulted in outcomes similar to children with less severe disease diagnosed postnatally. This same group compiled their data on complications of vesicoamniotic shunting (122). They compared 31 fetuses that underwent shunt placement to 31 whom did not. Forty-eight percent of the shunted fetuses suffered from mechanical complications—35% had migration out of the bladder, 23% developed urinary ascites (three from intraperitoneal shunt migration and four from bladder fistula), three had bowel herniation at the insertion site. Pre-term delivery occurred in 71% of cases, 32% had PROM, 6% had chorioamnionitis, and 6% died *in utero.* There were no peri-procedure deaths. Of the 16 nonshunted and nonaborted fetuses, 50% had pre-term delivery due to PROM (13%), chorioamnionitis (13%), and *in utero* death (25%).

In 2001, McLorie et al. (123) described the experience at Mt. Sinai Hospital and the Hospital for Sick Children in Toronto from 1989 to 1998. Of 89 fetuses with obstructive uropathy, nine underwent vesicoamniotic shunting. One fetus was electively terminated after shunting, and two with poor prognostic urinalysis died shortly after birth from pulmonary disease. Overall six of eight shunted fetuses (75%) survived, but two of six (33%) had renal failure, and three of six (50%) had bladder dysfunction requiring augmentation. The authors encountered similar complications as other authors including shunt migration with need for reinsertion and bladder prolapse in a fetus with prune belly syndrome. These long-term outcomes and those from Freedman (121) are summarized in Table 5.9.

In 2001, UCSF compiled their data on 14 patients with posterior urethra valves (PUV) treated prenatally (124) (Table 5.10). All had favorable urine electrolytes and only two had sonographic evidence of renal dysfunction. Interventions included cutaneous ureterostomies (1), bladder marsupialization (2), laser ablation of valves (2), and placement of vesicoamniotic shunts (9). Overall mortality was 43% (six fetuses). Five of the surviving eight (63%) have chronic renal disease and five have required urinary diversion due to poor urinary drainage. One surviving child has required an augmentation cystoplasty. Three have exhibited uninhibited bladder contractions, but normal bladder compliance and no vesicoureteral reflux. Two have decreased bladder emptying. These cases again strikingly illustrate the inability to accurately identify fetuses with reversible

Table 5.9 Long-Term Outcomes for Children After Vesicoamniotic Shunting (121,123)

	No.	%
Total number of children studied	20	100
Diagnoses		
Prune belly	8	40
Posterior urethral valves	8	40
Urethral atresia	2	10
Vesicoureteral reflux	1	10
Megacystitis	1	10
Renal function		
ESRD with transplant	7	35
Renal insufficiency	4	20
Normal function	9	43
Voiding function		
Incontinent	3	15
Continent	12	60
Not toilet trained yet	5	25
Urinary tract infections		
<3 in last year	10	50
>6 in last year	4	20
Prophylactic antibiotics	10	50
Growth		
Height <25%	14	70
Height <50%	9	45
Weight <25%	8	40
Respiratory function		
Asthma	2	10
Chronic bronchitis/frequent respiratory tract infections	4	20
Development		
Developmental delay	2	10
Speech therapy	3	15
Physical therapy	1	5
Total surgeries performed on these children	62	
Bladder augmentation	5	25
Renal transplant	7	35

Note: ESRD, end-stage renal disease.

renal disease. Intriguingly, 63% of survivors in this series had chronic renal disease which is higher than estimated to occur in prenatally diagnosed, postnatally treated PUV (125). This may indicate that prenatal treatment improved lung development which decreased perinatal mortality due to lung hypoplasia, but it did not prevent the renal disease. Therefore, more babies survived the immediate perinatal period only to be faced with chronic renal disease. The investigators suggested that the poor results may be due to diagnosis and treatment that is already too late to prevent irreparable disease. Also, the screening criteria used for determination of salvageable renal function remains controversial. Since four of five children with renal disease had no evidence by ultrasound of their renal disease, this should be used as a minor criterion to determine renal salvageability.

Table 5.10 Fetal Therapy for Posterior Urethral Valves (UCSF Experience 1981–1999) (124)

Case	Gest age at electrolyte	Na	Cl	Osmolarity	Gest age at operation	Technique	Survival	Cause of death	Serum Cr (age >12 m)	Renal transplant?
1	20	98	84	210	20	CU	C	PTL, resp failure		
2	18	95	82	205	18	BM	E		0.5	N
3	24	88	83	202	24	BM	E		2.2	Y
4	25	52	39	119	25	ABL	E		1.4	N
5	22	87	70	179	22	ABL	C	PTL, resp failure		
6	18	95	77	194	24	VAS	C	PTL, resp failure		
7	19	90	74	193	18	VAS	B	Terminated		
8	19	93	89	189	19	VAS	C	PTL, resp failure		
9	19	93	66	175	19	VAS (3)	E		1.0	N
10	24	85	70	170	24	VAS	E		3.1	List
11	24	81	64	170	24	VAS	E		3.3	Y (2)
12	25	92	81	103	25	VAS	E		2.0	N
13	28	73	72	172	28	VAS	C	PTL, resp failure		
14	30	70	68	158	30	VAS	E		0.5	N

Note: CU, cutaneous ureterostomy; VAS, vesicoamniotic shunt; BM, bladder marsupialization; ABL, *in utero* valve ablation; PTL, pre-term labor and delivery.
A, Died intra-operatively; B, Died *in utero* but not intra-operatively; C, Died within 30 days of birth; D, Died after 30 days old; E, Long-term survival.

Fetal urinary electrolytes were also considered to be only somewhat reliable because all fetuses in this study had "good" urine electrolytes. They concluded that this study did not support the ability of prenatal intervention to salvage renal function for posterior urethral valves, but that improvements in minimally invasive techniques and tocolysis may improve outcomes in the future.

The clinical outcomes of vesicoamniotic shunting for fetal obstructive uropathy demonstrates that there are some significant limitations. Although shunting does seem to improve the incidence of lung hypoplasia and perinatal death for patients with oligohydramnios, shunting has a high rate of complications including shunt migration and shunt obstruction. Shunting is only a temporizing measure, and the fetus requires another procedure(s) for definitive treatment. Finally, many survivors still suffer from renal damage and bladder dysfunction. As the bladder does not experience the developmentally important normal emptying and filling cycle, it shrinks and develops poor compliance (104). This "valve bladder" has decreased residual capacity which has led to some of the PUV children requiring bladder augmentation after birth. Postnatally it contributes to urinary tract infections which can worsen renal function (104).

A new approach to the treatment of obstructive uropathy utilizing cystoscopy appears promising. Quintero et al. (126) described the first case of fetal cystoscopy and fulguration of posterior urethral valves in 1995. The 19 week fetus was diagnosed with LUTO by ultrasound. Using an ultra-fine fiberoptic endoscope, fetal percutaneous cystoscopy was performed. The valves were electrocoagulated, and the distal urethra was immediately visualized. At 31 weeks' gestation, the patient suffered from PTL and gave birth to a baby boy. His urethra was patent as evidenced by spontaneous urine emission at birth, but he died 4 days later with pulmonary hypoplasia. This case demonstrated the feasibility of posterior urethral valve ablation *in utero*.

Creative variations on this technique have developed. Quintero et al. (127) has examined the use of fetoscopically guided cystotomy in two cases where safe percutaneous vesicoamniotic shunt placement was not possible. Hofman et al. (128) has described a fetoscopic approach in the antegrade placement of a transurethral stent which may avoid possible urethral and sphincter damage from laser ablation. Quintero et al. (129) has also used cystoscopy to guide a laser incision of a ureterocele causing bladder outlet obstruction and oligohydramnios. The right kidney was dysplastic by ultrasound. The oligohydramnios resolved. At birth, the right kidney was nonfunctional, but the left kidney's function was preserved.

As minimal access fetal surgical techniques evolve, patient selection criteria should be continually revisited and current algorithms revised. For example, asymmetric renal injury is currently not accurately assessed. The assumption of stable "normal" fetal urine composition is also inaccurate because it actually varies with gestational age (130). In addition, uniformly recognized outcome measures need to be adopted in order to better assess efficacy of these treatments. Freedman et al. (122) now contends that gross survival, postnatal survival, shunted survival, and nadir creatinine at one year are important parameters to use for comparison.

5. SACROCOCCYGEAL TERATOMA

Sacrococcygeal teratoma (SCT) is a tumor arising from extragonadal germ cells around the sacrum. It occurs in 1 out of 35,000 to 40,000 live births and four times more frequently in females (131). Nearly 90% of SCTs are benign (132). Neonates with SCT usually have an excellent prognosis after resection, but the fetus diagnosed with a large,

rapidly growing, solid, vascular SCT is at high risk. The fetus with a large vascular SCT is at risk for hydrops, placentomegaly, and high-output cardiac failure with subsequent rapid fetal demise due to "vascular steal" from high blood flow through the tumor (133). Other complications include tumor rupture, PTL, and dystocia (134–136). Additionally, mothers may develop a hyperdynamic "mirror syndrome" similar to pre-eclampsia that mimics the fetal condition.

The goal of fetal intervention is to interrupt the high flow circulation through the tumor. Historically, this was accomplished by open hysterotomy with tumor debulking or complete resection. In 1989, the first case of attempted *in utero* resection was reported by Langer et al. (137). The 22 week gestation fetus had a large SCT, hydrops, and placentomegaly. The tumor was debulked and its major blood supply interrupted successfully, but the fetus was delivered prematurely at 26 weeks and died from pulmonary immaturity. The first successful case was reported by Adzick et al. (138) in 1997. A 25 week gestation fetus underwent successful resection with resolution of hydrops. The fetus was delivered at 29 weeks, underwent resection of the remaining tumor mass and coccyx, and did well. In 2000, a unique case was described of an SCT fetus with rectal atresia discovered at the time of surgery. The SCT was resected, and a pull-through anorectoplasty and a bilateral ureterostomy performed. The fetus survived and delivered at 31 weeks (prompted by chorioamnionitis), but died several days postnatally from iatrogenic atrial rupture from a central venous catheter. The experience at UCSF and CHOP (137–142) has demonstrated the feasibility of open surgery, but it continues to be limited by the problems inherent to open fetal surgery accomplished via hysterotomy.

Potentially promising, less invasive approaches have been attempted to minimize these complications. In the context of predominantly cystic, avascular tumors, cyst aspiration to reduce uterine irritability or maternal discomfort has been used with some success (140). Cyst aspiration at near delivery has helped prevent tumor rupture (140) and even allowed for a vaginal delivery (143). Some groups have placed cyst-amniotic shunts to alleviate obstructive uropathy due to the tumor's mass effect (144,145). These cases demonstrate the nonvascular steal morbidity of fetal SCT which relates to tumor size with potential for resultant PTL and/or dystocia.

For large, predominantly solid, highly vascular tumors in fetuses with impending heart failure, minimally invasive approaches have included tumor embolization, balloon occlusion, sclerosis, endoscopic snaring of the tumor neck, laser ablation, thermocoagulation, and RFA. Tumor embolization, balloon occlusion, sclerosis, and endoscopic snaring have been attempted by a few groups without success (141).

Hecher et al. (146) has described the successful use of fetoscopically guided Nd:YAG laser treatment to reduce blood flow to the tumor in a 20 week gestation fetus with polyhydramnios but no hydrops. Three weeks after surgery, there was bleeding into the cystic portion of the tumor requiring intra-uterine blood transfusion and repeat laser coagulation. In this case, the maternal morbidity was minimal, and the baby was successfully delivered at 37 weeks and underwent subsequent surgical resection. This fetus, however, did not meet standard criteria for fetal intervention (i.e., hydrops), thus the survival may or may not have been due to the therapy. Lam et al. (147) attempted thermocoagulation of the tumor neck in one fetus but resulted in fetal death on postoperative day two. The cause of death was not evident, but may have been due to microbubbles from thermocoagulation, hyperkalemia from tissue necrosis, or hyperthermia from hemolysis.

Paek et al. (148) attempted percutaneous coagulation using RFA of large SCT tumors in four fetuses (Table 5.11). Two fetuses died due to hemorrhage after ablation of a large percentage of the tumors. The other two fetuses survived with delivery at 28 and 31 weeks after having only the tumor necks ablated. There was minimal maternal

Table 5.11 RFA Experience (148)

Case	Gest age at operation	Gest age at delivery	Outcome
1	20	20	Died (*in utero)*: hemorrhage into tumor
2	21	28	Alive: perineal necrosis
3	19	31	Alive: gluteal necrosis
4	22	25	Died postnatally: hemorrhage into tumor

morbidity but the neonates had gluteal and perineal necrosis requiring additional operations. Ibrahim et al. (149) reported the long-term sequelae in a newborn (case 3, Table 5.11) after *in utero* RFA of an SCT. The child was born at 31 weeks' gestation with a large soft-tissue defect over the left hip, exposure of a dislocated hip, and loss of sciatic nerve function. These experiences indicate that a more focused field of ablation using a probe that limits collateral heat production is required.

6. MYELOMENINGOCELE

Myelomeningocele, or "spina bifida," is a devastating birth defect affecting 5 children per 10,000 live births or 1500 infants per year (150) at a cost of approximately $200 million annually (151). It is the most common congenital malformation of the central nervous system. Although typically nonlethal, this neural tube defect may result in a spectrum of morbidity including somatosensory loss, paresis, neurogenic sphincter dysfunction of bowel and bladder, musculoskeletal deformities, sexual dysfunction, Arnold Chiari malformation, hydrocephalus, and impaired mental development. Eighty-five percent of patients require lifelong cerebrospinal fluid decompression with mortality in the first two decades of life approaching 30% primarily due to shunt complications (152). The clinical effects of a myelomeningocele are believed to result from a two "hit" theory: a defect in neurulation which causes the neural tube defect and myelodysplasia from subsequent exposure of the spinal cord to the intrauterine environment causing chronic chemical and mechanical trauma (153).

An increasing body of experimental data suggests that fetal therapy focusing on alleviating the traumatic insult to the spinal cord may be of benefit. A variety of animal experiments involving mice, rat, rabbit, sheep, pig, and monkey have been promising (154–157). These experiments showed that surgical creation of spinal dysraphism replicates the neurologic sequelae of myelomeningocele and immediate surgical repair allows for normal development. In addition, experiments in sheep by Paek et al. (158) demonstrated that hindbrain herniation may be prevented with *in utero* repair. Fetoscopic repair in sheep by Copeland et al. (159) showed that a minimally invasive approach may be feasible.

As the risks to the mother and fetus were originally believed to outweigh the benefit of an open repair, a minimally invasive approach was first attempted. Four fetoscopic repairs were attempted at Vanderbilt University Medical Center (160). The technique involved a maternal laparotomy, placement of three ports, aspiration of the amniotic fluid, and replacement with CO_2 for the duration of the procedure. A maternal skin graft was then fixed over the defect with a combination of fibrin glue and sutures. The uterine cavity was refilled with either amniotic fluid or sterile saline. One fetus died intraoperatively from PTL. One died a few weeks later after getting PTL from amnionitis.

The other two delivered near term, but the skin graft could not be identified and the defect was essentially unrepaired.

The UCSF group also reported their attempts at fetoscopic repair (161). In one case, an alloderm patch was placed fetoscopically, but the case was converted to open due to placental bleeding. At delivery, the patch was partially pulled away. The baby required re-repair and a VP shunt. In the second case, the defect was repaired with a two-layer closure. The mother delivered pre-term due to spontaneous rupture of membranes, and the baby required re-repair and VP shunt due to CSF leak. The baby later died from urosepsis. The third case was converted to open due to an anterior placenta. The fetus, however, was spontaneously aborted 2 weeks later. Table 5.12 summarizes the experience in human fetoscopic repair of myelomeningocele.

The fetoscopic approach to meningomyelocele repair was then abandoned and open repair was again contemplated. Advances in tocolysis and management of PTL, coupled with the fact that the meningomyelocele fetus is otherwise healthy, have possibly diminished the risks to both mother and fetus. The first open repairs were done in 1997, and in 2000, Bruner et al. (162) compared the initial endoscopic cases with the initial open cases. The Bruner study concluded that the hysterotomy approach was technically superior because of shorter operating times, no fetal mortality, shorter neonatal hospital stay, and a suggestion of better functional outcomes.

Since 1997, nearly 200 open repairs have been performed. The open repairs have followed the standard technique of laparotomy, hysterotomy, neurosurgical dissection, three-layer closure with or without lumbar-peritoneal shunt placement and occasionally amnion patch placement. Sutton et al. (163) has shown MRI evidence that the incidence of hindbrain herniation is improved by intrauterine repair, but this did not correspond to a demonstrable reduction in neurologic symptoms or hydrocephalus. Others studies report that leg function and bladder function are not improved in intrauterine repair over conventional therapy (164,165). Recently, in a large retrospective review, Tulipan et al. (166) compared all intrauterine meningomyelocele repairs performed at VUMC and CHOP from 1997 until 2000 with conventionally treated patients from 1983 to 2000. The study demonstrated that fetuses repaired before 25 weeks' gestation have a 50% reduction in the need for ventriculoperitoneal shunts compared with historical controls. Even with these impressive results of decreased shunts rates, the fetal and maternal morbidity (11%) and fetal mortality (4%) are still significant for the treatment of a nonlethal disease (167). As minimally invasive techniques and instrumentation improve, these techniques will be revisited for myelomeningocele treatment (168).

A randomized, prospective trial with three participating centers, Vanderbilt University Medical Center, CHOP, and UCSF, has been underway for the past 2 years to definitively compare fetal myelomeningocele repair with postnatal repair. The primary outcome variable being studied is the need for a ventriculoperitoneal shunt at 1 year of age.

7. TENSION HYDROTHORAX

Fetal tension hydrothorax is an uncommon fetal lesion which occurs in 1 out of 15,000 pregnancies (169). It has a male predominance and is typically right-sided (170). It is caused primarily by leakage of lymph or secondarily by generalized hydrops or certain anatomic abnormalities. Congenital hydrothorax has a 57–100% perinatal mortality (114) with the main cause of death being pulmonary hypoplasia due to compression (171). Intrauterine death is usually caused by fetal hydrops due to mediastinal shift

Table 5.12 Outcome of Fetoscopic Repair of Myelomeningocele (160–162)

GA at procedure	Lesion (US)	Technique	GA at delivery/outcome
VUMC			
22 3/7	L4-S3, mild VM, AC	Maternal STSG, fibrin glue	35 week 1 day: planned C-section, lesion covered by thin translucent membrane, neonatal closure and VPS, mild somatosensory deficit at 30 months
23 6/7	L3-S2, mild VM, AC, bil talipes	Maternal STSG, fibrin glue	24 week 5 day: amnionitis, death in delivery room, lesion uncovered, graft not attached
22 4/7	T12-S5, mild VM, AC	Maternal STSG, fibrin glue, absorbable sutures	28 week 1day: disruption of membranes, PTL, C-section, lesion covered with thin translucent membrane, neonatal closure and VPS, mild somatosensory deficit at 6 months
24 3/7	T12-S3, hemivertebra L-3, mild VM, AC, bil talipes	Maternal STSG, fibrin glue, absorbable sutures	24 week 3 day: intraoperative uterine contractions, placental abruption, intraoperative demise
UCSF			
25	L2-S1, unilat VM, AC	One layer w/Alloderm patch with suture, converted to open to control placental bleeding	35 week: planned C-section, patch partially pulled away from fetal skin, neonatal closure and VPS, somatosensory level to L4 by 1 year
24	L3-S, AC, bil talipes	Two-layer closure	31 week: spontaneous rupture of membranes, C-section, lesion with some CSF leak, neonatal repair and VPS, somatosensory level to L4, represented at 1 month with urosepsis with eventual demise
19	L3-S1, AC, bil talipes	Converted to open due to difficulty with fetal positioning, three-layer closure using chorioamnion patch, fetal ankle dislocation and laceration repaired, fetal bradycardia treated with epinephrine	21 week: premature rupture of membranes and spontaneous abortion of nonviable fetus

Note: GA, gestational age; US, ultrasound; L, lumbar; S, sacral; VM, ventriculomegaly; AC, Arnold-Chiari II malformation; VPS, ventriculoperitoneal shunt; STSG, split thickness skin graft.

resulting in compression of the heart and/or the vena cavae. Fetuses with hydrothorax that exhibit hydrops have a 76% mortality vs. a 25% mortality without hydrops (172).

Intrauterine therapy has focused on ameliorating lung compression, preventing or reversing hydrops, and improving postnatal pulmonary function. For effusions that are large and/or increasing in size, therapies have included percutaneous ultrasound-guided thoracenteses and thoracoamniotic shunts. The first cases of fetal intervention for pleural effusion were published in 1982. In 1992, Weber and Philipson (173) conducted an extensive review of 124 cases of fetal pleural effusions. They noted that poor outcomes were significantly associated with three risk factors: (1) <32 week gestational age at delivery, (2) presence of hydrops, and (3) no antenatal therapy. This same review found a mortality rate of 50% with no antenatal therapy, 42% with thoracentesis, and 22% with a shunt.

In another review, Hagay et al. (174) looked at all cases of fetal pleural effusion without hydrops at initial diagnosis. Of 82 cases reviewed, 54 did not undergo antenatal intervention and had a mortality rate of 37%, while fetuses that did undergo prenatal intervention had a comparable rate of 33%. In 1997, Wilkins-Haug and Doubilet (175) studied the treatment of unilateral pleural effusions. Both hydropic and nonhydropic fetuses had good outcomes with thoracoamniotic shunting. Those treated conservatively (all without hydrops) resulted in death in three out of ten.

In 1998, Aubard et al. (172) comprehensively reviewed 204 cases of primary fetal hydrothorax. The mortality rates with and without antenatal treatment were 57% and 78%, respectively. Those fetuses with hydrops had a 23.5% chance of survival with no treatment, a 10% survival with only thoracentesis, and a 66.6% survival with shunting. Without hydrops, fetuses had a 21.3% survival without treatment, a 60% survival with thoracentesis, and a 100% survival with shunting. On the basis of their findings, Aubard recommended a management algorithm for treatment of congenital hydrothorax. As most pleural effusions rapidly reaccumulate fluid, the invesitigators recommended thoracentesis only in fetus with acute signs of distress, in late gestation fetuses (>32 weeks), or in fetuses just prior to birth to optimize postnatal respiratory function. A shunt should be placed for fetuses that progressively worsen and are under 32 week's gestation or have failed thoracentesis. Aubard noted complications associated with thoracoamniotic shunts including shunt failure (11/80) and catheter migration/obstruction (10/80).

8. CONGENITAL HEART DEFECTS

Treatment of complex congenital heart defects (CHDs) and arrythmias is an exciting and promising new frontier in fetal surgery. CHDs are the most common congenital malformations affecting 8 in 1000 live births. Up to 20% of perinatal mortality from congenital malformations (176) and half of the deaths in childhood caused by malformations are due to CHDs (177,178). Many CHDs are successfully treated postnatally with good long-term outcomes. Unfortunately, some are impossible to correct at the time of birth and surgery is only palliative. These CHDs may be important targets for *in utero* treatment. Particular attention has been paid to severe aortic and pulmonary valve obstructions both of which can lead to severe dysfunction and malformation of the affected ventricle. The promise of *in utero* therapy is to treat the obstruction early enough in the disease process to prevent the ensuing ventricular hypoplasia. Several studies have reported on the progression of hypoplasia due to valvular stenosis (179,180).

Current standards for prenatal screening result in the majority of serious CHDs going undetected. The standard "four chamber" view by echocardiography, even with

additional views of the outflow tracts, identifies only gross structural abnormalities such as hypoplasia or vessel defects (181). Only real-time pulse-wave and color Doppler evaluation can detect valve stenosis and early degrees of obstruction prior to the evolution of hypoplasia (182), and these modalities are not used routinely during prenatal ultrasound screening.

Another barrier to *in utero* intervention is feasible access to the fetus. Innovative animal research has been performed to develop methods of fetal access. Fetal cardiac bypass has been studied in fetal sheep (183,184) and has been shown to be feasible after surmounting issues related to perturbation of placental circulatory homeostasis. Endoscopic techniques and advanced intra-amniotic imaging have been developed by Kohl (185–188) to perform balloon valvuloplasties in sheep through umbilical vessel access.

In human fetuses, the most common MAFS procedures for cardiac intervention have been ultrasound-guided balloon valvuloplasty in cases of severe aortic stenosis and pulmomary stenoses. In these cases, a needle is advanced percutaneously with ultrasound guidance through the maternal abdomen, into the amniotic sac and then through the fetal chest wall. It is placed into the obstructed ventricle, and then a coronary artery balloon catheter is advanced over a guidewire. Maxwell et al. (189) reported the first two balloon valvuloplasties in humans. A decade later, a total of 12 cases of aortic balloon valvuloplasties have been performed and reviewed by Kohl et al. (190). The outcomes of this initial clinical experience were poor. The mean gestational age at detection was 25.7 weeks and at the time of intervention was 29.2 weeks. Success was determined by echocardiographic relief of obstruction following the procedure. Seven fetuses (all without atresia) had successful valvuloplasties although only one survived. Of the five technical failures, only one survived with postnatal surgery. There was no PTL. The authors attributed the poor results primarily to the severity of cases, technical problems, high postnatal mortality, and complications ranging from fetal bradycardia and bleeding to difficulties with catheter introduction and withdrawal. The child who survived after successful valvuloplasty was doing very well at age four with trace mitral and aortic valve regurgitation, and an ejection fraction of 55% (191).

Recently, two cases of pulmonary valvuloplasty were reported by Tulzer et al. (192). Both fetuses (28 and 30 weeks gestation) had severe pulmonary valve obstruction (one, stenotic, and the other, atretic) and imminent hydrops. Both demonstrated more favorable results immediately postoperatively with decreased circulatory failure demonstrated by resolution of pericardial effusions and improved RV function. Both delivered near term gestation and required postnatal valvuloplasties and placement of systemic-to-pulmonary arterial shunts. All published balloon valvuloplasties to date are summarized in Table 5.13.

At the 2004 meeting of the Society of Maternal–Fetal Medicine, 15 unpublished cases of aortic balloon valvuloplasties were presented by a group from the Brigham and Women's Hospital and Children's Hospital of Boston (193). All were treated prior to 28 weeks gestation. The group's technical success rate was 66% (10/15) with a combined endoscopic/percutaneous approach (9/11) being more successful than a solely percutaneous approach (1/4). The only maternal complication was pulmonary edema requiring diuretics and supplemental oxygen. Given the minimal risk to the mother and the low risk of PTL, fetal balloon valvuloplasties may become more efficacious with better patient selection and improved technical methods.

Another potential target for *in utero* intervention is the pericardial teratoma. Pericardial teratomas are extremely rare neoplasms. They are usually large, multicystic, have associated pericardial effusions, and frequently cause cardiorespiratory failure due to tamponade or obstruction to blood flow (194). There have been multiple reports of fetal

Table 5.13 Fetal Balloon Valvuloplasties (190,192)

Case	Diagnosis	Gest age at diagnosis	Gest age at operation	Gest age at delivery	Technical success	Survival	Cause of death	Other
1	AV stenosis	30	31 33	34	Y	C	LV dysfunction	Balloon rupture Intermittent bradycardias
2	AV stenosis	32	33	38	Y	E		Balloon rupture Intermittent bradycardias
3	AV stenosis	30	30	35	Y	C	LV dysfunction	Balloon rupture Intermittent bradycardias
4	AV stenosis	29	32	32	Y	A	Sustained bradycardia	Balloon rupture Intermittent bradycardias Hemopericardium
5	AV stenosis	21	30	30	N	E		Balloon rupture Intermittent bradycardias
6	AV stenosis	24	26	37	Y	C	Postnatal post-op	
7	AV stenosis	26	28	38	Y	C	Postnatal post-op	
8	AV stenosis	27	27	38	Y	C	Postnatal intra-op	Balloon rupture
9	AV atresia	22	28	28	N	A	Hemothorax	Intermittent bradycardias Hemothorax
10	AV atresia	20	29	30	N	A	Chorioamnionitis	Intermittent bradycardias
11	AV stenosis with PV stenosis	24	27	27	N	A	Sustained bradycardia	Sustained bradycardia
12	AV stenosis with PV atresia	24	28	28	N	A	Sustained bradycardia	Sustained bradycardia Hemopericardium
13	PV atresia		28	38	Y	E		
14	PV stenosis		30	35	Y	E		

Note: A, Died intra-operatively; B, Died *in utero* but not intra-operatively; C, Died within 30 days of birth; D, Died after 30 days old; E, Long term survival; AV, aortic valve; LV, left ventricle; PV, pulmonic valve.

pericardiocentesis to stabilize fetuses at high risk for tamponade (194–198). There has been one attempt reported of *in utero* resection of a pericardial teratoma (199). A 24-week gestation hydropic fetus underwent open fetal surgery for teratoma resection. The tumor was removed, but the hydrops did not resolve. Three weeks later, the mother experienced severe pre-eclampsia which was deemed maternal mirror syndrome. Therefore, emergency C-section was performed, but the baby died immediately after birth.

Fetal cardiac arrythmias have also been treated by *in-utero* therapy. Fetuses with complete heart block have been treated unsuccessfully with ventricular pacemaker placement, both by open approach and by catheter approach (200–202). Maternal dexamethasone treatment has been shown to have some efficacy in fetuses with complete heart block (203).

9. PREMATURE RUPTURE OF MEMBRANES

One of the consistent complications following fetal interventions of any kind includes PROM. Iatrogenic PROM occurs in ~1.2% after amniocentesis, 3–5% after diagnostic fetoscopy, and 5–8% after operative fetoscopy (204). The perinatal mortality of iatrogenic and spontaneous PROM managed expectantly is 60% (205,206).

On the basis of the initial idea of a blood patch to treat spinal headache by Gormley (207) in 1960, Quintero successfully treated iatrogenic PROM with an intra-amniotic injection of platelets and cryoprecipitate (the "amniopatch") (208). The patient had been leaking amniotic fluid beginning the fourth postoperative day after fetoscopic umbilical cord ligation. She was expectantly managed for 3 weeks with continued leakage and reduction of amniotic fluid on ultrasound. One unit of platelets and one unit of cryoprecipitate were administered under ultrasound guidance. The leak stopped and the pregnancy continued with delivery at term. Another group reported success using whole blood to treat PROM in a patient after genetic screening amniocentesis (209).

In 2003, Quintero (210) reported a series of 28 patients treated with amniopatch. Patients with iatrogenic PROM between 16 and 18 weeks' gestation without evidence of infection were placed on bedrest and antibiotics for 1 week. If there were no spontaneous sealing of the membranes, autologous (if possible) platelets followed by cryoprecipitate were administered via amniocentesis into the largest amniotic fluid pocket. Initially one unit of each was administered, but subsequently one half unit of platelets was given with good results. This was done because some of the early fetal deaths were probably due to activation of a large number of platelets. Eleven had a large membrane detachment but no detectable leak, while the remaining seventeen had a gross leak. Amniocentesis was responsible for PROM in 10 (36%) patients. The average gestational age at delivery was 33.4 weeks. Membrane sealing occurred in 19 patients (67.9%).

Quintero et al. (210) then attempted to use the technique on women with spontaneous PROM. The first 12 patients did not respond to the treatment and continued to leak. To learn more about the membranes in spontaneous PROM, four women were studied endoscopically while undergoing the amniopatch procedure. These cases indicated that the membrane defect is usually located over the internal cervical os vs. elsewhere with iatrogenic PROM. This might be explained by the cervix failing to protect the membranes from ascending infection and/or gravity concentrating inflammatory agents in the lower portion of the membranes, thus weakening them. The membrane defect became larger with more time between PROM and intervention, and the edges became more rolled and less sharp. Perhaps intervention could be more successful in the early stages of spontaneous PROM.

In vitro and *in vivo* experiments with rabbits and sheep have demonstrated the feasibility of using an Nd:YAG laser to weld a collagen-based patch over the amniotic

membrane defect (210,211). The first human spontaneous PROM repair with this method was described in 2002 (212). PROM occurred at 16.5 weeks' gestation and severe oligohydramnios developed 2 weeks later. The fetus had a normal karyotype so that the "amniograft" procedure was performed. The patient did not leak for 2 weeks, but then had a recurrent leak. At this point, it was managed expectantly and the baby was delivered at 32 weeks by Cesarean section.

10. AMNIOTIC BAND SYNDROME

Amniotic band syndrome affects 1 in 1200 to 1 in 15,000 live births per year (213). It is a collection of acquired congenital malformations involving the limbs, craniofacial region, and trunk resulting from fibrous bands attached to the fetus causing constriction and deformation. These bands are thought to arise from rupture of the amnion and can result in digit, or limb amputations, facial clefting, and even death. Umbilical cord compromise by amniotic bands arising after uterine instrumentation can be fatal and may be targeted for intra-uterine release (214). Data from a sheep model of amniotic band syndrome suggest that fetoscopic release of these bands may prevent limb deformities (215,216).

The human experience in amniotic band release is limited to four cases (Table 5.14). Quintero et al. (217) performed the first fetoscopic release of amniotic bands in two patients. The first fetus was at 21 weeks gestation with evidence on ultrasound of a band constricting her left upper extremity causing abnormal deviation and distal edema. She was deemed at risk of arm amputation. The fetus had bilateral cleft lip and normal karyotype. It was planned to cut the band with scissors under fetoscopic guidance, but when uterine bleeding caused the second port to be removed, ultrasound guidance was used. Band release resulted in an immediate improvement in the angulation of the arm, and the edema improved over time. The second case involved a fetus with a band constricting the ankle. A YAG laser fiber was used to disrupt the band. There was marked improvement to the foot. It developed normally and full functional recovery was expected.

In 2003, Keswani et al. (218) attempted fetoscopic release of amniotic bands on two patients with isolated limb constrictions. One fetus had a circumferential band of the right wrist which was released with an Nd:YAG laser. The wrist recovered normal blood flow and was viable but suffered from secondary lymphedema. The second case involved bands to the right upper and lower extremities released with laser. Both extremities were viable at birth, but the upper extremity was atrophic and subsequently amputated for prosthesis compatibility.

11. GASTROSCHISIS

Gastroschisis is a condition affecting 1 out of 10,000 live births (219). It is a condition in which the bowel remains exteriorized through a paraumbilical defect. After delivery, the bowel must be surgically replaced into the abdomen. Survival rate is 90%, but at least 25% of the babies suffer from perivisceritis. These babies, typically, have a prolonged stay in the NICU and hospitalization averaging 20–80 days with a delay to enteral feeding of 25 days (220).

Animal models of gastroschisis have led to the proposal that gastrointestinal waste in the amniotic fluid can contribute to the intestinal lesions of the fetus with gastroschisis (221). In 1998, Aktug et al. (222,223) demonstrated that exchanging the amniotic fluid with saline or saline plus dextrose decreased intestinal lesions in a chick gastroschisis model. Improvement was also seen with amnioexchange in a lamb model (224).

Table 5.14 Experience With Fetoscopic Release of Amniotic Bands (217,218)

GA	Lesion (US)	Technique	GA at delivery/outcome
22	Bil cleft lip, bands to face and left arm, left forearm edema, arterial blood flow ok	Initially two ports but due to bleeding second port removed, scissors used under ultrasound guidance	39 week: Bil cleft lip, craniofacial cleft, right microphthalmia, left arm—minimal scarring, radial paresis and mild hypoplasia
23	Band constricting left ankle, distal edema, minimal blood flow	Two ports–one port due to bleeding, scissors attempted—no success, YAG laser	34.5 week: PROM at 31 weeks, z-plasties of amniotic band remnants, expected full functional recovery of foot
23	Circumferential band around left wrist, distal edema, compromised blood flow via single artery	600-micron endostat with Nd:YAG laser	33 week: Normal blood flow to left wrist but persistent lymphedema. Viable hand but limited range of motion secondary to scarring from reconstruction for lymphedema.
20	Circumferential band around right wrist, distal edema, compromised perfusion. Intraoperative findings also included bands around right calf and right thigh.	400-micron endostat with Nd:YAG laser	Atrophic, malformed, viable right hand—later amputated for prosthesis. Normal right lower extremity.

Note: GA, gestational age; US, ultrasound; Nd:YAG, neodymium-yttrium aluminum gamet; PROM, premature rupture of membranes.

The first case of human amnioexchange was reported in 1998 by Aktug et al. (225). Four amnioexchanges were performed by an ultrasound-guided percutaneous approach for a fetus with a gastroschisis. After delivery, the baby's abdomen was closed primarily with low intra-abdominal pressure, was fed by day 5, and discharged by day 8.

Luton et al. (226) compared 10 fetuses that underwent amnioinfusion with 10 who did not. Their data suggest that amnioinfusion decreased inflammation of the bowel and allowed for immediate and easier primary closure of the defect. The treated babies required less days of ventilation and hospitalization and less time to enteral feeds. These data were not statistically significant, but suggest a trend that may or may not be substantiated as more cases are performed in a prospective and randomized manner.

12. POTENTIAL FUTURE APPLICATIONS OF MINIMAL ACCESS FETAL SURGICAL TECHNIQUE

Until very recently, fetal surgery has been a treatment option only for fetuses with other lethal conditions. However, as advances in fetal minimal access technique evolve, a wider

variety of maladies, including nonlethal, but highly morbid conditions may become more appropriate target lesions. Just as advances in prenatal imaging provided a better understanding of the natural history and pathophysiology of the full spectrum of current fetal anatomic lesions, many advances in the molecular understanding of disease and our ability to detect them earlier may lead to a variety of *in utero* cell and gene therapy correction strategies. With the cataloging of the human genome leading to better genetic characterization of disease, advances in DNA microarray technology, new methods for obtaining fetal DNA, and the ability to diagnose virtually all human genetic disease prior to birth may be within grasp. These technologies taken together may thus establish *in utero* molecular manipulation as a treatment option. The advantages of an antenatal treatment of genetic disease may exploit the dissemination effect of either stem cell or gene expansion in the highly proliferative tissues during fetal development (227).

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6
The Role of Minimal Access Surgery in Pediatric Trauma

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1. INTRODUCTION

Multiple diagnostic modalities exist to ensure the prompt and accurate assessment of the injured patient. In abdominal trauma, these include physical examination, computed tomography (CT), ultrasound, and diagnostic peritoneal lavage (DPL). The use and sequence of these methods depend on the patient's hemodynamic status, the mechanism of injury (blunt or penetrating), and the potential for significant associated injuries. Although each modality has added to the quality of care in trauma patients, the incidence of negative and nontherapeutic laparotomies continues to range between 11 and 34% (1,2). Minimal access surgery (MAS) may lead to prompt diagnosis and therapy, thus decreasing the incidence of unnecessary laparotomies, length of hospital stay, overall costs, and unnecessary morbidity to the patient.

Patients who present to the emergency department with hemodynamic instability or clear evidence of significant intra-abdominal injury merit urgent laparotomy. However, the need for exploratory surgery in the stable patient following either blunt or penetrating injury remains unclear. Mandatory laparotomy for penetrating trauma leads to a negative laparotomy rate of up to 37% (1,3). This results from the fact that many stab wounds never penetrate the peritoneum and 20% of gunshot wounds are tangential, without intra-abdominal injury. In hemodynamically stable patients, local exploration of stab

wounds revealing fascial penetration leads to a 50% negative laparotomy rate (4); whereas, positive DPL leads to unnecessary laparotomy rates of 20–37% (5), the use of abdominal CT following stab wounds has helped to reduce the nontherapuetic laparotomy rate of 18% (4).

In blunt abdominal trauma, DPL has been the standard for diagnosis for more than three decades. DPL is a sensitive, safe, and inexpensive procedure, which can be done promptly in the emergency department under local anesthesia. The accuracy of DPL >95%. However, this high sensitivity has led to nontherapeutic laparotomies following blunt trauma in 13–27% of cases (5). Many positive lavages reflect minor bleeding from the liver or spleen, which could be managed nonoperatively. This is particularly true in children, in whom nonoperative management of solid organ injury, even in the presence of free intraperitoneal blood, is often successful. In addition, DPL is unreliable in diagnosing injuries to the diaphragm or retroperitoneum. Abdominal CT offers the advantage of specific anatomic definition of the site of bleeding, as well as a clear view of the retroperitoneum. However, visualization of injuries to the diaphragm, hollow viscera, and pancreas remains a limitation.

2. ROLE OF MINIMAL ACCESS SURGERY

The precise role of MAS in trauma has yet to be defined. Prompt and accurate MAS could contribute significantly to the care of trauma patients by decreasing the rate of negative and nontherapeutic laparotomies (i.e., operations where the injuries identified can be managed equally well nonoperatively). Another role for MAS is in the diagnosis of injuries to the diaphragm and hollow viscera, organs traditionally difficult to assess without open exploratory surgery. Finally, as instrumentation improves and experience accumulates, the role of MAS could evolve not only as a diagnostic tool, but also as a potential therapeutic modality. Multiple studies have been reported by examining the role of MAS in the workup of the trauma patient. Herein, we review several large prospective studies evaluating MAS in both blunt and penetrating trauma victims. Note that all of these studies have involved adult trauma patients. The literature concerning MAS for pediatric trauma is limited to small case reports, which will be summarized briefly.

In 1993, Fabian et al. (6) reported a prospective analysis of 182 abdominal trauma patients; 90% of whom suffered penetrating wounds. Hemodynamically stable stab wound patients with violation of the anterior fascia by local wound exploration were candidates for study, as were individuals with gunshot wounds thought to be tangential to the abdominal wall. These patients would traditionally have undergone laparotomy, but first underwent MAS. Laparotomy was avoided in 55% of the penetrating injury patients by identifying no peritoneal violation by laparoscopy. Those undergoing negative MAS had significantly fewer complications and shorter hospital stays than a historical cohort following negative laparotomy.

In another prospective study, Salvino et al. (7) compared the use of MAS versus DPL in 75 patients with blunt or penetrating trauma and found MAS to be especially useful in the assessment of stab wounds. Diaphragm lacerations were diagnosed laparoscopically in three patients with normal DPL findings. These patients were taken to laparotomy for repair of the diaphragm. Moreover, MAS was successfully performed under local anesthesia in the emergency department in 93% of the patients, significantly reducing the time and resources required for the procedure. Among the blunt trauma patients, MAS offered no advantage over DPL as a primary assessment tool.

In 1995, Carey et al. (2) reported on 35 blunt and penetrating trauma patients, all meeting criteria for exploratory laparotomy. In this group, MAS had 100% sensitivity and 88% specificity in determining the need for therapeutic laparotomy. Nontherapeutic laparotomy was avoided in 68% of the patients. Another prospective study by Brandt et al. (4) showed that MAS was 100% accurate in determining the need for therapeutic laparotomy following blunt or penetrating trauma in 21 patients. Sosa et al. (8) prospectively studied 121 patients with gunshot wounds and no obvious signs of peritoneal penetration and found that MAS had 100% sensitivity and 99% specificity in determining peritoneal penetration and need for laparotomy. Most importantly, MAS had a 100% negative predictive value; there were no missed injuries.

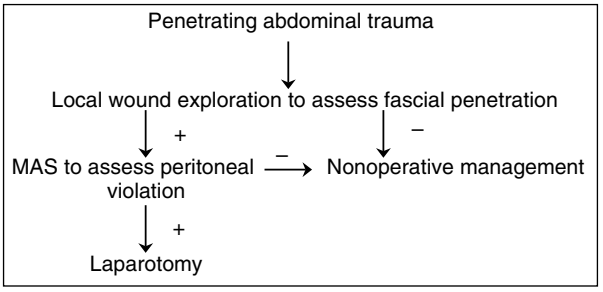
In a collective review of 11 reports totaling 355 blunt injury patients, the sensitivity and specificity of MAS for predicting the need for therapeutic laparotomy was 94% and 98%, respectively (9). Following positive DPL or CT scan, MAS was especially useful in reducing nontherapeutic laparotomies by establishing whether bleeding had stopped or solid organ injury could be managed nonoperatively.

The enthusiasm and early success of MAS in abdominal trauma patients must be balanced by the risk of missed injuries. Elliott et al. (10) prospectively studied 47 trauma patients, all undergoing MAS prior to planned laparotomy. Laparoscopy had 96% sensitivity and 100% specificity in determining the need for laparotomy; however, only 57% of the injuries found at laparotomy were seen through the laparoscope. The majority of missed injuries were to hollow viscera. This study confirms the value of MAS in determining the need for open surgery, but raises concerns about using laparoscopy alone as a therapeutic modality.

Several groups have reported their experience with MAS in small numbers of injured children. Chen et al. (11) used laparoscopy to evaluate six children (two blunt and four penetrating injuries) and thoracoscopy in two children. The authors accurately identified all injuries using MAS, avoided laparotomy in four patients, and successfully repaired diaphragm lacerations in two children. There were no complications. VanderKolk et al. (12) performed laparoscopy in four children with suspected seat belt injury, on the basis of CT findings of free fluid or mesenteric thickening without solid organ injury. Intestinal perforation is one of the injuries associated with lap belt injury, and one for which CT scan is unreliable. These authors identified contusions of the intestine, gastric perforation, and mesenteric laceration treated with laparoscopic clips. They reported no missed injuries and no morbidity with the use of MAS. One case report describes the successful use of MAS for identification and repair of a jejunal perforation in a 4-year-old with a lap belt injury (13). Hasegawa et al. (14) reported the feasibility and safety of MAS in five children with persistent abdominal pain after blunt trauma. They successfully diagnosed injuries to the duodenum, pancreas, and spleen in three patients who subsequently underwent therapeutic laparotomy.

Laparoscopy has also proven to have a role in the identification of suspected injuries to the diaphragm in both penetrating and blunt trauma (7,15,16). In a prospective study of 75 trauma patients evaluated by both DPL and MAS, 23 patients had negative DPL results. Of these, 20 were successfully managed nonsurgically and three had surgery for diaphragmatic lacerations identified by MAS and missed by DPL (7). Diaphragmatic lacerations are notoriously difficult to ruleout with conventional imaging. When clinical suspicion of an injury is high, laparoscopy is an excellent tool for definitive diagnosis.

Several conclusions about the role of MAS in trauma can be reached, on the basis of the studies discussed earlier (Fig. 6.1). Laparoscopy can play a significant role in reducing the rate of unnecessary laparotomies. In patients with penetrating injuries to the abdomen, mandatory laparotomy for fascial penetration is no longer necessary. Diagnostic MAS can



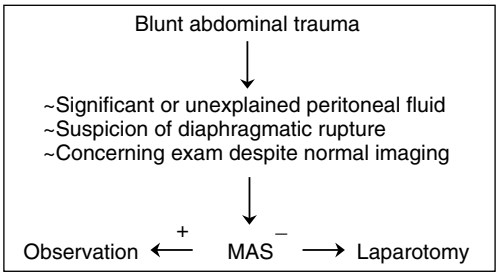


Figure 6.1 Algorithm for the use of MAS in the assessment of the hemodynamically stable pediatric trauma patient.

be used as a primary method to accurately identify peritoneal penetration and associated intra-abdominal injury, thereby accurately predicting the need for therapeutic laparotomy. For blunt trauma victims, such as hemodynamically stable children following lap belt injuries, or other crush injury to the abdomen with positive findings on DPL and/or CT scan, laparoscopy can serve as an important diagnostic adjunct prior to planned laparotomy. This algorithm may diminish the incidence of nontherapeutic laparotomies by identifying patients with solid organ injuries that can be safely managed without operation and also accurately diagnosing hollow visceral injury.

The question of whether laparoscopy can be used in the treatment of traumatic injuries remains unanswered. To be used therapeutically, MAS must be capable of diagnosing all injuries needing repair. However, MAS is reported to have low sensitivity ($<50\%$) for the identification of perforation of hollow viscera (10). This sensitivity will vary greatly among surgeons, depending on individual experience with running the bowel laparoscopically. In addition, the use of higher resolution video equipment and newer instrumentation will facilitate a more thorough and reliable look at all the viscera. At the present time, aside from the occasional case report, there are no data available to support the routine use of therapeutic laparoscopy in trauma.

3. TECHNIQUE OF TRAUMA LAPAROSCOPY

The child is positioned supine and belted securely to the bed. Following placement of nasogastric and urinary catheters, a 3–5 mm trocar is placed transumbilically using either the closed or open technique. The abdomen is then insufflated with carbon dioxide to a pressure of 8–15 mm Hg, depending on the size of the patient. At this point, close communication with the anesthesiologist is critical, as both hemodynamic compromise and tension pneumothorax or pneumomediastinum may occur. If hemodynamic instability develops,

the insufflation pressure can be decreased and volume status re-assessed. Rarely, a thoracostomy tube may be necessary or conversion to an open procedure.

Pneumoperitoneum is well tolerated in healthy subjects, carbon dioxide insufflation in hemodynamically compromised trauma patients can cause vascular compression with metabolic acidosis, increased airway pressures, hypoxemia, and respiratory acidosis. The metabolic and hemodynamic impact of pneumoperitoneum was recently studied in a controlled hemorrhage rat model (17). As the amount of hemorrhage increased, the safe pneumoperitoneum pressure decreased. Excessive pressures led to metabolic acidosis and hypotension. The trauma surgeon and anesthesiologist must be aware of the potential ill effects of CO_2 pneumoperitoneum in these patients.

A 3, 5, or 10 mm 30° laparoscope is then inserted and a thorough inspection of the peritoneal cavity performed. The parietal peritoneum is examined for any signs of penetrating injury. Note should be made of free fluid (blood, succus entericus, bile) in the peritoneum, especially in the pelvis and paracolic gutters. Intraperitoneal blood needs to be thoroughly cleared to avoid missing any significant injuries. Two additional ports are placed under direct vision in the right and left mid-abdomen for use of a suction/irrigation device, retractors, and atraumatic graspers to examine the bowel. A systematic inspection of all the abdominal contents is then undertaken. Sequentially, rotating the bed from Trendelenburg to reverse Trendelenburg, and from right to left, is critical to allow all quadrants to be thoroughly inspected. The posterior surface of the diaphragm can be hard to visualize, although steep reverse Trendelenburg with use of a 30° scope can facilitate this. The lesser sac is accessed via a clear area through the gastrocolic ligament. If an entrance wound suggests possible retroperitoneal injury, then the colon is mobilized and rotated medially for visualization. Thorough exploration of the entire small bowel, the posterior surface of the colon, and the retroperitoneal organs can be difficult, with the potential for missed injuries in these areas (10).

One should bear in mind during MAS that the goal is to determine the need for laparotomy. Therefore, the presence of significant ongoing bleeding, intraperitoneal intestinal contents, or any injury requiring laparotomy eliminates the need for further laparoscopic inspection (9). There is currently no prospective evidence to support the safety of therapeutic laparoscopic procedures in pediatric trauma, although technology and expertise are accumulating.

4. THE ROLE OF THORACOSCOPY IN THORACIC TRAUMA

Thoracic injury directly accounts for 25% of trauma-related deaths and is second only to head injury in causing pediatric trauma deaths (18). Although the indications for emergency thoracotomy in adults are clear, hemodynamically stable patients who are initially observed may have significant sequelae of missed injuries. Similarly, most children with thoracic trauma do not need thoracotomy; yet earlier diagnosis and treatment of those patients with significant injury would improve outcome. Much as the role of video-assisted thoracic surgery (VATS) is continually expanding in elective surgical conditions so too is it the role of VATS in the diagnosis and treatment of the trauma patient. Graeber and Jones (19) have summarized the details of VATS equipment and techniques in a recent review.

The management of patients with penetrating thoracic wounds who are hemodynamically stable may be subjective and imprecise. Jones et al. (20) re-introduced thoracoscopy for the evaluation of stable patients with penetrating thoracic wounds and persistent hemorrhage. Thirty-six patients had rigid thoracoscopy in the operating room

using local anesthesia. The authors reported that management of 44% of their patients was modified as a result of the thoracoscopic findings.

The widespread use of CT has better defined thoracic fluid collections that frequently persist after trauma. Yet, the indications for evacuation of these persistent collections are unclear. Heniford et al. (21) recently reported the use of VATS in 25 patients (15 penetrating and 10 blunt) with a retained thoracic collection defined as any persistent intrathoracic material, infected or not, that was unable to be drained by a chest tube within 72 h. The findings included hemothorax in 19 patients and empyema in six. Patients were more likely to have sterile hemothoraces and shorter hospital stay, if VATS evacuation was performed within 7 days of injury. Patients with delayed VATS were more likely to have empyema and require conversion to open thoracotomy for adequate drainage and decortication. Sosa et al. (22) reported an 89% success rate (24 of 27 patients) in evacuating residual hemothorax or empyema after injury.

Villavincencio et al. (23) reviewed 28 studies with a combined total of more than 500 patients undergoing VATS after trauma. Diagnostic VATS was used primarily for continued chest tube bleeding, suspected pericardial penetration, and evaluation of the hemidiaphragm. Thoracoscopy was found to be accurate in 98% (188 of 191 patients) patients for diagnosis of diaphragm injury. Prospective studies evaluating patients with anterior thoracoabdominal wounds have confirmed the accuracy of thoracoscopy in identifying diaphragm injury. The therapeutic indications for VATS included control of persistent chest tube bleeding and evacuation of retained intrathoracic collections. The success rate of VATS was 90% (89 of 99 patients) for evacuation of hemothorax, 86% (19 of 22 patients) in the evacuation of empyemas, and 82% (33 of 40 patients) in controlling chest tube hemorrhage. The authors concluded that unnecessary thoracotomy or celiotomy was avoided in 323 of 514 (62%) patients because of VATS findings and interventions. Risks included procedure-related complications in 11 of 514 (2%) patients and a missed injury rate of 0.8% (four of 471 patients).

Uribe et al. (24) prospectively evaluated the benefit of thoracoscopy in 28 patients with penetrating thoracoabdominal wounds. All patients were hemodynamically stable, had no indications for emergency celiotomy, and demonstrated thoracic injury on chest radiography or physical examination. All thoracoscopy procedures were performed in the operating room under general anesthesia. Diaphragmatic injury was confirmed in nine (32%) of 28 patients, and all were repaired at celiotomy. Eight of these nine patients had associated intra-abdominal injuries requiring repair including one colonic and three gastric injuries. These patients had prompt abdominal exploration because of the thoracoscopic findings. In addition, 19 patients were spared exploratory celiotomy. There were no procedure-related complications. Thoracoscopy appears to be particularly useful in patients treated at centers that mandate celiotomy for penetrating thoracoabdominal wounds. Our recommendation includes initial observation and proper monitoring of asymptomatic patients with thoracoabdominal wounds followed by VATS in those at high risk for diaphragm penetration, if issues such as a full stomach or alcohol/drugs would be less likely to complicate the anesthetic/surgical procedure. Oschner et al. (25), in a prospective evaluation of 14 patients with penetrating thoracoabdominal trauma, also found thoracoscopy to be very sensitive and specific in the diagnosis of diaphragmatic injury.

5. SUMMARY

Trauma remains the leading cause of death in children. An expeditious and accurate assessment of a patient's injuries is critical for ensuring the best possible outcome.

MAS has been shown to be a safe, effective, and potentially cost-saving means of evaluating abdominal and thoracic trauma with the potential to decrease the incidence of unnecessary open explorations and to minimize the occurrence of delayed diagnoses.

The American Pediatric Surgical Association (APSA) Center for Outcomes and Clinical Trials has established a Trauma Study Group including more than 50 pediatric trauma centers. A multi-center, prospective trial concerning the role of MAS in trauma coordinated by APSA would be ideal in answering specific questions in an expeditious manner and could attract extramural funding.

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7
Minimal Access Surgery for Pediatric Cancer

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1. INTRODUCTION

The use of thoracoscopic and laparoscopic procedures in children has been broadened to include their use in the diagnosis and treatment of malignancies. In adult surgery, the use of minimal access surgery (MAS) is included in almost all malignancies. In pediatric malignancies, solid viscus tumors are more common than hollow viscus tumors. The size of solid viscus malignancies in children limits the use of therapeutic MAS. Therefore, minimally invasive diagnostic procedures are more common in children than therapeutic procedures for malignancy.

Current diagnostic and therapeutic procedures include:

1. thoracoscopic lung biopsies,
2. thoracoscopic mediastinal biopsies,
3. laparoscopic biopsy for diagnosis,
4. laparoscopic exploration for a "second look",
5. laparoscopic oophoropexy,
6. laparoscopic adrenalectomy, and
7. laparoscopic retroperitoneal lymph node sampling.

Although there may be other topics which may be relevant to the role of MAS in children with cancer, it was the authors' intent to limit the scope to this chapter to those topics where evidence-based information existed in the English literature.

Prior to considering MAS for pediatric cancer, the potential short-term and long-term benefits must be reviewed. The short-term benefits are similar to many procedures in MAS (e.g., less pain, shorter recovery period, etc.). In cancer, however, it is the long-term benefits that are more important, specifically recurrence rates and survival. There has been a significant amount of basic science that has been carried out to investigate the role of MAS and its potential to either positively or negatively alter the outcome of patients with cancer. It is worth reviewing this topic briefly to justify the role of MAS in cancer in children using basic science information before proceeding on with a discussion of clinical studies.

Basic science investigators have used animal models to demonstrate two major ways that surgery alters tumor growth. The first way involves observations made about the direct effects of surgery upon tumor growth. Some investigators have shown that open laparotomy is associated with accelerated tumor growth and others have demonstrated that open laparotomy results in an increased rate of metastatic tumor formation (1–5); all of these studies were compared to sham anesthesia control groups. Other investigators of examined this direct effect in rodent models of laparoscopy, focusing upon the role of CO_2 pneumoperitoneum (1,6–10). All of these investigators have demonstrated an increase in the incidence of metastases and tumor size; however, these increases were smaller than those observed with open laparotomy.

The second way involves observations made about the indirect effects of surgery upon tumor growth. Open laparotomy has been shown to decrease tumor cell apoptosis and to increase tumor cell proliferation rates (11,12). Some investigators have postulated the existence of laparotomy-related plasma soluble factors to explain these observations (13,14). It has also been noted that open surgery leads to a period of immunosuppression in animal models and human studies. Changes in immunosuppression were assessed with various measurements, including serum cytokine levels, lymphocyte proliferation rates, delayed-type hypersensitivity response rates, and neutrophil function (15–18). Changes in these levels have been similarly assessed in laparoscopic procedures. Many authors have shown significantly less immunosuppression with laparoscopy when compared with laparotomy in rodent models and human studies (19–25). In some of these studies, the changes in immunosuppression were brief and were only noted in the early postoperative period.

The findings of the direct and indirect effects of laparoscopy upon tumor growth compared to open laparotomy support the potential role of MAS in the treatment of children with cancer. There is good basic science data to suggest that MAS may confer a survival benefit upon those children needing surgery for cancer compared to those undergoing open surgical procedures. It seems, therefore, appropriate to consider the current

diagnostic and therapeutic MAS procedures in children with cancer in the remainder of this chapter.

2. THORACOSCOPIC LUNG BIOPSY

The first use of thoracoscopy in children was reported by Rodgers et al. (26). Equipment was modified for small biopsies in pediatric patients to evaluate various intrathoracic lesions and perform limited evaluations of empyema (26,27). In 1990s, more advanced procedures were performed in children (28–31). Now thoracoscopy in infants and children is becoming routine for benign and malignant lesions.

Waldhausen et al. (32) published a study which reviewed their experience with MAS and clinical decision-making with pediatric cancer patients. Forty-seven children underwent thoracoscopy for tissue diagnosis of a newly-discovered mass for the evaluation of a residual mass or new lesion found during surveillance studies. Forty of the lesions were pulmonary and seven were mediastinal. In ten of the pulmonary lesions, computed topography (CT)-guided needle localization was required as the lesions were too deep within the lung parenchyma or too small to be seen. Four of the procedures (9%) were converted to open thoracotomies because of an inability to visualize the lesion. Two of the patients (4%) had a complication including parenchymal injury during port access and a pneumothorax after chest tube removal. Importantly, there were no incorrect decisions made on the basis of the tissue from the procedures (100% accuracy). The investigators thought that the benefits of the thoracoscopic procedure were (1) alleviating the need for open thoracotomy and thus avoiding the associated morbidity of this procedure, (2) hastening of the recovery time, and (3) improved visualization of the pleural surfaces. Limitations where thoracoscopy was not useful included (1) medial-located pulmonary lesions (where CT-guided needle localization could not be used), (2) children who had uncorrectable coagulopathies, and (3) children in whom an ipsilateral peumothorax could not be created safely. Overall, thoracoscopy was felt to be accurate and safe. The study did not report any port site recurrences. In addition to obtaining a diagnosis of a suspicious pulmonary lesion with thoracoscopy, a percutaneous approach can be used. Lesions, which are ≥ 1.5 cm in diameter, can be safely and accurately biopsied percutaneously using image guidance (33). Some investigators feel that thoracoscopic lung biopsy (TLB) in children should be used in cases in which the percutaneous biopsy has been inaccurate or when peripheral or pleural based lesions are <5 mm in diameter.

In a study from our institution (34), TLB was compared to percutaneous lung biopsy (PLB) in children. A total of 28 TLB and 35 PLB were retrospectively reviewed. Over 80% of these biopsies were done for the evaluation of possible metastatic disease. All TLB yielded adequate tissue; however, in 20% (7/35) of the PLB patients, the amount of tissue obtained was inadequate for diagnosis. In five of these patients who required repeat biopsies a diagnosis of metastatic disease was made. This is in contradistinction to the 100% accuracy rate obtained with TLB.

Eight patients (28%) required conversion to a thoracotomy after TLB was unsuccessful. Reasons for conversion included: (1) the lesions could not be identified, (2) adhesions were too dense, (3) adequate tissue was not able to be sampled, and (4) the lesions needed to be completely resected for therapeutic reasons. Although morbidity was listed as 18% in the TLB group, it pertained only to five cases of prolonged air leak, all of which resolved. No patient undergoing PLB required a chest tube or had a significant pneumothorax or hemothorax. There was no morbidity from PLB. This study re-emphasized the conclusions of Waldhausen et al. including the high accuracy of TLB and its low

complication rate. A significant weakness of the study was its retrospective status; as such, it could draw few if any conclusions in the comparison of TLB to PLB.

Other studies have examined length of hospital stay, including time requirement for chest tube drainage. Rothenberg (35) found in 88 thoracoscopies in children with benign and malignant conditions that the average hospital stay was 1.1 days coincident with length of chest tube placement. In a study by Fan et al. (36) comparing open to thoracoscopic and transbronchial biopsy in pediatric patients, they found that the duration of an indwelling chest tube in TLB was 0–30 h and the median hospital stay was 36 h. In the later study, interstitial lung disease only was included.

In summary, on the basis of these studies, a few conclusions can be made. First, thoracoascopic biopsies are accurate, have a low complication rate and appear to have a faster rate of recovery. Secondly, the initial approach to a pulmonary lesion may be with a percutaneous approach where the biopsy is for diagnostic purposes and the lesion is >5 mm in diameter. Thirdly, in the case of a percutaneous biopsy, there is a relatively high chance that a second biopsy may have to be performed to obtain a definitive diagnosis. Although thoracoscopy is effective for diagnostic purposes, its role in the therapeutic realm remains untested. Presently, there is no role for thoracoscopic surgery in the treatment of children requiring pulmonary resections for multiple metastatic lesions, including osteogenic sarcoma and peripheral nerve sheath tumors. In these tumors, it is imperative that all palpable lesions be removed. There is no means to locate these lesions such that thoracoscopy can be effective; they are often missed on CT scan imaging (37) and only noted on direct palpation. As the precise excision of these metastatic lesions will directly influence the patient's survival, they should always be addressed with a thoracotomy.

3. THORACOSCOPIC MEDIASTINAL BIOPSIES

More than 50% of all childhood lymphocytic lymphoma present with an anterior mediastinal mass (38). Greater than one-third of non-Hodgkin's lymphoma (NHL) presents with primary disease in the mediastinum (38). Thoracoscopy has been found to be very accurate in diagnosing mediastinal masses (32,39–41). Many studies confirm the diagnostic accuracy and low morbidity of thoracoscopy for diagnosis of mediastinal masses in children. In Waldhausen's series of 62 minimally invasive procedures in children, seven patients had biopsies for or resections of mediastinal masses. There were no major complications and all biopsies were diagnostic (32).

Cirino et al. (40) reviewed thoracoscopic management of mediastinal tumors in adults and children. Between 1983 and 1999, 73 patients underwent thoracoscopy for the treatment of mediastinal masses. Thirty-three were diagnostic and 40 were therapeutic. The age range was 2–81 years (11 patients were <12 years of age). The airway management of all children required bronchial blockers or double-lumen endotracheal tubes. Definitive histological diagnosis was obtained in all cases. The diagnoses obtained included NHL, Hodgkin's lymphoma, seminoma, malignant thymoma, teratoma, inoperable schwannoma, normal thymus, and lung cancer. The tumors were located throughout the mediastineum: 18 tumors (24.6%) in the anteriosuperior mediastinum, 28 tumors (38.3%) in the middle mediastinum, and 27 tumors (37.1%) in the posterior mediastinum. Four patients had superior vena caval syndrome and two were dyspneic.

Conversion to thoracotomy was necessary in nine patients: four of these necessitated thoracotomy due to technical difficulties and the other five required thoracotomy for therapeutic reasons. No patient required conversion because of bleeding. Overall, complications occurred in 9.6% of patients. No tumor implants were noted at trocar sites with a

follow-up of 80 ± 42 months (range 1–186 months); 85% of patients were satisfied with the cosmetic result. Four patients (5.4%) died within 30 days after the diagnostic procedure secondary to their primary disease.

In an earlier experience, Dmitriev and Sigal (41) reviewed thoracoscopic surgery in the management of mediastinal masses. Twenty-eight patients aged 3–71 years were assessed and treated between 1993 and 1994. Four patients had signs of vena cava compression and all procedures were performed with single-lung ventilation. Thoracoscopy was diagnostic in 14 cases and therapeutic in 14 cases. In cases of malignant lymphoma, frozen-section examination yielded definitive diagnosis in all cases. One case was converted to an open thoracotomy for therapeutic resection, when malignancy was suspected. Benign lesions were all completely resected thoracoscopically. One patient required open thoracotomy when electrosurgical damage to the aorta occurred during excision of a neuroma.

At St. Jude Children's Research Hospital, Rao (40) reviewed thoracoscopy in pediatric cancer patients. From 1991 to 1995, 64 patients underwent biopsy: 42 pulmonary masses, 11 mediastinal masses, and 11 underwent biopsy for leukemic pulmonary infiltrates. Two of eleven patients who underwent mediastinal biopsy required conversion to an open procedure because of insufficient tissue. Overall, 90% of patient diagnoses were successful, although 11 patients required conversion to an open procedure.

In the pediatric patients in these series, thoracoscopic biopsy of mediastinal masses was safe and effective (32,38,40,41). Thoracoscopic mediastinal biopsy is contraindicated, however, in children in whom carinal and subcarinal compression from the anterior mediastinal mass can be demonstrated. For these situations, percutaneous biopsy with local anesthesia only or 24 h of intravenous steroids are recommended (42–45). Alternatively, a Chamberlain procedure in a semiupright position under local anesthesia, with spontaneous ventilation, can be helpful in patients in whom a needle diagnosis for Hodgkin's disease (HD) is unable to be obtained (43). A single case has been reported in which radiation was administered to a limited area of the tumor before the initial biopsy was performed. A diagnosis was established on the basis of tissue from the shielded area after the main mass had decreased in size (46). To minimize morbidity and mortality in those children undergoing a biopsy of a mediastinal mass the following have been recommended: a tracheal diameter of at least 50% of normal and a peak expiratory flow rates of at least 50% of predicted value (43). Deviation from these standards may result in loss of the airway or hemodynamic collapse while undergoing induction for general anesthesia with subsequent mortality.

In reviewing the role of thoracoscopy in the diagnosis of mediastinal masses, it is important to note that there have been no studies comparing percutaneous biopsies of mediastinal masses to thoracoscopic biopsy in children. This is clearly a study that needs to be undertaken in the future.

3.1. Future Possibilities

Thoracoscopic resection of mediastinal masses in children has been limited. However, there have been reports of resection of benign neurogenic tumors of the posterior mediastinum in adults and a few children (47,48). In a study from Hong Kong, 23 patients between 1990 and 1998 underwent video-assisted thoracic surgery (VATS) for resection of posterior neurogenic tumors (48). Age ranged from 14 months to 70 years and operation time from 30 to 120 min. Four patients required conversion to open thoracotomy to allow for complete tumor resection. Tumor size ranged 0.7–13 cm in diameter. Median hospital stay was 2 days (range 1–9) and median chest tube time was 1 day (range 1–4). Seven

minor complications occurred. Diagnoses included Schwannoma (11), Neurofibroma (8), Granular cell (1), Ganglioneuroma (1), Ganglioneuroblasotoma (1), and Neuroblastoma (1). No patient had elevated hormones suspicious for malignancy.

Thoracoscopic resection of suspected benign neurogenic tumors appears to be safe and effective in adults. In the future, this modality should be considered in children in the management of benign, posterior neurogenic tumors; its safety and effectiveness will have to be confirmed by careful study.

3.2. Complications of Thoracoscopic Procedures for Malignancy

3.2.1. Port Site Recurrence

Port site recurrences after thoracoscopic procedure for malignancy have been reported in one case report of an 18-year-old (49) and two pediatric patients (51). The 18-year-old female presented with pulmonary metastasis from osteogenic sarcoma two years after local control of the tumor. Four months after a standard wedge biopsy of two pulmonary lesions, a mass at the site of the posterior thoracoscopic port was identified. Resection of this area revealed osteogenic sarcoma. In the authors' experience (in a not yet published report), two patients were treated at St. Jude Children's Research Hospital who had port site recurrences. One patient had a locally aggressive and subsequently metastatic pleuropulmonary blastoma and the other had a malignant thymoma (50). Both patients were referred after tumors had ruptured at the first thoracoscopically attempted diagnostic biopsy. Of note both of these tumors are chemo-radio-resistant and both of these patients died of metastatic disease despite very aggressive local surgical excision, chemotherapy and radiotherapy.

At thoracoscopy caution should be exercised when the mediastinal or pulmonary mass does not appear to be limited to the anteriosuperior mediastinum or does not appear typical of lymphoma. Tumors of mesenchymal origin (such as leiomyoma, rhabdomyosarcoma, extraosseous Ewing's, epitheliod sarcoma, and undifferentiated sarcoma) or thymomas may be difficult to completely resect because of invasion into critical neighboring structures (51). In the case of these tumors, the presence of microscopic residual disease may necessitate radiation therapy and/or chemotherapy (51).

3.2.2. Repeat Thoracoscopy

Thoracoscopic operations on re-operated chests have been studied in adults and children by Yim et al. (52). From 1992 to 1996, 2477 patients underwent VATS. Forty patients aged 9–78 years had prior thoracotomy (22 patients), VATS (17 patients) and one median sternotomy. Four of the twenty two patients who underwent an initial thoracotomy procedure and 12 of the 17 patients who underwent an initial VATS procedure had benign disease. Median hospital stay was 5.1 ± 3.2 days (range 0–17). There was no mortality or intraoperative complications. Two patients required conversion to thoracotomy and one patient required a 2-unit blood transfusion. Although adhesions were noted in all patients, in only two patients (5%) was it necessary to abandon the procedure. Operative techniques included avoiding old port sites and entering the pleura using a "clamp and finger" technique (as in placement of a chest drain). A pleural space was created by gentle blunt finger dissection before insertion of the port.

Of the 40 re-operative VATS procedures, only six were pulmonary wedge resections done to exclude metastasis or recurrent malignancy; of these, one required conversion to thoracotomy compared to 1 of 15 which required conversion to a thoracotomy when re-operative VATS was done for a benign condition. This suggests that re-operative

thoracoscopy done for the evaluation of recurrent pulmonary malignancy should be approached with caution. The authors concluded that VATS on re-operated chests was feasible and not associated with a higher morbidity even though it may be technically more difficult.

Results in children should be similar although this has not been well studied. Only two patients in this study (ages 9 and 15) were children; both children had repeat wedge resections for metastatic sarcoma. There have been no formal studies in the English literature examining the role of re-operative thoracoscopy in pediatric cancer.

4. LAPAROSCOPIC BIOPSY FOR DIAGNOSIS

Minimal access surgery has been found to be an ideal way to obtain biopsy specimens in children with cancer (32,39). This is the most common indication for laparoscopic operations in pediatric malignancy. Liver biopsies, kidney biopsies, and biopsies of indeterminate masses have been safely and accurately carried out in children (32,39,53). The difference in accuracy and morbidity in laparoscopic vs. percutaneous image-guided biopsy in children with malignancy has not been comparatively studied. In the study by Saenz, 93 minimal access procedures performed between 1990 and 1997 were analyzed. Laparoscopic liver biopsy (21), diagnostic laparoscopy (4), cholecystectomy (4), oophoropexy (3), and kidney biopsy (1) were included. There were two complications after laparoscopy (4%) and six patients (13%) required conversion to an open procedure (39).

Laparoscopic or open biopsy of hepatoblastoma, neuroblastoma, or Wilms' tumor using a cup forceps, despite thrombocytopenia in some patients, does not necessitate a blood transfusion (39); however, it is for this concern of potential hemorrhage that percutaneous image-guided core biopsy have been preferred for these and other intra-abdominal malignancies. A prospective study comparing these two modalities has not yet been completed but is necessary to determine any differences in complications or diagnostic accuracy between laparoscopic and percutaneous biopsy of intra-abdominal masses.

In 1990, a study compared CT lymphography and staging laparotomy in children with Hodgkin's disease and found that laparotomy affected the stage of the disease in 37% of cases (46). Laparoscopy can be used in Hodgkin's disease both before and after neoadjuvant or adjuvant therapy to assess extent of disease. The use of laparoscopy for this indication is probably limited as the quality of CT scan imaging has become so accurate in the staging of this disease (54).

5. LAPAROSCOPIC EXPLORATION FOR A SECOND-LOOK OPERATION

Peritoneal metastasis to the anterior and lateral abdominal wall can often be missed by CT and magnetic resonance imaging (MRI). As such, laparoscopy has been found to be beneficial in some second-look procedures (32,39). Waldhausen et al. describes six patients who underwent second-look operations using laparoscopy (32). Five patients had a diagnosis of lymphoma and one had a diagnosis of germ cell tumor. There are no studies that specifically examine the role of second-look operations in children with cancer. Because of this, it role remains presently undefined.

6. LAPAROSCOPIC OOPHOROPEXY

Long-term survival of patients with HD emphasizes the importance of anticipating and preventing long-term complications of the treatment of HD. Abdominal radiation in an inverted "y" field produces a high dose of radiation to the unprotected ovaries. This may result in loss of ovarian function. The severity of the loss is related to age of the patient and the amount of gonadal exposure (55,56). Loss of ovarian function may result in menopausal symptoms and is associated with osteoporosis. In prepubertal girls, secondary sexual characteristics may not develop (55,56). Oophoropexy can spare ovarian function in many cases and has been recommended since the 1960s as a means of preserving ovarian function (57).

Oophoropexy is done by placing the ovaries in the midline, preferably behind the uterus and out of the radiation field. Hemoclips are placed at the final site of the ovaries so that their position can be verified radiographically. Before laparoscopy, the advantage of oophoropexy was studied (57). Between 1970 and 1984, 17 women underwent ovarian translocation. There were six complications including salpingo-oophorectomy and ovarian cysts, and endometriosis; all procedures were done as outpatients. Eight of these seventeen became pregnant. Two of the pregnancies resulted in spontaneous abortion. Of the six near- or full-term pregnancies, five were uneventful. One mother delivered at 32 weeks gestational age.

More recently, laparoscopic oophoropexy (LO) has replaced the open technique. Williams et al. (58) reviewed the outcome of 12 patients (age 21–36) who underwent LO after pelvic irradiation for HD between 1989 and 1995. Two were excluded from analysis because of death and a second malignancy. Of 10 women who underwent LO for HD, five had evidence of normal ovarian function. Two of the five required repeat oophoropexy at 5 and 6 months. Of those five, four underwent successful pregnancies and one underwent hysterectomy for abnormal bleeding. Of the five women who had premature ovarian failure after LO, four underwent six or more cycles of chemotherapy and one received 3500 cGy boost to her pelvic primary site. Therefore in cases of advanced HD in which multiple courses of chemotherapy are required, permanent loss of ovarian function is nearly certain. In stage I or II HD, LO prior to pelvic irradiation yields excellent results in the preservation of ovarian function with minimal increased risk of recurrent HD (58).

Although many centers are performing LO in pediatric patients with HD, long-term results are not available. Presumably, results in the pediatric population will be as good or better than in adult women because of younger age of the ovaries at the time of oophoropexy.

Technically, one can sew the ovaries individually to the posterior surface of the uterus or sew them to each other posterior to the uterus. It is unclear which method results in better fixation of the ovaries. With either method, hemoclips on the ovaries at the site of the oophoropexy are necessary to monitor possible migration.

7. LAPAROSCOPIC ADRENALECTOMY

Laparoscopic adrenalectomy in children has been reported in benign and malignant diseases such as ganglioneuroma, pheochromocytoma, and neuroblastoma (59). Mirallie et al. reported a limited group of six children undergoing laparoscopic adrenalectomy for various benign diagnoses as ganglioneuroma, paraganglioma and pheochromocytoma, and malignant diseases as neuroblastoma and pheochromocytoma with lymph node involvement.

Another study specifically focused upon laparoscopic resection of early stage neuroblastomas. Yamamoto et al. (60) published a small series of children who had their neuroblastoma detected by mass screening techniques. Their selection criteria were very strict, limiting their series to only three eligible patients. All patients had (1) stage I disease, (2) a lesion which measured <20 mm in diameter, (3) slightly increased vanillylmandelic acid of homovanillic acid, and (4) normal serum markers of lactate dehydrogenase, neuron-specific enolase and ferritin. In addition, the resected specimens all had favorable pathologic features, including no N-*myc* amplification, aneuploid DNA pattern and favorable Shimada classification. There were no complications and all of the patients were alive with no evidence of recurrence at a follow-up of 17–22 months.

It is difficult to draw many conclusions about the role laparoscopic adrenalectomy in the management of adrenal malignancy based on these two small series. Specifically, the pathologic behavior of neuroblastomas detected on mass screening is unique compared to those that are noted on clinical assessment. Those detected on mass screening have a very high overall survival rate of 97%; this includes a number of tumors (27%) which are advanced-stage neuroblastomas (61). Such superior clinical results are the exception among most other neuroblastomas and not indicative of their behavior. The generalizability of laparoscopic adrenalectomy for the management of neuroblastomas cannot be made with this study; the applicability of this technique remains to be determined for those neuroblastomas that are clinically detected.

8. LAPAROSCOPIC RETROPERITONEAL LYMPH NODE SAMPLING

Juvenile yolk sac carcinoma is the most common testicular neoplasm in childhood. In adulthood, pure yolk sac tumor of the testes is very rare. The juvenile type presents before 2 years of age and demonstrates metastatic disease in $<20\%$ of cases. Metastases rarely develop only to the retroperitoneum and therefore, these infants require chemotherapy rather than retroperitoneal lymph node dissection (62).

Nonseminomatous testicular carcinoma can occur in the late teen years. In 25–30% of patients retroperitoneal metastasis are present that were not diagnosed with imaging techniques (63). In one of the largest studies, Janetschek reviewed the long-term outcome and efficacy of laparoscopic retroperitoneal lymph node dissection (LRLND) for clinical stage I nonseminomatous testicular carcinoma. Seventy-three consecutive patients between 1992 and 1999 underwent LRLND. Patient's age ranged from 15 to 51 years. Mean operative time was 228 min in the most recent 28 cases. Conversion rate to open procedure was 2.7%. Mean hospital stay was 3.3 days. In the last 44 patients, there was one minor (2.3%) and no major postoperative complications and the ability to have normal ejaculation was preserved in all patients. Mean blood loss was 156 mL (range 10–350 mL). There was one retroperitoneal relapse due to false-negative histological findings, but there were no other relapses within a follow-up of 43.3 months. This compares favorably to a relapse rate of 10% in other studies of stage I lesions done in an open fashion (63).

Other authors have reported successful LRLND in adults (64,65) but none have been reported to date in children. These reports show a shorter hospital stay, increased blood loss in the early portion of the learning curve and a conversion rate of 10% or less. There has been no increase in relapse rates seen using the laparoscopic approach although no controlled randomized studies are available in the English literature.

8.1. Complications of Laparoscopic Procedures for Malignancy

8.1.1. Port Site Recurrence

A number of studies have tried to address this question from a basic science point of view, utilizing porcine and murine models (66,67). Yamaguchi et al. (67) showed that the intraperitoneal concentration of hyaluronic acid was significantly higher after CO_2 pneumoperitoneum, compared to open laparotomy. This rise in hyaluronic acid was felt to be associated with port-site metastases. Schneider et al. (66) examined ways of minimizing the incidence of port-site metastases. They found that they could significantly reduce the incidence of these metastases by improving the quality of their surgical technique, including such protective measures as trocar fixation, prevention of gas leaks, rinsing of instruments with povidine-iodine, minilaparotomy protection, rinsing of trocars before removal, peritoneal closure, and rinsing of all wounds with povidine-iodine.

9. RANDOMIZED CLINICAL TRIALS

A limitation of all of the studies involving the clinical application of MAS for the management of children with cancer is their design. Specifically, none of them are randomized trials and therefore suffer from the innate weaknesses of nonrandomized studies. To address this concern, in 1996 multi-institutional, cooperative group, randomized trials were developed through the surgical sections of the Children's Cancer Group (CCG) and the Pediatric Oncology Group (POG). Unfortunately, the trials were closed 2 years after they started because of a lack of patient accrual. These failures illustrate a number of major problems in trying to study MAS in pediatric cancer management and trying to determine its optimal use. A study was published which critically examined these trials and their failures in an attempt to prevent them in the future (68). Using a questionnaire, the authors polled the surgeons and their institutions involved in the CCG and the POG. They made a number of significant observations. These included: (1) $<25\%$ of the potential protocols were submitted for IRB approval, (2) $\sim 40\%$ of surgeons were not actively performing MAS at their institutions, (3) not all surgeons received the study protocols in a timely fashion, (4) almost 50% of surgeons did not know who would submit the protocol for IRB approval, and (5) a preconceived surgeon bias for either MAS or traditional open approach. These issues strike at a recurrent theme in clinical studies in surgery. McLeod (69) has noted that surgeons have traditionally placed more emphasis upon case series than on objective data, as can be obtained in a randomized controlled trial. As such, few randomized controlled trials have been completed in surgery. This is potentially a fatal flaw which will prevent the appropriate study of MAS in pediatric cancer. It is clear that surgeons are going to have to change the way they do business, if MAS in pediatric cancer is going to move forward and improve the quality of care of these children.

10. SUMMARY

MAS in the diagnosis of children with cancer is safe, effective, and accurate and is comparable to open surgical techniques. There is very good basic science data which suggests that MAS may cause less tumor growth and metastases and less immunosuppression, compared to open laparotomy. There has been a significant problem with trying to translate

these basic science observations into successful randomized, controlled trials. It is clear that some prospective trials are necessary to determine the diagnostic accuracy and outcomes of laparoscopic and thoracoscopic techniques in children with cancer. These trials need to examine long-term patient survival, which should not be compromised for the current, perceived short-term benefits of MAS.

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8

Complications of Pediatric Minimal Access Surgery

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1. INTRODUCTION

Minimal access surgery (MAS) requires creation of a working space, the insertion of a telescope for visualization and additional ports for therapeutic instrumentation. Since the early 1990s, the number and variety of minimal access procedures performed by pediatric surgeons has grown exponentially. More advanced procedures have become feasible with the major advances in miniaturized video equipment and instrumentation. MAS is now firmly established in the armamentarium of modern pediatric surgical practice. In the United States, >80% of pediatric surgeons perform MAS (1).

Enthusiasm for MAS must be tempered; however, by an appreciation for its complications and limitations. As Tam stated, MAS only provides an alternative method of performing the same operation as open (1). No additional lives are saved. The benefits are measured in terms of quality of life. Current literature details reports of minimal access approaches to traditional open operations. Few procedures have been critically evaluated in a prospective randomized controlled trial, but initial studies suggest better outcomes secondary to reduced hospitalization and earlier return to normal activity.

Despite the adoption of MAS by pediatric surgeons, most literature reporting complications originates from the older gynecology literature (2). Pediatric patients are particularly susceptible to complications due to their thin abdominal wall, proximity of organs to the surface, and their square abdomen resulting in an intraabdominal liver, spleen, and bladder (3). The overall mortality rate from MAS in children has been quoted at 0.1%, and the complication rate at 1–2%, but this is age and procedure dependent (3–5). The Italian society of videosurgery in infancy (SIVI) reported an overall complication rate of 4.6% in 1689 minimal access procedures (6). In 574 pediatric laparoscopic procedures reported by Chen et al., the conversion rate was 2.6% and the complication rate was 2% (5). All of the intraoperative complications were early in the author's experience.

Complications of MAS can be divided into three categories: general complications common to all MAS procedures, energy source complications, and procedure-specific complications (7). This chapter will review general complications and the complications related to energy sources. Procedure-specific complications will be discussed in subsequent chapters. A discussion of complications specific to thoracoscopy and retroperitoneoscopy is also included, and the chapter concludes with measures to reduce complications. Most of the evidence presented is drawn from the gynecology literature because it is the most comprehensive. Data from both the general surgery and pediatric surgery literature will be presented when available.

2. GENERAL COMPLICATIONS

2.1. Access

MAS requires obtaining access to a body cavity (i.e., abdomen or chest) and creation of a working space. This section will focus primarily on access complications during

abdominal surgery. Visceral injury during abdominal access and creation of pneumoperitoneum is divisible into vascular, gastrointestinal, and genitourinary injuries (2). Overall, these are rare events, but they are a major reason for morbidity, mortality, and conversion to an open approach.

2.1.1. Open vs. Closed Laparoscopy

Abdominal access may be obtained by the insertion of a cannula under direct vision (open) or blindly into the abdomen (closed). There are four methods to create pneumoperitoneum: (1) blind Veress needle insertion, followed by blind insertion of a primary trocar, (2) blind insertion of a primary trocar without prior Veress needle insertion or establishment of pneumoperitoneum, (3) open insertion of a cannula without pneumoperitoneum, (4) the use of an optical trocar with or without pneumoperitoneum. Each technique has its proponents and major injuries have been reported with all of them (8–13).

Veress needle insertion with pneumoperitoneum and subsequent blind trocar insertion is the most common technique employed. The Veress needle consists of a blunt-tipped spring loaded inner stylet and sharp outer needle. The stylet retracts during passage of the needle through abdominal layers. The stylet does not lock once it protrudes, therefore, it can penetrate intraabdominal structures because the stylet will again retract on contact.

Proponents of this approach claim the pneumoperitoneum creates a space between the organs and the abdominal wall decreasing the risk of injury; however, 50% of major vascular or intestinal injuries occur with the Veress needle technique (14). There is enough literature to suggest that pneumoperitoneum does not prevent major visceral injury. The distended abdomen is difficult to grasp and to elevate during trocar insertion and the pressure of the pneumoperitoneum is not sufficient to resist trocar forces and prevent contact with underlying viscera (15,16). In a large Japanese series of 15,279 laparoscopic procedures, 156 (1.02%) needles and trocar complications were reported (17). The incidence of major vascular injury was 10/156 (0.07%) and gastrointestinal injuries occurred in 11/156 (0.07%). The authors concluded open access techniques were superior.

Direct trocar insertion without prior pneumoperitoneum avoids the difficulty of grasping and elevating an abdominal wall made tense with insufflated gas. Subcutaneous emphysema is also prevented. The trocar has a safety shield with a sharp tip. The shield automatically locks once the peritoneal cavity is entered to prevent visceral injury. Three randomized controlled trials with a total of 664 patients have evaluated Veress needle access to the direct trocar (8,9,18). No advantage to the direct trocar technique has been found. Subcutaneous emphysema was seen in the Veress needle group. The only major complication (vascular injury) was in the direct trocar group.

Open laparoscopy requires a tiny abdominal incision, usually at the umbilicus. Insertion of the primary trocar under direct vision increases the certainty of the pneumoperitoneum and allows correct anatomical repair of the fascial defect. Theoretically, open access should decrease the risk of visceral injury to zero, but injuries have still been reported, albeit considerably less. Penfield (12) reported a complication rate of 0.2% and a bowel injury rate of 0.06% in 10,840 open laparoscopic procedures. The mechanism for visceral injury is often poor visualization and the need for blunt dissection with a finger or an instrument to enter the peritoneal cavity (2).

The optical trocar was introduced in 1993 by Melzer et al. (19) in Germany. An endoscope is inserted into the trocar, therefore, the fascia is cut under direct vision. Manufacturers have each developed their own version of the optical trocar. Olympus ("Optical scalpel", Melville, NY) requires a special scope and cannula. United States Surgical

Corporation ("Visiport", Norwalk, CT) created a hollow cannula with a transparent tip. A 10 mm camera is inserted to the end and a stainless steel blade is triggered to protrude 1 mm. Ethicon ("Optiview", Endo-Surgery Inc., Cincinnati, OH) employs a nonbladed obturator with a clear tip that comes to a point. There are no published controlled trials of these trocars.

2.1.2. *Vascular Injuries*

Vascular injuries are a major cause of mortality (15%), second to anaesthetic complications (20). They are most common after insertion of the Veress needle or the primary trocar after insufflation (21). The reason is the close proximity of the anterior abdominal wall to retroperitoneal vascular structures (usually <2 cm). The distal aorta and right common iliac artery are the most vulnerable (2). Injuries may be recognized by the presence of hemoperitoneum or retroperitoneal hematoma. The patient may also exhibit systemic signs such as hypotension, tachycardia, or an increase in $ETCO_2$ from embolization of CO_2 gas.

Levinson published the first report of vascular injury in 1974 (22). MacDonald et al. (23) published the first review of 400 cases in 1978 and reported the incidence of major vascular injury at 0.5%.

In the gynecology literature, vascular injuries occurred in 0.26% of cases in both a large Canadian series ($n = 136{,}997$) and a American series ($n = 37{,}000$) (14,15). Two large French retrospective studies reported a smaller incidence of vascular trauma (0.02–0.04%) (24,25). Major vascular injury has been reported in 0.03% of laparoscopic cholecystectomies and 0.03% of laparoscopic hernia repairs (26–28).

The true incidence of vascular injury is probably higher than what is reported because many injuries are not reported, omitted or miscoded in retrospective reviews. It is also possible that series from experienced surgeons underestimate the rate of vascular morbidity and the figures are not generalizable to all surgeons.

The factors responsible for major vascular injury include surgeon inexperience, failure to sharpen the trocar, failure to elevate the abdominal wall, perpendicular insertion of the trocar/needle rather than into the pelvis, forceful thrust, and inadequate incision size (2).

Minor vascular injuries represent damage to mesenteric, omental or abdominal wall vasculature (inferior epigastric artery). These injuries are often the reason for conversion, transfusion, or re-operation. Minor vascular trauma occurred in 0.7% of laparoscopic hernias ($n = 10{,}837$) (26–28) and 2/362 (0.5%) laparoscopic fundoplications (29).

Cutting trocars are more likely to cut an abdominal wall vessel, whereas conical trocars push the vessel aside (30). Flute injuries in the side of arteries continue to bleed as the vessel cannot spasm.

Safe placement of secondary trocars can be achieved by insertion of the cannula under direct vision with transillumination of the abdominal wall to visualize the vasculature, the use of small trocars, an adequate abdominal incision, and not angling the trocar towards the midline (2). Bleeding from an abdominal wall vessel can be controlled by applying direct pressure with the cannula, ligation of the blood vessel (laparoscopically or open), or by tamponade with a foley catheter. All trocar sites should be inspected after the cannulas are removed to ensure hemostasis.

2.1.3. *Bowel Injuries*

Intestinal injuries are the third cause of death after vascular injury and anaesthetic complications (20). They often go initially unrecognized and the delay can lead to significant

morbidity and medico-legal action. One-third of injuries occur during access (50% from the umbilical trocar and 17% from the secondary trocar) (25) and two-thirds are secondary to dissection, electrocoagulation, or bowel grasping (31).

The gynecology literature contains many large studies from several countries that provide an estimate of intestinal injury. A series of 37,000 gynecological laparoscopies from the US reported a bowel injury rate of 0.16% (14). This is consistent with the injury rate of 0.18% in a Canadian series ($n = 136{,}997$ laparoscopies) (15) and 0.16% in a French study ($n = 29{,}966$) (31).

Bowel injury occurred in 0.05–0.3% of adult laparoscopic cholecystectomies (32–35). The incidence is as high as 0.66% in laparoscopic fundoplication, but this data includes injury to the esophagus and stomach which are being directly manipulated (2).

Fewer reports exist in the pediatric surgical literature. The rate of intestinal injury secondary to trocar access is 1–7% in operations for cryptorchidism (20). Valla et al. (36) reported two intestinal injuries in 465 childhood appendectomies and Chen et al. (5) presented two esophagotomies (0.3%) in 574 laparoscopic procedures.

2.1.4. Bladder Injuries

Bladder injuries occur during insertion of a suprapubic trocar over a distended bladder. Hence, the bladder should be drained prior to cannula insertion. This is especially relevant in pediatric surgery where the bladder is an intraabdominal organ due to the shallow pelvis. An injury may be detected by gaseous distension of the urometer bag during the operation or by instillation of indigo carmine into the bladder in more subtle cases (37).

A Canadian survey of gynecologists reported eight bladder injuries in 136,997 laparoscopic procedures. Four occurred with the Veress needle, two with the primary trocar, and two with the secondary trocar (15). In laparoscopic hernia repair, at least nine reports of bladder injury exist secondary to dissection or electrocoagulation (26–28,38–43).

Three to five millimeter dome injuries should resolve with an indwelling catheter for 7–10 days (44). Larger defects require two layer closure and an indwelling catheter.

2.2. Pneumoperitoneum

2.2.1. Insufflation Gases

Carbon dioxide is the most commonly used gas to create a working space for MAS. Nitrous oxide and helium have also been evaluated.

Carbon dioxide has the benefit of being rapidly absorbed because of its high blood solubility. This results in fast resolution of the pneumoperitoneum and a low risk of gas embolism. It is also odorless, does not support combustion and is inexpensive (45,46). The disadvantage of CO_2 is hypercapnea, acidosis, and peritoneal irritation from the conversion of CO_2 to carbonic acid.

Helium does not lead to hypercapnea, but its poor solubility increases the risk of gas embolism (47).

Helium, CO_2, and an abdominal lifting device were evaluated in a rat study by Hazebroek et al (48). Brown Norway rats received either helium or CO_2 at an intraabdominal pressure (IAP) of 6 or 12 mmHg for 120 min. A fifth group of rats received an abdominal lifter to create working space. Rats randomized to CO_2 insufflation at both 6 and 12 mmHg showed significant acidemia and hypercapnea. Rats in the helium and abdominal lifter group had no change in serum pH or CO_2. Neither the abdominal pressure (AP) nor gas type affected the mean arterial pressure.

Nitrous oxide induces less peritoneal irritation than CO_2, is also inexpensive and has both analgesic and anaesthetic properties. Nitrous oxide is slightly less soluble than CO_2, but the major concern over its use has been its combustibility. Combustion only occurs in the presence of volatile gases such as hydrogen or methane. These gases are present in the colon and are generated by bacteria. They are not located in other regions of the intestinal tract unless bacterial overgrowth is present (46). The quantity of hydrogen and methane gas can be decreased with a preoperative bowel cleansing that uses nonfermentable substrates (magnesium citrate, polyethylene glycol). The fear over combustibility is actually based on limited evidence such as two case reports from Sri Lanka and Egypt (49,50). In both of these cases, combustion occurred after all electrical current had stopped, therefore, it is unlikely ignition of combustible gas with laparoscopic electrosurgical devices took place. As a result, Tsereteli et al. (46) conducted a prospective randomized double-blinded study comparing nitrous oxide and CO_2 pneumoperitoneum (CDP) for laparoscopy. Patients receiving CDP ($n = 51$) demonstrated an increase in the mean $ETCO_2$ level despite an increase in minute ventilation. Nitrous oxide patients ($n = 52$) suffered less pain at 2 h, 4 h, and 1 day postoperatively based on mean scores from a 10 point visual analog scale. Despite the subjective difference in pain with NO_2, there was no difference in postoperative narcotic or Toradol use. There were no complications in either group. The authors concluded that NO_2 should replace CO_2 as the first choice for gas insufflation, but recognized the difficulty in overcoming the fear over NO_2 safety.

2.2.2. Gas Embolism

Gas embolism is a rare, but potentially lethal complication. The incidence of gas embolism is hard to determine. With the use of transesophageal echocardiography (TEE), gas embolism has been detected in 6–69% of patients undergoing laparoscopic procedures, but these were clinically insignificant. Gas enters the blood stream when a Veress needle or trocar punctures a blood vessel, a tension pneumothorax forces air into an injured vessel or by the venturi effect across an open blood vessel (45). Consequences depend on the amount, rate, and nature of the gas (51). Canine studies have demonstrated that large air boluses (3–8 cc/kg) cause an "air-lock" with obstruction of the right atrium and ventricle. A slow infusion of gas (0.3 cc/kg per min) can be absorbed across the alveolar membrane without untoward effect. Higher infusion rates cause bubbles to lodge in the pulmonary arterioles. This results in clumping of polymorphonuclear leukocytes, platelet aggregation, and initiation of the coagulation cascade. Release of inflammatory mediators causes pulmonary vasoconstriction, bronchospasm, and right heart failure (52). Portal venous gas embolism can lead to release of emboli into the hepatic portal circulation and result in delayed manifestations (53).

Patients may manifest abrupt oxygen desaturation, cyanosis, hypotension, and a "millwheel" murmur. Patients who are able to shunt from right to left through a septal defect, patent foramen ovale or patent ductus arteriosus are at risk of a paradoxical embolus and a cerebrovascular accident. The immediate treatment is to stop insufflation of gas and release the pneumoperitoneum. The patient is placed in the left lateral decubitus position with steep Trendelenburg and the patient is hyperventilated to promote CO_2 removal. The air within the right atrium or ventricle can be aspirated through a central venous catheter (7).

2.2.3. Pneumothorax

Pneumothorax during MAS may result in abrupt oxygen desaturation and elevated peak airway pressures. Patients are at risk of tension pneumothorax due to the high pressures

involved. Patients require the insertion of a large bore angiocath into the thoracic space and the insertion of a chest tube.

The etiology may represent dissection of peritoneal gas into the mediastinum via tissue planes or a diaphragmatic defect. Air can breach the mediastinal pleura posterior to the root of the heart (54,55). Perforation of the falciform by a trocar may also permit gas to enter the mediatinum through the caval orifice. Dissection in the esophageal hiatus during a fundoplication or Heller myotomy can cause pneumomediastinum or pneumothorax. In addition, pneumothorax from barotrauma or rupture of congenital bullae may occur secondary to elevated ventilation pressures induced by the IAP.

2.2.4. Other

Pneumoperitoneum may also result in other complications such as subcutaneous emphysema (0.4%) (usually a result of improper placement of the Veress needle), bradyarrhythmias (0.01%) (that are vagally mediated and occur with peritoneal stretching from rapid insufflation), postoperative shoulder pain (0.04%) (as CO_2 is metabolized to carbonic acid causing peritoneal irritation), and hypothermia (if gases are not warmed) (1,56).

2.3. Cardiopulmonary

The physiologic effects of CDP, IAP and body position on the hemodynamics of infants and children are not well known. The cardiovascular effects of pneumoperitoneum in adults are better understood and include increased MAP and systemic vascular resistance (SVR) (57–59), increased afterload and right and left ventricular filling pressures (60–62), increased end-tidal CO_2 (57,61), and decreased cardiac index (CI) (58,59).

Cardiopulmonary physiology in infants and children is not simply a smaller version of adult physiology. Children represent evolving systems with age-dependent maturation. Much of the literature presents conflicting conclusions. The results are confounded by the anesthetic technique, surgical technique and stimulation, the participants's fluid status, cardiovascular physiology, and co-morbid disease (61). In addition, the methods chosen to measure cardiac function will influence the reliability of results. Type II statistical error secondary to small sample size is also a factor.

2.3.1. Carbon Dioxide Pneumoperitoneum

The systemic absorption of gas from pneumoperitoneum is dependent on its solubility, the magnitude of the IAP, and the duration of surgery (45). Hypercarbia has a direct action on the cardiovascular system and an indirect action through the sympathetic nervous system. The CO_2 effects that may act as stressors on a patient's physiologic reserve include acidemia, tachycardia, catechol release, decreased threshold for arrhythmias, and increased sensitivity to vagal stimulation (63,64). Manifestations include tachycardia, arrhythmia, increased cardiac output, and elevated pulmonary and SVR (65).

The factors accounting for hypercarbia include absorption across the peritoneum and the change in respiration and cardiac output induced by elevated IAP. Diminished diaphragmatic excursion results in smaller lung volumes, lower functional residual capacity, and ventilation–perfusion mismatch. These respiratory changes may be exacerbated by the Trendelenburg position (45). Adult studies suggest an increase in minute ventilation of 15–30% is required to compensate for hypercarbia (61). In a study involving laparoscopy in 65 neonates between 2 and 30 days of life, all patients developed hypercarbia (mean CO_2: 52.8 ± 6.21 mmHg) when fixed minute ventilation was used.

Minute ventilation had to be increased 30–40% to keep the $ETCO_2$ normal when an IAP of 8 mmHg was used (66).

Carbon dioxide is buffered and is absorbed into muscle and bone, however, it will be gradually excreted postoperatively which results in increased ventilation requirements after surgery. This may be detrimental to the patient if they are unable to increase their minute ventilation due to residual anaesthetics, narcotics, or poor diaphragmatic excursion (67).

2.3.2. Intraabdominal Pressure

Elevated IAP effects hemodynamics by altering SVR, venous return, and myocardial performance (45). The effect on venous return and cardiac output depends on the magnitude of the IAP. When IAP is <20 mmHg, venous return increases because of the augmented blood return from the splanchnic circulation. At an IAP >20 mmHg, venous return is impaired resulting in a drop in preload and an increase in SVR. As a result, cardiac output drops, but the MAP may remain constant because of the rise in SVR (45,68).

The rise in central filling pressures with pneumoperitoneum reflects to some extent, the transmission of the elevated IAP to the thorax (68). The question is whether the increased filling pressures are a true reflection of the volume status of the patient or whether the pressures represent visceral pressure on the diaphragm. In a canine experiment, intrathoracic pressure was measured with an intrapleural catheter. At high IAP (20, 30, and 40 mmHg), a substantial rise in intrathoracic pressure was seen that paralleled the rise in central venous pressure (CVP). Hence, when elevated filling pressures are witnessed on a patient during surgery, caution must be used in interpreting the results and taking action on a number that may not accurately reflect changes in filling volume. CVP and pulmonary capillary wedge pressure should not be used as indices of filling conditions during pneumoperitoneum. When intensive monitoring is needed, TEE is more accurate (69).

There is growing data regarding the maximum IAP for paediatric MAS. Sakka et al. (70) evaluated the effect of IAP (6 and 12 mmHg) on hemodynamics in eight children aged 2–6 years. All patients were ASA I and had $ETCO_2$ held constant with adjustment of the minute ventilation. TEE was used to evaluate cardiac function. They found no change in hemodynamics at 6 mmHg, but a 13% drop in CI at a pressure of 12 mmHg. They concluded that healthy children could tolerate MAS at 12 mmHg, but children with underlying cardiopulmonary disease may need more invasive monitoring. Gueugniaud et al. (71) reached a similar conclusion in a study involving 12 ASA I children aged 6–30 months. At an IAP of 10 mmHg and constant $ETCO_2$, the authors found a 30% decrease in cardiac output, but there were no negative consequences.

2.3.3. Body Position

During MAS, patients are positioned to produce gravitational displacement of the viscera away from the operative site (45). Intuitively, changes in body position should effect hemodynamics, but the literature is very inconsistent. Odeberg et al. (61) evaluated the effect of IAP (0 and 11–13 mmHg) and body position (Trendelenburg and reverse-Trendelenburg) on the hemodynamics of 11 adults. They discovered that ventricular filling pressures and mean pulmonary airway pressures depend on body position and reflect the influence of gravity. For patients in the Trendelenburg position, there was an increase in preload and afterload suggesting an additive effect between pneumoperitoneum and body position. When patients were placed in reverse-Trendelenburg, they demonstrated an increase in afterload, but normal filling pressures. Here, the

influence of position counteracted the increased filling pressures otherwise seen during pneumoperitoneum.

In a porcine model comparing CO_2 and helium pneumoperitoneum to open surgery or an abdominal lifter, Horvath et al. (68) reported that when animals in the abdominal lifter group were placed in Trendelenburg, they displayed a twofold increase in caval pressure (68). The authors concluded that position alone can increase IAP without pneumoperitoneum.

It is possible the effect of body position varies with patient age. Fujimoto et al. (66), presented a series of laparoscopic procedures in neonates and concluded that body position had no effect on the degree of hypercapnea at an IAP of 8 mmHg.

2.3.4. Evidence from the Hospital for Sick Children

Recently, a prospective randomized controlled trial was performed at our institution to evaluate the effects of CO_2 levels, body position, and IAP on hemodynamic parameters of infants (<12 months). A total of 16 infants were randomized to a normocarbic ($ETCO_2 < 38$ mmHg) or a permissive hypercarbic ($ETCO_2$ up to 60 mmHg) group. Any infant receiving laparoscopy within the first year of life was eligible for enrollment, but children with congenital heart disease, lung disease, altered fluid balance, emergency surgery, history of prematurity, or history of cardiac medication were excluded.

All patients received a standardized anaesthetic technique with rocuronium (0.6 mg/kg per hr), fentanyl (1 μg/kg per hr), sevoflurane (3–6% induction), and isoflurane (1% maintenance). Monitoring included $ETCO_2$, noninvasive blood pressure, ECG, SaO_2, respiratory rate, peak airway pressure, gastric balloon catheter for pressure verification, and a TEE probe.

A 9 mm paediatric TEE probe was inserted transorally. The data was videotaped and evaluated by a blinded cardiologist. Data was analyzed using multiple linear regression to account for the complex interrelationship between variables.

Baseline measurements were taken with an IAP of 0 mmHg and in three different body positions: horizontal (0°), Trendelenburg (10°), and reverse-Trendelenburg (15°). The IAP was increased to 10 mmHg and then to 15 mmHg and the cardiopulmonary measurements were repeated.

There were eight patients in each group and the groups were similar with respect to age, sex, and weight (Table 8.1). End-tidal CO_2 was significantly higher in the hypercarbic group. Cardiac workload is determined by changes in preload, afterload, contractility, and heart rate. An increase in any of these will increase myocardial work and oxygen consumption. The hemodynamic effects of laparoscopy are summarized in Table 8.2.

Position had no effect at all on hemodynamics. This finding is contradictory to data generated from studies involving older children and adults. It may reflect immaturity of the autonomic nervous system as infants spend their time horizontal or crawling. They lack the hemodynamic response to upright positioning (72).

IAP of 0 and 10 mmHg had no effect on hemodynamics. At a pressure of 15 mmHg, however, preload increased due to an increase in venous return from the splanchnic bed. Contractility increased likely due to optimal stretching of myocardial fibers (Starling's forces). This translated into increased cardiac output. On the basis of our data an IAP of 10 mmHg is optimal for infants to avoid increased myocardial oxygen consumption.

Normocarbia did not alter cardiopulmonary function. Minute ventilation needed to increase 15–30% to maintain the $ETCO_2$ at normal levels. Hypercarbia had the largest effect on hemodynamics and the greatest impact on myocardial work. Carbon Dioxide acts directly on the heart and indirectly through the sympathetic nervous

Table 8.1 Patient Demographics

	Normocarbic ($n = 8$)	Hypercarbic ($n = 8$)
Age (months)	3.7 ± 0.9	3.6 ± 1.5
Sex (M:F)	7:1	7:1
Weight (kg)	6.5 ± 1.0	5.3 ± 1.0
Mean AWP (mmHg)	18.5 ± 4.8	20.5 ± 4.0
Respiratory rate	20 ± 6	20 ± 5
$ETCO_2^*$ (mmHg)	36.8 ± 2.5	42.6 ± 8.0

$^*p < 0.01$.
Note: AWP, air-way pressure.

system. Hypercarbia causes tachycardia that raises cardiac output (CO = HR × SV). Rapid heart rate allows less ventricular filling time and a resulting drop in preload. Carbon dioxide also causes peripheral vasodilation that decreases peripheral resistance and MAP. The drop in MAP reduces outflow resistance (afterload) as demonstrated by a decrease in wall stress. Contractility rises to compensate for the decreased afterload and also by a direct stimulatory effect of CO_2 on the heart. There may be maturational factors at play that make the immature heart of the infant more sensitive to changes in afterload. The vigorous changes in contractility appear to dampen as a child ages.

All infants tolerated laparoscopy without difficulty, but caution is needed when treating children with co-morbid disease or diminished cardiac reserve. Patients with co-morbid disease may benefit from laparoscopic surgery because it is less invasive and promotes a rapid recovery. However, laparoscopic surgery may cause significant physiologic disturbances. Improved understanding of the potential problems will allow appropriate anaesthetic and surgical management to improve safety.

2.4. Other

2.4.1. Wound Complications

Wound infections are less common after MAS and have a reported incidence of 0.4–2% (33). The most commonly infected port sites include the umbilical trocar site (because the skin is more contaminated with bacteria to begin with) and any trocar site where an inflamed organ is removed from. Systemic sepsis secondary to a port site infection is very rare (0.08%) (73). Good skin preparation, prophylactic antibiotics, and the use of a specimen bag to remove an organ are all measures to help minimize the risk of wound infection.

Table 8.2 Effect of Laparoscopy on Hemodynamics

	Preload	Contractility	Afterload	HR	MAP	CI
Position	↔	↔	↔	↔	↔	↔
Pressure	↑	↑	↔	↔	↑	↑
Normocarbia	↔	↔	↔	↔	↔	↔
Hypercarbia	↓	↑	↓	↑	↓	↑

Note: ↑ and ↓ indicate $p < 0.01$; ↔ indicates $p > 0.01$.

Trocar site hernias occur in 0.77–3% of all laparoscopic procedures (74,75). Chen et al. (5) reported hernias in 2/574 (0.3%) laparoscopic cases and Plaus reported trocar hernias in 4/110 (3.6%) laparoscopic cholecystectomies (76). It is standard to close all 10 and 12 mm trocar sites to prevent hernia formation, however, trocar hernias have been reported at 5 mm port sites in small children. Many surgeons now routinely close 5 mm fascial defects.

Laparoscopic surgery has the advantage of producing fewer adhesions, however, postoperative bowel obstruction may still occur. The incidence of bowel obstruction was 0.14–0.77% in two large series of laparoscopic cholecystectomy (74). Cadiere et al. (77) noted a single case of bowel obstruction after 156 paediatric Nissen fundoplications and Thom Lobe's group reported obstruction in 2/574 laparoscopic procedures, but in both series the follow-up was brief (5). Trocar sites are the most common location for bowel obstructions to occur after laparoscopic surgery. Outward pressure as cannulas are removed permit entrapment of small bowel at port sites. This is facilitated by the high IAP relative to atmospheric pressure (2). To decrease the chance of bowel entrapment, port sites should be removed under direct vision, cannulas should not be removed with their valve open, and the abdominal wall should be shaken to allow bowel to drop away (2).

2.4.2. Positioning

During MAS, patients are positioned to produce gravitational displacement of the viscera away from the surgical site. The principles to prevent injuries secondary to poor positioning are similar to open surgery. Operative time may be increased early in a surgeon's experience and as more complex minimal access procedures are attempted. Therefore, patients could be at higher risk of pressure injuries or neuropraxias if positioning and padding are inadequate.

2.4.3. The Risk to the Surgeon

All surgeons are at risk of exposure to blood-borne infections (HIV, Hepatitis B and C). The overall risk is low, but needle sticks or splashing of body fluids onto mucous membranes (eyes, nose, and mouth) or open skin wounds can lead to seroconversion. MAS has at least the theoretical advantage of decreasing the likelihood of accidental exposure to blood products. In open general surgery, glove perforation occurs in 20–30% of cases. Seventy-six percent of these perforations occur during wound closure (78,79).

No data comparing the relative risk of accidental body fluid exposure following MAS or open surgery is available. In theory, the risk is lower with the smaller incisions and fewer instruments on the operating field, however, there may still be potential for transmission of infectious agents during decompression of the pneumoperitoneum.

MAS often requires the insertion of trocars into fixed locations that can result in surgeons having to operate or hold a camera in uncomfortable positions for prolonged periods of time. Berguer et al. (80) surveyed members of Society of American Gastrointestinal and Endoscopic Surgeons (SAGES) and found of 149 responders, 8–12% of surgeons experienced frequent neck and back pain after laparoscopic procedures (80). The same authors also assessed muscular workload using an ergonomics workstation comparing a hemostat to a laparoscopic grasper. The laparoscopic grasper was associated with greater total and peak muscle effort in the forearm and thumb muscles.

An ergonomic evaluation of surgeons' axial skeleton and upper extremity movements during laparoscopic surgery compared with open surgery concluded that laparoscopy was associated with more static posture of the neck and trunk and more frequent awkward movements of the upper extremity (81). Many reports also exist of digital

nerve injuries and of repetitive-use injuries in laparoscopy (82–85). Ergonomic changes to the operating room environment (such as dedicated endosurgical suites or robotics) and instrument design should help decrease physical stress.

3. ENERGY SOURCES

Energy sources are used for dissection, mobilization, and coagulation of tissues. In addition to equipment failure, these instruments can cause complications at a reported incidence of 1–2/1000 operations (86). The majority of injuries are unrecognized at the time and may present 3–7 days later. Electrocoagulation and the harmonic scalpel are the two most common energy sources used in MAS.

3.1. Electrocoagulation

Electrocoagulation forms a coagulum by heating tissues to denature protein. Electrons are transferred to the tissue, resulting in the excitation of the electron orbitals of molecules. As the electrons return to the resting state, heat is given off, which dessicates and burns tissues. Heat increases the tissue temperature to $>150^{\circ}C$, which is much $>50–60^{\circ}C$ the coagulation temperature of protein.

The amount of heat produced by current is inversely proportional to the electrode surface area. With monopolar cautery, one electrode is large (ground-plate) and the other one is small. The small electrode controls the current density. Current enters via the active electrode and exits by the return electrode. There is greater penetration of current density which is effective for hemostasis, but it is also more prone to complications. Bipolar cautery is considered safer because the current flows between two forceps localizing the tissue effect. Each electrode has the same surface area, therefore, only the tissue between them is damaged. Bipolar cautery functions at a lower current, which decreases nerve and muscle stimulation and prevents arcing, but it makes a poorer dissector.

The flow of electric current is along the path of least resistance. This characteristic is what leads to complications. There are three types of electrosurgical injuries: direct coupling, capacitance coupling, and insulation failure (87,88).

Direct coupling results from inadvertent contact between a viscus and a metal probe that is touching the activated electrode. This may go undetected. Capacitance coupling occurs when a cannula with a metal sheath becomes charged, but it is covered by a plastic guard that prevents its discharge through the abdominal wall. The charged metal releases energy when it contacts a viscus. Insulation failure permits passage of energy to the abdominal wall or a viscus. This is seen in older instruments with worn insulation.

3.2. Harmonic Scalpel

The mechanism of protein denaturing is different with the harmonic scalpel. A vibrating blade moves 80 μm in distance 55,000 times per second. Protein is denatured as hydrogen bonds break from the transfer of mechanical energy (vibrational) to the tissue. Tissue becomes "welded" together. Blood vessels are sealed by coaptive coagulation.

The advantages are the production of less heat ($<80^{\circ}C$) resulting in a cooler blade. Tissue becomes dessicated, but there is no burn, char, smoke, or odor. There is no risk of electrical injury because no current flows through the patient. In addition, there is no risk

of remote injury because cutting and coagulation only occur between the blades when they are activated and in pressured contact with tissue.

4. COMPLICATIONS OF THORACOSCOPY

Jacobaeus introduced thoracoscopy almost a century ago (89), but it was not until the invention of improved instrumentation over the last decade that thoracoscopy became popular. Traditionally, thoracoscopy was used to treat empyema, lung blebs, and perform lung biopsies. Today, resection of mediastinal masses, tracheoesophageal fistula repair, and anatomic lung resections are a few of the more complex procedures being performed.

The contraindications to thoracoscopy are similar to that of laparoscopy, but also includes patients who are unable to tolerate single lung ventilation or have had extensive pleurodesis (90).

Overall, thoracoscopy is well tolerated. The mortality rate is between 0.8% and 1.0% and is usually related to the underlying condition. Hemorrhage (0.3–0.8%), prolonged respiratory failure requiring ventilation (3.3%), gas embolus (1%), bronchopleural fistula/air leak, and conversion to open thoracotomy (7–16.5%) are also relatively uncommon events (90,91). The general complications discussed earlier also apply here, but there are a few complications specific to thoracoscopy.

To permit visualization and create a working space, most thoracoscopic procedures require ispilateral lung collapse. This can be achieved in several ways. Low flow, low pressure CO_2 is effective, but may induce hypothermia and tension pneumothorax, as a result of displacement of the very mobile mediastinum (4,5). Selective lung ventilation accomplished by mainstem bronchial intubation of the contralateral lung may cause bronchial injury from a relatively large endotracheal tube. Bronchial blockers (i.e., Fogarty catheter) placed when the patient is supine often become dislodged when the patient is repositioned and need to be re-checked prior to beginning the procedure. Double lumen endotracheal tubes can be difficult to insert. All these techniques are prone to failure and can cause premature lung re-expansion (92).

Thoracoscopic suctioning induces negative pressure and may lead to mediastinal displacement or lung re-expansion. An air entry vent can minimize this problem (45). Monopolar electrocautery may arc to the vagus nerve and induce cardiac arrhythmias or to the phrenic nerve and lead to diaphragmatic dysfunction. Inadvertent contact of any energy source with lung or the diaphragm may cause a pneumothorax or diaphragmatic hernia, respectively (92). Proper positioning and padding is important to avoid pressure or nerve injuries.

5. COMPLICATIONS OF RETROPERITONEOSCOPY

The urinary tract resides in the retroperitoneal space. It is desirable to approach the kidney posteriorly via an extraperitoneal route, to avoid potential complications that may arise from the transperitoneal approach (56). Like all MAS, the use of retroperitoneoscopy has dramatically increased over the last decade. In a survey of 24 urologic centres, 28% of urologic endoscopic procedures were performed retroperitoneoscopically in 1993, compared with 51% of cases in 1996 (93). In spite of this, retroperitoneoscopy has the disadvantages of a smaller working space, crowding of instruments, and a lack of anatomic landmarks.

The relative contraindications to retroperitoneal endoscopic surgery include (1) previous extraperitoneal surgery because adhesions may prevent surgical access by impairing CO_2 insufflation; (2) malignant renal tumors; (3) renal trauma because the perinephric hematoma absorbs much of the light and impairs visualization; and (4) xanthogranulomatous pyelonephritis or pyonephrosis with prior nephrostomy drainage because the whole perinephric space may be frozen and inaccessible (56).

Yeung (56) recently provided a nice summary of retroperitoneoscopy-specific complications.

5.1. Hypercapnia

The exact physiologic effect of CO_2 pneumoretroperitoneum is unknown. Wolf et al. (94) found greater absorption of CO_2 during retroperitoneoscopy and a higher risk for pneumothorax and pneumomediastinum. Mandressi et al. (95) found no difference in CO_2 levels and no side effects from retroperitoneal CO_2 insufflation. Presently, there is a comparative trial being performed at our institution to determine the physiologic effects of intraperitoneal MAS with retroperitoneal urologic MAS. Hopefully, this study will provide answers to some of the persistent questions.

5.2. Surgical Emphysema

During retroperitoneal MAS, a port may slip back into an intermuscular or subcutaneous plane. Emphysema is harmless if mild or well localized; however, extensive emphysema can hinder port insertion and sometimes leads to open conversion. In addition, pneumothorax or pneumomediastinum can develop.

5.3. Gas Embolism

This may result from placement of the Veress needle directly into a major vessel, or from the opening up of small veins in the retroperitoneal space during insufflation. The risk is likely greater if high insufflation pressures and fast flow rates are used. Fortunately, this an extremely rare complication.

5.4. Tension Pneumothorax

Development of the retroperitoneal working space may cause accidental disruption of the parietal pleura. Tension pneumothorax may evolve quickly if fast flow rates and high insufflation pressures are being used.

5.5. Access Failure

This is more common with retroperitoneal MAS. Failure to develop the working space can occur in patients with a history of previous surgery, pyonephrosis, and xanthogranulomatous pyelonephritis. Intermuscular balloon dissection may occur if the dilatation device is placed between layers of musculature rather than in the retroperitoneum. Development of the working space in the wrong plane can produce a complete loss of surgical orientation and lead to conversion.

5.6. Peritoneal Perforation

Perforation of the peritoneum permits CO_2 to enter the peritoneal cavity, diminishing the retroperitoneal working space and visibility. Insertion of the posterior trocar causes less indentation of the bulkier flank muscles than transperitoneal MAS; hence, it is more difficult to judge the exact site of trocar entry. This increases the risk of peritoneal perforation. Small perforations can be closed with a purse-string suture. The pneumoperitoneum can be decompressed by Veress needle insertion under video guidance, or if the perforation is large, the hole can be enlarged to create a single working space. The intraabdominal organs should also be inspected to exclude any iatrogenic injury.

5.7. Bleeding/Vascular Injury

Excessive bleeding during MAS renal surgery is usually from tears to the renal vein or one of its branches, or from an accessory or polar renal artery. The combination of light absorption from blood and a small working space increases the risk of complications. In the small series of pediatric urologic retroperitoneal operations published by El-Ghoneimi et al. (96) two of eight partial nephrectomies required open conversion because of bleeding from polar arteries.

6. REDUCING THE CHANCE OF COMPLICATIONS

MAS requires the acquisition of new skills. It is true; therefore, that more complications may be anticipated early in a surgeon's experience. This is not the rule; however, as Calvete et al. (97) determined when they found an equal distribution of complications between junior and senior surgeons. This finding may reflect more complex cases being attempted by more experienced surgeons. To prevent injury, a surgeon must be able to perform the operation and be aware of where and when problems may arise. Attendance at MAS courses and/or mentoring by a more experienced colleague should be encouraged. It is important to remember that when an experienced surgeon performs an operation for the first time, they too are considered inexperienced.

Complications secondary to equipment failure should be preventable. With increasingly complex instrumentation, the chance of malfunction exists and this is sometimes unavoidable. It is ideal; however, to discover problems before the operation commences, but this requires operating room personnel who are adequately trained. Surgical staff should be comfortable with both assembly and disassembly of all equipment. With new equipment, education should take place before it is used on a patient and the salesperson should be available to help troubleshoot. To change the culture in the operating room, surgeons may need to educate staff about why a particular technique is beneficial for patients to increase awareness and understanding (92). Integrated surgical suites may help to minimize many of the problems encountered with equipment set-up.

Complete preoperative evaluation and sound indications for surgery are obvious measures to reduce complications. The same contraindications to MAS also apply to open surgery. Patient-related factors, such as illness severity, coagulopathy, hemodynamics, nutritional status, and immunosuppresion, need to be optimized preoperatively, if possible. In addition, close communication with the anaesthetist needs to be maintained throughout the procedure.

As Lobe has stated, a close collaborative relationship with industry is important to ensure the needs of the surgeon are being addressed (92). This is especially true during the

research and development phase of new instrumentation. It is the responsibility of the surgeon to notify the company when complications arise.

7. CONCLUSION

The advantages of MAS include less postoperative pain and wound complications, shorter hospitalization, faster return to activities, and better cosmesis. Modern technology has vastly improved the safety and efficacy of MAS and the availability of smaller instruments have expanded the application of this modality to children and infants (5). The literature is filled with enthusiastic reports of technical triumphs, but proponents of MAS must remember that it is still invasive with occasional technical mishaps. Surgeons need to acquire the necessary skills and training to minimize morbidity.

Advances in pediatric surgery often follow those made in adult general surgery. With the recent explosion of MAS in children, pediatric surgeons have the responsibility to objectively evaluate their patient outcomes with properly conducted prospective controlled trials. As we are all aware, infants and children are not small adults and we should no longer rely on data extrapolated from adult studies. MAS has been a tremendous advance in pediatric surgery with the potential to improve the lives of some of our smallest patients. Improved technology with careful application and evaluation should continue to lead to improved results.

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Specific Disease and Procedures in Pediatric General Surgery

9

Minimal Access Surgical Approaches to Childhood Hepatobiliary and Pancreatic Disorders

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Pediatric surgical experience in minimal access hepatobiliary and pancreatic surgery has been limited. This is partly due to the generally advanced laparoscopic skills necessary to undertake many of the operations, a factor that will be remedied as pediatric surgeons gain expertise in these techniques. A second factor is the limitations in instrumentation that most pediatric centers are faced with. The complexity of the hepatobiliary and pancreatic anatomy, together with the relatively small working space in children, requires smaller instrumentation and more advanced technology that may be unavailable to the pediatric surgeon. It is likely that tools, such as the surgical robot, will greatly increase the facility

with which minimal access hepatobiliary surgery can be performed. Furthermore, the development of radiologically guided procedures as an alternate form of minimal access surgery may obviate the need to perform some laparoscopic procedures.

Despite the obvious challenges, a number of surgeons have utilized minimal access techniques for hepatobiliary and pancreatic surgical diseases. This chapter reviews the literature on the approaches that are currently used to treat pediatric hepatobiliary and pancreatic surgical disorders.

1. BILIARY SYSTEM

1.1. Cholecystectomy

One of the first minimally invasive operations to be performed in children, as in adults, was cholecystectomy. The safety and efficacy of this procedure in the pediatric population is well established (1). A common cause of cholelithiasis in children is hemolysis secondary to sickle cell disease and other hemoglobinopathies (2–4). Because of their significant co-morbidity, the complexity of perioperative and intraoperative care of sickle cell patients is considerably greater than that of the general population.

Postoperative acute chest crisis is a common cause of morbidity and mortality in these patients. This and other problems have led surgeons to limit cholecystectomy to asymptomatic patients only (5). With laparoscopy as a less invasive modality, it was assumed that the incidence of chest crisis would decrease, and hence the indications for cholecystectomy in children with sickle cell disease were widened to include those with asymptomatic cholelithiasis. More recently, however, the literature suggests that laparoscopy does not decrease the incidence of acute chest crisis when compared with open approaches (6). For this reason, operations on sickle cell patients, such as laparoscopic cholecystectomy, should be performed at a tertiary care pediatric center with expertise in these complex patients. The management of asymptomatic gallstones in these children remains controversial.

1.2. Choledocholithiasis

The prevalence of choledocholithiasis is increased in patients with sickle cell disease (25%) (4) when compared with the general population (10%) (7). The pediatric surgeon must have a high degree of suspicion for the presence of common duct stones based on details of the history (pale stools, dark urine, pancreatitis) or laboratory investigations (elevated transaminases, conjugated hyperbilirubinemia, elevated alkaline phospatase, hyperamylasemia), although it is not uncommon for these features to be normal despite the presence of stones in the duct. Furthermore, the sensitivity of ultrasound for detecting choledocholithiasis is at best 55% (8).

The surgeon managing a child with choledocholithiasis must decide whether the child needs imaging of the bile duct and whether exploration of the bile duct should be carried out concurrent with the laparoscopic cholecystectomy or at another time. Although most surgeons will take a selective approach for investigating the bile duct in patients with idiopathic gallstones (9), routine cholangiography is recommended for children with hemoglobinopathies (10).

A number of minimal access techniques have been developed to manage suspected common duct stones in patients undergoing laparoscopic cholecystectomy, including preoperative endoscopic retrograde cholangiopancreatography (ERCP), laparoscopic common duct exploration, postoperative ERCP, and magnetic resonance cholangiopancreatography

(MRCP). There is controversy over which modality is most appropriate, and their use is subjected to surgeon preference (1). Preoperative ERCP +/− sphincterotomy has the benefit of reducing the need for intraoperative cholangiogram or duct exploration. If preoperative ERCP is unsuccessful, the surgeon is then prepared to perform imaging and exploration of the duct. Using this approach, however, some patients are subjected to an unnecessary ERCP. Intraoperative cholangiogram to confirm the presence of stones followed by laparoscopic duct exploration is another option. Although the feasibility (11) and efficacy (12) of laparoscopic duct exploration in children have been established, the procedure is technically very challenging and limited to surgeons with advanced laparoscopic skills and tools. A third option is to perform intraoperative cholangiogram and follow with ERCP +/− sphincterotomy for patients with large ductal stones that cannot be washed into the duodenum at the time of cholecystectomy. The benefit is that some patients will be saved an unnecessary ERCP and its inherent morbidity (13). However, laparoscopic cholangiography has false-positive rates as high as 25% (12) and, if postoperative ERCP is unsuccessful, the patient will be subjected to a third operative procedure (open duct exploration) to remove the stones.

Although choice of management strategy depends in great part on available expertise, some strategies have economic benefits over others. Using decision modeling to determine the cost effectiveness of the various approaches, Urbach et al. (14) determined in adults that the most cost-effective strategy was to perform laparoscopic common duct exploration at the time of cholecystectomy. In the absence of such expertise, selective postoperative ERCP following intraoperative cholangiogram was preferred to routine preoperative ERCP unless there was a very high suspicion of ductal stones. Preoperative ERCP was, however, slightly more effective than laparoscopic duct exploration in preventing residual duct stones but this did not justify the cost disadvantage. In a randomized study, when comparing laparoscopic duct exploration with postoperative ERCP, the laparoscopic approach was as effective as the endoscopic approach in clearing the duct.

Magnetic resonance cholangiopancreatography is a noninvasive alternative to diagnostic ERCP. In patients with a medium to high suspicion of common duct stones, MRCP has been shown to have a 85–100% sensitivity and 96% specificity (15–17), particularly for stones >5 mm. This modality, however, is subject to variations in institutional experience, requires anesthetic or sedation in children and, unlike ERCP, has no therapeutic utility.

1.3. Approach to the Jaundiced Infant

When an infant presents with jaundice and when conjugated hyperbilirubinemia is confirmed, it is necessary to rule out surgical causes such as biliary atresia or choledochal cyst. The initial investigations include ultrasound to look for abnormalities of the gallbladder and bile ducts, ^{99m}TcHIDA scan to assess excretory function, and occasionally a liver biopsy to assess hepatic architecture. Although these investigations may lend support to a diagnosis of biliary atresia, they may also be positive in other conditions causing cholestasis. HIDA scan, for example, may be false-positive in the cases of bile duct paucity (Alagille's syndrome), alpha-1 antitrypsin deficiency, cystic fibrosis, and neonatal hepatitis (18). To confirm the diagnosis, a cholangiogram is typically performed through a small right subcostal incision. If the cholangiogram shows open ducts, the drainage procedure is abandoned and the patient is closed, otherwise the incision is enlarged for a biliary drainage procedure. Four distinct minimal access methods have emerged for definitive diagnosis of biliary atresia: laparoscopic cholangiogram, ERCP, MRI cholangiography, and percutaneous image-guided cholangiography.

Of note, all of these methods, while potentially avoiding a laparotomy, require general anesthesia.

1.4. Laparoscopic Cholangiography

In 1971, Gans and Berci (19) were the first to allude to the use of laparoscopy as a diagnostic tool for the jaundiced infant in their review article that set the stage for pediatric laparoendoscopy. They noted that in addition to liver biopsy, laparoscopic tools could potentially be used to inject the gallbladder with radioopaque material to visualize the biliary tree. They followed in 1973 with a description of four infants in whom they used laparoscopy to visualize the gallbladder and liver (20). In the absence of a gallbladder, they converted to an open procedure. In the presence of a shrunken gallbladder, they obtained a liver biopsy to rule out neonatal hepatitis. Once again, they suggested that a transhepatic cholecystodochogram could help in defining the ductal architecture, but did not actually report on its use. In 1977, Leape and Ramenofsky (21) described laparoscopy in four infants suspected of having biliary atresia in whom three were found to have an atretic gallbladder. A fourth patient had a distended gallbladder, percutaneous puncture revealed white bile, and cholangiography failed to show filling of the hepatic duct. In 1980, Hirsig and Rickham (22) described the use of laparoscopic cholangiogram in three patients using a fine trocar to puncture the gallbladder directly, followed by placement of a catheter through the trocar. Biliary atresia was excluded in all three infants.

These initial reports have led to two larger series of laparoscopic assessment of infant jaundice. In the first, Hay et al. (23) used the technique in 33 patients over a period of 4 years. All patients had nonexcretory HIDA scans. In the 12 patients in whom a gallbladder was visualized, an 18Fr cannula was passed transcutaneously and transhepatically to enter the gallbladder by way of the gallbladder fossa. A cholangiogram showed patent bile ducts in all patients, although two of the patients had hypoplastic ducts. Results of the cholangiogram then determined whether patients went on to percutaneous liver biopsy or laparotomy with portoenterostomy. The mean operative time was 15 min for laparoscopy and cholangiogram and 25 min for liver biopsy.

In a second study, Senyuz et al. (24) performed laparoscopy on 24 jaundiced infants. Gallbladders were present in 17 patients who then underwent transcutaneous cholecystocholangiogram directly into the gallbladder using a 16Fr cannula. Ten of these patients had filling of the proximal and distal ducts, and a needle liver biospy was carried out. Bile duct filling was not seen in seven patients, all of whom were confirmed to have atretic ducts at laparotomy and underwent a portoenterostomy. At laparoscopy, seven patients were found to have a totally atretic gallbladder, and these infants also underwent portoenterostomy for biliary atresia.

Laparoscopy appears to provide a useful adjunct to the investigational armamentarium for the jaundiced infant, facilitating both evaluation of the bile ducts and liver biopsy. As an added benefit, the same anesthetic can be used to perform laparoscopic portoenterostomy (discussed subsequently) by the surgeon who wishes to attempt such a procedure.

1.5. ERCP and MRCP

ERCP has been described as an alternate approach to the jaundiced infant, which potentially avoids the need to enter the peritoneal cavity (25). In one series (26), the procedural completion rate was 86%, and all patients who did not have visualization of the common bile duct at ERCP were confirmed to have biliary atresia at laparotomy. The procedure required general anesthetic and scope time ranged from 30 to 60 min. MRCP offers the

least invasive assessment of the bile ducts. The procedure has been shown to be feasible in infants (27) with a high diagnostic accuracy (28).

There are a number of limitations to both ERCP and MRCP at the present time. ERCP in children can be quite challenging given the low lumen-to-tube diameter ratio and difficulty in cannulating the papilla. MRCP is limited by availability of this technology and expertise in performing the imaging. Furthermore, patient movement as a result of inadequate sedation, respiration, and bowel peristalsis can significantly affect the quality of the study. Neither modality provides for an option to biopsy the liver, which would necessitate a second procedure.

1.6. Percutaneous Cholangiography

Initial attempts at percutaneous transhepatic cholangiography met with poor success due to the lack of intrahepatic ductal dilatation in infants with biliary atresia (29,30). This approach has been supplanted with ultrasound-guided percutaneous cholecystocholangiography. In the first report of the application of this technique in jaundiced infants, Treem et al. (31) used an anterior transcutaneous, transhepatic approach to enter the gallbladder neck with a 22-guage needle. The liver parenchyma served to tamponade the bile leak from the gallbladder. A cholecystocholangiogram was performed under fluoroscopy in this manner in four infants ranging in age from 4 to 10 weeks and in weight from 2.1 to 6.5 kg. One patient was confirmed to have biliary atresia and went on to portoenterostomy. The remaining patients had good filling of their ducts and went on to diagnoses of intrahepatic ductal paucity, neonatal hepatitis, and pigment gallstones from bacterial cholangitis.

The experience at our own institution (Peter Chait, personal correspondence) has consisted of 19 infants mean age of 8.3 weeks and mean weight of 4.0 kg. Eight of the patients had atretic ducts, whereas 11 had patent ducts. When compared with open cholangiogram, percutaneous cholecystocholangiogram took significantly less anesthetic time (mean of 95 min when compared with 154 min) and the infants took significantly less time to regain full feeds (1.3 vs. 2.5 days). Liver biopsies were performed simultaneously, and no complications were encountered.

1.7. Biliary Atresia

When a diagnosis of biliary atresia is confirmed, early and expeditious surgical management is necessary to avoid progression of hepatic fibrosis. This involves prompt diagnosis followed by resection of the atretic bile duct system and Roux-en-Y portoenterostomy to provide a biliary drainage conduit for the microscopic biliary radicles at the hilar plate. This operation, first developed by Kasai and Suzuki (32), is traditionally performed through a wide subcostal incision. Although technically challenging, attempts have been made to perform this operation laparoscopically.

Esteves et al. (33) reported two successful cases of laparoscopic hilar dissection and portoenterostomy. The Roux limb was created extracorporeally through an umbilical incision. Dissection was carried out using a specially designed laparoscopic monopolar fine needle cautery. The anastomosis was created using 5–0 polyglactin sutures tied both intra- and extracorporeally. Potential benefits to a laparoscopic approach include the avoidance of a large and painful incision, high magnification view of the hilar dissection, and avoidance of excessive scar tissue, which may later interfere with hepatic transplant.

1.8. Choledochal Cyst

Choledochal cysts may present in infancy with obstructive jaundice, acholic stools, and hepatomegaly or later in childhood with abdominal pain, palpable mass, and jaundice. The vast majority (90%) of choledochal cysts are saccular or fusiform dilatations of the extrahepatic biliary duct (34). Surgical management has evolved, and optimal management currently consists of cyst resection with a biliary enteric drainage procedure using a Roux-en-Y hepaticojejunostomy.

There are only a few reports of a laparoscopic approach to type I choledochal cyst, and these were in adolescent or in adult patients (35–38). This is not surprising given the dense anatomy and unforgiving dissection of the portal region, as well as difficulties with adequate exposure. After cyst resection using a combination of sharp and blunt dissection, as well as endostaplers, a stapled or sutured retrocolic Roux limb is constructed. The Roux limb can be made intracorporeally with endostaplers and suturing or by way of a minilaparotomy. Dissection is made more difficult if previous biliary procedures, such as cholecystectomy, have been done or if there have been previous bouts of cholangitis. It is likely that the technical difficulty of this procedure would be magnified in the smaller working space of a more typical pediatric patient. Nevertheless, the feasibility of biliary surgery has been demonstrated, and technological advances, such as the surgical robot, will serve to decrease the difficulty of this and similar procedures (39,40).

Laparoscopic resection of the much rarer type II or diverticular cyst has also been described (41). A combination of blunt and sharp dissection was used to excise the cyst. A clear channel connecting the cyst to the bile duct was not found, and intraoperative cholangiography did not demonstrate a leak. In situations where there is a true connection to the bile duct, it would be necessary to place an endoscopic loop suture at the base of the cyst prior to excision or, if the cyst is already excised, to suture the resultant defect without causing ductal narrowing.

2. PANCREAS

2.1. Pancreatic Pseudocyst

Pancreatic pseudocysts in children are usually the result of pancreatic trauma. Some of these lesions resolve spontaneously and do not recur, and therefore there should be an initial trial of monitoring with serial ultrasound to look for resolution. In a minority of cases, pseudocysts become large, symptomatic, and persistent. These persisent cysts may be associated with significant pancreatic ductal disruption and stricture (communicating pseudocyst).

Before management of persistent pseudocysts can be initiated, it is important to obtain clear anatomic delineation of the disease. Contrast enhanced CT scan of the abdomen shows the structural relations of the cyst. Management of a pseudocyst that is intimate to the duodenum or in the pancreatic tail differs from the much more common mid-body lesion. A distal lesion may necessitate a distal pancreatic resection with or without splenic preservation. Endoscopic retrograde cholangiopancreatography may be necessary if a disruption and/or proximal stricture of the pancreatic duct is expected. A communicating pseudocyst with a proximal pancreatic duct stricture may form a persistent fistula when drained.

Surgical management has classically involved some type of internal drainage procedure such as cystgastrostomy or Roux-en-Y cystjejunostomy, and requires that the

cyst be large and thick walled. Minimal access approaches to management of pseudocysts include laparoscopic, endoscopic, or image-guided modalities. Endoscopic management is best suited for lesser sac lesions that are intimate to the posterior gastric wall and are amenable to catheter drainage into the stomach. Image-guided therapy utilizes percutaneous drains to treat the cysts.

Some authors have advocated percutaneous drainage as a first line therapy for pseudocysts in the pediatric population when conservative management has failed (42). External percutaneous drainage has been shown to be safe and effective (43,44) in children, although the failure rate in some adult series with long-term follow-up is reported as 78% (either failed to resolve or recurred) (45). In making this comparison, it is important to note the difference in etiology of pseudocyst between young and old, with adults suffering primarily from alcoholic or gallstone pancreatitis and children from traumatic or infectious pancreatitis. In the case of a pseudocyst associated with pancreatic transection, such a procedure runs the risk of creating an external pancreatic fistula. There are data, however, to suggest that transection injuries may resolve with conservative management, including nasogastric decompression, total parenteral nutrition, octreotide, and percutaneous pseudocyst drainage (46). A second risk of drainage is that of introducing infection into the cyst, which is associated mainly with prolonged duration of catheter drainage (47). Percutaneous drainage does not allow for biopsy of the cyst, although this is of minimal concern in the pediatric population.

Park and Heniford (48) give a thorough description of the options in laparoscopic management of pseudocysts. These include variations of endoscopic and/or laparoscopic transgastric cystgastrostomy, as well as laparoscopic cystjejunostomy. Endoscopic drainage involves gastroscopic insertion of a catheter through the posterior gastric wall and into the cyst. The cyst must be relatively thin walled to allow piercing. The procedure is limited by the inability to clear necrotic debris, catheter blockage and dislodgement, bleeding, and cyst infection due to inadequate drainage (49).

2.2. Laparoscopic Pancreatectomy

Indications for pancreatectomy in children are limited. In infants, persistent hyperinsulinemic hypoglycemia of infancy (PHHI) sometimes necessitates near-total pancreatic resection for control of hypoglycemia. In older children, acute traumatic transection, insulinoma, and the relatively rare cystic neoplasm may be indications for resection.

Persistent hyperinsulinemic hypoglycemia of infancy presents with irritability and seizure. Lack of early treatment can result in permanent brain damage, and failure of medical management with agents, such as diazoxide, glucagon, and octreotide, necessitates pancreatic resection. Surgical management has involved 95% pancreatectomy, leaving a rim of pancreatic tissue abutting the second part of the duodenum, although focal disease defined by preoperative pancreatic venous sampling may be amenable to lesser resections (50). In older children, differentiation must be made from insulinoma. Laparoscopic pancreatic resection is feasible (51) and has been attempted for this indication (52). This case study, however, limited the extent of resection from the pancreatic tail to the mesenteric vessels, and the patient subsequently required an open near-total pancreatectomy. This speaks to the difficulty of laparoscopically resecting pancreas in the vicinity of the common bile duct and superior mesenteric vessels. It is foreseeable that pancreatic venous sampling in conjunction with laparoscopic ultrasound may allow for identification of distal focal disease or insulinoma that are more suitable for laparoscopic distal pancreatectomy or enucleation (53).

Blunt traumatic transection of the pancreas has also been treated with laparoscopic distal pancreatectomy in a child, with good results (54). There are some, however, that promote a nonoperative approach to such injuries (55).

3. LIVER

3.1. Laparoscopic Hepatectomy

There is very little in the literature on laparoscopic resection of liver lesions in infants and children. This is not surprising as primary liver tumors comprise $<3\%$ of tumors seen in children, of which less than one-third are benign (56). Hepatocellular carcinoma and hepatoblastoma account for 90% of malignant tumors, whereas benign tumors consist mainly of vascular lesions such as hemangioma and hemangioendothelioma.

Only those hemangiomas causing significant pain or disability should be considered for resection. Laparoscopic resection of hemangioma has been reported in the adult literature. In a multicenter European experience (57), 13 patients with hemangiomas ranging in size from 2 to 14 cm underwent laparoscopic resection. Of these, six patients had wedge resection, three had segmentectomy, one had left lateral segmentectomy, and three had major hepatectomy. A five trocar approach was used, and the tumor was localized using laparoscopic ultrasound. Dissection was carried out using an ultrasonic dissector or crushing forceps. Intraparenchymal hemostasis was maintained using clips, electrocautery, and vascular staplers. Argon beam coagulator was also used on some patients to coagulate the raw surface. None of the patients required blood transfusion. The specimens were sometimes crushed for removal with an endobag and this did not hinder pathologic identification. In this report, the authors suggested that lesions adjacent to the hepatic veins or the cavohepatic junction and those centrally or posteriorly located in the right lobe may not be good candidates for resection as laparoscopic hepatic mobilization and control of large intrahepatic vessels are difficult to attain.

Treatment of hepatic hemangioendothelioma consists mainly of steroids (58) or chemotherapy (59), with surgery reserved for intractable cardiac failure or coagulopathy (60). When pharmacologic treatment of hemangioendothelioma fails, minimally invasive therapy includes hepatic artery embolization (61), but laparoscopic approaches to resection have not been attempted because of the significant risk of bleeding.

Other lesions that are potentially resectable laparoscopically include symptomatic hamartomas and congenital cysts. Simultaneous resection using ultrasonic dissector of a 3 cm diameter congenital cyst with repair of a Morgagni hernia in a 5-month-old infant has been reported (62). In general, unroofing and/or marsupialization of these cysts are possible laparoscopically, and complete resection is probably not necessary. To date, there are no reports of laparoscopic resection of malignant pediatric liver lesions.

3.2. Staging Laparoscopy

Laparoscopy for hepatic evaluation in cases of intraabdominal malignancy appears to be a very useful technique. In conjunction with laparoscopic ultrasound, thorough investigation of the liver can be conducted to determine the presence and extent of hepatic metastases and biopsies taken. This information can be vital for further operative planning and help guide adjuvant therapy. In adult patients with hepatocellular carcinoma, use of staging laparoscopy with laparoscopic ultrasonography helped avoid exploratory laparotomy in $>60\%$ of patients with unresectable disease (63). This increased the resectability rate at laparotomy from 74% to 88%. The procedure was less accurate than laparotomy for

assessing the presence of tumor thrombi and extent of invasion into adjacent organs. The angle and direction of laparoscopic ultrasound was limited by port placement, particularly when dealing with bulky tumors >10 cm in diameter. Procedural morbidity was negligible. The safety and efficacy of laparoscopic liver biopsy for the diagnosis and evaluation of pediatric malignancies have also been established (64).

3.3. Hydatid Cyst

Hydatid cyst disease of the liver is endemic in many parts of the world including Mediterranean countries, the Baltics, South America, Australia, and the Middle East (65). Although uncommon in North America (apart from Northern Canada), the prevalence of the disease is likely to rise with increasing immigration from endemic areas. The laparoscopic management of this disease is now well described (66–68). The general technique involves injection of a scoliocidal agent under laparoscopic visualization followed by aspiration of cyst contents and wall, with meticulous attention to the prevention of spillage. To avoid this risk of uncontrolled spillage during laparoscopy, Bickel et al. (69) have recommended the use of transparent cannulas measuring up to 30 mm that can be placed transabdominally and into the cyst under laparoscopic visualization through which controlled cyst aspiration can be performed.

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10
Laparoscopic Splenectomy

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The laparoscopic technique for splenectomy has ascended to a prominent, and at some institutions, preferred technique since the initial report of Delaitre and Maignien (1). Frequently cited advantages of laparoscopic splenectomy (LS) include decreased pain, less occurrence of intestinal ileus, shortened hospital stay, and improved cosmesis. The disadvantages identified include longer operative times, a steep learning curve, and difficult splenic removal in cases of splenomegaly. Some authors have also questioned the efficacy of accessory spleen detection with the laparoscopic technique and the potential for splenosis if capsular disruption occurs (2,3).

Unfortunately, the data on LS comes primarily from case reports, large single and multiinstitutional series, and case controlled series. There are no prospective randomized trials. In many of the case controlled series, open splenectomy (OS) predominates in the early portion of the time period and LS over the later time period, thus, raising questions about comparisons of pain control techniques and length of stay. As with other procedures, surgeons have eliminated practices such as the routine use of nasogastric tubes and lengthy periods of withholding oral intake after many open procedures, thus, decreasing the length of stay for open procedures during the same time period that laparoscopic procedures have become more popular.

This chapter evaluates the existing data concerning LS and OS and provides the reader with an evidence based approach to issues regarding splenectomy including technical factors, operative time, conversion rates, detection of accessory spleens, pain medication usage, length of stay, operative complications, and late sequelae. Adult series are cited selectively; however, all comparative data comes exclusively from pediatric series.

1. TECHNICAL FACTORS AND CONVERSION RATES

The technique of LS has evolved over the past 10 years and during this time various modifications and instruments have been utilized (Table 10.1). This particular technique may affect the efficacy of the procedure, conversion rate, occurrence of capsular disruption, and the detection of accessory spleens. Splenectomy in children is usually required for hereditary spherocytosis, idiopathic thrombocytopenic purpura (ITP), sickle cell disease, and other less common conditions including lymphoma. In addition, the laparoscopic approach has been utilized for the excision of splenic cysts, splenopexy for wandering spleens, and partial splenectomy for trauma (4–6). Although splenomegaly is often noted with hereditary spherocytosis and sickle cell anemia, the spleens are still typically smaller than that of adults with splenomegaly and are usually removable with the standard devices.

Splenic artery embolization was widely utilized in the initial reports (7) in order to lessen blood loss and decrease splenic size; however, it has been associated with pain, abscess, and retroperitoneal necrosis (8). Complications related to embolization in one study occurred in 5 of 26 patients (9), and others (10) have found that it did not lower the risk of bleeding or transfusion requirement in their patients. Poulin et al. (9) in an update, currently recommends splenic artery embolization for spleens between 20 and 30 cm in length and OS for spleens >30 cm, although others only utilize it in spleens >25 cm (8). The splenic artery may also be ligated through the lesser sac in case of splenomegaly and some authors feel this is as effective as embolization (11). Embolization has not been widely utilized in children because even the largest spleens usually fit in the retrieval bag.

Table 10.1 Reported Adjuncts for LS

Splenic artery embolization
Approach
Anterior
Lateral
Wall-lifting device
Immobilization with suction cup retractor
Hand-assisted procedures
Clip appliers
Endoscopic linear staplers
Ultrasonic dissector[a]
Removal techniques
Endocatch II bag[b]
Liposucker
Accessory incision
Splenic coring

[a]Ultrasonic Harmonic Scalpel, Ethicon Endosurgery, Inc., Cincinnati, OH.
[b]United States Surgical Corp, Norwalk, CT.

The anterior approach was initially utilized at several institutions; however, most currently utilize a lateral approach with three to four trocars as depicted in Fig. 10.1 (12). Gossot et al. (13) describe the evolution of their technique from an anterior approach to a hand assisted and then to a lateral approach identifying advantages of less blood loss and shorter operative time with the lateral approach. Poulin et al. (9) also noted that the change from an anterior to a lateral approach allowed them to avoid splenic artery embolization in most instances.

With the introduction of mini-laparoscopic instruments, it is possible to reduce the trocar size to 2–3 mm at the sites where retractors and dissecting forceps are utilized (Fig. 10.1). Yuan and Yu (14) reported utilizing one, 12 mm umbilical port (for camera, endoscopic stapler, and retrieval bag) and three, 2 mm ports (for retraction, dissection, and for the 2 mm camera). The author's institution has found the 3 mm instruments more useful than the 2 mm instruments which appear to lack the ability to elevate the spleen and retract adjacent structures. If the camera is at the umbilical site and ultrasonic dissector or clip applier are utilized, a 5 mm trocar is needed at one site, usually the left lower quadrant. The introduction of the ultrasonic scalpel (Ultracision Harmonic Scalpel, Ethicon Endosurgery, Inc., Cincinnati, OH) has been one of the most important technical advances and the 5 mm diameter size allows it pass through a smaller trocar than the original device. This allows division of splenic ligaments and small vessels (short gastrics) without the need for introducing clip appliers and scissors, thus decreasing operative time.

The diameter of the EndoCatch II device (United States Surgical Corp, Norwalk, CT) is 15 mm and a similar sized umbilical trocar allows splenic removal and "hides"

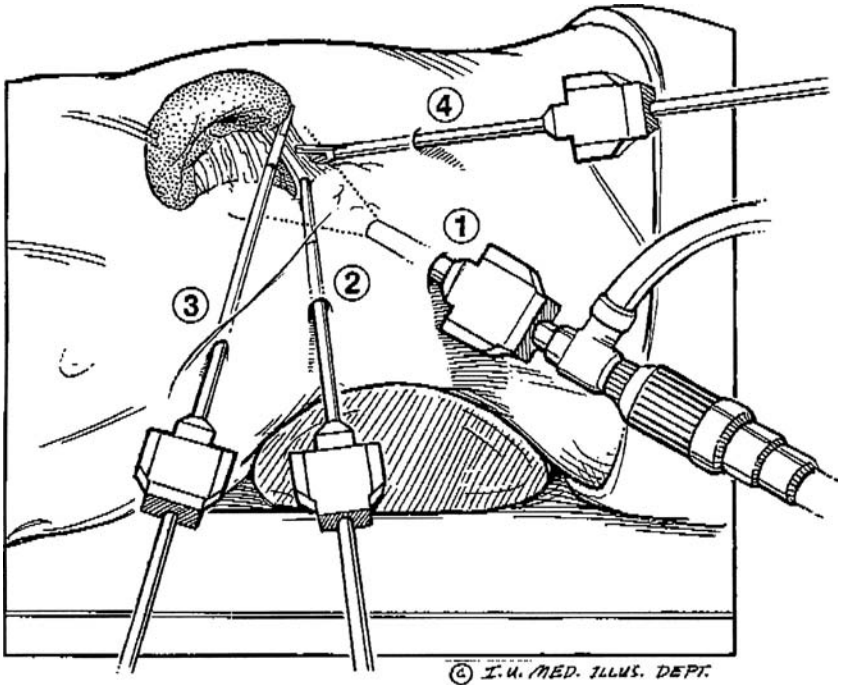


Figure 10.1 The lateral approach with a common location of trocars. The author utilizes a 3 or 5 mm trocar at the upper midline sites (2) and (3), a 5 mm at trocar (4), and a 10, 12, or 15 mm umbilical trocar at the umbilicus (1).

most of the largest incision in the umbilicus. This bag has a diameter of 13 cm (5 in.) and depth of 23 cm (9 in.) allowing removal of most pediatric spleens. Smaller spleens will fit in the bags which are packaged in 10 mm devices (Endopouch, Ethicon Endosurgery, Inc., Cincinnati, OH), allowing a smaller umbilical port. The authors institution has not found it necessary to "double bag" the spleen; however, some institutions have experienced bag disruption and currently utilize a second bag. (15) Esposito et al. (16) reported 4 cases of bag rupture out of 46 cases, although the type of bag was not specified. Various techniques have been utilized to assist spleen fragmentation and removal from the bag, including introduction of a finger, ring forceps, morcellator, or use of a liposucker (17). In addition, Hebra et al. (18) described a technique in cases of splenomegaly of coring out the spleen prior to placement in the bag.

Other operative adjuncts have included the use of a wall-lifting device with wires to provide exposure and avoid pneumoperitoneum (19), an atraumatic suction cup grasper to manipulate the spleen (20), an umbilical tape to elevate and immobilize the hilum (21), and an accessory incision to remove the spleen or to allow a hand-assisted procedure in cases of splenomegaly (22–23). An accessory incision has not been widely utilized in children and would likely neutralize the positive effect of less pain medication, improved cosmesis, and shorter length of stay associated with LS. Gigot et al. (24) noted that the addition of this incision in adults prolonged the hospital stay when compared with those without an accessory incision (7.9 ± 5.1 days vs. 4.7 ± 2.8 days, $p < 0.03$).

The technical aspect of some studies raise serious questions about the application of LS. Sandoval et al. (25) in a series of 11 cases reported one child with sickle cell anemia who underwent a splenectomy and cholecystectomy with addition of a left lower quadrant incision to remove a 558 gm spleen. The procedure took 295 min and the child was hospitalized for 5 days because of atelectasis. Patton et al. (26) performed a laparoscopic cholecystectomy and splenectomy with a procedural time of 12 h. These patients may have been better served with an open procedure.

Conversion rates are generally lower in pediatric series than in adult series (Table 10.2), although several recent large adult series have a very low conversion rate. Conversion is most frequently required for bleeding or splenomegaly. Several series have noted higher conversion rates in their initial experience. Yee et al. (31) in a series of adults and children converted 4 of their initial 11 (36%) but none of their next 14. Parks et al. (8) in the largest reported series converted 3 of their initial 40 (7.5%) but only 2 in the next 163 (1.2%). These rates are lower than those of most adult series which include larger spleens associated with malignancy. Berman et al. (32) noted a

Table 10.2 Accessory Spleen Detection and Conversion Rates of Several Large Pediatric and Adult Series

Series	Year	Peds/adult	N	Accessory spleen (%)	Conversion rate (%)
Gigot et al. (24)	1995	Adult	50	14	10
Friedman et al. (7)	1997	Adult	63	19	7
Katkhouda et al. (27)	1998	Adult	103	17	3.9
Minkes et al. (28)	2000	Peds	35	29	2.8
Park et al. (8)	2000	Both	203	12	3
Walsh et al. (29)	2001	Both	150	—	1.3
Esposito et al. (16)	2001	Peds	54	13	1.9
Rescorla et al. (30)	2002	Peds	112	17	2.7

conversion rate of 41% in adults with malignancy compared with a 3% conversion rate in benign conditions.

2. ACCESSORY SPLEEN DETECTION

Table 10.2 provides the accessory detection rates in several large series of both adult and pediatric LS, whereas Table 10.3 provides the comparative rates of accessory spleen detection in pediatric series with OS and LS. As noted, the detection rates are comparable in most series and the cumulative percentage of the comparative series are not statistically different. Accessory spleens have been missed with both OS and LS leading to failure of resolution or recurrence of thrombocytopenia in ITP and in hereditary spherocytosis of recurrent anemia. Walters et al. (38) reported recurrence of ITP 5 years after splenectomy in a woman with a missed accessory spleen.

Missed accessory spleens may achieve large size in cases of hereditary spherocytosis and present clinically after an initial response to splenectomy. Crawford et al. (39) reported a 31 year interval in a man with hereditary spherocytosis and recurrent anemia. The mass which appeared most consistent with a splenic remnant was 12 cm in its largest dimension and weighed 359 gm. The laparoscopic removal of missed accessory spleens has been reported with good success by several authors (40–41).

Some authors have questioned the ability of laparoscopy to detect accessory spleens. Taragona et al. (3) reported the details of 9 patients who failed to improve out of a group of 48 consecutive LS with an initial accessory spleen detection rate of 12.5%. The nine failures underwent heat damaged red blood cell scintigraphy with the detection of residual splenic function in three. Computed tomography (CT) scans confirmed accessory spleens in two, and in the other, scintigraphy demonstrated multiple hot spots in the splenic fossa which the CT confirmed as splenic implants. This latter patient had required conversion to open surgery and may have had splenic capsular disruption. None of the three, however, had confirmatory surgical exploration to excise the residual splenic tissue.

Gigot et al. (2) identified residual splenic function in 9 of 18 patients after LS despite an operative accessory spleen detection rate of 41%. Only three of these had a clinical recurrence and only one underwent exploration with removal of an accessory spleen. They also noted that the rate of residual splenic tissue was 75% in those with capsular disruption vs. 36% in those without parenchymal injuries.

Table 10.3 Comparative Data of Pediatric LS and OS

		Accessory spleen (%)		OR time (min)		Length of stay	
Series	*N*(OS/LS)	OS	LS	OS	LS	OS	LS
Farah et al. (33)	20/16	25	0	96	138	4.9	3.6
Waldhausen and Tapper (34)	10/10	20	20	90	211	3.3	2.6
Moores et al. (35)	20/12	35	50	84	173	4.7	3.3
Rescorla et al. (36)	32/50	25	18	83	115	2.5	1.4
Reddy et al. (37)	29/16	7	6	168	237	3.0	1.3
Minkes et al. (38)	17/35	12	29	—	—	4.0	1.8
Total		26/128 (20.3)	28/139 (20.2)				

Several authors initially utilized preoperative scans including CT and scintigraphy to detect accessory spleens; however, these tests have been abandoned by most. Katkhouda et al. (27) performed routine preoperative CT scans on their initial 32 patients and detected only two of seven (28.6%) accessory spleens and therefore abandoned the practice. Gigot et al. (2) noted that a careful laparoscopic exam yielded a higher rate of accessory spleen detection than combined preoperative CT and damaged red blood cell scans. Despite the failure of several studies to demonstrate effective preoperative accessory spleen detection, some authors recommend use of preoperative scans (16,42) although they fail to provide evidence of its value. In addition, some have utilized intraoperative scintigraphy for localization and the confirmation of complete excision of accessory spleens (43).

Although it is difficult to determine an acceptable rate of accessory spleen detection, some series had low numbers. Morris et al. (42) identified one accessory spleen in 16 LS (6%) performed in adults with ITP. Two presented with recurrent thrombocytopenia and after radionuclide scans identified accessory spleens, underwent successful laparoscopic removal. The technique in which splenectomy is performed may also affect the detection rate. A technique which divides the short gastric and hilar vessels together (31), with several applications of the stapling device, without opening the lesser sac, may miss accessory spleens located in this area. The available data would suggest that a careful laparoscopic exam is equal to an open laparotomy for the detection of accessory spleens.

3. OPERATIVE TIME

Nearly every study has demonstrated increased operative time for the laparoscopic technique (Table 10.3) and several have noted decreased operative time with increasing surgeon's experience. Gigot et al. (24) noted that operative time for surgeons performing their first 10 LS averaged 214 min, whereas after this, averaged 165 min. Rege and Joehl (44) demonstrated the learning curve in a teaching environment with increased success (95% vs. 65%), shorter operative time (97 vs. 195 min), and shorter LOS (1.5 vs. 2.5 days) associated with increased experience as well as technical advances such as the increased use of the ultrasonic scalpel and a change to the lateral position. A pediatric series of 112 LSs noted shorter operative time (98 vs. 115 min) in their first 50 when compared with the remainder of the series ($p = 0.055$) (30). This is still longer than the 83 min for OS reported by the same group (36). Longer operative time results in higher operating room and anesthesia charges which in most series have been balanced by a shorter length of stay.

4. PAIN

Relatively few studies report pain medication use, however, most of these demonstrate lower narcotic use with LS when compared with OS (Table 10.4). The most commonly reported factor is total narcotic dose and some reports also include the time duration of intravenous narcotics. This is similar to adults studies (46) which have noted equal quality of life and adequacy of control of hematologic disease with less pain experienced by the laparoscopic group. A potential shortcoming of a comparison of pain associated with LS and OS is that OS predominates in the early portion of the time period of some studies and may not reflect the use of patient controlled analgesia techniques which have decreased narcotic use in patients with open procedures.

Table 10.4 Comparative Studies of Narcotic Use with LS and OS in Children

		Morphine equivalent dose (mg/kg)			
Series	Year	*N*(LS/OS)	LS	OS	
Moores et al. (35)	1995	12/20	0.66	0.79	NSD
Beanes et al. (45)	1995	7/14	0.34	0.40	NSD
Curran et al. (15)	1998	7/7	0.18	0.8	$p < 0.05$
Rescorla et al. (36)	1998	50/32	0.239	0.48	$p = 0.006$
Reddy et al. (37)	2001	16/29	0.15	1.5	$p < 0.001$

Note: NSD, no significant difference.

5. LENGTH OF STAY

Nearly every study has identified a shorter length of stay for LS when compared with OS (Table 10.3). Length of stay contains significant variability and the length of stay for LS in some series is longer than the length of stay for OS in other series. Shimomatsuya and Horiuchi (47) in an adult series from Japan had an 8.9 day length of stay with laparoscopic and 15.2 day with OS, clearly higher than most other series. Geiger et al. (48) performed OS through a muscle, splitting lateral incision in 39 children with an average operative time of 98 min, and an average LOS of 2.7 days. This length of stay is less than that of LS in some pediatric series (Table 10.3), although it is similar to the 2.5 day stay reported by the author's institution in 32 open splenectomies (36). In that series a comparative case controlled study noted a statistically significant shorter length of stay with LS.

The underlying diagnosis also appears to affect the hospital stay. Donini et al. (49) noted shorter stays in patients with ITP and Hodgkins compared with other hematologic diagnoses. A study from the author's institution noted a longer length of stay in children with sickle cell disease compared with ITP or spherocytosis (2.5 vs. 1.4 days) (36). This difference may be related to splenic size as well as the effect of the primary disorder on other organ systems such as the occurrence of acute chest syndrome in sickle cell disease. The benefits of laparoscopy also exist with splenomegaly. Berman et al. (32) in an adult series noted shorter stay for LS vs. OS (4 vs. 6 days) in patients with splenomegaly related to malignancy.

Although adult series report length of stay data, back to work time is not reported. An unreported effect of surgery in children is the return to the child's full activity as well as the effect of the child's period of home rehabilitation on the ability of parents to return to work. Full activity is difficult to define in children as return to full activity in a 1 year old is different than that of a 10 year old, where return to school could be considered analogous to adult series reporting return to work. One pediatric series (34) noted children with LS returned to full activity 1–5 weeks faster than those with the open procedure. They also noted that parents who had undergone splenectomy themselves felt that the laparoscopic procedure was better than their own OS and if given a choice would choose laparoscopy.

6. COST

Evaluation of cost data is difficult because of the differences in various hospital methods of calculating cost and charges. Most studies which break down the cost note higher operative expenses for laparoscopy balanced by lower costs related to a shorter length of stay.

Table 10.5 Comparative Studies of Cost in LS and OS

Series	Year	Peds cost			*p*-Value
		N(LS/OS)	LS (US dollars)	OS (US dollars)	
Janu et al. (50)	1996	14/47	7,223	4,081	<0.05
Farah et al. (33)	1997	16/20	13,410	14,405	
Waldhausen and Tapper (34)	1997	10/10	13,033	7,106	
Curran et al. (15)	1998	7/7	10,899	8,275	
Rescorla et al. (36)	1998	50/32	5,713	6,566	
Reddy et al. (37)	2001	16/29	4,950	5,400	

Table 10.5 lists pediatric series with comparative cost data. As noted, there is no clear cost savings for either procedure. Waldhausen and Tapper (34) had the greatest discrepancy reported in cost and in their series, the operative cost of laparoscopy was over three times than that of open, perhaps because of the relatively long operative time (211 min) in this early experience. Although procedure cost is an important factor, as the cost is not clearly higher for LS, it is probably not an important factor in deciding the method of splenectomy.

7. COMPLICATIONS AND LONG-TERM SEQUELAE

Perhaps the most significant aspect of a comparative study is the evaluation of complications. Clavien et al. (51) presented a classification system of surgical complications in an attempt to stratify the severity of complications and allow comparative analysis (Table 10.6). Tables 10.7A and B report the complications for pediatric series of LS and OS from 1995–2002. As seen in Table 10.7C, there is no statistical difference between the rate of Grade I and Grade II or total complications between OS and LS. One potential limitation of this could be under-reporting of Grade I complications which would include atelectasis, gastric distention requiring nasogastric tube, and fever with temperatures $>38.5^{\circ}$C on two consecutive days, although under-reporting should equally affect both groups. These results are similar to adult series. Targarona et al. (57) in a group of 122 LSs had a complication rate of 18% with all Clavien type I and II. Forty-three percent of their complications were technically related. Espert (58) had a complication rate of 16% with laparoscopic and 28% with OS (NSD). Friedman et al. (7) in the largest comparative adult series (63 LS, 74 OS) noted an equivalent rate of Grade I and II complications (5–9%) between OS and LS but higher Grade III (4% vs. 0%) and Grade IV (3% vs. 0%) in OS compared with LS.

Table 10.6 Clavien Classification of Surgical Complications

Grade I	Minor, complication resolves spontaneously or with minor bedside procedure
Grade II	Potentially life threatening, usually require intervention. Iatrogenic injuries. Complications resulting in doubling of hospital stay
Grade III	Residual or lasting disability. Iatrogenic organ resection
Grade IV	Death as a result of the complication

Table 10.7 Complications Associated with LS, OS, and Comparison of Cumulative Complication Rate

Series	Year	*N*	Clavien grade I	II	III	IV
(A) LS						
Beanes et al. (45)	1995	7	1	1	0	0
Moores et al. (35)	1995	12	1	0	0	0
Yoshida et al. (52)	1995	8	0	0	0	0
Fitzgerald et al. (12)	1996	18	0	1	0	0
Janu et al. (50)	1996	14	0	1	0	0
Esposito et al. (53)	1997	8	0	0	0	0
Farah et al. (33)	1997	16	1	4	0	0
Schleef et al. (21)	1997	11	0	0	0	0
Waldhausen and Tapper (34)	1997	10	0	0	0	0
Curran et al. (15)	1998	7	0	1	0	0
Liu et al. (54)	2000	11	3	0	0	0
Minkes et al. (28)	2000	35	0	3	0	0
Park et al. (55)	2000	59	0	3	0	0
Reddy et al. (37)	2001	16	0	1	0	0
Rescorla et al. (30)	2002	112	4	8	0	0
Total		344	10 (2.9%)	23 (6.7%)	0	0
(B) OS						
Beanes et al. (45)	1995	14	0	2	0	0
Moores et al. (35)	1995	20	1	1	0	0
Yoshida et al. (52)	1995	11	0	0	0	0
Janu et al. (50)	1996	47	6	1	0	0
Esposito et al. (53)	1997	8	1	0	0	0
Farah et al. (33)	1997	20	1	5	0	0
Waldhausen and Tapper (34)	1997	10	0	0	0	0
Curran et al. (15)	1998	7	0	0	0	0
Geiger et al. (48)	1998	39	0	1	0	0
Rescorla et al. (36)	1998	32	1	0	0	0
Al-Salem (56)	1999	115	7	5	0	0
Minkes et al. (28)	2000	17	0	1	0	0
Reddy et al. (37)	2001	29	1	2	0	0
Total		369	18 (4.9%)	18 (4.9%)	0	0

(C) Comparison of cumulative complication rate

	Open $N = 369$	Laparoscopic $N = 344$	*p*-Value
Grade I	18	10	0.18
Grade II	18	23	0.30
Total I and II	36	33	0.94

Although there have been no Grade III or IV complications in the pediatric laparoscopic series reported to date, there have been serious complications in adult reports. Bernard et al. (59) in a series of 20 patients had two serious complications: one death from intraoperative hemorrhage and one delayed the presentation of a diaphragmatic hernia and gastric perforation. Although most splenectomies in children are performed

for benign conditions, adults often have lymphoma, myelodysplastic syndrome, leukemia, or HIV associated thrombocytopenia, a fact which undoubtedly affects morbidity and mortality. The operative complications in adults with OS are also significant and appear related to the disease process. A recent report (60) of 83 OSs in adults noted a direct correlation between splenic weight and blood loss. They observed a postoperative complication rate of 27% and mortality of 6%; however, in patients with myelofibrosis there was a 50% complication rate and a 21% mortality rate. This is similar to a somewhat earlier series of 142 OSs in adults noting a 22% complication rate and 6% mortality rate (61). Berman et al. (32) in a multiinstitutional review reported 22 patients undergoing LS for malignancy observed longer operative time (conversion rate, 41%); however, noted similar morbidity, mortality, and transfusion requirements compared with OS. They also noted shorter length of stay (4 vs. 6 days) with laparoscopy. Few reports directly address splenomegaly in children. In North America, splenomegaly is usually associated with spherocytosis and sickle cell anemia, whereas, in other parts of the world thalassemia is a much more common cause. Al-Salem (56) in a recent report of 115 children undergoing OS included 20 with massive splenomegaly ($\geq$1000 g) and noted no mortality; however, there was a higher morbidity (10%) with massive splenomegaly compared with a 6.3% rate for splenomegaly.

8. CONCLUSIONS

The available evidence in the literature, although limited, indicates that LS is comparable to OS for control of disease including detection of accessory spleens. A careful search must be made for accessory spleens and capsular disruption must not occur. The operative time for LS is longer than that for OS and, although decreasing with increased experience, is not equivalent to OS. The complication rate for LS is low and comparable to the low rate for OS. Although most studies have not evaluated procedural pain, of those patients evaluated in comparative studies, less narcotic use has been observed with the laparoscopic technique. LS is associated with a shorter length of stay than OS. There is insufficient data available to draw conclusions about return to full activity; however, one could infer that this is improved because of shorter hospital stay and decreased pain medication usage. The cosmetic appearance is improved in LS compared with OS, although this subjective factor has not been studied in any fashion. The laparoscopic technique has been validated as an acceptable approach for splenectomy and in many aspects is superior to the open technique.

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11

Laparoscopic Adrenalectomy in Children: An Outcomes Analysis

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1. INTRODUCTION

Laparoscopic adrenalectomy has been adopted as the standard of care in adults and is becoming a standard of care in children. It was first described by Gagner in 1992 (1). Since then, there have been several comparisons between open and laparoscopic adrenalectomy in adults (2–16). These studies are all retrospective or case–control studies. There are no prospective randomized studies. Despite this limitation, they show that patients undergoing laparoscopic adrenalectomy benefit from decreased hospital stay, less pain, less morbidity, faster return to activities, and decreased overall costs. Currently, there are not many studies in children; although two of the earlier studies include a few pediatric patients (2,16). The studies in children (17–20) show similar results to those of the adults.

2. TECHNICAL CONSIDERATIONS

The unique anatomy of the adrenal gland lends it to multiple operative approaches. This complicates direct comparison of the various techniques. Traditional open approaches are anterior transperitoneal, flank transperitoneal, flank retroperitoneal, and posterior retroperitoneal. Each approach has its unique advantages. The flank and posterior approaches are

generally less invasive; however, the anterior approach provides generous exposure for malignancies and large tumors. Likewise, these four approaches may be used laparoscopically. There are no comparisons of all of these techniques in one study. The differences in pain, hospital stay, cost, and morbidity among the three open approaches are greater than the differences among the laparoscopic approaches, though there are no comparisons among the various laparoscopic approaches in children.

The author prefers to use a modified lateral transperitoneal approach. This technique provides generous exposure of the adrenal gland and the peritoneal contents. The patient is placed in a modified lateral decubitus position. The distance between the iliac crest and the costal margin is increased either by placing a rolled blanket under the operative side or by flexing the table (Fig. 11.1). The advantage of this technique is that you may easily manipulate the patient during the procedure from a true lateral position to the supine position. The right adrenal is approached using four trocars: umbilical, mid-clavicular, anterior axillary, and mid-epigastric. A fifth trocar may be needed for retraction. The mid-epigastric port is used for a liver retractor. The camera is placed in the mid-clavicular port, and the surgeon operates through the anterior axillary and umbilical ports. The adrenal is approached after mobilizing the duodenum and the right lobe of the liver. The vena cava is identified, and the adrenal vein can be easily isolated and ligated with clips or impedance feedback bipolar electrocautery device (Ligasure, Valleylab, CA, USA). Once the vein is ligated, the gland is dissected medially to laterally, dividing the remaining vessels with clips or electrocautery.

The modified lateral approach to the left gland is performed with three or four trocars. After an initial trocar is placed in the umbilicus, trocars are placed in the mid-clavicular and anterior axillary lines. The camera is then moved to the mid-clavicular trocar. A fourth trocar may be placed in the mid-epigastrum for retraction, if necessary. The splenic flexure of the colon is mobilized. The inferior pole of the spleen is then freed from its retroperitoneal attachments, and Gerota's fascia is entered. Larger tumors

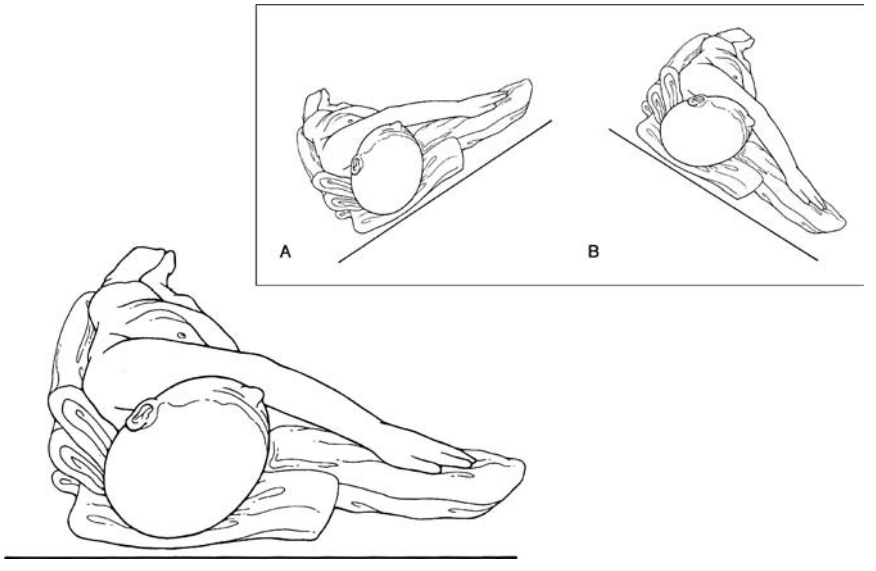


Figure 11.1 Modified lateral decubitus position set up by placing a roll under the patient's shoulder and hip. (A) Table tilted right resulting in a relatively supine patient position. (B) Table tilted left resulting in the lateral decubitus position.

will displace the spleen superiorly and anteriorly. It may be necessary to mobilize the lateral attachments of the spleen in smaller tumors to gain access to the gland. The spleen and the tail of the pancreas are rotated medially. The key to the dissection is to exploit the plane between the kidney and the adrenal gland. The posterior and lateral attachments of adrenal gland are left alone, and the dissection is continued anterior and inferior to the gland to expose the adrenal vein. The vein is then ligated as mentioned previously, and the remainder of the adrenal gland dissected free using electrocautery for hemostasis.

The retroperitoneal approaches to the adrenal glands are described elsewhere (21). These approaches are best for small tumors. There are theoretical advantages to the retroperitoneal approach because of the fact that the peritoneal cavity is not violated. This has not been substantiated in any outcomes study. Yoneda et al. (22) compared the lateral transabdominal approach with a posterior retroperitoneal approach in sixteen patients. The only statistically significant difference he found between the two approaches was a longer operative time for the retroperitoneal approach (129 vs. 269 min). There were no differences with respect to blood loss, time to oral intake, or time to ambulation. It does not appear that there are any clinically relevant benefits to the more technically challenging and longer retroperitoneal approach.

3. ADULT EXPERIENCE

In 1999, a review article by Smith et al. (23) declared laparoscopic adrenalectomy the new gold standard in adults. As of that time, there had been nearly 600 cases of laparoscopic adrenalectomy reported in the literature. It is clear from these articles that laparoscopic adrenalectomy is safe and efficacious in benign disease including pheochromocytoma. There are little or no data for laparoscopic adrenalectomy for malignant disease. There are scattered reports of patients with malignant disease treated with laparoscopic resection; however, there are no long-term results relating to recurrence rates. Most authors recommend that malignant disease should be resected by an open technique. This may be different in the pediatric population as it relates to neuroblastoma because of the biological nature of the disease.

Prior to laparoscopy, the posterior approach to the adrenal glands was gaining favor as a minimally invasive approach to adrenal disease. In studies comparing open posterior vs. laparoscopic adrenalectomy, Thompson et al. (12), in a case–control study of 100 patients, and Dudley and Harrison (16), in a retrospective study of 46 patients, demonstrated a shorter hospital stay in the laparoscopic group although the laparoscopic group had a longer operative time. Acosta et al. (24), in a study of 59 patients, demonstrated no significant differences in hospital stay or operative time in patients undergoing unilateral adrenalectomy. They noted significantly longer operative times in patients undergoing bilateral adrenalectomy. The clinical significance of longer operative times is not clear.

In summary, the adult literature supports laparoscopic adrenalectomy for benign disease. There are several authors who have removed malignant tumors, although there is no long-term follow-up.

4. PEDIATRIC EXPERIENCE

Seven published articles were reviewed (17–21,25,26). Of the seven articles, four are case reports. In all, 30 patients are reported (Table 11.1). In this analysis of the pediatric

Table 11.1 Summary of Reports of Pediatric Adrenalectomy

	Number of patients	Mean age (years)	Mean tumor size (cm)	Number converted to open
Shanberg et al. (17)	3	NR[a]	≤ 5	1
Pretorius et al. (18)	1	12	4	0
Clements et al. (19)	1	12	4×3.5	0
Schier et al. (20)	1	14	NR[a]	0
Mirallie et al. (21)	6	9.5	4	2
Radmayr et al. (26)	1	8	NR[a]	0
Miller et al. (25)	17	9.8	4.8	1

[a]NR, not reported.

literature, there were four conversions (12.5%) to open procedures. The reasons for conversion to an open procedure included poor control of the right renal vein (17), tumor thrombus in the right renal vein secondary to an adrenal carcinoma (25), and two conversions for difficulty in gaining exposure on the left side. Laparoscopic adrenalectomy was performed for a variety of diagnoses (Table 11.2). The most common was pheochromocytoma (11), and all were cured of their disease. There were four adrenalectomies performed for malignant disease (three neuroblastomas, one adrenocortical carcinoma). There have been no local or distant recurrent tumor metastases in those patients with neoplasms. The postoperative stay for all patients ranged from 1 to 6 days. Most patients were discharged on the second postoperative day. There were no major complications reported.

These studies represent a small number of pediatric patients with a distribution of disease that is somewhat different from the adult studies although malignant disease was included. This is likely because of the biological nature of the common pediatric malignancy of the adrenal gland, neuroblastoma, when compared with adrenal carcinomas, which are more common in adults. Localized small neuroblastomas are not invasive, and tumor "spillage" is not an issue. Adult surgeons are also less likely to encounter ganglioneuromas. It is reasonable to assume that the morbidity of laparoscopic adrenalectomy is relatively independent of age for similar diagnoses. It also appears, from both the adult and the pediatric literature, that the recovery time after laparscopic adrenalectomy is relatively rapid. Although the number of pediatric patients is small, it seems to follow that the conclusions of the large adult studies appear to be valid for the pediatric population. This assumption may not be valid for laparoscopic adrenalectomy for malignancy, as the nature of pediatric adrenal malignancies is so different from adult adrenal malignancies.

Table 11.2 Diagnoses (Number) of Pediatric Patients Undergoing Laparoscopic Adrenalectomy Based on Reported Literature (17–21,25,26)

Pheochromocytoma (9)
Ganglioneuroma (9)
Cushing's disease (6)
Adenoma (5)
Neuroblastoma (1)
Adrenocortical carcinoma (1)
Paraganglioma (1)

5. CONCLUSION

Laparoscopic adrenalectomy appears to have the same benefits in children as it has in adults. This procedure is becoming the standard of care in children with diseases of the adrenal gland. It has been successfully demonstrated that laparoscopic techniques may be used to treat multiple diseases of the adrenal gland in children, including pheochromocytomas and malignancies. It is reasonable to extrapolate from the adult studies that children undergoing laparoscopic adrenalectomy have a shorter hospital stay and less operative morbidity than those who undergo an open procedure. The current pediatric reports support the conclusions of the adult literature. Long-term results appear to be satisfactory following laparoscopic adrenalectomy in children for a variety of diagnoses, although more long-term follow-up and outcome analyses are needed to solidify these conclusions.

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12

Outcomes Following Laparoscopic Pyloromyotomy for Infantile Hypertrophic Pyloric Stenosis

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Infantile hypertrophic pyloric stenosis (IHPS) is a common surgical condition encountered in the neonatal period, with an estimated incidence of two to three cases per 1000 live births (1). The surgical technique of extramucosal pyloromyotomy for the treatment of IHPS was first described by Ramstedt in 1912 (2). This operation is technically straightforward, associated with minimal morbidity, and remains the treatment of choice for IHPS nearly a century after it was first described. Treatment of IHPS with laparoscopic pyloromyotomy (LP) was first performed in France nearly 15 years ago, and a number of surgeons from around the world have since reported their experience using this approach (3). Proponents of LP have cited improved cosmesis, quicker return to full feeding, and a shorter hospital stay when compared with open pyloromyotomy (OP) (4–6). Although a number of early reports have suggested the superiority of LP over OP, other studies have found the two approaches to be similar with respect to these outcomes (7–10). Furthermore, concerns have been revised regarding the safety of LP, particularly with the reported increased risk of (unrecognized) mucosal perforation (9,11). At present, there remain unanswered questions regarding the safety, efficacy, and magnitude of benefits for LP when compared with the conventional OP approach. The purpose of this chapter is to clarify outcomes relating to postoperative recovery, efficacy, and complication rates for LP compared with OP through a review of the available clinical evidence.

1. LITERATURE REVIEW

At the conception of this chapter (November, 2004), 10 clinical studies were identified from the National Library of Medicine's Pubmed database comparing outcomes between LP and OP (Table 12.1) (4–13). These studies reported collectively on 290 patients who underwent LP and 403 patients who underwent OP. For the purpose of minimizing the influence of selection bias on outcome measures, only studies reporting comparison data between LP and OP were included in this review (studies reporting on outcomes of LP or OP alone were excluded). Seven of the 10 clinical reports were retrospective cohort studies (7–13), two were prospective cohort studies (5,6), and only one study was a randomized clinical trial (4). Outcomes of interest were those pertaining to operative time, postoperative recovery (time to full feeds and length of hospital stay), efficacy (rates of conversion for the LP group and rates of inadequate pyloromyotomy for either group), and safety (perioperative and postoperative complications). There were no significant differences in baseline demographic or electrolyte parameters between infants treated with LP and those treated with OP in any of these studies. A formal meta-analysis was not attempted for any outcome measures owing to the heterogeneous and predominantly retrospective nature of the studies included in this review.

2. DURATION OF SURGERY

The duration of surgery was reported for both groups in all 10 studies, although not all studies clearly described how this outcome was defined (e.g., total anesthetic time, time from incision to dressing, etc.). The range of operative duration for LP was 19–50 min, whereas the range for OP was 19–33 min (Table 12.1). Two of the studies found a significant difference suggesting LP was the quicker procedure (both studies being prospective, including the RCT), whereas two other studies found a significant difference favoring the open approach. The other six studies found no significant difference between the outcomes. The absolute difference in mean operative time was negligible in most studies, with only two of the eight studies reporting a difference of >5 min. Interestingly, the collective results from these studies did not suggest a trend of decreased operative time for LP with either larger experiences (assumption of greater experience with LP) or more contemporary experiences (assumption of better equipment to facilitate LP).

3. POSTOPERATIVE RECOVERY

Eight of the 10 studies reported the length of postoperative time before full feedings were tolerated (Table 12.1). Patients treated with LP were able to tolerate full feedings sooner than those treated with OP in all eight studies, although a significant difference favoring LP was found in only two studies (both of these were prospective, including the RCT). The time to achieve full feeds was quite variable across the eight studies, ranging from 4 to 35 h in the LP group and from 9 to 61 h in the OP group. Despite this apparent heterogeneity, there was a consistently small (and likely negligible) difference in the time to achieve full feeds between groups in each study. A difference of 5 h or less was observed in six of the eight studies, reporting this outcome (including the RCT).

With respect to postoperative feeding outcomes, any generalizations must be made in the context of important observations regarding the methodology used in these studies. Although all eight studies provided adequate details regarding postoperative feeding

Table 12.1 Demographic and Perioperative Outcome Data from 10 Clinical Studies Comparing LP and OP for the Treatment of IHPS

			Patients (n)		OR time (min)		Time to full feeds (h)		Length of stay (h)		Conversion rate (%)
Study	Year	Type	LP	OP	LP	OP	LP	OP	LP	OP	LP
Greason et al. (10)	1995	RC	14	11	25	26	19	23	NR	NR	0% (0/14)
Scorpio et al. (6)	1995	PC	26	37	29	27	NR	NR	**41**	**64**	NR
Ford et al. (9)	1997	RC	33	51	41	28	32	41	NR	NR	3% (1/33)
Greason (4)	1997	RCT	10	10	**19**	**24**	**4**	**9**	23	25	0% (0/10)
Bufo et al. (12)	1998	RC	29	125	26	30	14	15	**26**	**34**	NR
Sitsen et al. (11)	1998	RC	26	21	**32**	**19**	NR	NR	70	74	8% (2/26)
Fujimoto et al. (5)	1999	PC	29	30	**27**	**32**	**35**	**61**	NR	NR	3% (1/29)
Campbell et al. (8)	2002	RC	65	52	38	33	19	20	31	28	8% (5/65)
Caceres and Liu (7)	2003	RC	28	28	36	33	24	27	60	62	7% (2/28)
Hall et al. (15)	2004	RC	39	38	**50**	**30**	24	26	48	48	8% (3/39)

Note: NR, not reported, Data in bold indicate that a significant difference was found for that outcome in the referenced study ($p < 0.05$). Of note, no statistical analysis of outcome measures was attempted in the study by Ford et al. (9).

protocols, there was little, if any, detail given to how protocols were modified when feeding intolerance was present (or how intolerance was defined for this purpose). Furthermore, there was significant heterogeneity in the postoperative feeding protocols across different studies. This was not only the case with respect to when and by what modality feeding would begin, but also how feedings would be advanced over time.

The total length of hospital stay (LOS) was reported for both groups in seven of the 10 studies. The duration of hospitalization was found to be shorter for the LP group in five of these studies, although this difference was significant in only two studies. For the three prospective studies, one found a significant difference favoring the LP group, whereas the other two (including the RCT) did not find a difference in this outcome. In general, differences in the duration of hospitalization were minimal (and likely clinically negligible), with most studies reporting differences of 4 h or less between groups. As was the case with postoperative feeding data, significant variability was observed between studies with respect to LOS (range for LP: 23–70 h and range for OP: 28–74 h).

Several observations regarding the methodology used in these studies must be kept in mind when drawing conclusions from LOS data. It is noteworthy that none of the studies clearly described their discharge criteria or who was responsible for determining when these criteria were met. There likely exists significant heterogeneity between different centers and over different time periods with respect to such practice patterns, and this may explain much of the observed variability. Although the number of studies in this review is relatively few, it was interesting that there appeared to be no correlation between a quicker return to full feeding and a shorter hospital stay. In both studies reporting a significantly shorter period of time to full feeds in the LP group, the total LOS was not significantly different between groups. Conversely, the length of time to achieve full feeding was not significantly different between groups in the two studies, demonstrating a shorter LOS for the LP group.

4. EFFICACY

For the purpose of this review, a pyloromyotomy was considered technically successful when an interval procedure was not required for inadequate pyloromyotomy. Intraoperative conversion to an open procedure due to a complication or the inability to perform the operation safely was also considered a technical failure for the LP group. Of the 290 patients who collectively underwent LP in this review, 18 (16%) patients from six different studies were considered technical failures by these criteria. The individual failure rates for each of the eight studies reporting efficacy data ranged from 0% to 5%. Of the 18 technical failures for LP, 14 (5%) were due to intraoperative complications requiring an open conversion (most commonly due to a mucosal perforation) and four (1%) were due to inadequate pyloromyotomy. Of the collective 403 patients who underwent OP as initial treatment, only one patient (0.2%) required a reoperation for inadequate pyloromyotomy.

5. COMPLICATIONS

Data regarding complication rates for LP and OP were available from all 10 studies (Table 12.2). None of the studies was able to demonstrate a significant difference in overall complication rates between groups. With respect to intraoperative complications, mucosal lacerations appeared to be more common in the LP group and serosal lacerations were more common in the OP group (although neither significantly so). Of the nine

Table 12.2 Complication Profiles for 10 Published Studies Comparing LP and OP for the Treatment of IHPS

			Perioperative complications						Postoperative complications					
	Total complications		Mucosal perforation		Serosal laceration		Inadequate PM		Protracted emesis		Wound infection		Other	
Study	LP	OP	LP	OP	LP	OP	LP	OP	LP	OP	LP	OP	LP	OP
Greason et al. (10)	0/14 (0%)	1/11 (9%)	0	0	0	0	0	0	0	0	0	1	0	0
Scorpio et al. (6)	1/26 (4%)	3/37 (8%)	0	0	0	0	0	0	NS	NS	0	2	1	1
Ford et al. (9)	7/33 (21%)	5/51 (10%)	3	3	0	0	1	1	NR	NR	1	1	2	0
Greason (4)	0/10 (0%)	1/10 (10%)	0	0	0	1	0	0	0	0	0	0	0	0
Bufo et al. (12)	1/29 (3%)	4/125 (3%)	0	0	0	0	0	0	NR	NR	0	2	1	2
Sitsen et al. (11)	9/26 (35%)	5/21 (24%)	3	2	0	0	2	0	2	2	2	1	0	0
Fujimoto et al. (5)	1/29 (3%)	5/30 (17%)	1	0	0	3	0	0	SD	SD	0	2	0	0
Campbell et al. (8)	11/65 (18%)	6/52 (12%)	1	0	0	0	1	0	4	3	3	0	2	3
Caceres and Liu (7)	1/28 (4%)	4/28 (14%)	0	0	0	0	0	0	1	3	0	1	0	0
Hall et al. (13)	6/39 (15%)	10/38 (26%)	1	2	1	1	0	0	2	1	1	4	1	2
Total incidence	37/290 (13%)	42/403 (11%)	9 (3%)	7 (2%)	1 (<1%)	5 (1%)	4 (1%)	1 (<1%)	9 (3%)	9 (2%)	7 (2%)	14 (3%)	7 (2%)	8 (2%)

Note: NR, not reported; Numbers in the column under "protracted emesis" indicate the number of patients in the study who were deemed to have abnormal postoperative emesis (the criteria for this diagnosis were variable and often ambiguous across studies). SD, a significant difference was found in this study favoring the LP group, although this was based on comparing the proportions of all feedings associated with emesis. NS, reported in the study as "not significant," although the actual data were not available.

duodenal perforations in patients treated with LP, three were not detected intraoperatively. These patients underwent reoperation following the development of peritonitis, but ultimately did well and suffered no long-term morbidity. All seven of the duodenal perforations noted in the OP group were detected and managed during the initial procedure. The overall incidence of perforation was relatively rare, however, occurring in $<3\%$ of cases in both treatment groups. Inadequate pyloromotomy requiring reoperation was four times more common in the LP group, although this complication was also a fairly rare event (occurring in $<1\%$ of cases in both groups).

Postoperative complication profiles were very similar between groups with respect to wound infections and non-wound related complications (pneumonia and other respiratory complications). Of the eight studies reported on postoperative emesis, only one found a significant difference in favor of the laparoscopic group (5). However, this study compared the overall proportions of feedings associated with any degree of emesis, which may not in itself be clinically significant. Of the six studies (including the RCT) which examined postoperative emesis in the context of a clinically relevant endpoint (e.g., delayed hospitalization or need for further work-up), none found a significant difference between those treated with LP or with OP. In general, comparing the incidence of clinically relevant emesis between studies was difficult given the variable and often ambiguous definition of this complication.

6. CONCLUSIONS

From the currently available data, we conclude that both operative approaches appear to provide a safe and efficacious treatment for IHPS. There appear to be no clinically relevant differences in operative time, measures of postoperative recovery, or complication profiles between the two operations. Serious complications such as duodenal perforation were fairly uncommon in both groups, although it is important to note that all three unrecognized perforations occurred in the LP group. It has been proposed that the loss of tactile feedback and direct visualization in the laparoscopic environment may increase the risk of duodenal perforation and at the same time decrease the surgeon's ability to recognize this complication when it does occur. This underscores the importance of adequate training (and mentorship) to successfully negotiate the learning curve for LP.

The importance of experience in achieving satisfactory outcomes for LP was recently highlighted by van der Bilt et al. (14) in their review of 182 LPs performed over a 9 year period. Although this was not a study comparing LP with OP (therefore not included in this review), this study represents the single largest experience of LP to date. The authors compared 36 LPs performed between 1993 and 1996 with 146 LPs performed between 1996 and 2002. They found a significant difference in complication rates between these two periods, with the incidence of mucosal perforations decreasing from 8.7% to 0.7% and the incidence of inadequate pyloromyotomy decreasing from 8.7% to 2.7% over time.

The conclusions of this review mirror those offered by Hall et al. (15) in their recent meta-analysis of studies comparing LP and OP. Eight of the studies examined in this review were the focus of their analysis, and they found no statistically significant differences in operative time or complication rates. They did report a statistically significant difference favoring the laparoscopic group with respect to time to full feeds and LOS, although they did not comment on the magnitude of treatment difference for either outcome. As discussed previously, the absolute difference in these outcomes was negligible in most studies, and a statistical difference, if one exists, does not necessarily translate into a clinically relevant difference. Furthermore, we caution the use of meta-analysis

for data sets which are predominately retrospective and uncontrolled. Using such techniques to analyze data in which the component studies may already contain significant bias could potentially provide even less valid conclusions.

On a final note, the results of this review highlight the fact that very little high-quality evidence exists to base treatment decisions in pediatric surgery. As is the case for most interventions in our field, the available data are limited to predominantly retrospective, single-institutional case-series experiences. From these observations, several pediatric surgeons have proposed that a formal multiinstitutional randomized trial be conducted to discern the treatment differences between LP and OP (6,9,15). These differences, if they do exist, are likely to be quite small and with perhaps little clinical relevance (e.g., showing a statistical difference in operative time by a few minutes or time to discharge by a few hours). Furthermore, any differences in outcomes such as cost and analgesia requirements (which have not been appreciably examined in any study to date) are also likely to be very small. As such, a formal trial may not be justified from the standpoint of practicality, and multicenter efforts should probably be conserved for more complex pediatric surgery anomalies.

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13

Laparoscopic Fundoplication in Infants and Children

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1. INTRODUCTION

Since the first laparoscopic fundoplication in 1991 (1), the frequency with which complex minimally invasive procedures are being performed has increased at an unprecedented rate. Fortunately, the art of pediatric surgery has kept pace with these surgical techniques primarily due to the advancements made in smaller operating telescopes and improvements in minimally invasive instruments developed specifically for the body cavities of infants and children.

Infants and children are commonly affected by symptomatic gastroesophageal reflux (GER). The ability to differentiate pathologic GER from "physiologic" vomiting in infancy is sometimes difficult. Pathologic GER can present as a single or as multiple clinical conditions resulting in significant morbidity or possible near-fatal events. Physiologic vomiting, likely due to an incompetent lower esophageal sphincter (LES) mechanism, is normal during the first year of life (2). However, physiologic vomiting is a self-limited

process which usually resolves by 18 months of age and usually does not affect the growth or development of the infant.

Due to swallowing dysfunction, pediatric patients may require a temporary or permanent route for enteral nutrition, which is usually accomplished with a gastrostomy. When these patients are considered high operative or anesthetic risks, or are neurologically impaired, they should undergo a standard evaluation for GER prior to creation of a gastrostomy to assure that there is no need for simultaneous fundoplication.

When pathologic GER exists, laparoscopic Nissen fundoplication (LNF) is effective in treating children with pathologic GER (3–6). LNF, with or without concomitant gastrostomy, is becoming the preferred surgical approach in infants and children requiring surgical correction of GER.

2. PATHOPHYSIOLOGY OF GER

The mechanisms responsible for GER are not completely understood. It has been well established that prolonged exposure of the esophageal mucosa to gastric acid and other secretions induces injury. The extent of this damage depends on the susceptibility of the esophageal mucosa to injury, the length of time the refluxed material is present within the esophagus, and the volume of acid refluxed into the esophagus. In normal individuals, an antireflux mechanism exists at the gastroesophageal junction, creating a barrier to GER and subsequent injury. This barrier is composed of several factors that have been extensively studied (Table 13.1).

The most important component of this barrier is the LES. Developmentally, the LES arises from the inner circular muscle layer of the esophagus, which is asymmetrically thickened in the distal esophagus. This thickened muscle layer creates a high pressure zone which can be measured manometrically. In addition, this muscular thickening extends onto the stomach more prominently on the greater than lesser curvature (7). The phrenoesophageal membrane, arising from the septum transversum of the diaphragm, and the collar of Heveticus, originating from the oblique muscle fibers, hold the LES in position. The result is an LES that lies partially in the chest and partially in the abdomen. This positioning is important for the normal barrier function against GER. Esophageal manometry can identify this transition (which is known as the respiratory inversion point) from the thoracic esophagus to the abdominal esophagus.

Table 13.1 Factors Which Provide Protection Against Gastroesophageal Reflux

Barriers to GER	Reason for failure
Lower esophageal sphincter	Malposition (intrathoracic in location)
	↓ Pressure (<6 mmHg)
	↓ LES length (<2 cm)
	↓ LES abdominal length (<1 cm)
	Transient relaxations
	Abnormal smooth muscle function
Intra-abdominal esophageal length	Short length (<3 cm)
Angle of His	More obtuse angle (gastrostomy)
Esophageal motility	Inefficient clearance
	Poor smooth muscle function (e.g., scleroderma)

Note: GER, gastroesophageal reflux; LES, lower esophageal sphincter.

The LES is an imperfect valve that creates a pressure gradient in the distal esophagus. The ability to prevent GER is directly proportional to the LES pressure and its length, provided that LES relaxation is normal. In one study, LES pressures >30 mmHg prevented GER as documented by a 24 h pH study, while pressures between 0 and 5 mmHg correlated with abnormal pH studies in $>80\%$ of patients (8). In normal subjects, GER is more likely to develop if the LES pressure and length fall below the 2.5th percentile, which is 6 mmHg at the respiratory inversion point or an overall LES length of 2 cm, of which 1 cm is intra-abdominal GER (9). As noted previously, the LES is relatively fixed across the esophageal hiatus by its surrounding attachments. Malposition of the LES, which can occur with a hiatal hernia or abnormal development, results in loss of the protective function of the LES and GER can occur. Finally, LES relaxation occurs with esophageal peristalsis initiated by the swallowing mechanism. This relaxation must occur and is normal. Inappropriate LES relaxations, referred to as transient LES relaxations, have been shown to occur sporadically, unassociated with the swallowing mechanism. Interestingly, when children with symptoms of GER were studied with pH and manometry simultaneously, reflux episodes rarely correlated with decreased LES pressures. Rather, the majority of reflux episodes occurred during transient LES relaxations, and no reflux episodes were identified during LES relaxation after swallowing with normal peristaltic sequence (10,11). There continues to be growing support for identifying these LES relaxations as a primary mechanism of GER.

In summary, the barrier function of the LES is imperfect but highly effective. Short LES length, abnormal smooth muscle function, increased frequency of transient LES relaxations, and LES location within the chest can contribute individually or in combination leading to LES failure and GER. Disruption of the LES from a hiatal hernia, abnormal LES function, or previous esophageal surgery can all lead to poor function and subsequent GER.

Another barrier to the development of symptoms of GER is the intra-abdominal length of the esophagus (12). Although no absolute effective intra-abdominal esophageal length has been identified that prevents GER, correlation between several lengths and GER has been identified. In one report, an intra-abdominal length of 3–4.5 cm in individuals with normal abdominal pressure provided LES competency 100% of the time (8). A length of 3 cm was sufficient in preventing reflux in 64% of individuals, while <1 cm of intra-abdominal esophagus resulted in reflux in 81% of patients. This becomes very important when considering surgical correction for GER. Failure to mobilize adequate esophageal length for intra-abdominal positioning can lead to less than successful results or recurrent GER.

A third barrier to reflux is the angle of His, which is the angle at which the esophagus enters the stomach. The usual orientation is that of an acute angle which creates a flap valve at the gastroesophageal junction. Although the actual functional component of the angle of His is not well known, it has been shown to provide resistance to GER. Experimentally, when this angle is more obtuse, GER is more prone to develop. Conversely, accentuation of the angle inhibits GER (13). The ability of the angle of His to prevent GER may be diminished as a result of abnormal development or may be iatrogenic as occurs after gastrostomy placement.

The final factor involved in the pathophysiology of GER is the ability of the esophagus to effectively clear luminal contents. When impaired esophageal motility is present as a result of either abnormal smooth muscle function, impaired vagal stimulation or with obstruction, gastric acid, which has been refluxed into the esophagus, is not moved caudally into the stomach in a timely manner. This prolonged exposure can lead to esophageal mucosal injury as well as can potentiate the motility disturbance due to vagal and/or smooth muscle inflammation or injury.

3. CLINICAL MANIFESTATIONS

There is a great deal of variability in the clinical presentation of GER in infants and children. The surgeon must be cognizant of the patient's age and associated medical conditions when considering if the symptomology is a manifestation of pathologic GER. The frequency of symptoms seen in infants requiring surgical intervention was assessed recently by Tovar et al. (6) (Table 13.2).

Persistent regurgitation remains the most common symptom of GER of infants (14). Persistent regurgitation as a result of GER can lead to complications. Significant malnutrition and failure to thrive can result from insufficient caloric intake due to vomiting. In infants, however, vomiting can be "normal." This benign variant of vomiting, known as chalasia of infancy, is seen early in life, usually during burping, after feeding, or when placed in the recumbent position (15). Chalasia of infancy does not interfere with normal growth or development and rarely leads to other complications. It is a self-limited process with most infants transitioning to being asymptomatic by 2 years of age or near the time of initiating solid foods (16). No treatment is indicated in this instance and no diagnostic evaluation is necessary.

In infants, another presenting symptom is irritability due to pain. Painful esophagitis can be the result of the acid refluxate. Discomfort leads to crying despite consoling measures (17,18). Occasionally, small volumes of feeds briefly assists in alleviating pain; however, this is not a lasting effect (12,13). In contrast to infants, children with pathologic GER more often present with pain. As in adults, the pain is retrosternal in nature, often described as heartburn. Long-standing GER with esophagitis can lead to chronic inflammation or even ulcer formation with eventual scarring and stricture formation. Dysphagia is the end result due to the narrowed esophageal lumen. Obstructive symptoms and pain are the two most common associated complaints when an esophageal stricture is present (19,20).

Barrett's esophagitis is a premalignant condition in which metaplasia occurs in the esophageal squamous epithelium leading to replacement with columnar epithelium. In adults, it is thought to be the result of chronic esophageal injury by gastric acid reflux (21–23). Fortunately, it is rare in infants and children. However, when it does develop, serious complications often result. In addition to the increased risk for adenocarcinoma, ~50% of these patients will develop stricture and may well develop ulcers (23,24). Aggressive GER management, along with vigilant long-term surveillance, must be pursued in an effort to minimize these often difficult and possibly fatal complications.

Respiratory symptoms are commonly seen in infants and children. Delineating the role of GER as an etiologic entity for ongoing respiratory complaints in these patients can be difficult because of the variability of the symptoms often seen with other pulmonary diseases. Chronic cough, wheezing, choking, apnea or near sudden infant death syndrome (SIDS) can all be symptoms attributable to GER. Recurrent bronchitis or pneumonia can

Table 13.2 Frequency of Symptoms in Children Undergoing Operation (6)

Symptom	Percentage (%)
Regurgitation	81
Pain or dysphagia	30
Respiratory disease	41
Hemorrhage	7

occur from aspiration of refluxate (26–29). Acid stimulation within the esophagus causes vagally mediated laryngospasm and bronchospasm, which clinically presents as apnea or choking, or mistakenly as asthma (30,31). Esophageal inflammation, as seen with esophagitis, likely enhances this mechanism (32,33). Hemorrhage is uncommonly the presenting symptom of GER. Esophagitis, gastritis, and ulcer formation can lead to hematochezia or melana in a small percentage of infants or children (6).

Regardless, the patients' symptoms remain the most important factor in determining the surgical treatment of GER. The responsibility lies with the surgeon to evaluate these symptoms in relation to GER and have an accurate diagnosis prior to offering surgical intervention.

4. DIAGNOSTIC EVALUATION

Once the clinical history raises the suspicion of GER as the etiologic cause of the patient's complaints, the diagnostic evaluation should be initiated. Upper gastrointestinal radiography is the most frequent initial study employed. Evidence for reflux is often seen on the exam; however, the presence or absence of reflux is an extremely poor indicator of GER as a cause of the patient's symptoms. The contrast study is most useful for delineating the anatomy of the esophagus and esophagogastric junction as well as evaluating esophageal clearance. In addition, it crudely assesses esophageal and gastric motility and can identify the presence of esophageal strictures or webs. Distal obstruction, such as duodenal obstruction (web, stenosis), antral web, or malrotation, is also excluded as a cause of the symptoms.

Twenty-four hour pH monitoring is the gold standard for establishing the diagnosis of GER, especially in infants and children whose history is unclear. It provides the diagnosis of GER in cases where clinical history cannot be obtained or is confusing, such as a patient presenting with respiratory symptoms only. The study is performed by placing an electrode $\sim$2–3 cm proximal to the GE junction and measuring the pH in the distal esophagus. Although initially developed in adults, its use in children is now accepted and invaluable (34,35). The accuracy of the exam is dependent on the cessation of all antireflux medication. Proton pump inhibitors should be withheld for seven days and histamine receptor blockers are stopped 48 h prior to the exam. A reflux episode is considered to have occurred if the esophageal pH is recorded as <4. Ideally, the exam should occur over an uninterrupted 24 h period. The pH is continuously monitored via the esophageal electrode while the patient's position (upright and supine) and activities (awake, asleep, eating) are recording simultaneously. The final score is calculated based on the percent of the total time that the pH was <4, the total number of reflux episodes, the number of episodes lasting longer than 5 min, and the longest reflux episode (36,37). A normal range of values has been established, which is easily reproducible and reliable (38).

Although essential in adults, esophageal manometry is infrequently utilized in the pediatric population. When employed, the study measures the motility of the esophagus and the pressure at the LES via a multiport pressure transducer placed in the esophagus and transversing the LES. The clinical data accumulated in adult patients has revealed several important points that are likely referable to infants and children with GER. First, it has been shown that esophageal clearance of refluxed gastric contents is accomplished primarily by pharyngeal swallowing rather than by secondary and tertiary peristalsis as previously believed (39). In addition, through the use of concomitant 24 h pH study and esophageal manometry, it has been shown that there is a direct relationship between worsening esophagitis secondary to GER and deterioration of esophageal motility.

Manometric evaluation has been particularly useful in documenting abnormal distal esophageal motility in infants following repair of esophageal atresia with tracheoesophageal fistula (40). Hopefully, as technology continues to provide more appropriately sized instruments for sophisticated manometric studies in infants and children, the usefulness and feasibility of such studies will increase our knowledge of the physiology and abnormalities associated with GER in this population.

Endoscopic evaluation of the esophagus and stomach is occasionally used in the diagnosis of GER in infants and children. Hematemesis, dysphagia, or irritability in infants or dysphagia with or without heartburn in children should prompt esophagogastroscopy to determine if esophagitis is present. Complications of GER, including ulcer formation, esophageal stricture, and Barrett's esophagus, are also diagnosed during endoscopic examination. Mucosal biopsy may be necessary to stage the severity of esophagitis or to histologically exclude dysplasia or malignancy in Barrett's esophagus (41,42).

The relationship between delayed gastric emptying and GER in infants and children has been extensively studied and continues to be one of the more controversial aspects of antireflux surgery. The presence of delayed gastric emptying is assessed preoperatively using radionucleotide scanning via a technecium-99-labeled meal. Unfortunately, the presence of delayed gastric emptying preoperatively has not been shown to significantly improve with the addition of an emptying procedure during the antireflux procedure (43). In fact, one study evaluating patients with delayed gastric emptying undergoing fundoplication showed significantly improved gastric emptying for both solids and liquids after fundoplication *without* the addition of an emptying procedure (44). The population of neurologically impaired patients with GER has been shown to have delayed gastric emptying more often than neurologically normal children (45,46). Conflicting studies regarding the benefit and complication rates for these patients undergoing emptying procedures at the time of their fundoplication have been reported (46–48).

Currently, our evaluation of GER includes upper gastrointestinal contrast series and 24 h pH monitoring of all patients suspected of having GER. Esophagogastroscopy and esophageal manometry are employed only when circumstances suggest that the information they will provide will dictate changes in the operative management. An example of this situation is the patient with symptoms of GER but a normal pH study. The presence of esophagitis or other complications of GER would prompt surgical intervention. We do not usually employ preoperative gastric emptying studies primarily due to the improvement that has been seen and reported in gastric emptying after fundoplication (44). If symptoms of delayed gastric emptying persist after antireflux surgery, gastric emptying studies can be performed with a subsequent emptying procedure if necessary. This scenario has not occurred in our experience.

5. TECHNIQUE

The patient is placed on the operating room (O.R.) table. We utilize a robotic telescopic device (AESOP, Intuitive Surgical, Sunnyvale, CA) for operational control of the telescope and attached camera. Because of this usage, we have not found an appropriate location to secure this device if an infant is placed sideways across the O.R. table in a fashion similar to that used for a laparoscopic pyloromyotomy. Therefore, both infants and older children are positioned at the foot of the bed and AESOP is secured to the table at the level of the patient's left shoulder. It is placed in -1 tilt toward the foot of the bed and the lower limit of the robotic device is set. The patient is secured to the operating room table as the table will be placed in reverse Trendelenburg position for the

operation and the patient's bladder is emptied using a Crede maneuver. The abdomen is then prepped and draped from the patient's nipples to the pubic bone and laterally to the mid-axillary lines.

The operation is performed in a similar fashion, whether the patient is an infant or an adolescent. (The primary exception is the use of a 5-mm instrument for elevation of the left lobe of the liver in the older patient, whereas a 3-mm forceps is satisfactory for this purpose in an infant.) A 5-mm incision is made in the umbilical skin and the dissection is carried through the umbilical fascia with the cautery. Using either cautery or blunt dissection with a hemostat, the peritoneal cavity is entered and a 5-mm expandable sheath (U.S. Surgical, Norwalla, CT) is introduced into the abdominal cavity. A 5-mm cannula with a blunt trocar is then placed through this expandable sheath. The blunt trocar is removed and pneumoperitoneum is created to a maximum pressure of 15 mmHg (although 8–10 mmHg may be necessary in certain patients with congenital heart or respiratory disease). Diagnostic laparoscopy is then performed. We have adapted a technique in which the instruments are placed through the skin and the abdominal wall without the use of cannulas, much in the fashion similar to that used for laparoscopic pyloromyotomy (Fig. 13.1). Therefore, using a #11 B-P blade (Becton-Dickinson, Franklin Lakes, NJ), a stab incision is made in the right upper epigastrium through which a 3-mm locking grasping forceps in an infant and a 5-mm locking grasping forceps in an adolescent is inserted. This instrument is then placed under the left lobe of the liver; it is attached to the peritoneal side of the diaphragm and secured in placed through the locking mechanism (Fig. 13.2). This instrument is then attached to the sterile sheets with a towel clip. Another stab incision with the #11 blade is then made just to the patient's right of midline in the upper epigastrium through which a 3-mm atraumatic grasping forceps is introduced. This forceps is used in the operating surgeon's left hand for



Figure 13.1 A 5-mm cannula is shown in the umbilicus of a 4-month-old infant. The 3 mm instruments for the laparoscopic fundoplication have been placed directly through the skin without cannulas.

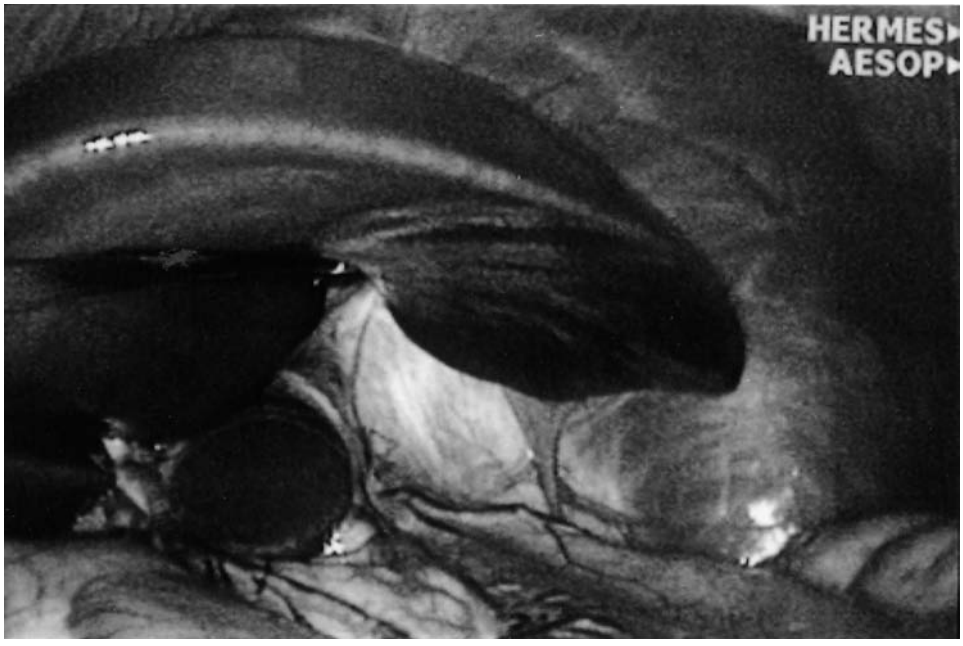


Figure 13.2 A grasping forceps is placed under the left lobe of the liver and the left lobe is elevated anteriorly exposing the esophageal hiatus.

retraction. A similar incision is then made to the patient's left of the midline almost directly across from the second incision. Through this incision, a 3-mm Maryland dissector is introduced, which is used by the surgeon as the main operating instrument. (If a gastrostomy is to be placed, it will be introduced through the abdominal wall at this site and, therefore, it is important to select the gastrostomy site prior to insufflating the abdomen). Finally, a fourth stab incision is created in the left lateral epigastrium, just beneath the left costal margin in the anterior axillary line. Through this incision, an atraumatic 3-mm grasping forceps is introduced, which is used by the assistant for retraction.

Having introduced all the instruments and elevated the lateral segment of the left lobe, the table is then turned in reverse Trendelenburg position to allow the colon and the small intestine to fall away from the area of dissection. A site is selected in the greater omentum at a point opposite the incisura through which an initial opening is made to start the ligation and division of short gastric vessels. From this opening, ligation and division of the short gastric vessels proceed using the cautery in a cephalad direction along the greater curvature of the stomach all the way to the esophageal hiatus. In an adolescent, the LCS (Ethicon Endosurgery, Cincinnati, OH) device is used for this purpose. Therefore, the patient's left epigastric incision, which is the main operating incision, should be 5-mm in size. The most cephalad short gastric vessels are usually intimately adherent to the spleen and require careful dissection. Having skeletorized the greater curvature of the stomach to the esophageal hiatus, the esophagus is separated from the patient's left diaphragmatic crus using cautery (Fig. 13.3). The retroesophageal space is then opened taking care not to injure the posterior vagus nerve that lies adherent to the posterior aspect of the esophagus (Fig. 13.4). Dissection then proceeds from the patient's left anteriorly over the esophagus and down the right diaphragmatic crus (Fig. 13.5). In addition, still using cautery, an opening is made in the gastrohepatic ligament to

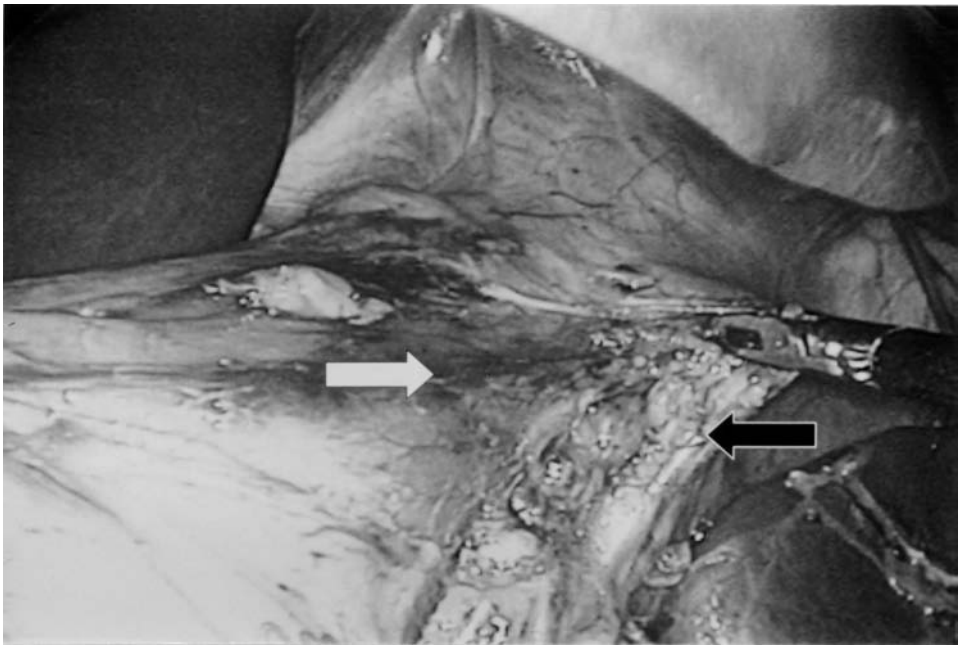


Figure 13.3 The short gastric vessels have been ligated and divided with the cautery. The esophagus (white arrow) is being dissected free from the patient's left diaphragmatic crus (black arrow).

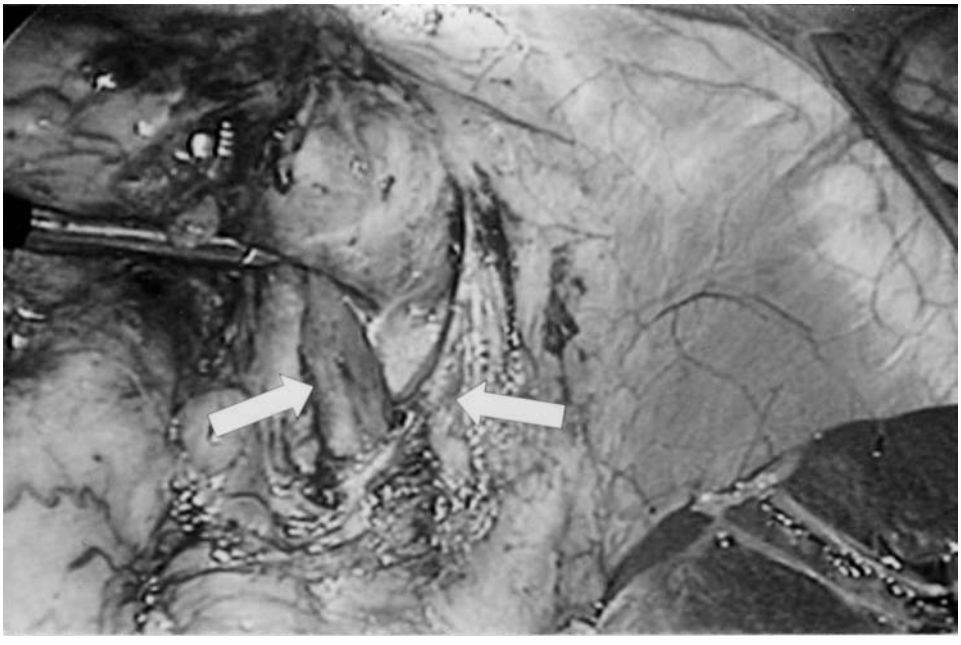


Figure 13.4 The retroesophageal space has been opened. Note the left and right diaphragmatic crura (white arrows) and a small hiatal hernia.

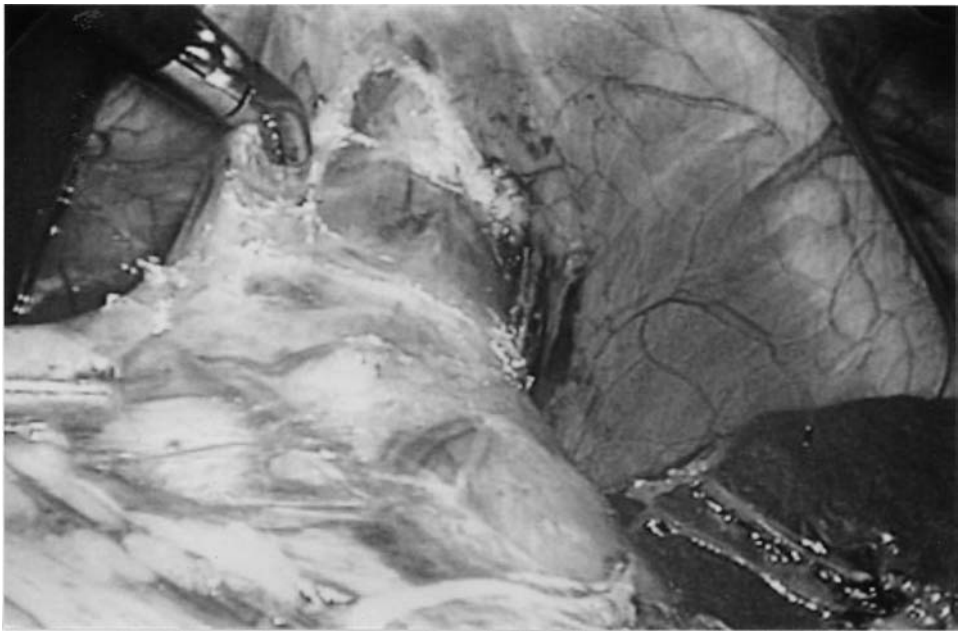


Figure 13.5 Incision in the phrenoesophageal ligament and peritoneum overlying the anterior esophagus.

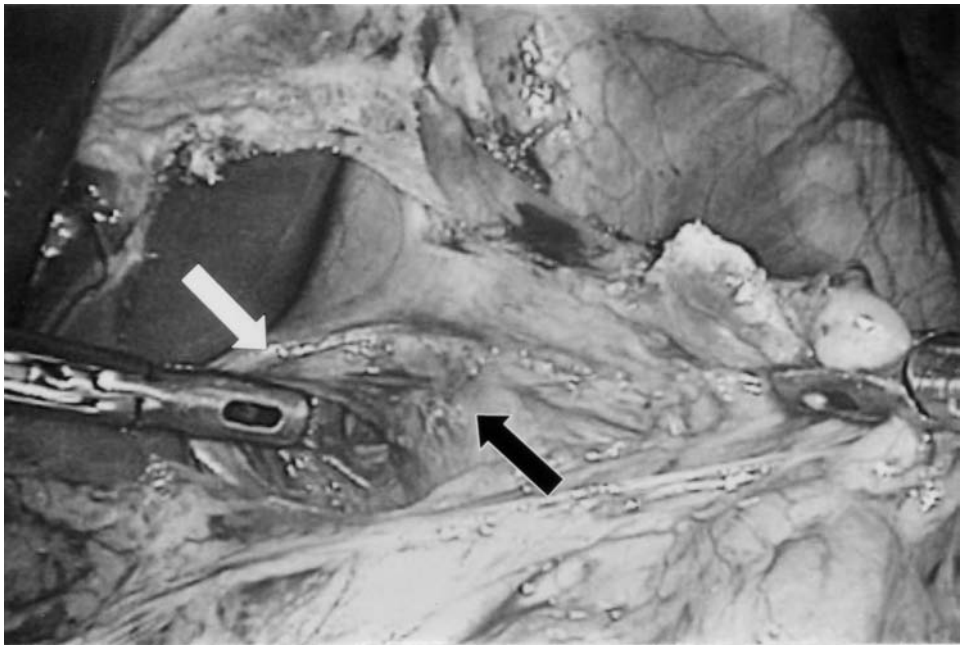


Figure 13.6 Attention is now turned to the patient's right side of the esophagus. The patient's right crus (white arrow) is being grasped and dissected free from the patient's right side of the esophagus (black arrow).

improve visualization. The right lateral aspect of the esophagus is usually intermittently adherent to the right diaphragmatic crus but, using careful dissection, it can be separated from the crus (Fig. 13.6). After this step, the entire esophagus has been freed from its diaphragmatic attachments and can be pulled further into the abdomen, thereby increasing the length of the intra-abdominal esophagus prior to fundoplication. Usually, at the very least, a small hiatal hernia is present and requires closure with a 2-0 silk suture that is tied intracorporally. Occasionally, a second suture is required either posterior to the esophagus or, sometimes, anterior to the esophagus if there is a large hiatal hernia. Once the esophageal hiatus is closed appropriately, a blunt esophageal bougie is then inserted by the anesthesiologist through the mouth and guided into the stomach. Careful attention is paid to the passage of the bougie through the esophageal hiatus to ensure that there is no evidence of angulation of the esophagus or narrowing from the crual sutures (Fig. 13.7). The bougie size is selected according to the patient's weight (Table 13.5).

At this point, the fundus of the stomach is then grasped by the atraumatic forceps in the operating surgeon's left hand which has been positioned through the retroesophageal space. Having grasped the fundus of the stomach, it is then brought through the retroesophageal space from the patient's left to the right (Fig. 13.8). The fundoplication is performed over the same indwelling bougie in the esophagus in order not to significantly narrow the esophagus with the wrap. Usually, three sutures are used to create the fundoplication with a small portion of the anterior esophageal wall incorporated with the most cephalad two sutures. The fundoplication is performed with 2-0 silk sutures, and they are also tied intracorporally (Fig. 13.9). Following creation of the fundoplication, the length of the wrap is measured and documented (Fig. 13.10). Usually, a wrap of 2 cm is sufficient to create an effective anti-reflux mechanism in infants and older children. The bougie is then

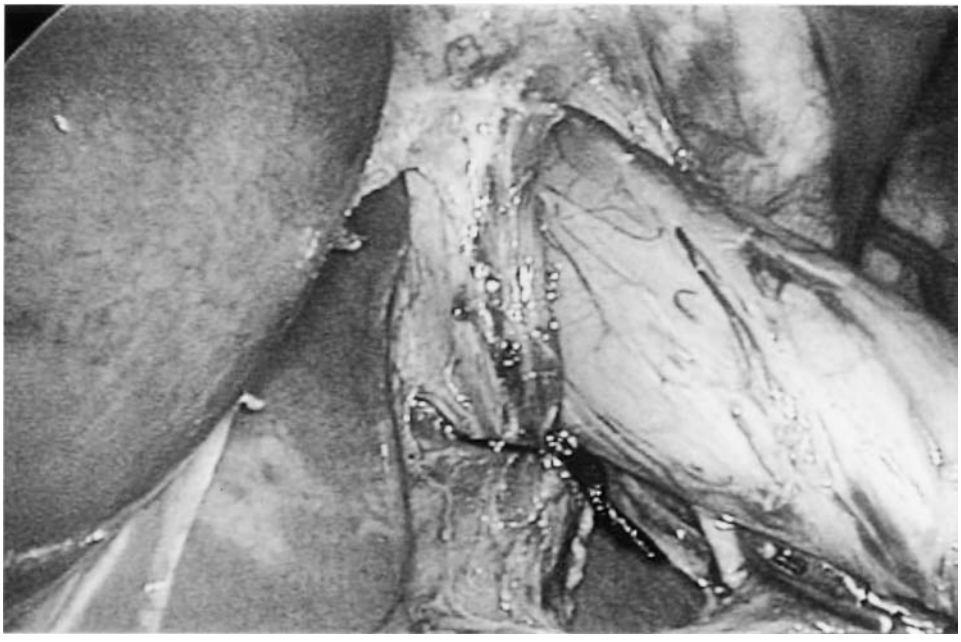


Figure 13.7 After the esophagus has been dissected free from its surrounding attachments, the small hiatal hernia is closed with a single 2-0 silk suture which is tied intracorporally. A blunt tip bougie has been passed into the esophagus and there is no evidence of residual hiatal hernia.

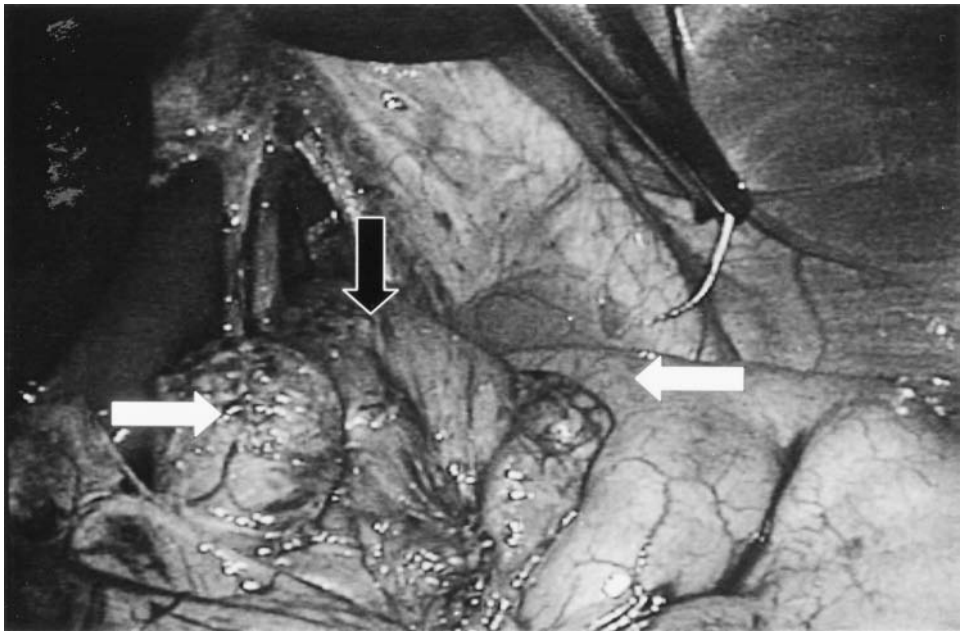


Figure 13.8 The fundus of the stomach has been passed through the retroesophageal space and is now ready to be sewn to itself in the standard fashion for a Nissen fundoplication. Note the esophagus (black arrow) and the fundus on each side of the esophagus (white arrows).



Figure 13.9 The completed fundoplication.

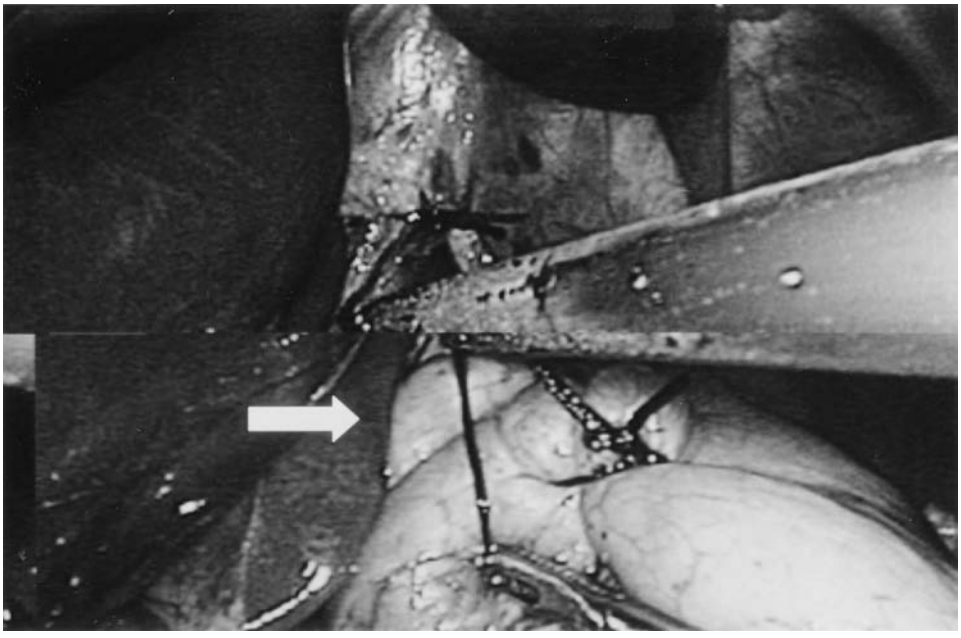


Figure 13.10 The length of the fundoplication is measured with a silk suture. In this patient, the length of the fundoplication measures 2 cm.

removed and the area of dissection carefully inspected. Assuming hemostasis is intact and there is no need for gastrostomy, the instruments are then removed; bupivicaine 1% without epinephrine is instilled into the incisions for postoperative analgesia, and the telescope and the umbilical cannula are extracted. The umbilical fascia is then closed with either 3-0 absorbable suture in an infant or 2-0 absorbable suture in an older child. The umbilical skin is closed with interrupted 5-0 catgut suture in an infant and 4-0 catgut suture in an older child. The other incisions are usually closed with steri-strips alone in an infant but may require a 3-0 or 4-0 absorbable suture placed in the anterior fascia in an older child (Fig. 13.11). In an older child, these incisions can be approximated with a 4-0 or 5-0 catgut suture as well.

6. GASTROSTOMY

If a patient requires a gastrostomy for enteral alimentation, the gastrostomy is positioned through the abdominal wall at the site of the left epigastric incision. This incision usually needs to be enlarged slightly with a hemostat by bluntly spreading the skin and soft tissues of the gastrostomy stoma. Also, the intra-abdominal pressure is reduced to $\sim$5 mmHg to reduce the distance and tension between the stomach and anterior abdominal wall.

A site is selected on the anterior aspect of the greater curvature of the stomach at the level of the incisura for placement of the gastrostomy. A 3-mm atraumatic forceps is introduced through the patient's left epigastric incision and the stomach grasped at this site (Fig. 13.12). Using the U-stitch technique described by Georgeson et al., two sutures are then placed through the skin and abdominal wall cephalad to the gastrostomy stoma, through the stomach and then out the abdominal wall and skin caudal to the gastrostomy (49,50) (Fig. 13.13). These sutures are usually 2-0 PDS but 0 PDS is

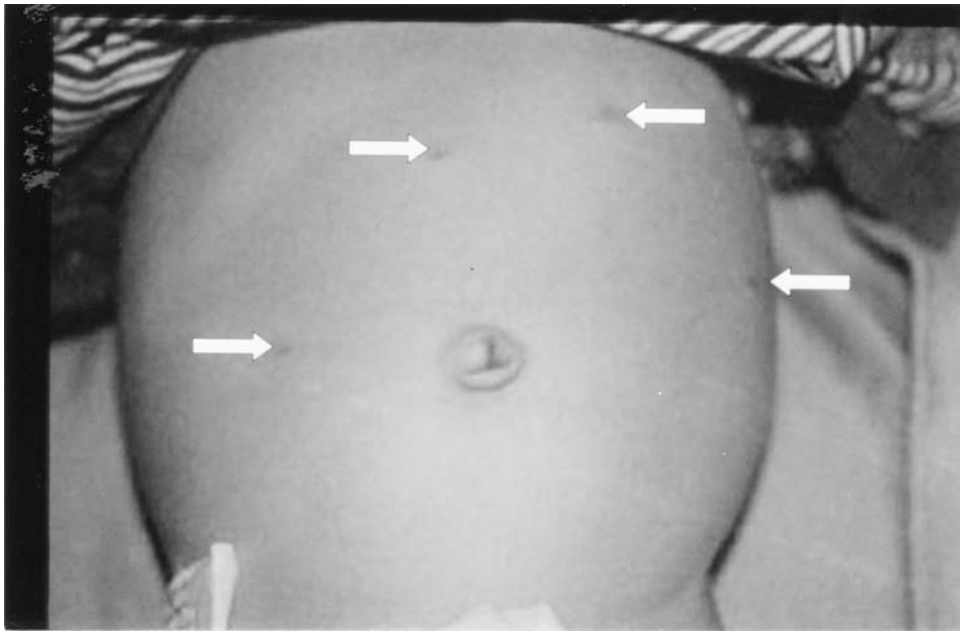


Figure 13.11 The appearance of the small scars (white arrow) from the laparoscopic fundoplication in a 4-month-old baby, 2 weeks after surgery.

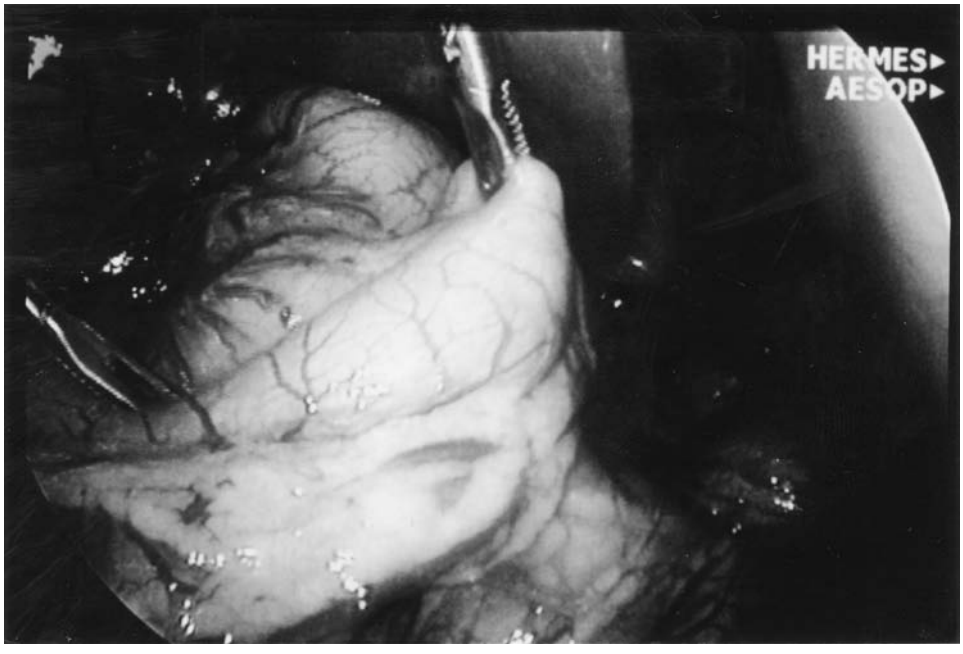


Figure 13.12 When performing the gastrostomy, a site is selected on the anterior aspect of the stomach near the greater curvature. The stomach is grasped with a atraumatic forceps inserted through the patient's left epigastric incision.



Figure 13.13 A needle is placed through the skin cephalad to the site of the gastrostomy, through the stomach medial to the planned gastrostomy and is shown exiting the abdominal wall inferior to the gastrostomy stoma.

necessary in older children due to the fact that it is attached to a larger needle. Once the stomach is secured on each side of the gastrostomy site, the Seldinger technique is employed by using the Cook Dilator Set (Cook, Inc., Bloomington, IN). With this technique, a needle is introduced through the incision and into the stomach in the center of the square formed by the two PDS sutures and a guidewire is inserted through this needle into the stomach (Fig. 13.14). The needle is then removed and dilators are placed over the guidewire and into the stomach, thereby dilating the skin, soft tissues as well as the gastrostomy (Fig. 13.15). The dilators used are 8, 12, 16, and 20 French (Fr.). Following dilation with the 20 Fr. dilator, the 8 Fr. dilator is placed through an appropriately sized Mic-Key button (Ballard Medical Products, Draper, UT), and the gastrostomy button and 8 Fr. Dilator are placed over the guidewire and into the stomach. The 5 cc balloon of the button is then inflated under direct visualization (Fig. 13.16). It is important to visualize the balloon being inflated inside the stomach to confirm its correct position. The sutures are then secured extra-corporally over the flange of the button, thereby attaching the stomach to the anterior peritoneum (Fig. 13.17). The 8 Fr. dilator and the guidewire are then removed. The other incisions are then closed in a fashion previously described for the laparoscopic fundoplication alone.

7. POSTOPERATIVE MANAGEMENT

The patients are transported from the operating room to the recovery room and then usually to a regular floor room. Intravenous fluids are initiated, but the patient is given liquids on the evening of the procedure. The next morning, either formula or a soft diet is initiated and the patient is usually discharged that afternoon. For patients requiring

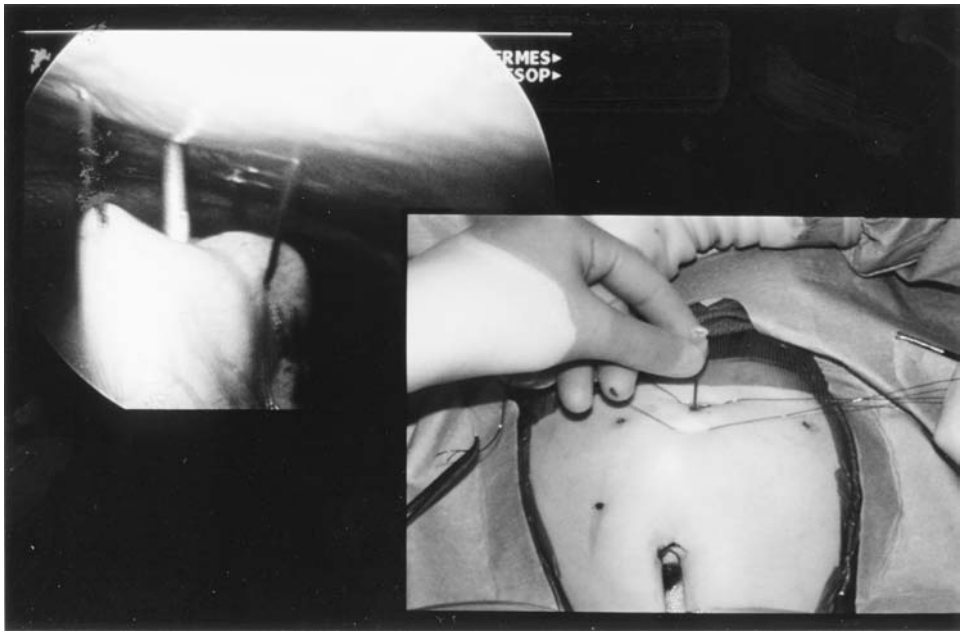


Figure 13.14 In the left upper aspect of the photograph, the U-stitches are seen holding the stomach near the anterior abdominal wall. A needle has been placed through the site of the gastrostomy through the stomach. An external view is shown on the right.

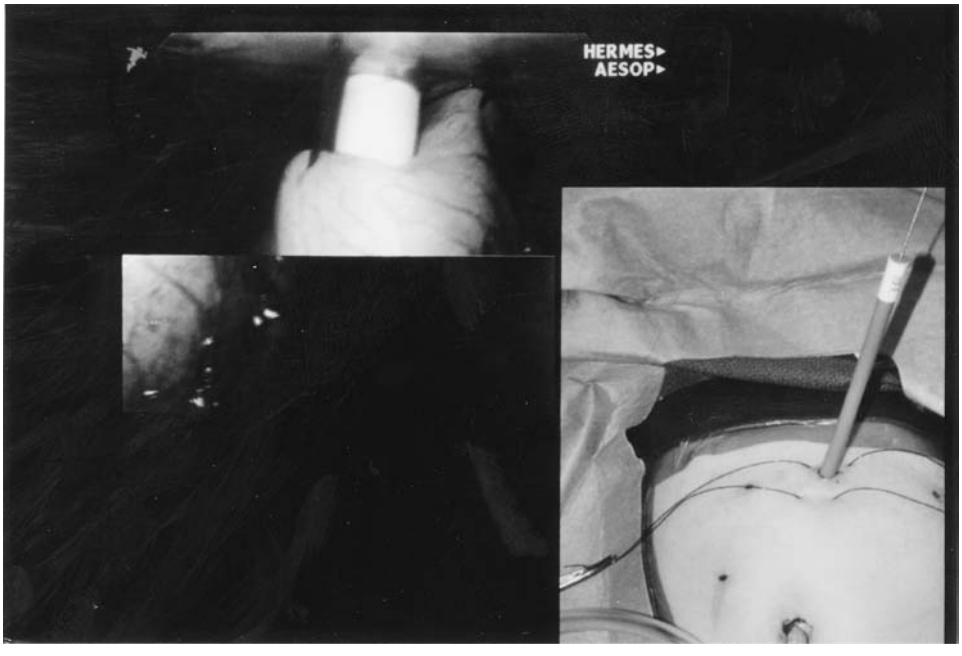


Figure 13.15 On the right, the guidewire and dilator are seen to pass through the skin and subcutaneous tissues at the site of the gastrostomy. On the left, the dilator is being introduced into the stomach through the gastrostomy.

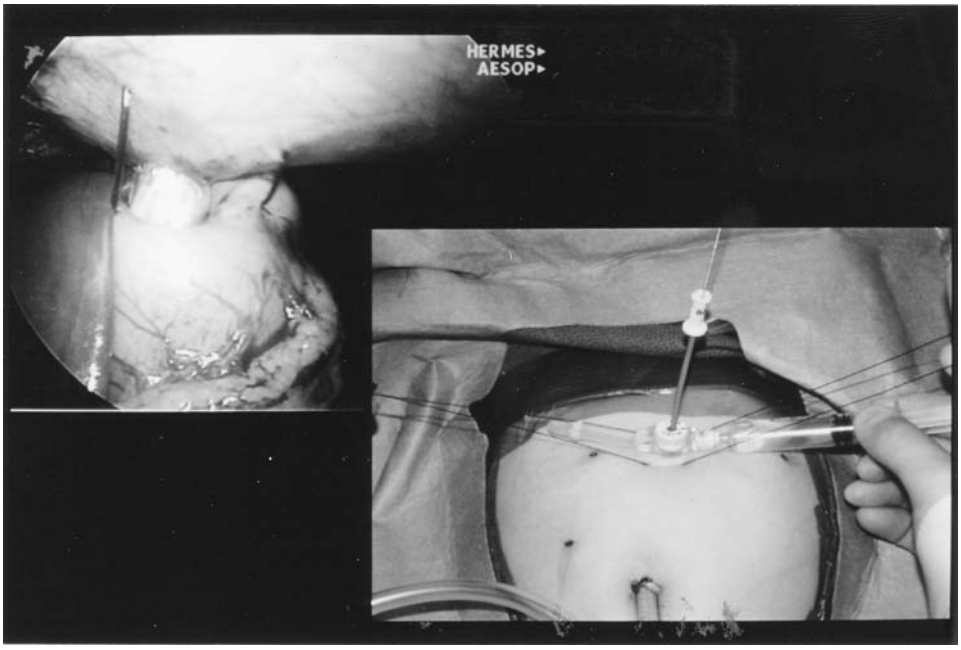


Figure 13.16 On the right, the guidewire, dilator, and gastrostomy button are seen exiting the gastrostomy stoma and the balloon of the gastrostomy button is being inflated. On the left, the button is seen entering the stomach at the site of the gastrostomy.

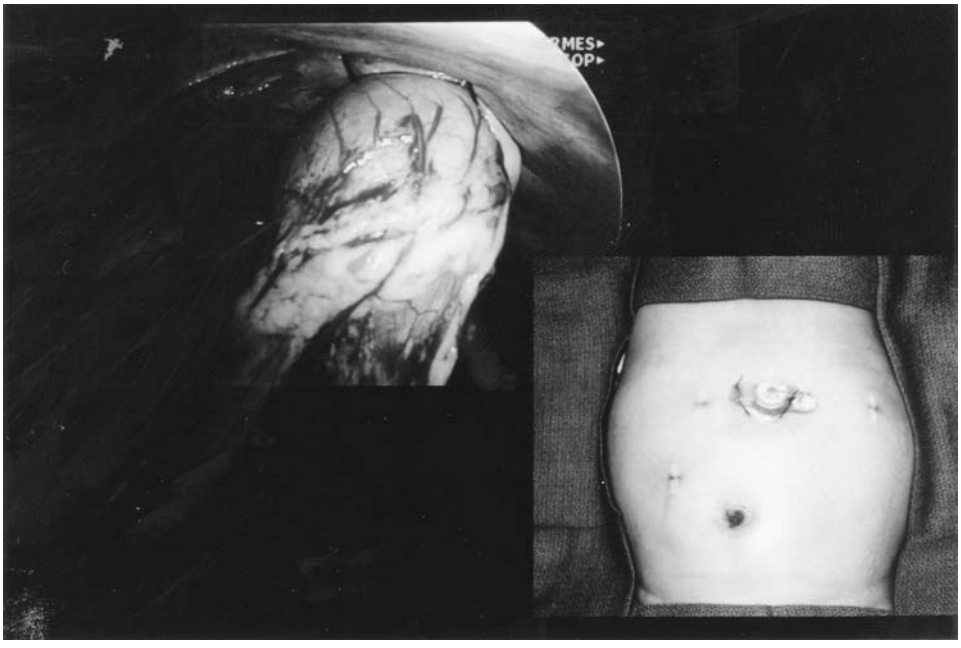


Figure 13.17 An intra-abdominal view of the stomach secured to the anterior abdominal wall at the site of the gastrostomy is shown on the left. On the right is an extracorporeal view of the incisions and the gastrostomy button, following laparoscopic fundoplication and gastrostomy.

gastrostomy feedings, they are usually initiated in a small volume and 50% strength the night of the procedure, and the patient is usually discharged the following day with instructions to advance the volume and strength over the next couple of days. As there appears to be some edema at the site of fundoplication for 2–3 weeks, patients who are able to take regular food are instructed to take soft foods with a consistency as in applesauce, mashed potatoes, or the like. There is a concern that an adolescent may take a large bite of meat and not chew it appropriately. In this case, the bolus may become lodged at the site of the fundoplication in the early postoperative period. If a gastrostomy is placed, the families are instructed to cut the sutures on the fifth postoperative day. All patients are usually seen in the surgery clinic two weeks following the operation and are then followed every six months for continued evaluation. All antireflux medications are stopped at the time of the clinic visit, if they have not been discontinued prior to this time.

8. RESULTS

Most long term studies reporting results after LNF stem from the adult literature and analysis reveal satisfactory results 1 year after LNF in 95% of patients undergoing fundoplication for GER and its complications. Failure rates for adults undergoing LNF are $\sim$1% per year. When failures occur, approximately one-third require a second fundoplication, while two-thirds can be managed with antireflux medications (9).

The evaluation of symptom control and satisfaction rates for children is not straightforward. Unfortunately, confounding variables such as continued antireflux medication on the part of the primary care provider, and individual comorbid conditions (i.e., neurologic impairment, congenital anomalies, and chronic lung disease), make establishing a benchmark for satisfaction very difficult. Therefore, it is perhaps best to evaluate the effectiveness of LNF in relation to recurrent GER as well as wrap failure (dysphagia, migration/breakdown). The adult literature clearly supports that the most common complaint postfundoplication is dysphagia, which persists for $>$1 month in $\sim$20% of individuals. This dysphagia is severe enough to require dilation in up to 40% of these patients. In our experience of over 200 LNF over the past 3 years, we have not had a patient develop persistent dysphagia requiring esophageal dilation. This dramatic divergence between adults and children is easily explained by the diet that each population consumes. Infants tend to maintain a diet of liquids and soft foods that pose little threat of lodging in the edematous distal esophagus at the level of the fundoplication. By the time they transition to a predominantely solid diet, similar to adults, the fundoplication has healed and all operative changes have resolved, hence little postoperative dysphagia.

The most common reason for wrap failure after LNF (regardless of age) is the "slipped Nissen" that occurs as a result of crural breakdown and migration of the wrap into the chest through the resulting hiatal hernia. Wrap migration occurs in up to 20% of adult patients; however, in our experience, it develops less frequently and others have reported wrap failure rates in 2–5% of children undergoing laparoscopic fundoplication (5,49). Multiple approaches to secure the wrap below the diaphragm, including securing the fundoplication to the diaphragm, incorporating the posterior aspect of the esophagus to the crural repair, and tacking the esophagus to the crura at multiple sites, have been tried in the adult population and have not proven to be more effective in preventing this complication. We currently anchor the esophagus to the crura at the 12, 3, and 9 o'clock positions in an attempt to maintain the esophagus in an intra-abdominal position and, hopefully, avoid migration. However, we have not evaluated this technique

in a scientific analysis. Wrap migration does occur more often in neurologically impaired children and is presumed to be a result of increased intra-abdominal pressure secondary to retching and often the underlying seizure disorder. Ensuring an adequate crural repair should be paramount in these children; however, regardless of efforts to prevent wrap migration, its development will likely continue to be higher in this subpopulation.

Recurrent GER has been reported to occur between 2% and 6% in pediatric patients and is dependent on the type of fundoplication created (56,57). Chung and Georgeson reported their results with both laparoscopic Nissen and Toupet fundoplications (49). In their series, Nissen fundoplication was superior to Toupet fundoplication with regard to symptom control. Of patients undergoing Toupet fundoplication, 6.1% developed recurrent symptoms, while only 3.5% of those undergoing LNF developed similar symptoms. For comparison purposes, failure rates for open fundoplication range from 20% to 47%, with the upper end of this range representing failures in children with neurologic impairment (51–55).

The functional results regarding motility, LES function, and evaluation for GER have been individually studied in the pediatric population. Okada et al. performed esophageal manometry preoperatively and at one month postoperatively in infants and children undergoing LNF (3). They found that LNF completely controlled symptoms and inhibited acid exposure $0 \pm 0\%$ of fraction time for $pH < 4$ in 24 h pH monitoring. Manometric examination revealed statistically significant increases in postoperative basal LES pressure and residual LES pressure at the nadir of swallow-induced LES relaxation, both functions of improved GER control. Evaluation of esophageal motility showed no change in motility patterns after LNF. Tovar et al., evaluated 27 patients prospectively undergoing LNF. Twenty-four hour pH monitoring performed preoperatively and postoperatively at 6 months after LNF revealed fewer total reflux episodes (70 vs. 19), fewer episodes lasting longer than 5 min (10.4 vs. 2.4), less percent of time $pH < 4$ (20.3 vs. 4.7 min), and an abbreviated longest episode (37 vs. 12 min) after LNF (6).

Table 13.3 summarizes the reports, to date, containing >100 pediatric patients undergoing laparoscopic fundoplication. The spectrum includes different fundoplication techniques such as Nissen, Nissen-Rosetti, Thal, and Toupet fundoplications. Regardless of the approach utilized, the recurrence rates, conversion rates, and operative time are remarkably similar between the different centers. The data for length of hospitalization and time to feeds are incomplete in several studies and appears to be institution-dependent as well as surgeon-preference regarding timing of initiating enteral alimentation after laparoscopic fundoplication. Complication rates range from 0% to 12.7% and usually developed early in the "learning curve" experience for each report. Four deaths (three postoperative and one operative) have occurred out of the 1582 patients (0.25%).

In review, advances in laparoscopy have been proven to be safe and provide superior cosmetic results to open approaches for GER. The effectiveness of laparoscopic fundoplication compared to open fundoplication in the pediatric population has only been studied from a retrospective approach, and these findings appear to support the laparoscopic technique as at least equal, and likely superior, to the open approach.

9. THE CHILDREN'S MERCY EXPERIENCE

Between November 1999 and March 2002, 154 patients underwent LNF at Children's Mercy Hospital for GER (62). All operations were performed as described in the procedure portion of this chapter. The mean age at the time of operation was 24.5

Table 13.3 Comparison of Large (>100 Patients) Studies of Children Undergoing Laparoscopic Fundoplication

Author	*N*	Recurrence (%)	Type	Complication	Conversion (%)	LOS (days)	TTF (days)	Op time (min)
Georgeson (4)	389	6.1 Toupet 3.5 Nissen	201 Toupet 188 Nissen	2 Deaths (1 Operative)	3.3	~3	N/A	~60
Esposito et al. (59)	289	2.10	148 N-R 141 Toupet	Intraop = 5.1% Postop = 3.4%	1.3	N/A	N/A	70
Montupet et al. (60)	284	2.10	Thal	Intraop = 0% Postop = 1%	N/A	N/A	~3	~60
Rothenberg (5)	220	3.40	Nissen	Intraop = 2.6% Postop = 7.3%	1.0	1.6	N/A	82
Allal et al. (61)	142	4.20	56 Toupet 83 Nissen	Intraop = 0.5% Postop = 2% 1 Death	2.1	3	N/A	105
Iglesias et al. (58)	104	2.90	Nissen	Major = 12.7% 1 Death	1.0	<10	~3	60
Ostlie et al. (82)	154	2	Nissen	Intraop = 0% Postop = 2%	0	2.8	~1	90

Note: LOS, length of stay; TTF, time to feed; Op time, length of surgical procedure.

Table 13.4 Comparison of Operative Times With and Without Robotic Telescopic Assistance (62)

Group	No. of patients	Mean operative time (min)
A	10	108
B	34	110
C	39	93
D	52	68
P-values A vs. B = 0.058, A vs. C < 0.05, A vs. D < 0.05		

Note: A = patients underwent fundoplication before the use of telescopic assistance (camera held by nurse); 10/99-4/00 B–D = patients underwent fundoplication with telescopic assistance over the following time period; B, 5/00-12/00; C, 1/01-5/01; D, 6/01-3/02.

months (range = 3 weeks–180 months). Mean weight was 10.6 kg with a range of 2.5–50 kg. Mean operative time for the entire group was 90 min. Since May 2000, we have employed robotic assistance as described. Interestingly, when comparing the operative times for infants that underwent LNF prior to robotic assistance to those where robotic assistance was employed revealed a statistically shorter procedure when AESOP was used (Table 13.4) (62).

Comparison of elective (130 patients) to non-elective (24 patients) LNF revealed longer hospitalizations in the non-elective group as would be expected. On average, non-elective patients remained hospitalized for 9.8 days, while mean time to discharge for elective patients was 1.7 days.

An effective fundoplication length and bougie size has never been established in infants and children. We have routinely measured the fundoplication length and recorded the bougie size in patients undergoing LNF. Employing a fundoplication length of $\sim$2 m and a gradated bougie size relative to the patients weight (Table 13.5), patients undergoing elective LNF have had resolution of their GER symptoms. There have been no cases of dysphagia requiring dilation, and only two patients have developed recurrent symptoms. One patient has required a second LNF, and the other has undergone repeat evaluation for GER showing a normal pH study and intact fundoplication on UGI.

Table 13.5 Gradated Bougie Size Relative to the Patients Weight

Weight (kg)	Bougie size
2.5–4	22–26
4–5.5	26–30
5.5–7	30–34
7–8.5	34–36
8.5–10	36–38
10–15	38–42

10. CONCLUSION

LNF, with or without gastrostomy, can be performed safely in infants and children by surgeons comfortable with advanced laparoscopic techniques. It is an excellent approach for pediatric patients who develop complications of GER or are resistant to medical therapy. The postoperative hospitalization and time to oral or gastrostomy feeding are short, and the outcome is comparable, if not superior, to open fundoplication.

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14
Gastrostomy, Jejunostomy, and Cecostomy

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1. INTRODUCTION

Access to the gastrointestinal tract is necessary for the care of many children with a variety of disorders. While short-term access may be accomplished with nasoenteric, oroenteric, or anoenteric tubes, the establishment of abdominal wall stomas facilitates care of children requiring chronic gastrointestinal access. Gastrostomies and jejunostomies are used commonly as portals for enteral nutrition, whereas cecostomies are used for antegrade enemas in children with constipation or fecal incontinence. The advancements in endosurgery have given surgeons powerful, minimal access tools for establishing entry to hollow visceral structures using a combination of gastroscopy, colonoscopy, and laparoscopy. This chapter examines existing outcome data for gastrostomies, jejunostomies, and cecostomies using minimal access surgery with a focus on the pediatric literature.

2. GASTROSTOMY

Gastrostomies are the most common method of providing long-term enteral nutrition in children who are unable to achieve adequate oral intake. A variety of conditions are indications for placement of gastrostomy tubes, including neurological impairment, failure to

thrive, primary aspiration, and severe gastroesophageal reflux disease (GERD). Traditionally, gastrostomy tubes have been placed by means of a laparotomy. However, the advent of endoscopy has given surgeons and gastroenterologist new tools to place gastrostomy devices with minimal abdominal incisions. Percutaneous endoscopic gastrostomy (PEG) was first described by Gauderer et al. (1) in both children and adults and has since gained immense worldwide popularity. An alternative technique for minimal access placement of gastrostomy tube is laparoscopic placement of gastrostomy tube (LAPGT). This has been described using either one or two ports by multiple authors in the last 10 years (2–13).

The method of placement has generally been based on referral patterns and operator's experience. Gasteroenterologists are trained in placing gastrostomy tubes using the PEG technique. Surgeons are trained to place gastrotomies with a laparotomy, and many are trained in the PEG and LAPGT techniques. Each of the different techniques has advantages and disadvantages.

2.1. Open Gastrostomy

The traditional approach for placement of a gastrostomy tube has been by laparotomy incision. Most surgeons use either a vertical midline incision or a transverse or oblique subcostal incision. The two most common methods for performing a gastrostomy are the Stamm gastrostomy and the Janeway gastrostomy. For the Stamm method, the stomach is tacked up to the abdominal wall with interrupted sutures and the gastrostomy tube is placed through a separate incision through a gastrotomy within two purse string sutures. The Stamm technique is favored by most surgeons. The Janeway method involves formation of a gastric tube with the creation of a nipple for continence. This technique is favored by some surgeons in children who require long-term gastrostomy feeds. The benefits of the open procedures are that they allow precise placement on the stomach with direct visualization of other visceral structures, minimizing the chance for inadvertent injury. Additionally, the placement of sutures from the stomach to the abdominal wall creates an immediate, secure attachment. In the setting of accidental tube removal in the early postoperative period, the likelihood that the stomach will fall away from the abdominal wall, resulting in free perforations of the stomach into the peritoneal cavity, is minimized. Also, percutaneous tube replacement through the previous gastrostomy site is possible in the early postoperative period with minimal worry of the stomach becoming detached from the abdominal wall. A gastrostomy can be placed by means of a laparotomy in instances when a PEG or LAPGT may not be appropriate such as in the presence of dense intra-abdominal adhesions. The one obvious disadvantage of using a laparotomy is the presence of an incision, both from the standpoint of cosmetics and postoperative pain. Surgeons traditionally placed a long tube, such as a Malencott® or Foley® catheter, at the initial operation, and subsequently replaced these with a low-profile device after 1–2 months, when the tract has matured. Many surgeons, including the author, now place a low-profile device primarily when placing a gastrotomy tube by a laparotomy. This low-profile device is colloquially termed as "button" and is made by several manufacturers including Kimberly–Clark (Mic-key®), Bard®, and Applied Medical Technology (Mini®). The remainder of the procedure is identical to the Stamm gastrostomy.

2.2. Percutaneous Endoscopic Gastrostomy

The PEG technique has gained worldwide popularity since its inception and is performed by both surgeons and gastroenterologists and has many benefits over gastrostomy

placement by laparotomy. Because many of the patients requiring a gastrostomy are malnourished or have chronic medical illness, operative risk is increased and wound-healing may present a significant problem. A benefit of PEG placement is minimal postoperative discomfort to the patient as well as no abdominal incision to heal; the only abdominal incision necessary is the one at the gastrostomy tube placement site. A PEG can generally be performed quickly with rapid postoperative recovery. In fact, many clinicians perform the PEG procedure without general anesthesia with a combination of local and sedation. Additionally, the endoscopist can visualize the mucosa of the esophagus and the stomach for potential abnormalities at the same setting.

Additionally, because of minimal manipulation of the peritoneal space, adhesive small bowel obstruction is virtually nonexistent after PEG placement. In fact, the more common cause of bowel obstruction following PEG seems to be from migration of part of the catheter into the small bowel (14,15).

Finally, several studies in the adult literature have shown that PEG placement may be cost effective compared with open operative placement. One retrospective review comparing gastrostomies placed by means of PEG ($n = 125$) and by Stamm technique ($n = 88$), showed that PEG was faster and cheaper by approximately US $1000 (16). Additionally, feeding time and complications were less in the PEG group than in the Stamm group. A prospective randomized study of 48 adult patients compared PEG and open gastrostomy (17). There was no difference in postoperative morbidity between the groups. Procedural cost was on average approximately US $250 higher for open gastrostomy, but statistical analysis was not performed to determine if this difference was significant. Similarly, initiation of tube feeds was successful in a greater percentage of patients in the PEG group than in the open group, but this difference did not achieve statistical significance. The authors commented that no definitive conclusions could be made regarding the superiority of one technique over the other although there were trends favoring PEG.

One potential disadvantage of the PEG is the possibility of injury to hollow viscous while performing gastroscopy, particularly to the esophagus. This complications is rare, occurring in one patient (0.6%) in one adult series of 168 patients (18), no patients in an adult series of 232 patients (19), and no patients in a series of 220 children (20). Another potential disadvantage is the possibility of injury to intra-abdominal visceral structures, as the peritoneal cavity is not directly visualized. Rather, placement of the gastrostomy tube is guided by transillumination to minimize the risk of visceral injury. The organ most frequently injured is the transverse colon, and the incidence of gastrocolocutaneous fistula during placement of a PEG in children was 2% in the series reported by Gauderer (20). Other visceral injuries reported have been injury of an arterial vessel requiring laparotomy in three of 232 adult patients (18). Also, as the stomach is secured to the abdominal wall only by the gastrostomy device, the risk of intra-abdominal gastric leak exists if the tube is inadvertently removed in the early postplacement period. This has occurred in 0.5–2.2% in adult series (18) and 0.5% in children (19, 20). In order to avoid some of these risk, some clinicians have combined PEG placement with laparoscopic guidance or used fluoroscopic guidance in conjunction with or instead of endoscopic guidance for gastrostomy placement (21,22).

The use of the endoscope through the mouth potentially leads to complications. The most common complication of PEG placement is wound infection or peristomal infection. Gauderer (20) reported a 1.8% PEG-site infection rate in his pediatric series. In adults, lesions obstructing the mouth or esophagus, such as head and neck or esophageal cancers prohibit gastroscopy, making PEG impossible. In rare cases, squamous cell carcinoma of the head and neck has been shown to metastasize to the gastrostomy site after

PEG placement, presumably from dragging tumor cells to the gastrostomy site by the endoscope (23). These circumstances rarely, if ever, occur in children.

A recent retrospective study investigated the feasibility and safety of PEG placement in infants weighing <3.5 kg (24). In this group, three of 26 (11.5%) developed wound infections and two of 26 (0.8%) required intervention for pneumoperitoneum as a result of the procedure. All gastrostomy tubes were successfully placed under general anesthesia. There were no deaths related to the procedure.

2.3. Laparoscopic Gastrostomy

Several groups have described various techniques for LAPGT using either one or two incisions (2–13). If two incisions are used, a port for a laparoscope is placed through the umbilicus. Once the stomach is visualized, a site on the abdominal wall conductive to gastrostomy placement is selected. Either a port is placed at this site or an instrument is placed directly through a stab wound. The stomach is brought up to the incision and a gastrostomy is then placed in this site. Figure 14.1 demonstrates one method for the two-incision method of LAPGT. In the single-port technique, a port is placed in the left upper quadrant where the gastrostomy will be placed. Either a telescope with a working port or a bronchoscope or an esophagoscope that can accommodate an instrument through the sheath is used to bring the stomach up to the incision for gastrostomy placement. Both techniques leave the gastrostomy site as the only visible incision, as the umbilical scar for the two-port technique can be hidden within the umbilicus. Another benefit of this technique is the ability to identify all visceral structures, thereby eliminating the potential for inadvertent injury, as may happen in PEG placement. Additionally, the gastrostomy can be placed precisely on the stomach, which may be beneficial, as will be discussed later in this chapter. Others have performed a continent Janeway gastrostomy using laparoscopic assistance (25).

A potential disadvantage of laparoscopic gastrostomy is the risk of visceral injury during initial trocar placement. Several large series have reported the incidence of visceral injuries during trocar placement. One study collected data prospectively on 14,243 patients with a mean age of 51.4 years who underwent laparoscopic procedures. There were 26 injuries resulting in an incidence of 0.18%. The most common injuries were to the small bowel ($n = 6$) and the liver ($n = 3$). Twenty-three injuries were identified and repaired intraoperatively with 18 of these needing conversion to a laparotomy. The remaining three patients were identified between 48 and 72 h postoperatively. Two underwent operative intervention and one underwent successful nonoperative treatment (26). The largest series in the pediatric literature reviewed 2157 children who underwent laparoscopic and thoracoscopic procedures with a total of 7117 cannulas placed. They reported no major vascular or visceral injuries. Complications included three cases of abdominal wall bleeding (1.4%) and one incisional hernia (0.5%). In this series, they used only radially expandable access devices for port placement (27).

Several retrospective reviews have been performed, analyzing the results of laparoscopic gastrostomy in children. Lee et al. (28) reviewed a series of 51 patients undergoing laparoscopic gastrostomy tube placement and reported no significant complications in this group. In an analysis of hospital charges in this series, LAPGT was approximately US $1050 more expensive initially than PEG for first operation. However, 13% of those children undergoing PEG required a second procedure in the operating suite to convert a gastrostomy to a low-profile device. In a study by Humphrey and Najmaldin (29), 28 children underwent laparoscopic gastrostomy, 16 in conjunction with a fundoplication. They reported no complications specific to the gastrostomies and reported an

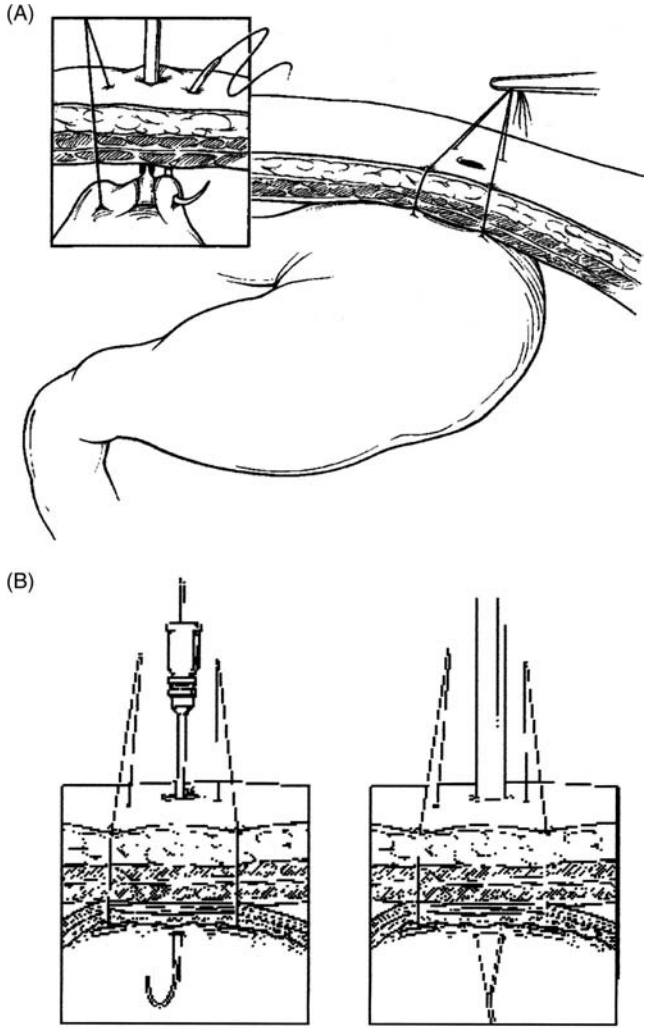


Figure 14.1 This set of pictures illustrates the method of laparoscopic gastrostomy tube placement as described by Georgeson. (A) demonstrates U-stitches placed around the gastrostomy site. (B) shows serial dilations of the gastrostomy tube tract with progressively layer dilators. (C) shows placement of the tube. [Reprinted with permission from Georgeson KE et al. Pediatr Endosurg Innov Tech 1998; 2(4):223–226.]

average operative time of 65 min in the 12 patients undergoing gastrostomy alone (29). Rothenberg et al. (30) reported their results on placement of low-profile gastrostomy "buttons" in 240 children. Forty-one patients underwent gastrostomy alone, whereas the remainder also underwent a concomitant fundoplication. They showed an average operative time, for a gastrostomy alone, of 15 min. There were five gastrostomy-related complications in the 240 children: two wound infections treated with oral antibiotics, two early tube dislodgements, and one required re-operation. These results are favorable to other large series reviewing gastrostomy placement by either PEG or open surgical techniques. One retrospective review compared open and laparoscopic gastrostomy and fundoplication (31). They found that while complications between the two groups were similar,

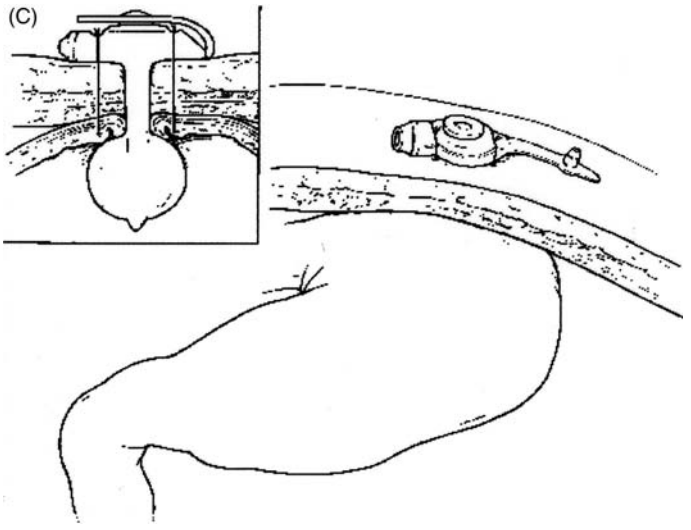


Figure 14.1 *Continued.*

patients undergoing laparoscopic procedures had a shorter hospital stay and quicker return to feeds postoperatively. One adult retrospective study compared PEG with LAPGT (32). Seventeen patients underwent PEG, whereas 14 patients underwent LAPGT. There was one death directly related to tube placement in each group. They reported no wound infections, gastrocolic fistulae, bleeding, stomal leaks, or visceral injuries in either group.

No single method of placing gastrostomy tubes is optimal in every circumstance. The current literature on gastrostomy placement consists largely of retrospective reviews of a single technique making rigorous comparisons of the different techniques difficult. Bearing this in mind, a prospective study comparing the different methods used children would be of great importance. The outcome data should include cost of the procedure, length to full feeds, amount of postoperative pain medications necessary, operative complications such as visceral injury and wound infections, short-term complications such as early postoperative tube dislodgment, long-term complications such as gastroesophageal reflux and gastric prolapse, and patient's and family satisfaction. The current decisions on which procedure is performed are usually based on existing resources and experience at a given facility; no existing data shows compelling evidence for choosing a given technique.

2.4. Gastrostomy Device Considerations

Patients and their families have consistently favored the use of low-profile gastrostomy devices or "buttons" such as made by Kimberly–Clark (Mic-key), Bard, and Applied Medical Technology (Mini), over the use of long tubes such as Malencott® and Foley® catheters. The longer catheters are technically easier to place than the button catheters when placed by means of laparotomy. The longer catheter can be changed to a button once the tract has matured; most surgeons favor waiting 1–2 months before doing this. The buttons are particularly favored by families of active children. Because the buttons lie nearly flush against the abdomen, clothes are easily placed over the buttons and the devices are more difficult to pull out inadvertently. The Mic-key buttons are particularly favored by many families. The connecting tube locks into place with the button and are

less likely to become disconnected, particularly with prolonged feeds such as continuous nighttime feeds. Several groups have reported successful results with primary low-profile gastrostomy placement using laparoscopic assistance in children (2,30,33). The author is unaware of studies that have compared different gastrostomy devices for family satisfaction. Such a study would certainly be an important undertaking.

Several other questions regarding gastrostomy tubes in light of differing methods of placement would benefit from critical examination, and one such question is inadvertent tube dislodgment in the postoperative period. Replacement in the early postoperative period may result in two potential significant complications: incorrect tube displacement and/or disruption of the gastrostomy site from the abdominal wall resulting in an intraperitoneal gastric perforation. The choices for tube replacement are: percutanceous tube replacement, percutanceous tube replacement with fluoroscopic confirmation, and intraoperative tube replacement with direct confirmation of correct placement by laparoscopy, laparotomy, or gastroscopy. If the tube has become dislodged before 1–2 weeks postoperatively, some practitioners will replace the tubes intraoperatively. In general, most practitioners will replace tubes percutaneously with fluoroscopy within 2–8 weeks postoperatively and percutaneously without fluoroscopy after 2–8 weeks postoperatively. After 8 weeks, the gastrocutaneous fistula tract is believed to be sufficiently well formed in order to replace tubes safely with low risk of complications. Placement of a gastrostomy by laparotomy likely would result in less incidence of intraperitoneal gastric leak as the stomach is generally sutured directly to the abdominal wall. Retrospective reviews of large series and meta-analysis of these series would probably suffice in determining the rate of incorrect replacement and intraperitoneal gastric leak at various time points postoperatively for both PEG and laparotomy-placed gastrostomies. Pofahl and Ringold (34) reviewed 197 patients undergoing PEG and identified six cases (3%) complicated by early tube dislodgment. The average time to dislodgment was 2.9 ± 1.3 days postplacement. Two patients underwent nonoperative tube replacement, two underwent observation followed by delayed repeat PEG, and one underwent observation followed by delayed LAPGT. One required emergent operation. Larger series of LAPGT are probably needed in order to determine which method of replacement is safe at various postoperative intervals. Determining the appropriate method of replacement of gastrostomy tubes would ensure safe replacement and could avoid unnecessary fluoroscopy and reoperation.

Another incompletely resolved question is the site of placement of the gastrostomy with respect to the stomach. Placing the gastrostomy in too close proximity to the pylorus may lead to obstruction of the pylorus by the tube particularly in small children and particularly with long-tube devices that may slip further into the stomach. Most surgeons place the gastrostomy in the mid-body of the stomach to avoid this potential complication. Placing a gastrostomy near the greater curvature in a patient without a fundoplication may exacerbate reflux by changing the angle of the gastroesophageal junction both in children and in experimental animals (35–41). Placement of the gastrostomy near the lesser curvature in children has been advocated by some as resulting in less gastroesophageal reflux (42–43). Further prospective studies comparing lesser curvature placement and greater curvature placement with rigorous quantitative analysis of postoperative reflux by pH probe would be an ideal method to examine this question. Exact placement may not be quantifiable using the PEG technique.

The question of potentially exacerbated GERD by gastrostomy alone has lead some surgeons to perform a fundoplication in addition to a gastrostomy in subsets of patients needing gastrostomy without quantifiable GERD. Frequently, these patients have severe neurologic impairment and are at high risk for GERD and aspiration. The author is unaware of any studies comparing gastrostomy alone to gastrostomy and fundoplication

in high-risk patients without quantifiable GERD. Careful preoperative workup and postoperative followup are particularly necessary in these children.

3. JEJUNOSTOMY

A subset of children who require enteral feedings may not tolerate gastrostomy feedings secondary to poor gastric emptying or refractory GERD. These patients may be considered for jejunostomy. As with gastrostomies, jejunostomy tubes may be placed by means of laparotomy, laparoscopy, or with endoscopic guidance.

Frequently, jejunal access is attained through a new or pre-existing gastrostomy, whereby a gastro-jejunal tube is passed into the stomach, through the pylorus and duodenum into the jejunum. This occurs in patients who are noted to have poor gastric emptying or GERD after placement of a gastrostomy as an alternative to pyloroplasty or fundoplication. Albanese et al. (44) retrospectively compared 112 neurologically impaired children with gastroesophageal reflux undergoing nissen fundoplication with gastrostomy tube placement ($n = 68$) and children having a percutaneous gastrojejunostomy ($n = 44$) to determine the optimal method for supplying enteral feeds in this population. They found that the group undergoing fundoplication and gastrostomy had a significantly higher incidence of major complications, although minor complications were much greater in the percutaneous gastrojejunostomy group. No deaths occurred in either group that was directly related to the procedure. They concluded that percutaneous gastrojejunostomy is a safe alternative for feeding the neurologically impaired child with gastroesophageal reflux.

In many patients, dual access to the jejunum and stomach is attained through a gastrojejunal tube to allow for gastric decompression with jejunal feeds. Bell et al. reported a 95% success rate of passing a jejunostomy tube percutaneously by means of a PEG site. Complications are frequent with these devices (45). DiSario et al. (46) reported a serious complication rate of 95% in jejunal tubes placed through a PEG-site complication. In this adult series of 20 patients, continued aspiration (67%) and tube failure secondary to occlusion, leakage, malposition, extrusion, cracking, kinking, or rupture (70%) were the most common. Shike et al. (47) reported a series of 150 adult patients undergoing endoscopic jejunostomy directly into the jejunum. They reported a success rate of 86% in 129 patients with a procedural complication rate of 8% including six wound infections, one colon perforation, one abscess, and one instance of postoperative bleeding. They had a low long-term catheter complication rate of 3%. Mean duration of catheter use was 113 days.

Laparoscopic jejunostomy has been advocated by several group in the last 10 years as an alternative to open or endoscopic jejunostomy placement (48–50). Hotokezaka et al. (49) retrospectively reviewed their adult series of jejunostomies placed with laparoscopic assistance. Four patients were converted to open laparotomy, whereas 28 were completed laparoscopically. Major complications including tube displacement, obstruction, and aspiration pneumonia occurred in 25% of patients. In the four patients with early postoperative tube dislodgment, three were replaced at the bedside, whereas one occurred laparotomy for redo jejunostomy. Tube feeds were begun on an average of 2 days in patients undergoing laparoscopic placement (49).

As with gastrostomy tube placement, no one method of jejunostomy placement has been shown in a rigorous fashion to be the best method. A prospective randomized study comparing open, endoscopic, and laparoscopic jejunostomy would be a useful one. Until then, clinicians should continue to use the techniques with which they are most comfortable.

4. CECOSTOMY

Fecal incontinence or constipation can be a devastating problem in children with anorectal anomalies, neurointestinal anomalies, or neurologic devastation. Chronic retrograde enemas are frequently used in the management of both. In 1990, Malone et al. (51) introduced the antegrade continent enema (ACE), in which a continent cecostomy is fashioned using the appendix. They reported improvement in 15 of 21 children (71%) with a high family satisfaction rate (51). The most significant complication has been that of stomal stenosis. Malone et al. reported a stomal stenosis rate of 24%. The procedure has quickly gained worldwide popularity. Lynch et al. (52) reported their series of 30 children undergoing laparoscopic ACE procedure. They showed a continence rate of 90% and a complication rate of 34% including 27% stomal stenosis (52).

Many patients with fecal incontinence have concomitant urinary incontinence and require the use of the appendix for a continent urinary stoma. Some surgeons have placed low-profile devices directly into the cecum either laparoscopically, by laparotomy, or by means of fluoroscopic guidance. Shandling et al. (53) reported their series of 15 patients who underwent fluoroscopically guided percutaneous insertion of a cecostomy tube. The group consisted of children with neurologic impairment or children with anorectal anomalies. Cecal access could not be gained in one patient because of the presence of a interposed, distended sigmoid colon. There were no significant complications in this group, and the procedure was performed under sedation. Duh and Way (54) reported the placement of a cecostomy with laparoscopic guidance in an adult patient with colonic pseudo-obstruction for colonic decompression. Yeung and Lund (55) reported placement of a laparoscopic cecostomy for a 26-year-old patient with fecal incontinence secondary to an anorectal malformation for antegrade enemas. While cecal catheters can be placed in a variety of minimal-access fashions, further investigation is needed in order to establish the optimal method.

5. SUMMARY

In the last 20 years, endoscopic and laparoscopic advances have led to a variety of minimal access techniques for stomas. However, few papers have rigorously studied the differences between the methods of placement. Most studies that have been published are retrospective reviews of a single technique in adult patients with comparisons to historical controls. The majority of these studies have shown minimal access techniques for stomas are generally safe and may be of benefit in terms of postoperative recovery and cost. Prospective randomized studies in children would be the optimal way to investigate the validity of these retrospective studies for the pediatric population.

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15
Achalasia

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Achalasia of the esophagus is a neuromuscular (1–7) disorder in which esophageal dilation and hypertrophy occur without organic stenosis. It is characterized by an increased resting pressure of the lower esophageal sphincter (LES), failure of complete relaxation of the LES in response to swallowing, and absence of normal esophageal peristalsis. The incidence of achalasia is one in 10,000 people (8,9). It is a relatively uncommon problem in children, with those under 15 years of age comprising $<5\%$ of the symptomatic cases (10–12). Presenting symptoms include regurgitation of food, dysphagia, and failure to thrive. Respiratory difficulties may also be present and are often the result of chronic aspiration.

1. DIAGNOSIS

The diagnosis is supported by radiographic, endoscopic, and esophageal manometric studies. Plain radiographs are rarely useful but can demonstrate an air–fluid level in the esophagus. A contrast esophagram will demonstrate a widened esophagus that tapers distally ("birds beak" appearance). With disease progression, the esophagus may assume a redundant, sigmoid appearance. Endoscopy is used to rule out other clinical entities that may present similarly. In some, failure of the gastroesophageal junction to dilate with insufflation has been noted (10). Esophageal manometry is the gold-standard method to

diagnose achalasia. Characteristics consistent with achalasia include: low amplitude tertiary wave peristalsis along the entire length of the esophagus (the only absolute criterion for diagnosis), increased resting pressure of the LES (usually two-times normal), and failure of prolonged relaxation of the LES in response to swallowing.

2. MANAGEMENT

The treatment of achalasia is geared toward symptomatic relief because the etiology is ill-defined. Management strategies include oral calcium-channel blockade, pneumatic dilation, botulinum toxin injection, and surgery. The adult literature is replete with prospective and retrospective studies (13–35) examining these various treatment strategies, all with relatively large numbers of patients. In contrast, there is a relative paucity of pediatric literature (Tables 15.1–15.3), with no prospective randomized trials.

2.1. Calcium-Channel Blockade

Nifedipine® decreases resting LES pressure and improves esophageal contractions. Case reports in children have demonstrated a positive response, but symptoms recur when the medication is discontinued. Side-effects are common and only two-thirds of the adult population with achalasia experience symptomatic relief (36).

2.2. Pneumatic Dilation

Table 15.1 examines the clinical outcomes of six studies, each with at least eight children (10,37–41). The percentage of patients with "good" results, defined as minimal residual or complete resolution of symptoms after dilation, is presented. With the exception of one study (38), there was a chronological improvement in outcomes, perhaps reflecting improvements in technique. Patients who required surgery in these studies had a mean of 2.8 (range 2–3) sessions of dilation prior to surgery. Long term follow-up of these patients, who did not have surgery at the time of publication, would be interesting, to see whether the favorable outcome with pneumatic dilation was long-standing, as the adult literature demonstrates that up to 50% of patients with initial good response to dilation have recurrence of their symptoms within 5 years of treatment (42).

Table 15.1 Comparison of Studies in Children with Achalasia Treated with Pneumatic Dilation

Author (Ref.)	Number of patients	Number (%) with good outcome	Mean number of dilations in those with good outcome	Number of patients with failed outcome	Mean number of dilations in those with failed outcome
Azizkhan et al. (37)	20	5 (25)	2.4	15	3
Boyle et al. (38)	10	8 (80)	1.9	2	3
Berquist et al. (10)	8	5 (63)	1	2	2.5
Nakayama et al. (39)	15	11 (73)	1.7	4	2.5
Perisic et al. (40)	12	10 (83)	1.1	2	2
Hamza et al. (41)	11	10 (90)	2	1	3

Table 15.2 Comparison of Studies in Children with Achalasia Treated with Open Esophagomyotomy

Author (Ref.)	Number of patients	Number (%) with good outcome	Follow-up in range of years
Buick and Spitz (5)	15	12 (80)	6.2 (mean)
Vane et al. (48)	21	18 (86)	1–14
Nihoul-Fekete et al. (49)	35	34 (97)	1–25
Emblem et al. (50)	12	10 (83)	0.8–11
Illi and Stauffer (51)	16	12/14 (86)	3–22
Myers et al. (52)	175 (world-wide survey)	110/154 (71)	1–20
Morris-Stiff et al. (53)	10	9 (90)	1–23
Karnak et al. (54)	20	14 (70)	0.16–16

2.3. Botulinum Injection

Administration of botulinum toxin into the gastroesophageal region has shown short-lived usefulness. On the basis of several studies (43–45), the effect after the first treatment was only for ~4 months. The temporary relief of symptoms afforded by the toxin injection allowed patients to gain weight before surgery or was reserved for poor dilation or surgery candidates. A major drawback of this intervention is the scar that is produced in the wall of the perisphincteric esophagus, which makes surgical myotomy quite difficult and, in many cases, leads to esophageal mucosal perforation during myotomy (an effect not seen with pneumatic dilation) (46,47) The evidence suggests that there is very little role for botulinum toxin injection in the pediatric population with achalasia.

2.4. Open Surgical Intervention

The surgical esophagomyotomy, or Heller myotomy, involves cutting a 6–8 cm length of esophageal wall (sparing the mucosa). It is performed on either side of the anterior vagus nerve. It is important to extend the myotomy across the gastroesophageal junction to avoid postoperative dysphagia, although an extended myotomy across this area may predispose

Table 15.3 Comparison of Studies in Children with Achalasia Treated with Minimal Access Esophagomyotomy

Author (Ref.)	Number of patients	Surgical procedure (number of patients)	Number (%) with good outcome	Follow-up in range of months
Waldhausen et al. (60)	8	Lap/fundo (8)	8 (100)	1–24
Esposito et al. (55)	10	Lap/fundo (10)	10 (100)	6–72
Rothenberg et al. (56)	9	Lap/fundo (5) Thorac (5)	7 (78)	Not available
Patti et al. (57)	13	Lap/fundo (13)	13 (100)	19 (median)
Mehara et al. (58)	22	Lap/fundo (18) Thorac (4)	22 (100)	1–54
Mattioli et al. (59)	20	Lap/fundo (20)	20 (100)	6–102

Note: Lap, laparoscopic; fundo, fundoplication; thorac, thoracoscopic.

to postoperative gastroesophageal reflux (GER). The Heller myotomy is performed via a left thoracotomy or a laparotomy. Recently, the procedure has been performed in both the hemithorax and the abdomen using minimal access techniques (discussed subsequently).

There are eight studies (5,48–54) in which patients underwent esophageal myotomy using either a thoracotomy (with or without an antireflux procedure) or a laparotomy (with or without an antireflux procedure) (Table 15.2). The patients in whom a transabdominal myotomy with an antireflux procedure was performed had the best outcome with a 91% symptom resolution rate. In those who underwent transabdominal myotomy alone, transthoracic myotomy with an antireflux procedure, and transthoracic myotomy alone, 25%, 50%, and 53% of children experienced complete symptom resolution, respectively. Recommendations from all of these studies include the transabdominal approach with a concomitant antireflux procedure for the treatment of achalasia in children. The antireflux procedure of choice is a partial (non-360°) wrap. Most commonly a Dor or Toupet fundoplication is used.

2.5. Minimal Access Surgical Intervention

There are six retrospective studies evaluating the clinical outcome after minimal access myotomy in children (55–60). Each study had at least nine subjects. Overall, the children had good to excellent outcomes in 78–100% of the cases. The type of procedure varied between studies, but the majority underwent laparoscopy with an antireflux procedure that was a partial fundoplication. The largest study, by Mehara et al. (58), had 22 children in a multicenter study with similar outcome among all patients. The mean duration of hospitalization was less for laparoscopic esophagomyotomy than for those converted to open esophagomyotomy or for thoracoscopic esophagomyotomy. The mean time to resumption of soft feedings occurred sooner after laparoscopic than after converted open or the thoracoscopic procedures. Recommendations from all of these studies include a concomitant partial antireflux procedure.

The two most common reasons for a poor outcome following myotomy are persistent dysphagia or GER. The occurrence of each of these may reflect differences in operative approach or technique. A transabdominal myotomy is likely to be carried not only across the LES but also down onto the cardia of the stomach. In those series where a transabdominal myotomy alone is performed, the rate of postoperative dysphagia is low and that of GER is high. The length of a transthoracic myotomy is relatively short compared with the abdominal approach. Thus, GER tends to occur less, whereas the rate of dysphagia is increased. In one series of adult patients undergoing thoracoscopic myotomies (61), a long myotomy was performed and resulted in excellent outcomes with respect to dysphagia, but 60% of patients experienced GER. Using the transabdominal approach with a partial fundoplication, the same authors decreased the postoperative GER rate to 10% (62).

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16
Laparoscopic Appendectomy

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1. INTRODUCTION

Appendectomy remains one of the most common procedures done for acute abdominal pain. Since MacBurney (1) described the classic abdominal wall incision in 1894 this has remained the classic approach to the inflamed appendix with low morbidity and mortality. The application of laparoscopy to appendicitis in adults followed Semm's initial description in 1983 (2). The first laparoscopic appendectomy in children was performed in 1988 (3). However, unlike laparoscopic cholecystectomy for gallbladder disease, it has not become the procedure of choice for appendicitis in children or adults, and the indications for this approach remain controversial.

Pediatric surgeons have been able to perform major operations through small incisions for many years. Pediatric surgeons predated adult surgeons in the application of many minimal access techniques (4–6). However, without the large volume of routine laparoscopic procedures, such as cholecystectomy, many pediatric surgeons have not had an elective surgical venue to expand their laparoscopic skills. This, in addition to the "after-hours" nature of appendicitis, has also discouraged pediatric surgeons from adopting the minimal access approach for appendicitis. Longer set up and operating times, as well as the unfamiliarity of the operating room staff in pediatric centers with minimal access techniques, has slowed the acceptance of laparoscopic appendectomy. However, as minimal access surgery experience in the pediatric age group has increased so has the use of these techniques in treating appendicitis.

2. DIAGNOSIS OF APPENDICITIS AND DIAGNOSTIC LAPAROSCOPY

The diagnostic dilemma of appendicitis remains a consistent issue for the pediatric surgeon at the bedside of the child with abdominal pain. The current armamentarium includes the history, physical findings, and laboratory results in addition to reliable diagnostic imaging techniques such as ultrasonography and CT scan (7).

Laparoscopy has also been used to diagnose the source of the abdominal pain and thus may reduce the chance of removing a normal appendix. This approach has been advocated especially in young women for whom the negative appendectomy rate is as high as 50%. With laparoscopy in fertile women, the negative appendectomy rate is five times less (8). Although there are no consistent data, some studies have suggested that the normal-looking appendix can safely be left in place at the time of laparoscopy (9–13), especially when other pathology is found (14). Others believe that the low risk of incidental appendectomy may be justified by the benefit of avoiding confusion if the patient returns in the future with recurrent right lower quadrant pain.

A number of studies in adult patients looked at diagnostic laparoscopy followed by open appendectomy compared to open appendectomy alone and found no advantage to this staged approach in terms of number of wound infections (15), intra-abdominal abscesses (15,16), or length of stay (15–17).

3. LAPAROSCOPIC APPENDECTOMY TECHNIQUES

Techniques described for laparoscopic appendectomy usually involve three trocars. However, methods have been described that utilize a single trocar to visualize and grasp the appendix (18). In this technique, the appendix is delivered extra-corporeally and the mesoappendix and the base of the appendix are dealt with as in an open appendectomy. In the standard three-trocar technique, an umbilical trocar is used for initial access to the abdomen. The size of the trocars depends upon the technique used for controlling the mesoappendix as well as the base of the appendix (18–21). Usually one 10 mm trocar for extraction of the appendix and two 5 mm trocars are required for the placement of instruments. Options for the positions of the other two trocars vary between the left lower quadrant and suprapubic area or the left lower quadrant with a right upper quadrant trocar (22,23). A 5 mm telescope is most flexible as it can be moved from one trocar to another. A 30° lens allows the surgeon to adjust the angle of view but does require a more experienced person to direct the laparoscope. Cautery and/or clips can safely control the mesoappendix and endoloops are commonly used for the base of the appendix.

In both adults and children, a linear stapling device (24–26) has been used to control the base of the appendix and the mesoappendix, but requires the use of a 12 mm trocar. This, in addition to the amount of room required for the instrument, makes it less suitable for smaller children (22). However, the advantages of this technique are speed and the ability to control a necrotic or inflamed appendiceal base more easily. The extraction of the appendix can be done by pulling the inflamed appendix into the 10 mm trocar and withdrawing the appendix and trocar as a unit from the abdominal wall, or alternatively by using a specimen retrieval bag.

One trial looked at various-sized instruments and compared open vs. laparoscopic vs. needlescopic laparoscopic appendectomy technique (using 2 mm instruments) (9). It demonstrated that the needlescopic procedure was feasible but did require the use of a 10 mm port for use of clips and for extraction of the appendix.

4. LAPAROSCOPIC APPENDECTOMY COMPARED TO OPEN APPENDECTOMY

A number of studies in children have compared laparoscopic and open appendectomy but suffer from being retrospective (27) or claim to be randomized but actually allocated according to parent preference (28), making analysis difficult. Other studies prospectively compared laparoscopic and open appendectomy but were not randomized as the performance of laparoscopy depended upon the availability of the laparoscopic surgeon (29). Many randomized studies excluded patients with diffuse peritonitis and abscesses (22). Large retrospective pediatric studies have reported good results with laparoscopic appendectomy but have no control group. One series of 1379 cases reported only 4 postoperative intra-abdominal abscesses (30). In this series there were nine conversions to laparotomy ($<0.7\%$). Conversion rates from laparoscopic to open have been noted to be higher with complicated appendicitis due to technical difficulties with completing the procedure (31).

An extensive meta-analysis by Sauerland et al. (32) and a more recent analysis in the Cochrane library (8) reviewed the current literature on the surgical approach to the patient with appendicitis (Tables 16.1 and 16.2). Other meta-analyses have concluded that laparoscopic appendectomy is a superior operation to open appendectomy (33). Many studies have been done but unfortunately many are poorly designed, not blinded, and do not follow the intention to treat guidelines. Other studies do not have proper control groups while others were not properly randomized. Sauerland excluded these studies after an extensive attempt to contact original authors in studies where study design was in question (8). Following this extensive review, this meta-analysis was left with 39 studies in adults and 5 studies in children (Tables 16.1 and 16.2) that compared laparoscopic and open appendectomy and fulfilled their criteria (8).

Table 16.1 Randomized Studies of Laparoscopic and Open Appendectomy in Children (8)

		Wound infection	IAA	Pain mean cm VAS	Operation time (min)	LOS (days)	Costs US$ (surgery/ total)	Time to RTNA (days)
Lavonius et al.	LA	0/23	0/23	—	42 ± 13	3	—	—
(22) ($n = 43$)	OA	0/20	0/20	—	34 ± 13	3.1	—	—
Lejus et al.	LA	0/32	0/32	3.0	54 ± 17	—	—	—
(34) ($n = 63$)	OA	0/31	0/31	2.9	39 ± 18	—	—	—
Lintula et al.	LA	0/30	0/30	2.8	34 ± 18	1.9	—	—
(35) ($n = 61$)	OA	3/31	0/31	3.5	30 ± 11	2.6	—	—
Little et al.	LA	2/44	1/44	—	75 ± 75	4	1480/8041	2
(23) ($n = 88$)	OA	1/44	1/44	—	51 ± 51	3.5	1240/7091	1
Yeung et al.	LA	0/91	0/91	—	60 ± 60	—	—	—
(36) ($n = 181$)	OA	9/90	0/90	—	60 ± 60	—	—	—
Overall	LA	2/220	1/220	NS	Tx-9 min longer	Tx-0.6 days less		
	OA	13/216	1/216					
Statistical effect		LA*	NS	NS	OA*	LA*	NS	OA*

Note: LA, laparoscopic appendectomy; OA, open appendectomy; IAA, intra-abdominal abscesses; VAS, visual analog scale; LOS, length of stay; RTNA, return to normal activities; LA*, significant effect favors laparoscopic appendectomy; OA*, significant effect favors open appendectomy; NS, not significant.

Table 16.2 Summary Data of Randomized Studies of Laparoscopic and Open Appendectomy in Adults (8)

	Wound infection	IAA	Pain mean cm VAS	Operation time (min)	LOS (days)	Costs US$ (surgery/ total)	Time to RTNA (days)
LA	86/2213	41/2239		14.3 min difference	0.7 day difference	Variable results	6 days difference
OA	161/2111	13/2134				Variable	
Statistical effect	LA*	OA*	LA*	OA*	LA*	Overall NS	LA*

Note: LA, laparoscopic appendectomy; OA, open appendectomy; IAA, intra-abdominal abscesses; VAS, visual analog scale; LOS, length of stay; RTNA, return to normal activities; LA*, Significant effect favors laparoscopic appendectomy; OA*, significant effect favors open appendectomy; NS, not significant.

Wound infections were approximately half as common in laparoscopic appendectomy (86 of 2213) as they were in open appendectomy (161 of 2111) in the 33 analyzable studies (8). This was a consistent and statistically significant finding amongst studies regardless of antibiotics used as well as specific techniques of the procedures. In children the five randomized studies (22,23,34–36) similarly showed a definite reduction in wound infection by laparoscopy (2 of 220 in laparoscopic vs. 13 of 216 in open). This result is somewhat skewed by the excessive result in Yeung's published abstract in which there were nine wound infections in the open group and none in the laparoscopic group (36).

Among the fifteen adult studies in which intra-abdominal abscesses could be analyzed, there was a three-fold increase in abscesses after laparoscopic appendectomy (13 of 2134 in the open group vs. 41 of 2239 in the laparoscopic group) (8). In children there were no intra-abdominal abscesses in four of the five randomized studies. Thus the risk of intra-abdominal abscess in children cannot be adequately assessed. A large retrospective series of pediatric patients showed only four intra-abdominal abscesses out of 1,379 appendectomy patients (30). All four occurred among the 221 patients with complicated appendicitis (30). Others however feel strongly that laparoscopy should not be used in cases of complicated appendicitis (12). A theoretical reason for increased intra-abdominal abscess formation in perforated appendicitis may be the insufflation of CO_2 that leads to spread of the localized infection around the peritoneal cavity (37), although this has not been proven. Animal studies have suggested that in the peritonitis model induced pneumoperitoneum leads to worsening of the peritonitis as well as more positive blood cultures (38,39). It may well be the simple fact that during laparoscopic appendectomy the manipulation of the appendix is all done intra-corporeally, while in an open appendectomy it is done extra-corporeally once the appendix is mobilized. Despite these data, advocates of a laparoscopic approach for perforated appendicitis suggest that laparoscopic visualization improves the surgeon's ability to effectively irrigate and suction the peritoneal cavity (40,41).

One retrospective pediatric study has suggested that laparoscopic appendectomy should be avoided in complicated appendicitis because of a higher incidence of intra-abdominal abscess (42). A small study, which was part of a larger randomized study, suggested that laparoscopic appendectomy was associated with more major complications when compared to open appendectomy (43). The laparoscopic and open groups were not comparable in this study, as there were more perforated appendicitis cases in the laparoscopic group and more cases of gangrenous appendicitis in the open group, suggesting a milder form of complicated appendicitis in the open group (43).

In the Cochrane study, operating time was increased by 14 min in the laparoscopic group compared to the open group and likewise there was an increase in the anesthetic and overall operating room time (8). These results varied among studies. Operating time was 9 min longer in children in the five pediatric randomized studies (22,23,34–36).

Pain was found to be reduced overall in accumulated data from 11 adult series of laparoscopic vs. open appendectomy (8). Pain was assessed in a blinded manner in two pediatric studies and was not found to be reduced after laparoscopic appendectomy compared to open appendectomy (34,35).

The length of stay varied among studies, but in no study was hospital stay longer after laparoscopic appendectomy. In one blinded pediatric study the laparoscopic approach shortened stay by 0.7 days (35).

The time to resumption of normal activities, work, and sports in adults was significantly reduced after laparoscopic appendectomy, although variability was noted (8). In children, only one study looked at return to full activities. In this study the laparoscopic group was slower to return to full activities (23).

In adults the overall cost of laparoscopic and open appendectomy were similar, as the extra operating room time was balanced by the earlier discharge and the early return to work with laparoscopic appendectomy (8). Cost was only assessed in one randomized pediatric series in which laparoscopic appendectomy resulted in statistically significant increases in surgical and anesthesia costs. However the overall costs for laparoscopic vs. open appendectomy were not statistically different (23).

The physiological effect of laparoscopic and open appendectomy was studied in a randomized pediatric study (44). The laparoscopic group had significant increase in the end tidal CO_2 with a 7-mmHg elevation in blood pressure. Intraoperative serum glucose was significantly higher in the laparoscopic group. The overall significance of this is not known but does suggest that open appendectomy should be considered in pediatric patients with compromised cardiopulmonary status.

5. APPENDICEAL MASS, LAPAROSCOPY, AND THE INTERVAL APPENDECTOMY

There is no evidence in the literature to support laparoscopic drainage of intra-abdominal abscesses instead of percutaneous radiological techniques. The laparoscopic technique leads to the risk of contamination of the peritoneal cavity by infection as the abscess cavity is disrupted laparoscopically.

Controversy abounds with respect to whether an interval appendectomy is necessary after a walled-off perforated appendicitis has been treated medically. Difficulty remains as to the natural history of this condition. Retrospective studies have suggested that about 30% of cases left without an appendectomy will develop recurrent symptoms during the first year after presentation (45,46). Some may use this statistic to proceed with interval appendectomy, while others say that there is a good chance that this will not cause any more problems and recurrent symptoms can be dealt with at that time with an appendectomy (45).

Others advocate a trial of antibiotics in all cases of perforated appendicitis followed by an interval appendectomy (47). For those who believe that interval appendectomy is necessary there is support in the literature for a laparoscopic approach (48). However, there are no randomized studies comparing laparoscopic approach to open approach for interval appendectomy.

6. IMPLICATIONS FOR CLINICAL PRACTICE

Clearly, laparoscopic appendectomy in the pediatric age group is becoming more widely accepted. Unfortunately, an extensive review of the current literature does not yield a clear answer to the question of whether a laparoscopic appendectomy is superior to the traditional open appendectomy for patients with either acute or perforated appendicitis.

Individual pediatric surgeons must decide where they are in their learning curve. Most surgeons who are publishing randomized trials on this subject are past their own learning curve. So how pertinent are these studies to the average pediatric surgeon's practice?

The lack of randomized controlled trials with researchers blinded to the surgical technique remains a problem in the search for an evidence-based decision regarding the use of minimal access techniques in the management of patients with appendicitis. Unfortunately, in non-blinded studies, bias may be introduced as the laparoscopic procedure is viewed as a more advanced procedure and caregivers and patients may believe that the laparoscopic group will do better.

It seems clear that laparoscopic appendectomy is a safe alternative to open appendectomy for the child with simple nonperforated appendicitis. For children with perforated appendicitis, it is unclear whether laparoscopic is any better than open appendectomy, or whether in fact it leads to more intra-abdominal abscess formation than the open technique. This question will require further studies in pediatric patients.

In summary, laparoscopic appendectomy cannot be recommended as the preferred approach to the child with appendicitis, but is a reasonable alternative with some potential advantages and disadvantages. Certain patients, such as adolescent females, obese children, and children with doubtful diagnosis, may represent groups to whom laparoscopic appendectomy may be particularly beneficial. One additional benefit of laparoscopic appendectomy is that it is a simple, common procedure, which helps individual surgeons and pediatric institutions to become proficient in laparoscopic techniques.

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17

Meckel Diverticulum, Duplications, Small Bowel Obstruction, and Intussusception

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1. INTRODUCTION

Minimally invasive surgical techniques are now standard treatment for commonly encountered diseases of childhood such as acute appendicitis and gastroesophageal reflux disease. As pediatric surgeons have become more comfortable with these techniques, they have been applied to a variety of more unusual disorders of the small intestine such as Meckel diverticulum, intestinal duplications, intestinal obstruction, and intussusception. This chapter will summarize the principles of laparoscopic treatment for a variety of small intestinal disorders, as well as the current available evidence for these principles.

2. MECKEL DIVERTICULUM

Meckel diverticulum represents the remnant of the most proximal portion of the yolk stalk, also known as the vitelline duct or omphalomesenteric duct. By 9 weeks gestational age, the yolk stalk usually involutes. Persistence of some portion of this structure results in a spectrum of congenital anomalies termed omphalomesenteric duct remnants. These include a true diverticulum (the most common anomaly), a fibrous cord, a fistula, cysts, and sinuses. Meckel diverticulum arises from the antimesenteric border of the ileum, between 40 and 100 cm from the ileocecal valve. It is a true diverticulum, containing all four layers of the bowel wall. It may be 1–10 cm in length and may have a wide or narrow base. The blood supply arises from the mesentery, where a single artery crosses the bowel from the mesenteric to the antimesenteric side to course along the longitudinal axis of the diverticulum.

2.1. Presentation and Indications for Surgery

Meckel diverticulum is found in 2% of the population. It is not known exactly how many of these will become symptomatic, although most reports estimate between 4% and 35% (1). More than 60% of those who develop symptoms are younger than 2-year-old (2). The most common presentations include bleeding, obstruction, inflammation, and umbilical drainage, and the incidence of each varies depending on the age of the patient.

Although most cases of Meckel diverticulum are discovered incidentally during abdominal exploration for some other reason, the most common symptom is painless rectal bleeding. This bleeding occurs at a mean age of 2 years, although it can certainly occur in older children and adults. The bleeding may be massive and require transfusion. Bleeding is due to ileal mucosal ulceration by acid secreted from ectopic gastric mucosa within the diverticulum. The bleeding site is usually at the junction of the two different mucosal surfaces.

The study of choice for the diagnosis of Meckel diverticulum in a patient with painless rectal bleeding is the technetium-99m pertechnetate radioisotope scan. This isotope localizes to gastric mucosa and in ectopic locations. The study takes ~60 min and has a false-negative rate of only 5–10%. Its sensitivity can be increased by the bladder catheterization and by the administration of pentagastrin and H2-blockers. The differential diagnosis of a positive study also must include an intestinal duplication, which may also contain ectopic gastric mucosa. If no other source of bleeding can be identified, upper and lower endoscopies, tagged red blood cell scan, or angiography, as well as laparoscopy or laparotomy, can be considered.

The next most common complication associated with Meckel diverticulum is intestinal obstruction, which occurs in ~25% of patients who present with symptoms. Obstruction may occur either by volvulus of the small bowel around an omphalomesenteric fibrous band attached to the umbilicus or to the mesentery or due to intussusception with Meckel diverticulum as a lead point. Meckel diverticulitis, with or without perforation, may occur in as many as 20% of symptomatic patients. Most are misdiagnosed as acute appendicitis.

2.2. Surgical Technique

All patients requiring operation for Meckel diverticulum should be adequately resuscitated and receive perioperative antibiotics prior to their operation. Resuscitation is particularly important in the cases of bleeding and may require transfusion. Operative principles vary with the indications for operation. In the case of bleeding from Meckel diverticulum,

principles include resection of the diverticulum with its ectopic gastric mucosa, and resection of any ulcerated intestine to prevent recurrent bleeding episodes. For patients with Meckel diverticulitis, the goal is simply to remove the inflamed diverticulum. In the cases of bowel obstruction, the obstruction must be relieved whether this involves resection of an intussuscepted diverticulum or correction of a volvulus from a Meckel band and resection of the band.

The indications for resection of an incidentally discovered Meckel diverticulum are not clear. A large retrospective, population-based study by Cullen et al. (3) concluded that Meckel diverticula discovered incidentally during operation should be removed for most patients, regardless of age. However, this remains an area of controversy.

Laparoscopic exploration of the abdomen is performed through the umbilicus. The incision should be 12 mm in length to accommodate an endostapler. Carbon dioxide insufflation is performed up to pressures of 10–15 mmHg, depending on the size of the child. Laparoscopic exploration of the peritoneal cavity is best performed with a 5 mm 30° laparoscope. One or two additional 5 mm ports are placed to triangulate a working space and to manipulate and resect the Meckel diverticulum.

The diverticulum typically has a prominent feeding vessel, which originates from the ileal mesentery, crosses over the ileum, and parallels the diverticulum for its length. When present, this vessel is divided with cautery, harmonic scalpel, or clips. In the case of Meckel diverticulitis or during resection of an incidentally discovered Meckel diverticulum, an endostapler is used to divide the base of the diverticulum in the transverse dimension, so as not to narrow the intestinal lumen. When the indication for resection is bleeding, some have advocated a similar approach, whereas others recommend a segmental bowel resection to minimize the risk of leaving behind ulcerated small bowel. Although a small bowel resection can be done intracorporeally, some surgeons prefer a slight enlargement of the umbilical incision, allows the diverticulum to be delivered so that an extracorporeal resection and anastomosis can be performed.

2.3. Evidence

There are no randomized controlled studies comparing open vs. laparoscopic resection of Meckel diverticulum. Almost every report in the literature is a series of a few cases (4–7). Outcomes were excellent in each series, and there were no reported complications, but because of the nature of the studies, no conclusions can be drawn to say that the laparoscopic approach is superior.

3. INTESTINAL DUPLICATION

Alimentary tract duplications are cystic or tubular structures lined by normal gastrointestinal mucosa and having intestinal smooth muscle in their wall. They occur anywhere from the mouth to the anus, but most commonly occur in the ileum and the jejunum. Their usual location is on the mesenteric side of the normal intestine, in contrast to omphalomesenteric remnants, which are antimesenteric in location. Duplications are often associated with other congenital anomalies, and multiple alimentary tract duplications can occur in the same patient.

3.1. Presentation and Indications for Surgery

The symptoms caused by intestinal duplications depend on the type and location of the duplication. The small intestine is the most common location, accounting for ~50% of

all lesions in large series (8). Of these, approximately two-third are ileal and one-third are jejunal. Noncommunicating cystic lesions are most common, but communication with the lumen may occur. If the communication occurs distally, drainage into the adjacent bowel will occur and the lesion may remain asymptomatic. If the communication is proximal, the distal end of the duplication becomes dilated and may cause obstruction, perforation, or volvulus. In addition, some duplications may contain ectopic gastric mucosa and, if communicating, it may result in bleeding through the same mechanism as Meckel diverticulum.

Symptoms include a mass discovered on physical examination or seen on a radiographic study, pain from obstruction, perforation, or peptic ulceration, vomiting, or bleeding. Because many patients present with a confusing clinical picture, ultrasound, CT, and contrast studies may all be helpful in the diagnosis of a small intestinal duplication. Recently, many enteric duplications have been diagnosed on prenatal sonography. Radioisotope scanning has been useful to help diagnose enteric duplications which contain ectopic gastric mucosa (9).

3.2. Surgical Technique

Surgical excision is the goal for the treatment of most small intestinal duplications. Many cystic duplications can be totally excised. If the duplication and the normal intestine share a common blood supply or muscular wall, the adjacent intestine must be resected with the cyst. Resection of the adjacent small intestine can usually be performed without jeopardizing the amount of functional small intestine, except for rare long tubular duplications, in which case the duplication may be partially resected, with its remaining mucosa stripped, so as to eliminate the risks of hemorrhage or cancer. Duplications of the duodenum present a unique challenge because complete removal may endanger the bile duct or the pancreas. These duplications are best managed by marsupialization of the cyst into the duodenum by an anastomosis of the common walls or by partial cystectomy and stripping of the mucosa of the residual cyst wall (10).

Minimally invasive treatment for small intestinal duplications is possible, although many cases require advanced techniques. Some patients will present with an acute abdominal problem and the diagnosis of an enteric duplication is not made until a laparotomy has already been performed. In those patients in whom the diagnosis is known or suspected preoperatively, the cyst can be decompressed with a needle and an entirely intracorporeal laparoscopic or laparoscopic-assisted resection may be performed. The technique is similar to that used when a bowel resection is needed for Meckel diverticulum.

For a long tubular duplication, mucosal stripping can be performed laparoscopically, but it is laborious and time consuming. If the diagnosis is made laparoscopically, the umbilical port site can then be slightly enlarged, the bowel brought out and the partial resection and mucosal stripping can be done outside the abdomen.

3.3. Evidence

Literature concerning laparoscopic treatment of intestinal duplications is scarce. In one series of 13 cases, two were treated by laparoscopic-assisted resection (11). Experience should increase as more surgeons utilize minimally invasive techniques as diagnostic tools.

4. SMALL BOWEL OBSTRUCTION

Postoperative adhesions form the basis for most cases of small bowel obstruction, although hernias, tumors, and inflammatory disorders are other important diagnostic

considerations. Patients present with crampy abdominal pain and vomiting and may have a history of previous abdominal surgery. They may have abdominal distension if the obstruction is in the distal small intestine. Plain abdominal films are frequently used to study patients who may have a small bowel obstruction. Dilated loops of intestine with air–fluid levels are characteristic features. CT scanning has also been utilized to help to make the diagnosis.

4.1. Surgical Technique

The abdomen is accessed through the umbilicus, usually using an open technique both because of the risk of adhesions and because of the intestinal distension. The first issue for the surgeon is whether an adequate working space can be developed with the pneumoperitoneum. If bowel distension or extensive adhesions precludes adequate visualization, the operation should be converted to a laparotomy. In addition, if frankly necrotic bowel is seen, most surgeons would convert to an open approach. If visualization is adequate, the point or points of obstruction are identified and adhesions are slowly and carefully divided. If a bowel resection is necessary, it can be done intracorporeally or using a laparoscopic-assisted approach.

4.2. Evidence

Laparoscopic adhesiolysis has been described in a number of case reports and series in both adults and children (12–14). Most authors have concluded that the laparoscopic approach is an excellent way to start, but that the procedure should be converted to open if visualization is inadequate or necrotic bowel is found. The results of laparoscopic adhesiolysis may be improved by using enteroclysis to identify the point of obstruction preoperatively (15).

5. INTUSSUSCEPTION

Ileocolic intussusception is a very common condition, which usually occurs in infants between 3 and 12 months of age. However, it can be seen in older and younger children as well. Many children have a history of a preceding viral illness. The etiology is not completely understood, but the most common explanation is that a viral illness results in hypertrophy of the Peyer patches within the bowel wall, which then act as a lead point for the intussusception. In some children, there is a "pathologic" lead point such as a polyp, tumor, Meckel diverticulum, or duplication.

Although the most common presentation is intermittent abdominal pain, intussusception may also cause rectal bleeding ("current jelly" stools) and intestinal obstruction. The diagnosis is suspected on the basis of the history and physical examination, and the definitive diagnosis is made either by contrast enema or more recently by ultrasound. Most intussusceptions can be successfully treated in the radiology suite using a barium or air enema, with larger series showing a success rate of $\sim$81% (16). Surgery is indicated for those cases that cannot be reduced hydrostatically or pneumatically, for cases in which perforation occurs during attempted reduction, or for recurrent cases where a pathologic lead point has been identified.

5.1. Surgical Technique

In some cases of failed reduction, the intussusception will actually have reduced itself between the radiology suite and the operating room due to reduction of edema and due to the effects of general anesthesia. The first goal of surgery is therefore to determine whether the intussusception is still present. This can be accomplished by inserting a small umbilical port to examine the ileocecal area. If the intussusception is no longer present, the port can be removed and the patient awakened. If the intussusception is still present, additional ports should be placed and a gentle attempt made to reduce the intussusception. Pneumaticically-assisted laparoscopic reduction has also been reported (17). If these maneuvers are unsuccessful, most surgeons would choose to make a right lower quadrant incision and complete the operation using an open technique. This might involve manual reduction or may require resection.

5.2. Evidence

There have only been a few series of patients undergoing laparoscopy for ileocolic intussusception (18–21). These reports confirm the safety of the technique and confirm its usefulness both as a diagnostic tool to ensure that the intussusception has not spontaneously reduced and as a technique for accomplishing reduction in some cases. However, most studies acknowledge that the need to convert to an open procedure is higher in this condition than for most other pediatric surgical diseases.

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18

Laparoscopic-Assisted Total Colectomy with Pouch Reconstruction

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1. INTRODUCTION

In general, children with ulcerative colitis and familial polyposis can be managed medically into adulthood. However, some children have unremitting bloody diarrhea even with maximum medical management. In addition, some children are growth retarded or have delayed puberty. The negative sequelae associated with the long-term use of corticosteroids is also an indication for total proctocolectomy in these children (1).

Restorative proctocolectomy with J-pouch pull-through is well tolerated by most children and adolescents (2). A laparoscopic-assisted technique is used for the total colectomy with the addition of a small suprapubic incision for the proctectomy and J-pouch formation. The author prefers a mucosectomy for the proctectomy in order to remove all of the rectal mucosa. The double-staple technique commonly favored for adult patients leaves more abnormal rectal mucosa behind. Throughout the lifetime of the child or the adolescent, there is potentially more time for the development of stricture or neoplasia in the native rectal remnant. The author uses a diverting ileostomy in most patients for 6–8 weeks to minimize the potential for anastomotic leaks in the pelvis.

2. OPERATIVE TECHNIQUE

The patient's colon should be cleansed mechanically before elective proctocolectomy. Broad-spectrum antibiotics are administered perioperatively. The patient is placed

supine in stirrups on the operating table. A urinary catheter is inserted by the surgeon after the patient has been prepped and draped. Four or five cannulas are placed as indicated in Fig. 18.1. Colon mobilization is begun in the rectosigmoid mesocolon just proximal to the rectosigmoid junction. The division of the mesocolon is best accomplished using an ultrasonic scalpel. The dissection is kept very close to the colon at all times to avoid encountering any large vessels which would not be well controlled by the ultrasonic scalpel. Alternatively, the dissection can be made more proximal in the mesocolon with the large vessels controlled by vascular staples or clips. However, the mesentery in these children is frequently thick owing to the fatty infiltration induced by the chronic use of corticosteroids. For this reason, keeping the dissection close to the colon is usually the safest technique. Once an opening is made completely through the rectosigmoid mesocolon, the dissection is continued proximally up to the terminal descending colon. Care must be taken to recognize the proximity of the left ureter to the sigmoid mesocolon. As long as the dissection is kept close to the colon, the ureter is usually well out of harm's way. At the level of the distal descending colon, the fusion fascia is divided. The author prefers to use scissors or a hook cautery for this division, as it is usually faster than the use of the ultrasonic scalpel. The dissection of the mesocolon is carried proximally to the level of the splenic flexure. As the surgeon approaches the splenic flexure, the mesocolic dissection becomes more difficult. It should be noted that the mesocolon can be

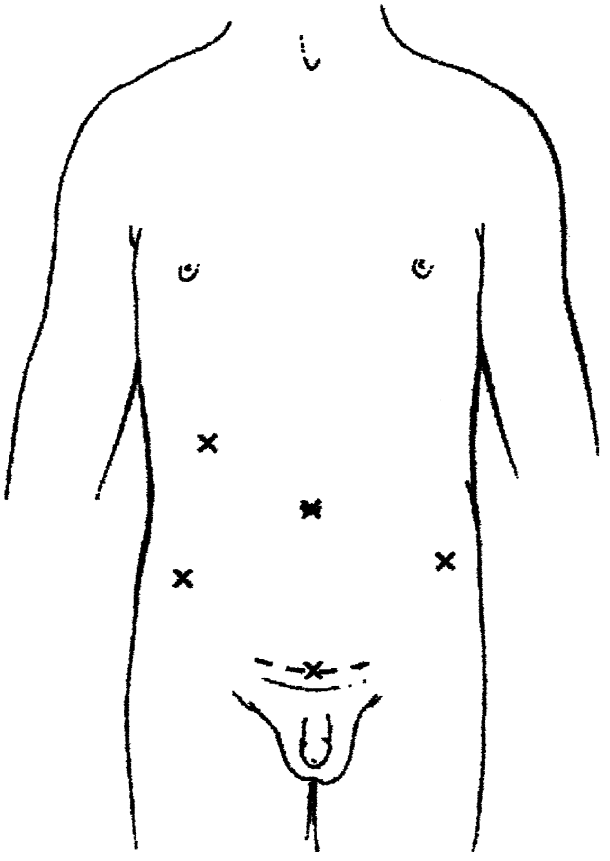


Figure 18.1 Trocar and incision sites used in laparoscopic-assisted proctocolectomy.

divided either from a medial approach or more commonly by pulling the colon medially and dividing the mesocolon lateral to the colon. In some situations, the medial approach is more efficacious, but in others the lateral approach can be useful and will speed the operation. When this mesenteric dissection becomes difficult and confusing, the gastrocolic ligament should be approached from the right side of the patient beginning at the level of the falciform ligament. Care must be taken not to divide both the gastrocolic ligament and the mesentery simultaneously as they are sometimes intimately attached to one another. The gastrocolic ligament is opened all the way to the lienocolic ligament. The lienocolic fibers are also divided. The mesocolon is then divided. The mesocolon can again be divided from below the colon inferiorly or by pulling downward on the colon and dividing the mesocolon above the colon. Both techniques can be useful and can aid in the more rapid resection of the colon. Once the splenic flexure of the colon has been amputated from the mesocolon, the surgeon changes position from the right side of the operating table to a point between the patient's legs, which are held in stirrups. The table should be tilted to the right during the dissection of the descending colon and splenic flexure. While dissecting the transverse mesocolon, the patient should be placed in a reverse Trendelenburg position to bring the small bowel and omentum toward the pelvis for better visualization of the transverse mesocolon. Some surgeons advocate retaining the omentum, but the author prefers to resect the omentum with the colon. The gastro- and hepatocolic ligaments are divided over to the hepatic flexure. When it is easily done, the transverse mesocolon is also divided. It is important to remember the pesky proximity of the duodenum to the hepatic flexure of the colon. The duodenum can be easily violated, if the surgeon is unaware of its adhesion to the transverse mesocolon. When the dissection of the transverse colon and hepatic flexure becomes difficult, the focus of the dissection is changed to the ascending colon. The appendix and ascending colon are mobilized away from the abdominal wall by dividing the fusion fascia. The terminal ileum is also mobilized away from the posterior abdominal wall at this point to make the eventual positioning of the J-pouch easier. It is expeditious to mobilize the terminal ileum and its small bowel mesentery away from the posterior attachment to the abdominal wall at this point. The fusion fascia of the ascending colon is divided. The ascending mesocolon is divided as close to the colon as possible. Every effort should be made to save the right colic and marginal arteries which are helpful in providing blood supply to the terminal ileum after the formation of the J-pouch. The proximity of the duodenum to the right transverse mesocolon should also be noted during this upward division of the ascending mesocolon (Fig. 18.2).

Once the entire colon has been devascularized and all of its intra-abdominal attachments have been divided, a suprapubic transverse incision is made into the peritoneal cavity. Injury to the bladder and the vas deferens should be carefully avoided. Once the peritoneal cavity is entered, the distal colon is divided with a gastrointestinal stapling device. The abdominal colon is removed through the suprapubic incision. The mesentery to the distal sigmoid colon and the proximal rectum are divided down to the peritoneal reflection of the pelvis. Saline with epinephrine solution is injected into the wall of the proximal rectum circumferentially. A plane is developed between the smooth circumferential muscle of the rectal wall and the submucosa. Dissection is continued down toward the anus from above in this plane using blunt and sharp dissection. A combination of a needle-tip electrocautery, fine-tipped scissors, and Kittner dissectors are used for this dissection. The mucosa should be stripped away from the muscular rectal wall with gentle upward traction applied to the mucosal sleeve to facilitate the dissection. Once the mucosectomy has been carried down to a point 4–5 cm from the dentate line of the anorectal junction, the transabdominal portion of the dissection can be discontinued. At this

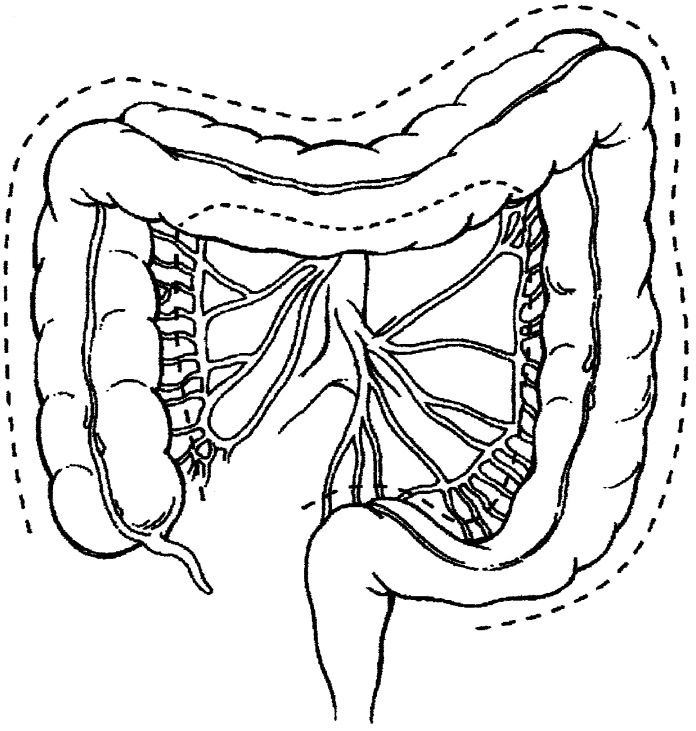


Figure 18.2 Laparoscopic detachment of the abdominal colon.

point, the J-pouch is constructed. The terminal ileum has previously been mobilized. The point on the terminal ileum for maximal length of the J-pouch is determined and is usually ~10–20 cm proximal to the ileocecal valve. A J-pouch is fashioned with each limb ~6–8 cm in length. The J-pouch is folded on itself and secured with a few basting sutures. Small incisions are made in the medial aspect of each proximal limb of the J-pouch to accommodate the stapling device. The spur is stapled and divided through these small incisions (Fig. 18.3). Two to three centimeters of distal spur is usually left after stapling from above. This distal spur is left in place and stapled from below. Often, this spur is left in place for ~8 weeks and is divided at the time of closure of the ileostomy, when the blood supply to the J-pouch is more secure and there is less upward tension on the J-pouch. The J-pouch is assessed for adequate length. In males, adequate length usually means the pouch can be brought out through the wound and over the pubis to the level of the scrotum. In females, the tip of the exteriorized J-pouch must reach the level of the clitoris to be considered long enough. Several lengthening techniques are useful. Transverse incisions in the anterior and posterior small bowel mesentery over the superior mesenteric artery and vein will usually allow several centimeters of increased length. In addition, division of restraining vessels can be accomplished if small bulldog clamps have been placed on the vessels and the remaining collateral blood supply is noted to be adequate. If there is any doubt about adequacy of blood supply, the bulldog clamp can be left in place during the transanal dissection of the remaining mucosal cuff and then re-evaluated. The posterior peritoneal attachment of the distal small bowel mesentery should also be divided on the right side beginning inferiorly and extending up to the duodenum. This release of the small bowel mesentery will usually provide another 1 or 2 cm

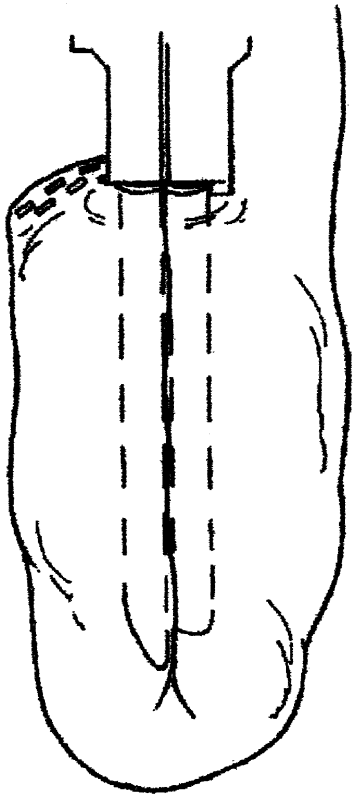


Figure 18.3 Formation of the J-pouch with division of the proximal spur.

of length to the pedicle. Another helpful maneuver is to divide the mesentery immediately adjacent to the inner portion of the J over a distance of ~3 cm. Separating the mesentery from the rounded portion of the J-pouch does not compromise the blood supply to the J-pouch because of adequate collateral inflow. This technique often allows an additional 1 or 2 cm of J-pouch length.

Once the length of the J-pouch is adequate, the patient's legs are flexed in the stirrups to allow for access to the anus. Retraction sutures are applied to the dentate line and attached to the buttocks 3 or 4 cm laterally in radial fashion. Six to eight sutures are placed radially so that they distract the anus for excellent visualization during the mucosal dissection and anastomosis. A circumferential incision is made in the mucosa near the top of the rectal columns in the transitional epithelium (Fig. 18.4). The surgeon should avoid leaving rectal mucosa behind because this mucosa can lead to the development of scarring and cancer over time. The mucosal sleeve should be cored out in a similar fashion to the dissection from above. The submucosa is infiltrated with saline and epinephrine solution. The circumferential incision is made. The edges of the proximal mucosa are secured together with multiple fine sutures, and the dissection is continued circumferentially between the circular smooth muscle of the rectal wall and the submucosa (Fig. 18.5). By continuing this dissection upward, the entire mucosal sleeve of the rectum is cored out. Once the transanal dissection meets the dissection already completed from above, the rectal mucosal sleeve is pulled out through the anus. It is possible to perform the entire submucosal sleeve dissection through the anus. However, the author has found this to be much

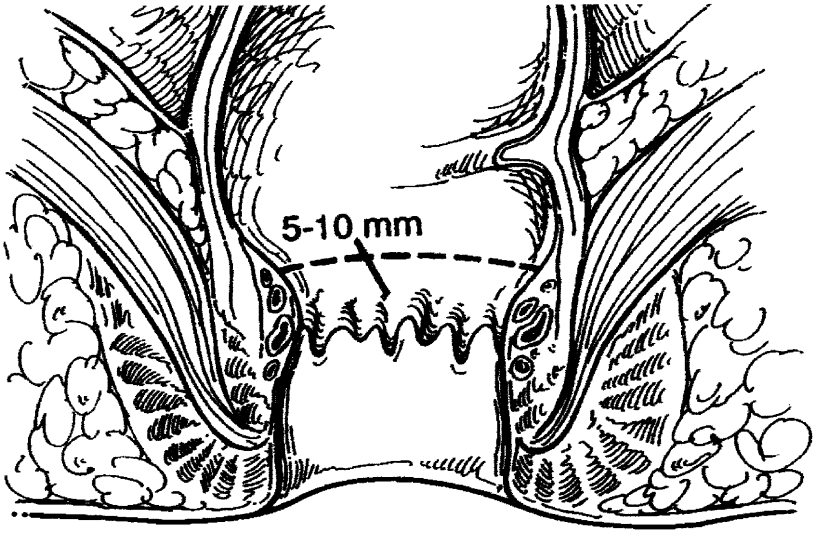


Figure 18.4 Circumferential incision in the anorectal transitional epithelium.

more difficult and time consuming. In addition, it requires significantly greater dilatation of the internal and external anal sphincters which may be deleterious to continence.

The remaining muscular sleeve may be divided posteriorly. This division of the sleeve is mandatory in the rectal dissections for Hirschsprung's disease, but is elective in patients with ulcerative colitis and familial polyposis. The J-pouch is pulled into

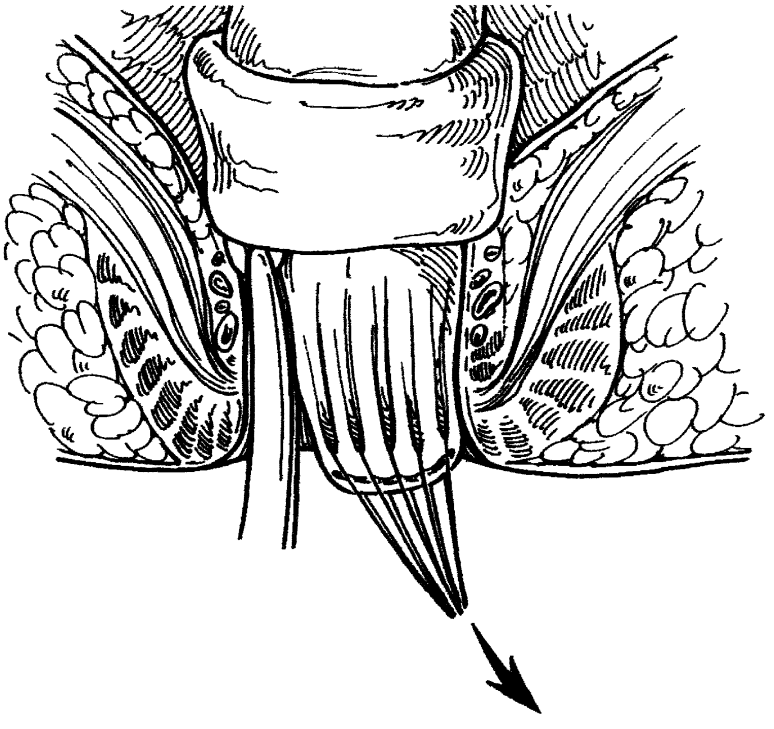


Figure 18.5 Transanal mucosectomy.

position through the muscular sleeve (Fig. 18.6). An anastomosis is made in a single layer by opening the distal portion of the J-pouch. The distal incision in the J-pouch should not be >2 cm as it tends to stretch during the formation of the anal anastomosis. As mentioned earlier, the remaining spur in the J-pouch is divided 6–8 weeks later when the ileostomy is closed (Fig. 18.7). In most cases, a protective ileostomy is to be performed so the anorectal anastomosis does not need to be water tight. Indeed, sometimes the anastomosis is very difficult because of tension on the J-pouch from above by a short mesentery. It is important to place at least 10 sutures circumferentially for this anastomosis, while avoiding tearing the wall of the J-pouch.

The surgeon's gown and gloves are changed. Transabdominal inspection of the small bowel and its pedicle should be performed carefully. The potential internal hernia behind the pedicle of the J-pouch should be closed with interrupted sutures taking care to avoid injury to the blood supply of the J-pouch. The pedicle should also be evaluated for twists. In addition, the pedicle should not be so tight that the blood supply is compromised. If so, further efforts to relieve the tension of the pedicle should be made.

A loop ileostomy is brought out onto the abdominal wall. Occasionally, the patient has such a short mesentery and such a thick abdominal wall that a loop ileostomy is impossible to perform. Under these circumstances, the stapled end of the distal ileostomy

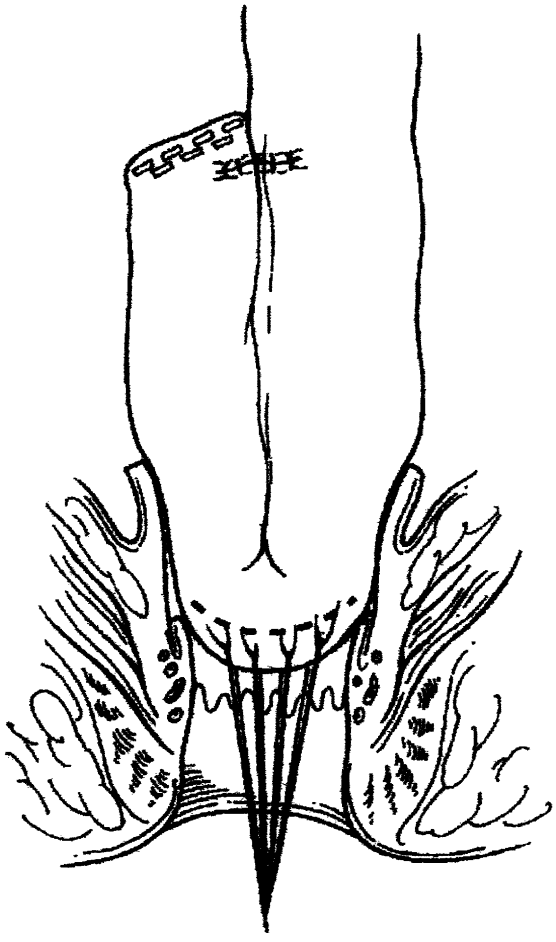


Figure 18.6 The J-pouch is pulled into position.

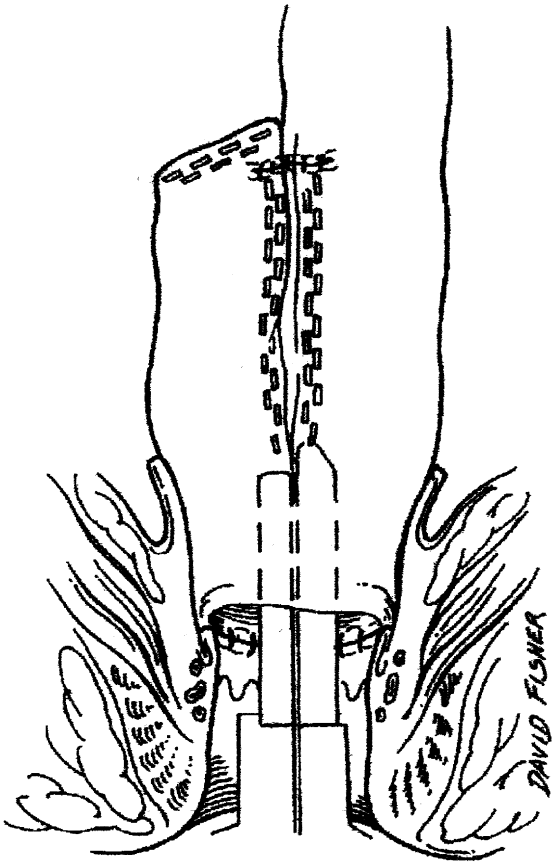


Figure 18.7 The distal spur is divided at the time the ileostomy is closed.

should be attached to the proximal limb of the ileostomy inside the peritoneal cavity so that it can be found easily for closure of the ileostomy in 6–8 weeks. Fascial attachment of the ileum to the anterior sheath should be performed to reduce the chances for a peristomal hernia. The ileostomy should be matured after the abdominal wounds have been closed.

The suprapubic wound and the trocar sites are closed. The skin of the suprapubic wound should be closed loosely to help prevent a wound infection. Broad spectrum antibiotics are continued for 3 days. Bowel function usually begins on the second or third postoperative day. Most patients are ready for discharge by the fifth or sixth postoperative day.

3. RESULTS

Total proctocolectomy with J-pouch reconstruction carries a relatively high morbidity rate both with the open and also the laparoscopic-assisted procedures (3,4). The advantages of laparoscopic proctocolectomy over open colectomy have been documented primarily in adult patients. Return of bowel function, advancement of an oral diet, and hospital length of stay generally favor the laparoscopic approach (3–6). Operative times are generally longer with the laparoscopic-assisted approach. Functional results including

continence, number of stools, and soilage are similar (3–6). Dunker et al. (5) has reported that body image after a laparoscopic approach is better than that after open surgery as demonstrated by a standardized image questionnaire. About 25 (25%) of the patients have complications with wound infection, small bowel obstruction, and ileostomy dysfunction being the most common. The author has performed 25 laparoscopic-assisted colectomies with J-pouch reconstruction in children and adolescents. The laparoscopic-assisted approach took 56 min longer than the open approach in the author's experience. One of the 25 patients has had to have the J-pouch resected and an end ileostomy performed when the patient developed obvious Crohn's disease in the pouch. Three of the patients are still in the early postoperative stage. Of the remaining 21 patients, two have occasional nighttime leakage but are continent in the day. This nighttime incontinence is of the urgency type in which the patient does not awaken rapidly enough to get to the commode. This symptom is usually ameliorated by refraining from eating and drinking at least 2 h before bedtime. The other 19 patients have satisfactory daytime continence and do not have nighttime incontinence. The author believes that the shortness of the J-Pouch (6–8 cm) is a significant factor in the success of this operation. On an average, the patients had two to six bowel movements per day. Most patients require Imodium during the first few years after pull-through but often stop taking it in later years without any significant increase in their stool output. All of these children have had excellent resolution of their growth and maturation problems after the discontinuation of steroids (2). Cosmetic results are better after the laparoscopic approach when compared with the open approach, although no body image instrument has been validated for children and adolescents.

4. CONCLUSIONS

Laparoscopic-assisted total proctocolectomy with J-pouch reconstruction is a useful therapeutic tool. Outcomes are excellent but complication rates approach 25% as in the open procedure. Excellent long-term outcomes are achieved with an average of two to four bowel movements per day and very satisfactory continence.

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19
Minimal Access Surgery for Hirschsprung Disease

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1. INTRODUCTION

Hirschsprung disease is characterized by the absence of ganglion cells in the myenteric and submucosal plexuses. Usually, the abnormality involves the rectosigmoid region, but in some cases it may extend proximally to involve the entire colon or even some of the small bowel. The absence of intrinsic innervation results in disturbed motility, with resultant neonatal bowel obstruction, constipation, or enterocolitis.

Since the pathophysiology of Hirschsprung disease was first described in the 1940s, there has been an evolution in the surgical management of this condition. Initially, a colostomy was used routinely and was then followed by an operation that involved resection of the aganglionic bowel and reconstruction using one of several "pullthrough" techniques, most commonly the Swenson, Soave, Duhamel, or Rehbein procedures. In some cases, the stoma would then be closed in a third operation.

Over the past 15 years, a number of authors have described one-stage pullthroughs for Hirschsprung disease (1–3). More recently, minimal access approaches to the one-stage pullthroughs have become popular. These consisted of pullthroughs utilizing laparoscopic abdominal and pelvic mobilization of the rectum (4–6) and a number of transanal approaches which do not include any intra-abdominal dissection (7–9). This chapter will

review the principles and current evidence for the use of minimal access surgery in the management of this disease.

2. IS A ROUTINE COLOSTOMY NECESSARY?

At the time of Swenson's first description of the surgical repair of Hirschsprung disease, many children presented with significant malnutrition, inflammation, and colonic dilatation. In these cases, a colostomy was a life-saving procedure and became the standard of care. Over the past 15 or so years, a number of authors questioned the need for routine colostomy and published series of cases repaired using a one-stage approach (10–12). These studies suggested that a one-stage approach was technically feasible and safe. A longer term study documented excellent functional results in children undergoing a one-stage endorectal pullthrough, although there was no control group that had undergone a two-stage approach (13). Subsequently, several comparative series were published, using historical controls. In these studies, there was general agreement that short-term outcomes were similar and that the use of a colostomy was associated with a specific set of complications (1,14). It is important to point out, however, that a one-stage pullthrough requires a judgment of the level of the transition zone based on frozen sections, which relies on the expertise and experience of the pathologist (15). This approach should not be used, if such a pathologist is unavailable.

3. LAPAROSCOPIC PULLTHROUGH

The first descriptions of a laparoscopic approach to pullthrough surgery were in animal models. Subsequently, technical descriptions of laparoscopic Duhamel and Swenson procedures were published (5,6). In 1995, Georgeson et al. (16) described a laparoscopic pullthrough which combined the rectal dissection done in the Swenson procedure with a mucosal dissection similar to a Soave. This operation has become the predominant laparoscopic pullthrough in North America.

There are few studies thus far which have critically evaluated the results of laparoscopic surgery for Hirschsprung disease. In a large multicenter report of 80 patients, the mean length of stay was 3.7 days, and the complication rate was comparable to those reported in the literature (17). Other single center series of the Georgeson operation and the Duhamel have also suggested that the laparoscopic approach is safe and associated with less pain, shorter hospital stay, and lower cost than open surgery (6,18,19). No prospective or randomized trials have been reported to-date.

4. TRANSANAL (PERINEAL) PULLTHROUGH

The transanal pullthrough developed as an extension of the laparoscopic approach and uses essentially the same anal dissection described by Georgeson. However, in a transanal pullthrough, there is no intra-abdominal dissection of the rectum, and potentially no incisions or port sites in the abdominal wall. Initial descriptions of the technique used a Soave approach with a submucosal dissection, coming through the muscular cuff at some point above the sphincters and continuing the dissection on the serosal surface of the rectum and more proximal colon until the transition zone is reached (7–9). More

recently, a transanal Swenson procedure has been described, where the full thickness of the rectal wall is divided just above the dentate line (Walton, unpublished data).

As with the laparoscopic approaches, there are few comparative series that critically evaluate the results of the transanal approaches. In several series comparing children undergoing a transanal pullthrough to those having a one-stage open Soave, the transanal approach was associated with less pain, shorter hospital stay, and lower cost (20,21). Functional outcome have also been shown to be similar to those undergoing an open operation (22). There have not been any prospective or randomized studies comparing the transanal and open approaches.

There have not been any studies that directly compare the laparoscopic technique with the transanal technique. Because both approaches have enthusiastic proponents, and the results with both operations are excellent, it is unlikely that such a study will be done.

5. IS ROUTINE IDENTIFICATION OF THE TRANSITION ZONE NECESSARY?

One of the issues that is often debated concerning the minimal access approach is the need to do biopsies to identify the transition zone, prior to beginning the anal dissection. Biopsies can be done laparoscopically, or in the case of the transanal procedure, through a small umbilical incision. The advocates of routine intra-abdominal biopsies express concern that an error in preoperative localization of the transition zone might result in an inappropriate operation being done for long-segment disease. For example, many surgeons prefer to do a Duhamel reconstruction for total colonic disease, and if the submucosal dissection had already been done before transition zone had been identified, the surgeon would be committed to a Soave procedure. Other surgeons have suggested that intra-abdominal biopsies be reserved for children who are at increased risk of long-segment disease, such as those with a positive family history or those with proximal or inconclusive transition zones on barium enema (20).

There are no comparative studies looking specifically at this question. It has been reported that 10–20% of infants with Hirschsprung disease will have no demonstrable transition zone on barium enema (23), and there is some evidence that the transition zone seen on the barium enema may be inaccurate in up to 10% of cases (24). As there does not seem to be any increased risk or difference in outcome with a biopsy through a laparoscopic approach or small umbilical incision, surgeons should probably be liberal in the use of intra-abdominal biopsies prior to beginning the anal dissection.

6. TIMING OF ONE-STAGE PULLTHROUGH

Many series have reported one-stage pullthroughs, done open or using MAS techniques, even in newborn infants. However, there are many surgeons who prefer to wait until the child has attained a certain weight, in order to improve visualization and make the operation technically easier (25). During this waiting time, the child is usually managed with rectal stimulation or irrigations to maintain evacuation of stools and prevent enterocolitis and is usually placed on either breast milk or an elemental formula to decrease stool output.

There are no controlled studies examining the need to attain a specific weight or to critically evaluate the safety of neonatal one-stage pullthrough. However, retrospective

studies have not noted different complication rates between the neonatal cases and the older ones. In addition, there have been occasional cases of enterocolitis occurring despite irrigations during the waiting period; some of which have proven to be fatal. It therefore appears that neonatal pullthrough is preferable in most cases, particularly in the hands of an experienced surgeon. However, this is a question which requires further study.

7. SUMMARY

Minimal access surgical techniques for the management of Hirschsprung disease are increasing in popularity. Most children can now be managed in a single operation, without a colostomy. The laparoscopic and transanal approaches have both been used successfully and are superior to open techniques by the virtue of decreased pain, earlier feeding, earlier discharge, and lower cost. Issues, such as which procedure produces the best long-term outcomes, what is the best timing of surgery, and whether routine intra-abdominal biopsies are necessary, remain controversial and will require further study.

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Minimal Access Treatment of Anorectal Malformations

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1. INTRODUCTION

To treat diseased or anomalous anatomy, pediatric surgeons have traditionally had to incise and dissect normal tissue planes to provide adequate surgical exposure. The application of minimal access techniques has revolutionized the performance of many routine pediatric surgical procedures by enabling the surgeon to access body cavities without significantly traumatizing intervening fascia, muscles, and nerves. The use of small puncture

wounds and trocars has resulted in fewer complications (e.g., wound pain, physiologic stress, infection, dehiscence, and incisional hernia) related to traditional open surgery (1–5). Moreover, laparoscopic magnification and illumination can enhance surgical exposure and anatomic visualization beyond that achieved with open operations.

The laparoscopically assisted anorectal pull-through (LAARP) for high anorectal malformation (ARM) employs fundamental concepts learned from decades of open surgical ARM repair and additionally incorporates modern technologic advancements in surgical instrumentation and technique (6). LAARP combines extraordinary anatomic exposure of an infant's deep pelvis with a reconstruction technique that minimizes surgical exposure-related trauma to important surrounding structures.

This chapter focuses on the historic background upon which this operation was conceived and the principles upon which it is based. The technical details of LAARP and a critical assessment of the merits of this procedure compared to other operations for high ARM are also presented. Outcome data are presently limited and will also be presented.

2. HISTORY OF SURGICAL REPAIR OF ARM

For centuries, surgeons have been faced with the challenge of how best to restore anorectal function in infants born without an anal opening. It has only been within the past 3 decades that significant strides have been made in understanding the anatomy and pathophysiology of the various types of ARM. Both Webster (7) and De Vries (8) have published comprehensive accounts of the surgical history of ARM. A brief overview of these accounts is important for understanding the current methods of treatment.

The earliest report of surgical treatment for ARM is from the 7th century when a blind perforation of the perineum and rectum was made with bistoury (long, straight blade), followed by anorectal dilatation. At that time, high ARM was essentially a lethal diagnosis. There was neither any concept of transperitoneal abdominal surgery nor any concept of decompressive colostomy. The goal was to relieve the bowel obstruction in the most expeditious fashion. Since surgeons were concerned about doing harm to muscles and nerves within the neonatal pelvis, no serious attempt at dissection deep into the perineum for the purpose of rectal mobilization and anorectoplasty was made for many centuries. Blind perforation was reasonably effective for patients with very low lesions, when the rectum ended within a centimeter of the perineal skin. However, for patients with higher ARM, this technique was largely a failure, resulting in strictures, wound infections, and septic death.

In 1835, Amussat realized that better exposure was needed to mobilize the bowel. He reported a perineal dissection of the blind-ending rectum high enough to bring the bowel down to perform a sutured proctoplasty between the rectum and perineal skin. The principles of strict midline dissection, avoidance of major pelvic floor disruption due to blind trocar passage, adequate rectal mobilization, and suture fixation of the bowel to the skin became the standard for many years to come. Although these principles helped to prevent many of the problems associated with severe strictures, the reconstructed rectum was not enveloped in any natural way by an intact, functional sphincter mechanism. Incontinence was therefore a common result.

With the availability of inhaled anesthesia and the application of Listerian principles by the end of the 19th century, dissection high into the pelvis could be accomplished more safely. In many cases, surgeons were able to adequately mobilize the bowel from a perineal approach. Laparotomy was also used (abdominoperineal approach) to enhance exposure of higher fistulae from the rectum to the genitourinary system.

In the first half of the 20th century, surgeons gradually became more comfortable with the abdominoperineal approach to high ARM, and colostomies were also sometimes performed to allow delayed reconstruction. Contrary to popular belief at that time, neither the puborectalis muscle (deep behind the bladder neck) nor the exact midline of the deep pelvis could be readily observed or used during reconstruction. Additionally, a strict midline perineal dissection without the benefit of a muscle stimulator was difficult to perform.

In 1953, Stephens (9) realized the critical contribution of the puborectalis muscular sling to postoperative continence, and endeavored to identify and preserve the integrity of this important structure when repairing high ARM. The addition of a sacral incision—to provide a better vantage point for identification of the puborectalis and dissection of the rectal fistula—was considered a major advance. The sacroperineal approach employed a right angle clamp placed through the trans-sacral wound to dissect a tunnel anterior to the puborectalis muscle. This dissection plane from above then joined a small midline perineal dissection, with the surgeon receiving tactile guidance by placement of a finger into the perineal incision. A laparotomy (sacroabdominoperineal approach) was only performed if deemed necessary based on rectal position or size. Emphasis was placed on preservation of the puborectalis muscle. The external sphincter was, however, largely disregarded because it was believed to contribute little to continence for patients with ARM. Many modifications of this approach were developed, including use of a mucosa stripping, endorectal pull-through (10).

Surgeons now still acknowledge the critical contribution of the puborectalis to continence, but also understand the need to place the bowel within the confines of the more caudal striated muscle complex and external anal sphincter (EAS) in order to achieve optimal anorectal function (11,12). Interestingly, Stephens readily acknowledged that in creating the tunnel for the bowel within the puborectalis sling, neither the puborectalis muscle nor the levator diaphragm was well visualized. Nevertheless, his premise was that the correct anatomic plane through the levators could be defined. This premise has been shown to be incorrect in many instances, and modern imaging techniques and operative revisions have documented the misplacement of the bowel through the levators in some patients who underwent abdominoperineal as well as sacroperineal pull-through (13,14). Although long-term follow-up (5–32 years) after such operations for high and intermediate ARM has shown that a majority of patients ultimately have socially acceptable continence, few are free of soiling between bowel movements (15). It is important to note that the number of patients who underwent these older procedures and who are available for long-term follow-up evaluation is only a fraction of all patients treated. Accurate assessment of outcome is thus quite difficult.

Difficulties encountered in anatomic visualization with the sacroperineal and sacroabdominoperineal operations led deVries and Peña to return to the prior concepts of high dissection from a posterior sagittal approach originally described by Amussat. They refined this technique by the addition of a muscle stimulator and performed an extensive, meticulous dissection from the perineum that was termed posterior sagittal anorectoplasty (PSARP) (11). This procedure represents one of the most important contributions in the history of treatment of ARM, and has now gained considerable acceptance worldwide. PSARP allows for the widest degree of surgical exposure of the high malformation of the rectum, with a midline incision from the base of the scrotum or introitus to the sacrum. Essentially all of the voluntary muscles of continence are identified in the midline. Specifically, the external anal sphincter, the vertically oriented striated muscle complex, and the levator ani muscles are seen and divided into two halves during PSARP. These facts also formed the basis for early criticisms of the technique (16). We

have learned now that when a strict midline dissection and precise reconstruction of muscles is properly performed, PSARP represents an elegant exposure and reconstruction of the pelvic anatomy. In most patients with high lesions, the bowel is found during PSARP immediately deep to the levator group. In patients with rectovesical fistulae, a laparotomy is additionally required because this type of fistula cannot be adequately exposed from below. It is clear that considerable mobilization of the rectum can be accomplished using this approach, with excellent visualization and protection of urogenital structures.

Clinical outcomes after PSARP vary depending upon the institutional experience with ARM (17) and the level of the malformation. Functional results after PSARP, as with many other procedures, are also dependent in part on technical factors at the time of operation. Specifically, the dissection must "remain strictly in the median plane," with precise and meticulous reconstruction (18). This goal can be a technical challenge in that some patients may not have sufficient symmetric striated muscle bulk to guide a strict midline dissection, even with optical magnification and electrostimulation.

In an impressive long-term follow-up of nearly 1200 of Peña's patients with all varieties of ARM, total continence (voluntary bowel movements and no soiling) was seen in 39% of all patients (12). Subgroup analysis demonstrates that essentially all patients with a perineal fistula were totally continent. Total continence was seen in only 55% of girls with a vestibular fistula, 31% of boys with a rectobulbar urethral fistula, 20% of boys with a rectoprostatic urethral fistula, and none of the patients with rectovesical fistula. The extent to which the muscle-splitting dissection ultimately impairs overall anorectal function is not clear. Follow-up has also shown that constipation is also seen in some patients after PSARP. Whether this aspect of anorectal dysfunction is related to the scarring after perineal dissection or more likely to intrinsic dysmotility of the colon is not certain.

A problematic issue pertaining to patients with high ARM is determining the inherent potential for continence in each individual patient. In light of this, surgeons have been unable to know with any certainty what goals are realistic and achievable for continence in individual patients. The potential for continence varies greatly depending on the degree of development of pelvic and perineal anatomy, and the general health status of the patient. The ultimate outcome is also likely to be dependent on some factors that are surgically correctable (e.g., proper positioning of the bowel within the levators and striated muscle complex) and on some factors that are not surgically correctable (e.g., intrinsic bowel dysmotility, pelvic muscular hypoplasia or asymmetry, abnormal pelvic innervation). It seems reasonable to assume, however, that if an anatomic reconstruction of the ARM (such as that which results after PSARP) could be achieved without surgical trauma to the continence mechanisms (e.g., pelvic nerves and musculature), the clinical outcomes might then approach each individual's maximal potential for continence.

3. ANATOMY AND PHYSIOLOGY

The muscles responsible for continence are the striated muscle of the pelvic diaphragm and the associated striated and smooth muscle groups constituting the anal sphincters (19). Although these muscles act as a single entity, they have slightly different individual functions. For instance, the upper sphincter group is made up of portions of the levator ani, the iliococcygeus, and pubococcygeus muscles. The levator group supports and envelopes the rectum distally and can be readily seen by MRI (20). The pubococcygeus muscle has a thicker and more vascular anteromedial component, termed the puborectalis muscle,

which has a critical role in defecation and continence. This muscle has both a sensory (detection of rectal distention) and a motor function (constriction around the bowel). It also has both slow twitch (red muscle) fibers that maintain a tonic contractile force on the anorectal junction and a fast twitch (white muscle) component that can be used to accentuate this anorectal angle. The puborectalis muscle forms a funnel-like sling around the distal rectum, producing an antero-cephalad elevation. This elevation results in an anorectal angle that opposes propulsive forces from above (21,22). Since the puborectalis is a voluntary muscle, it is an important mechanism by which conscious control over defecation is exercised (23). Caudally the puborectalis and pubococcygeus essentially form a continuum with the deep and more superficial portions of the EAS—together termed the striated muscle complex (24). The continuum of striated muscle complex compresses and elevates the anal canal under conscious control to efficiently collapse the lumen and control fecal flow.

The anatomic orientation of the EAS fibers has been controversial. The EAS has been described as a strictly parasagittally oriented muscle with no circular component in either normal individuals or patients with ARMs (18,24). Yet another view holds that the EAS is organized as a U-shaped loop around the anal canal (25). MRI, endorectal ultrasound, and actual histological sections from cadaveric specimens, however, clearly show that the EAS surrounds the anal canal. It consists of circular skeletal muscle fascicles that pass circumferentially around the bowel and are continuous anteriorly and posteriorly, just as is seen with the adjacent internal sphincter (26–28). At attachment points with the anococcygeal ligament posteriorly and with the bulbospongiosum muscle (male) or perineal body (female) anteriorly, there are many fascicles that cross the midline (28). There is a subcutaneous division of the EAS as well as deeper components, which is continuous with the more vertically oriented muscle complex. Differences are likely to exist between the normal EAS and the EAS found in patients with ARM. Regardless of the anatomic organization however, the action of the external anal sphincter is to provide temporary voluntary constriction of the anal canal in order to resist passage of material through the canal.

The internal anal sphincter is a thickening of the inner circular smooth muscle of the bowel wall, which encircles the upper two-thirds of the anal canal. This sphincter is tonically contracted and provides an involuntary high-pressure zone around the anal canal that prevents leakage of gas and mucus. The internal anal sphincter is thought to be important for prevention of soiling, and thus provides an important mechanism for continence. In patients with high ARM, a normal internal anal sphincter cannot be demonstrated in the distal rectal pouch. However, in the distal fistula of high ARM, muscle fibers with internal sphincter-like properties have been found (29). This has led some surgeons to suggest that the fistula be used for anal reconstruction (30). However, the beneficial contribution of the fistula for long-term continence has not been universally accepted (18,31).

Normal continence is the result of a poorly understood, complex interplay between anatomic structures and physiologic forces in the pelvis. The major factor controlling fecal continence is the influence of pelvic muscles on the anatomic configuration of the distal rectum and anal canal. After the rectum has been filled by the sigmoid, contraction of the rectum results in evacuation of the rectal lumen. The recto-anal inhibitory reflex refers to relaxation of the internal sphincter in response to rectal pressure. This relaxation helps to open the anal canal to allow passage of fecal material. Momentary simultaneous contraction of the external sphincter allows time for a conscious decision to be made regarding defecation. As cited, in high ARM, development of a normal internal sphincter does not occur. The amount of internal sphincter present after reconstruction (measured by recto-anal inhibitory reflex) is variable, but is frequently not found (31,32). Patients with

ARM thus have a greater reliance on the striated muscle for maintenance of continence and preservation of the integrity of these muscles during reconstruction is an important principle. Specifically, the recto-puborectalis reflex (contraction of the puborectalis after contraction of the rectum) becomes a very important voluntary mechanism to guard against involuntary opening of the rectal neck if defecation is not socially appropriate. Patients with high ARM may have an abnormal sacra and may lack the muscle mass, innervation, and therefore the strength and sensation of a normal puborectalis and EAS. These factors certainly contribute to the difficulties that these patients experience with fecal continence.

In summary, defecation and continence require proper sensory and motor innervation, coordinated voluntary and involuntary muscle contraction and relaxation, and appropriate bowel motility. In patients with anorectal malformations, the muscles, sensory elements, and nerves responsible for continence are also severely affected. It is these factors, rather than the technique of surgical reconstruction, that are ultimately the most important predictors of clinical outcome in these patients.

4. MANAGEMENT AND CLASSIFICATION

Upon diagnosis of an ARM, a complete maternal and prenatal history is obtained. The physical examination and a short period of observation should provide the physician with sufficient information as to the type of defect present and the best initial management. Additionally, a number of diagnostic procedures are performed to determine the presence of associated anomalies. Echocardiography is performed to exclude cardiac malformations. A tube is passed down the esophagus to exclude esophageal atresia. A renal ultrasound is obtained to determine whether abnormalities of the urinary system exist, since these are seen in $\sim$50% of patients with ARM. Plain radiographs of the spine are performed to exclude coexisting vertebral abnormalities, since the condition of the lumbosacral spine has a significant bearing on future fecal as well as urinary continence. Finally, spinal ultrasonography or MRI is routinely performed to exclude tethered cord and occult spinal dysraphism.

Anatomic classification of malformations has traditionally been based on clinical presentation and radiographic studies (33). Emphasis has been placed on whether the termination of the bowel was above or below the levator muscles, since this has an important bearing on operative planning. Agenesis of the rectum resulting in a rectal pouch that terminates above the levator muscles is considered a high ARM. Defects resulting in a rectal pouch that passes through the levators are considered low ARMs.

Currently, a low lesion (anal agenesis with rectoperineal fistula) for either sex is typically diagnosed by physical examination alone, but this interpretation may be verified with a cross-table lateral view of the prone infant, or by using perineal ultrasound. These ancillary studies support the diagnosis of a low ARM if the rectal gas pattern or rectal wall is found within a centimeter of the perineum. Low ARMs are treated by limited posterior sagittal anoplasty in the newborn period (34). In cases where a perineal fistula is not readily appreciated, a urinalysis may show evidence of meconium and should be obtained. If a newborn with imperforate anus is observed for 24 h and is judged to not to have a low ARM or if a cloaca, vestibular fistula, or urinary fistula is clearly diagnosed during the period of observation the infant is treated as if a high malformation exists. A colostomy is conventionally performed prior to definitive management. For some patients, exact classification is not made until the distal colostogram is performed. Males are commonly found to have a rectourethral fistula, a rectovesical fistula, a high ARM without a patent

fistula, or rectal atresia. Females are commonly found to have a rectovestibular fistula, a cloacal malformation, high ARM without fistula, or rectal atresia.

5. RATIONALE FOR LAARP

Increasing experience in laparoscopic dissection of the rectosigmoid colon in neonates with Hirshprung's disease led Georgeson et al. (35) to realize that the laparoscope could provide significantly improved exposure (compared to that of the laparotomy) for dissection of the rectum and associated genitourinary fistula from above in patients with ARM. This technique avoids the need for an extensive dissection from the perineum (6). Magnified laparoscopic visualization with high intensity illumination focused directly on the surgical field of view offers an anatomic exposure that is often superior to that achieved with open surgery.

The sheathed Veress needle trocars that are now available are capable of sequential radial expansion to a 5, 10, or even 12 mm diameter. The use of these devices has resulted in a safe, noncutting method to create an anatomic tunnel in the pelvis without the traumatic disruption of muscle associated with other methods of tunneling. These technologic innovations have facilitated the development of LAARP. With LAARP, surgeons have an option of treating high malformations like low ARM, essentially doing a fistula transfer with only a limited perineal dissection.

5.1. Single-Stage Approach

For several years surgeons have recognized the safety and excellent results of single-stage endorectal pull-through for Hirshprung's disease in neonates (35). Using LAARP, some high ARMs can also be reconstructed in a single stage (initial anorectoplasty alone), since no large perineal wound is created. If the single stage LAARP approach is used for high ARM, caregivers must be diligent with dilation protocols to prevent stricture or sepsis of the neoanus. This method obviates additional operation(s) and allows the neonate to experience the sensation of stool passage earlier in life.

Several reasons have been enumerated for not proceeding with neonatal primary reconstruction for high ARM (36). These include: (1) The risk of infection and dehiscence. After LAARP, the risk of infection and dehiscence is reduced, since there is only a small perineal anastomosis, but no extensive skin wound or dissected and reconstructed deeper tissue planes; (2) the need for preoperative distal colostogram. The preoperative distal colostogram has been used extensively to determine whether a strictly posterior or a combined anterior and posterior approach will be necessary. Alternatively, when considering use of LAARP, the major preoperative consideration is ruling out a perineal fistula (that can usually be done by physical examination with or without perineal ultrasound) that might best be approached with a limited perineal procedure. Once a low lesion is ruled out, high fistulae are easily defined using a combination of cystoscopy and laparoscopy. Distal colostogram can provide supplemental anatomic information, but is not considered necessary preoperatively; and (3) injury to adjacent structures. Concerns have also been raised about finding and damaging other unexpected structures if a newborn is subjected to a deep perineal exploration for a high ARM without prior colostomy. However, at the time of LAARP, the surgical dissection starts from above and moves methodically from "known to unknown" with excellent illumination and magnification, and with confidence in identification of all relevant adjacent structures. Cystoscopy can provide additional information and infrared marking of the urethra is another adjunctive technique (see

subsequent paragraphs). For these reasons, the LAARP can be safely performed in newborns with high ARM, as either a single- or two-stage procedure as outlined earlier.

5.2. Two-Stage Approach

LAARP can also be performed in two stages: initial anorectoplasty with simultaneous colostomy, followed by reversal of colostomy at 4–6 weeks of age. This approach may become the preferred method because it allows the neonate to experience normal defecation within the first 4–6 weeks of life and allows for initial healing of the anorectal anastomosis prior to active stooling through the neoanus.

5.3. Three-Stage Approach

Until experience with LAARP is acquired, the three-stage approach is recommended. This approach consists of initial colostomy then anorectoplasty at 4–6 weeks of age, followed by reversal of colostomy 4–6 weeks later. This approach is particularly beneficial to surgeons who are gaining experience with LAARP. A colostomy is initially performed, and a distal colostogram and cystourethrogram are subsequently carried out to provide anatomic details. The LAARP is performed as an elective procedure (as early as 1 month of age) and the neoanus can heal with little risk of septic complications at the anastomosis. After the caregivers have become comfortable with dilations of the neoanus, the colostomy is reversed (as early as 2 months of age).

6. TECHNIQUE OF LAARP

6.1. Preparation

If the patient has had a prior colostomy, a mechanical and antibiotic bowel preparation is carried out, and the terminal rectum is irrigated through the mucus fistula with a saline solution + 100 mg% neomycin and erythromycin or betadine/saline. The patient is positioned supine, transversely across the end of the table. The torso, pelvis, and lower extremities lie flat on several folded sheets to elevate the body. This allows the neck to extend and protects the head and endotracheal tube while covered during laparoscopy. This positioning also allows the surgeon and assistant maximal access to the patient for both laparoscopic and perineal parts of the procedure. The antiseptic skin preparation is performed from nipples to toes. If LAARP is performed as a primary procedure without prior colostomy, a cystoscopy should be done immediately prior to LAARP to detect coexisting major urologic abnormalities, and to define the level of urinary fistula. The bladder is decompressed by transurethral catheterization; this prevents postoperative urinary retention and also aids in identification of the urethra during laparoscopic dissection. It may also be beneficial to illuminate the urethra within the lumen by using a urethral catheter capable of emitting an infrared signal (Stryker Endoscopy, Cupertino, CA). A detection filter in the camera must also be used, and visualization of the urethral position is enhanced.

6.2. Trocar Placement

There are many possible variations on trocar placement, but the following strategy has been useful. The surgeon stands at the patient's head and assistant stands at the end of the table to the side of the patient. Pneumoperitoneum is created using a Veress needle inserted into the right upper quadrant (Fig. 20.1). The site is infiltrated with lidocaine

and a 5 mm incision is made. The fascia is grasped between two hemostats and elevated to allow the Veress needle to be inserted safely. After 8–10 mmHg of pneumoperitoneum is established, this site is used for placement of a 5 mm radially expandable trocar (Step SL75, Innerdyne Corporation, Salt Lake City, UT). Laparoscopy is performed through the right upper quadrant site using a 4 or 5 mm, 30° angled telescope. Smaller 3.5 mm trocars can be placed in the left upper quadrant and left lower quadrant. Another 5 mm trocar is placed in the right lower quadrant. This site is later used for the 5 mm bipolar electrocoagulating shears, the endoloop, or the clip applier.

6.3. Dissection

Laparoscopic rectal dissection begins distally at the peritoneal reflection. This dissection is facilitated by cephalad traction applied to the bowel wall by the assistant's grasper in the left lower quadrant. The most distal mesorectum is opened and divided using hook cautery. Bipolar shears are used throughout the remainder of the dissection to prevent thermal injury to the rectal wall, since viability of the pulled-through bowel will be dependent upon the intramural blood supply. As the rectum tapers into the fistula distally, a meticulous and deliberate dissection toward the termination of the fistula must be performed. When the fistula has been sufficiently cleared of surrounding tissue, it is ligated and divided (Fig. 20.2). This may be accomplished by placing an endoloop around a locking grasper and grasping the fistula near its termination. The fistula is next sharply divided proximal to the grasper. The endoloop can then be slipped off of the grasper and secured around the distal fistula stump. Alternatively, the fistula can be ligated with a 5 mm titanium clip at the termination and sharply divided.

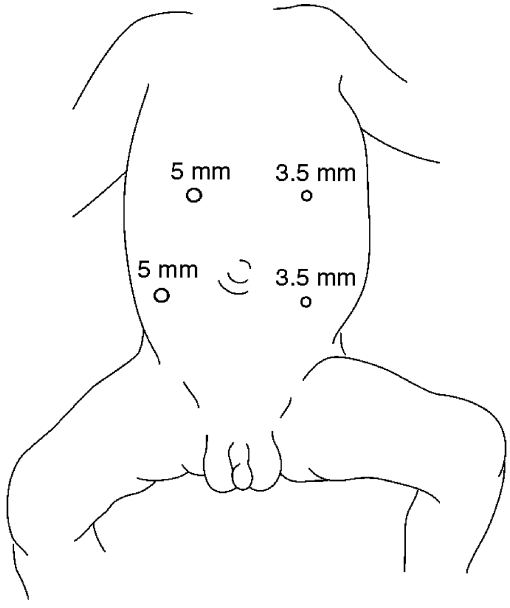


Figure 20.1 Laparoscopic port placement. A total of four laparoscopic ports are placed for LAARP. The camera is usually placed in the RUQ 5 mm site, while the RLQ site is used for the bipolar shears. The surgeon's left hand uses a grasper in the LUQ, and the assistant's grasper is placed into the LLQ.

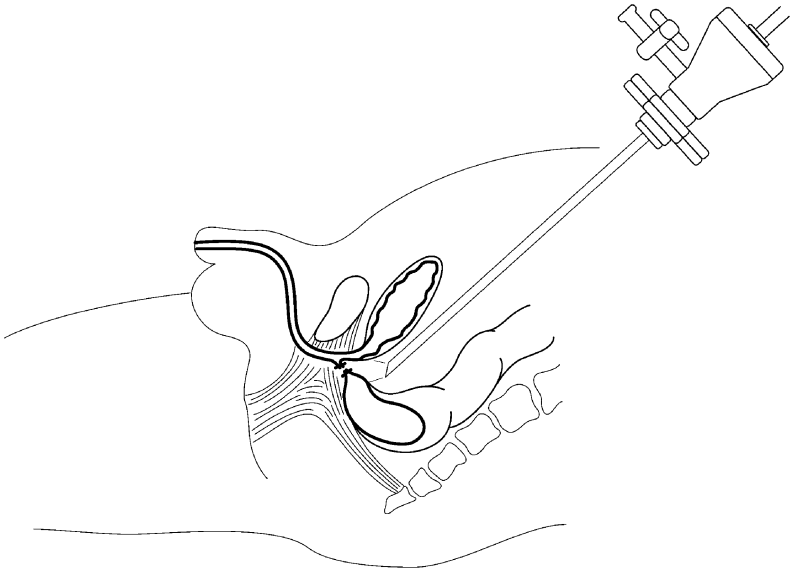
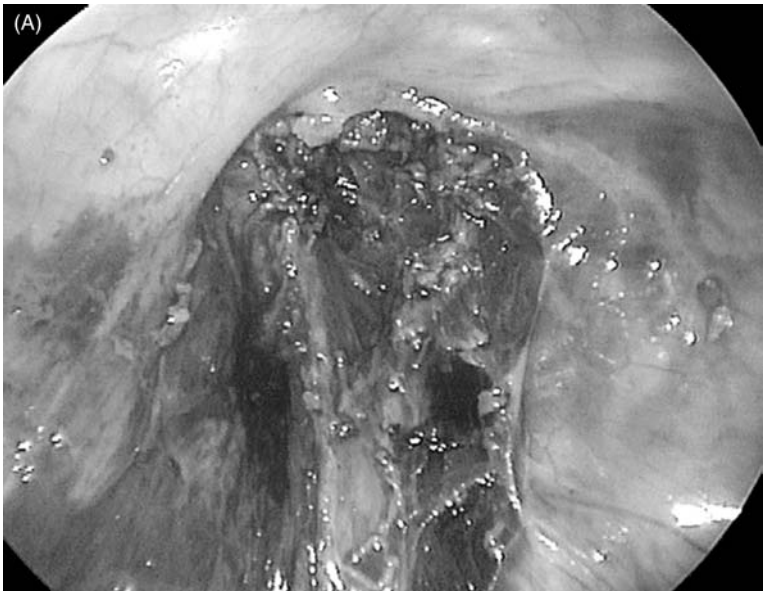


Figure 20.2 Divided rectourethral fistula. The angled laparoscope is used to maximal advantage by peering around the bladder neck at the rectourethral fistula in this male patient with a high ARM.

The rectum is brought out of the pelvis and into the abdomen to facilitate close inspection of the pelvic floor musculature (Fig. 20.3). To accomplish this exposure, the 30° angled telescope is advanced just beyond the sacral prominence, with the end of the lens directed upward so that the surgeon is looking "around the corner" of the bladder neck. This is a surgical exposure that would be difficult if not impossible without laparoscopy. In some patients, visualization of the entire pelvic diaphragm and puborectalis muscle is possible. In other patients, clear visualization may be difficult. This is due to either a variable amount of endopelvic fascia left *in situ* on the pelvic floor after the rectal wall dissection or to the fact that the levator muscle mass is poor in some patients. The distal end of the divided fistula gives the surgeon clear identification of the midline [Fig. 20.3(C)].

6.4. Pull-Through

Next the legs are elevated, the hips flexed, and the feet held together over the face to exposure of the perineum (Fig. 20.4). The hip-flexed position in normal individuals is analogous to the "squatting" position that straightens the anorectal angle to facilitate defecation. Thus, in a patient undergoing LAARP, this position should also straighten the vertical muscle complex to improve alignment of the perineal anal site, the vertical muscle complex, and the puborectalis sling. The MRI finding of an appropriate anorectal angle in the supine sagittal plane after LAARP suggests that this occurs (personal observation). The muscle stimulator (Peña Muscle Stimulator, Radionics Corporation, Burlington, MA) is used to determine the center of the sphincter complex—the optimal site for the neoanus. The area of maximal stimulated contraction and upward pull denotes the region where the highest density of striated sphincteric muscle is located. A hemostat is used to make a 1 cm vertical midline incision in the perineum. The intrasphincteric plane is bluntly dissected from below for a short distance without dividing the sphincter. This dissection seems to



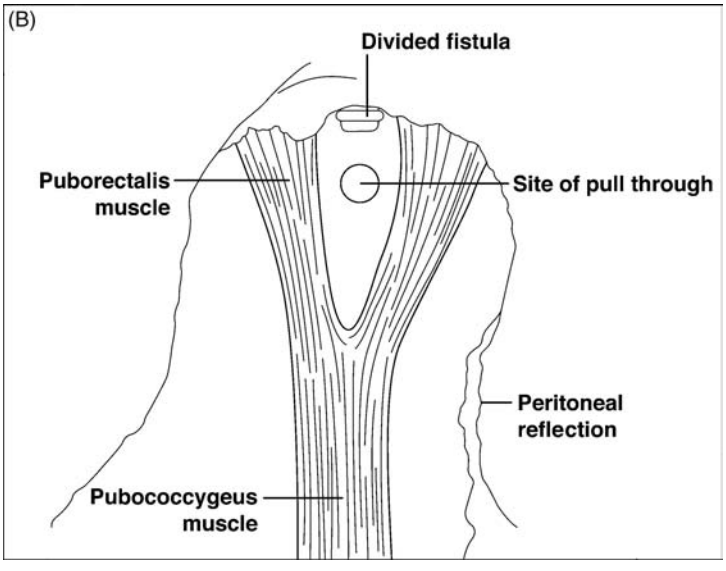


Figure 20.3 Composite of laparoscopic view in the pelvis after division of the rectourethral fistula. *Panel A*: Laparoscopic image of the puborectalis sling after division of the fistula and retraction of the rectal pouch out of the pelvis. *Panel B*: Line drawing with anatomic labels of the laparoscopic image in panel A. *Panel C*: Laparoscopic image of another patient (with a rectovesical fistula). The anterior extensions of the puborectalis are labeled "A" and "C," while the posterior communication in the midline is labeled "B." The grasper was used to "palpate" the muscle bundles, and identify the levator hiatus (labeled "*") where the pull-through rectum will be placed. The PDS suture occluding the bladder neck end of the fistula is also seen between the two bellies of the puborectalis.

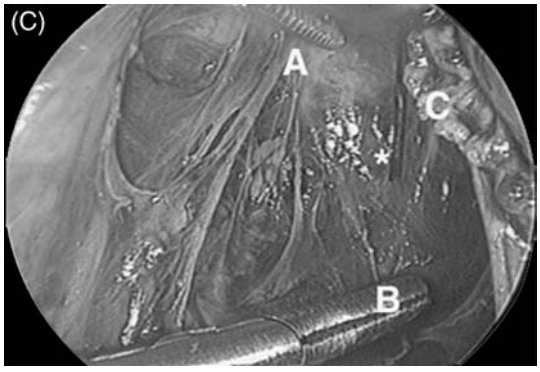


Figure 20.3 *Continued.*

be facilitated by the tension created in the pelvic diaphragm by the pneumoperitoneum, and to some degree by the backlighting provided by the laparoscope.

Using laparoscopic guidance, the low profile Veress needle with radially expanding sheath (Step, Innerdyne) is then placed into the perineal wound, and passed through the midline intrasphincteric plane, advancing between the two bellies of the pubococcygeus muscle. The angle of insertion should be approximately 45° from the horizon (coronal plane) in the hip-flexed position. The needle must enter the pelvis in the midline through the levator sling, just posterior to the urethra. Due to the conical nature of the pelvic diaphragm, the sheathed needle is easily deflected from the midline to the side while advancing through the muscle complex. This deflection is readily apparent from the laparoscopic vantage point. When the deflection occurs, the needle is removed and reintroduced until it is placed accurately between the "V" of the puborectalis sling (Fig. 20.3). Proper placement of the needle and the creation of the pull-through tunnel are possible only if the

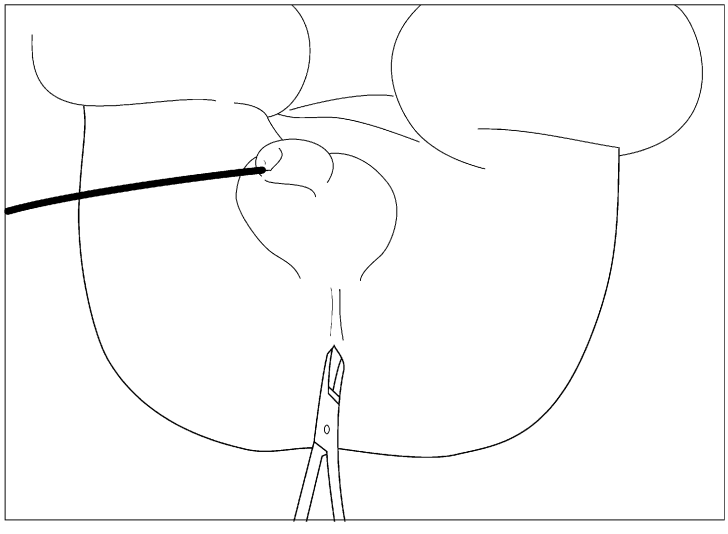


Figure 20.4 Perineal dissection. The patient is in an exaggerated lithotomy position with a towel roll beneath the sacrum to provide optimal exposure to the anal site. A 1 cm incision has been made, based contraction seen with electrostimulation. A hemostat is used for blunt dissection of the EAS fibers in the midline, followed by passage of the sheathed Veress needle trocar.

surgeon is simultaneously watching the laparoscopic image. This is the key difference between this technique and the blind techniques used in the past, where a surgeon's intuition rather than objective visual guidance was used in tunneling through the pelvis.

Technologic advancements in laparoscopic surgery enable the pediatric surgeon to look around the pubic arch and to see a magnified view of the pelvic floor of the infant. When the laparoscope is controlled robotically, the image is steady and can be adjusted simply by voice command of the surgeon working at the perineum.

6.5. Anoplasty

After accurate placement of the sheathed needle through the anal site and puborectalis sling, the needle is removed from the sheath. The tract is dilated gently in a stepwise fashion, first to 5 mm and then to 10 mm diameter without cutting any of the delicate fibers of the muscles of continence. The rectum is grasped with a Babcock clamp and gently guided through the tract downward to the perineum, retracting it out with the trocar as a unit (Fig. 20.5). The rectal fistula is next incised to expand the opening in the rectum to $\sim$1 cm in diameter. The anastomosis between rectum and perineal skin is next completed with circumferential, interrupted 4-0 polyglycolic acid sutures. The rectum is retracted cephalad laparoscopically and secured to the presacral fascia at a convenient location using 2-0 silk sutures, in order to prevent prolapse and to lengthen the skin-lined anal canal (Fig. 20.6). The neoanus is sized with Hegar dilators.

Dilations begin 2–3 weeks postoperatively, with the Hegar dilator size noted intraoperatively. The colostomy is closed once satisfactory healing has occurred and an adequate orifice is present.

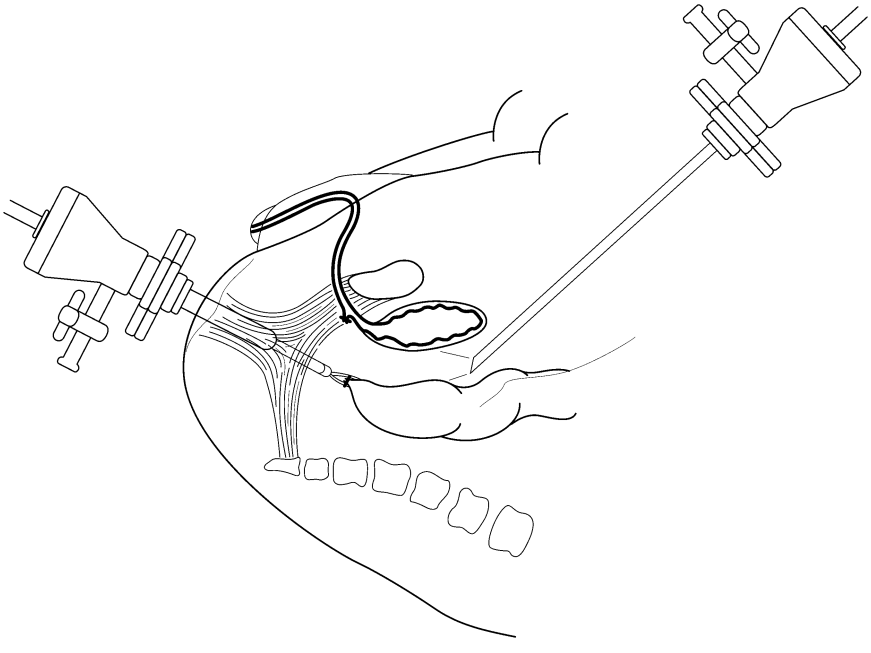


Figure 20.5 Perineal pull-through. The transsphincteric, translevator tunnel has been created with the radially expandable Step trocar. The rectal fistula has been grasped and is being transferred down to the perineum for anorectoplasty.

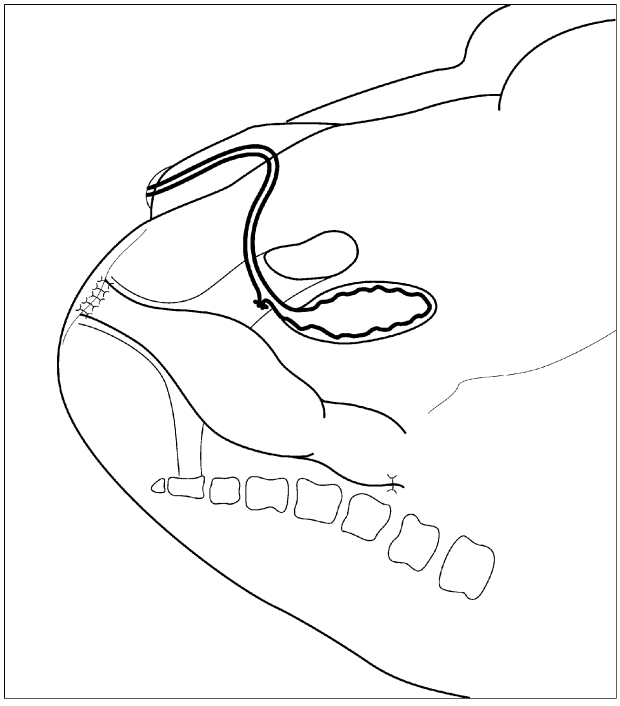


Figure 20.6 Postoperative appearance. The sutured anorectoplasty has been performed and the pelvic fixation suture is shown between the bowel and the presacral fascia.

7. POTENTIAL COMPLICATIONS

As with any operative procedure, there are many real and theoretic pitfalls to be avoided. LAARP is not necessary for patients with rectoperineal fistula. It has not been used with cloacal malformations, and has no particular advantage for rectovestibular fistulae. LAARP is most useful in males with rectourethral and rectovesical fistulae, and in females with high rectovaginal fistulae. The more important preoperative consideration is ensuring that no perineal fistula exists; in such cases, only a limited PSARP is needed.

Although evidence is mounting for the safety of laparoscopic surgery in infants <5 kg (35,37–39), the effects of pneumoperitoneum on cardiopulmonary performance must be considered carefully prior to any neonatal laparoscopic procedure. Insufflation pressures should be maintained at 6–10 mmHg to prevent the predictable pressure-related depression of venous return and cardiac output, which have been documented in animals as well as in children (40). These lower insufflation pressures also minimize the hypercapnea and increase airway resistance which can be seen with pediatric laparoscopy (41). Associated conditions must also be taken into consideration. Patients considered least suitable for laparoscopic procedures are those with complex congenital heart disease resulting particularly in right heart pump failure, patients with marked pulmonary dysfunction, premature infants, and those with marked abdominal distension.

After repair of high ARM, short-term urinary retention should be expected, and the urethral catheter should remain in place for 2–3 days. It is far safer to leave the catheter in place for several days than to remove it prematurely and risk the possible need to re-catheterize after repair of a rectourethral fistula has been performed.

The placement of trocars in neonates must be done using the technique most familiar to the surgeon, since penetrating injuries to abdominal and retroperitoneal viscera can result if appropriate care is not taken during placement (42). Dissection of the colon should begin at the peritoneal reflection, since ischemia of the rectum can result if dissection is begun more proximally. Before pull-through, the length of dissected rectum can be tested by moving it into the deep pelvis and assessing the amount of tension on the bowel. More sigmoid mesocolon can then be divided using hook electrocautery if more length is needed.

During division of the rectal fistula, it is important to stay on the plane of the longitudinal layer of the bowel wall since important pelvic autonomic nerve supply to the bladder and penis pass in close proximity and can be injured when dissecting the rectourethral fistula. This dissection is considerably easier to perform for the higher rectobladder neck and recto-vaginal fistulae than is the more meticulous dissection needed to avoid urethral injury at the termination of rectourethral fistulae. For rectourethral fistulae, it is also important that the fistula is completely and securely ligated to prevent postoperative pelvic sepsis and recurrence of rectourethral fistula. Occasionally, it is necessary to up-size the right lower quadrant trocar to 10 mm to apply a larger clip across a broad-based fistula. Also, a rectourethral fistula may be encountered more distally than expected. If it cannot be safely ligated and divided laparoscopically, it may be necessary to extend the perineal incision *anteriorly* and proceed with a midline dissection to find and ligate the fistula (modified Mollard approach).

8. DISCUSSION/OUTCOME

Developmental neurobiologists have increasingly asserted that normal infant development is dependent on sensory input that provides necessary neural activity to shape the developing cerebral cortex (43). In the 1960s, Wiesel found that neonatal visual deprivation led to failure of development of the visual cortex such that even if vision was later restored, permanent cortical blindness resulted (44,45). More recently, others have found that cortical growth and blood vessel density in the mammalian brain is significantly greater in regions of chronically increased neural activity (46). One rationale for repairing high ARM during the neonatal period has been to give the infant the earliest sensorineural experience possible (47,48). If a critical early phase of cortical synaptic remodeling and growth occurs with continence pathways as has been documented in the visual system, then the infant with high ARM should benefit from procedures that offer the earliest experience with proper anorectal function. LAARP is an attractive alternative procedure that can be performed early in infancy and thus allows for earlier anorectal functioning.

LAARP combines principles outlined by many of the pioneers in surgical care of ARM. Before the 1970s the major problem with repair techniques for high ARM was that the puborectalis was not well seen during reconstruction (16). After this time, repair techniques that were capable of exposure of the puborectalis required either an extensive anterior perineal dissection (49) or an extensive posterior perineal dissection combined with division of the puborectalis (11). Georgeson's most important contribution in developing the LAARP is the unparalleled endopelvic visualization and ability to accurately target the center of the puborectalis sling without an extensive perineal dissection or

division of sphincter components critical for proper anorectal function. Accurate placement of the rectum has been documented after LAARP by MRI, which clearly demonstrates the pull-through rectum in the midline between the two bellies of the puborectalis sling and EAS, with an appropriate anorectal angle (personal observation).

Patients who have undergone LAARP should be followed on a regular basis. The first patient who underwent LAARP at the Children's Hospital of Alabama was born at 30 weeks' gestation with complex VACTERL malformations (VSD, horseshoe kidney, tethered spinal cord, severe caudal regression with absent right leg, partial sacral agenesis, lumbar vertebral anomalies, scoliosis, hip dysplasia, vaginal duplication, grade 3 vesicourethral reflux). The LAARP was performed in 1995 and she was (not unexpectedly) still totally incontinent after age five. She underwent a continent appendicostomy and with the use of nightly antegrade colonic enemas, her outcome has been excellent.

It is difficult to make any judgments about fecal continence in patients with high anorectal malformations before 3 or 4 years of age, since it is common for this group of patients to experience delayed toilet training. Only three other patients who have undergone LAARP (one rectobulbar, age 43 months; one rectoprostatic, age 46 months; and one rectovesical fistula, age 43 months) have reached the age of 3 years. All of these boys have abnormal sacra and none has yet demonstrated fecal continence, although toilet training has been initiated. Caregivers have however reported that these patients show facial expression and/or activity interruption when having a bowel movement, suggesting some degree of sensation of the event. While this has been considered a good prognostic sign for later continence, it is still too early to make any conclusive statements about functional outcomes of LAARP. Since March 1999, 12 patients (10 male, 2 female) have undergone LAARP at the University of California, San Francisco (54). All but two male patients had fistulae to either the bladder neck or the prostatic urethra. Nine procedures were two-staged repairs, two were a single-stage procedure, and one was a three-staged repair. There were no intraoperative complications and no postoperative strictures after an outpatient regimen of dilations of the neoanus. Not described in their report but known by personal communication is a patient who had developed a urethral diverticulum 4 years after the division of the rectoprostatic fistula. He presented with recurrent urinary tract infections and a large diverticulum resembling an enlarged utricle was diagnosed by cystourethrogram. Presumptively it resulted from division of the fistula remotely from the urethra. It was successfully repaired with an open, transcystic procedure. The longest follow-up in this series is 3 years. Two children are taking oral medications for constipation. Continence has yet to be assessed. No child developed significant rectal mucosal prolapse in the follow-up period.

The developmental mutations that lead to ARM during early embryogenesis are only now being elucidated (50–52). While the potential for preventative antenatal genetic interventions are not inconceivable, medical progress for these birth defects currently depends on refinements in postnatal management. For patients with high ARM, the overall goal is to treat the anomaly so that the best anorectal function and socially appropriate continence can be achieved. In addition to the obvious imperforate anus and rectal fistula, these patients suffer from a spectrum of other problems related to the anomaly. These problems often include neurogenic bladder, inadequate levator and sphincter musculature, abnormal pelvic sensory and motor nerve supply, and intrinsic dysmotility of the hindgut. Unfortunately, currently there is no specific remedy for these structural and functional abnormalities. The clinical problems that result from these abnormalities can be effectively treated with a bowel management program (53), and socially acceptable control of incontinence can often be achieved. The technical goal of surgery for the infant born with a high ARM is therefore to correct the gross anatomic anomalies in

such a way that each patient's maximal anorectal functional potential is preserved. Innovations in minimally invasive pediatric surgery have provided the tools and techniques to accomplish this surgical goal while minimizing collateral surgical injury to surrounding structures. A prospective, controlled clinical trial with long-term postoperative evaluation of anorectal function will be needed to determine the optimal technique for reconstruction of the patient with high ARM.

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21
Laparoscopy for Ovarian Pathology

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1. INTRODUCTION

The evaluation and treatment of the child with an adnexal mass has changed considerably in recent years. The routine and liberal use of prenatal screening ultrasound and advances in technology have resulted in the detection of a number of masses, particularly cysts, that previously might have gone undetected (1–5). Increased sophistication of postnatal imaging modalities, including the use of doppler ultrasound, computed tomography, and magnetic resonance imaging have allowed more precise and specific characterization of adnexal masses prior to surgery (4,5). As laparoscopic techniques and equipment have become increasingly suited to the pediatric patient, increased treatment options have brought with them a new set of controversies. Principles derived from a better understanding of adnexal masses in children can make it possible to make good use of still emerging laparoscopic approaches.

In this chapter, we discuss the characteristics of adnexal masses in children, suggest a diagnostic approach, and then propose a therapeutic algorithm, paying particular attention to the expanding role of laparoscopy.

2. THE NATURAL HISTORY OF OVARIAN MASSES

An increase in the utilization and accuracy of diagnostic modalities is changing our understanding of and approach to adnexal masses. Prenatal ultrasounds frequently detect masses that would have heretofore gone undetected (2,6). Increasing utilization of ultrasound and computed tomography (CT) to evaluate abdominal pain and suspected urologic abnormalities is leading to an increase in the discovery of asymptomatic masses of all types in older girls. It is this change in technology and utilization that, while increasing our understanding of these lesions, is also contributing to a wide variation in the reported incidence and nature of these lesions in the literature (1–5).

Prior to the almost routine use of ultrasound, most ovarian masses were thought to be solid, and a substantial number of them were believed to be malignant in children (7–10). Furthermore, adnexal pathology in the earliest years of life was thought to be infrequent. In a recent review, however, non-neoplastic ovarian cysts were predominant (11) (Table 21.1). We will briefly discuss the natural history of adnexal masses by lesion and by age.

2.1. Cystic Ovarian Lesions

The cause of ovarian cysts varies with patient age. In the perinatal period, most cysts are non-neoplastic simple follicular cysts and are a result of stimulation of Graffian follicle development by maternal (estrogen), fetal (FSH), or placental (hCG) hormones (1,12). Although most of these simple cysts resolve in the neonatal period (3,7,13), these cysts can cause ovarian torsion with loss of the gonad, adhesion formation, and other symptoms (1,5,11,14,15). Complex ovarian cysts in the neonatal period are less common, but can occur (11). In a review by Croitoru, 8 of 9 complex neonatal cysts were associated with

Table 21.1 Pathologic Findings from 106 Operations on 102 Patients

Pathology	No. of patients	Percent of total
Non-neoplastic	49	46.2
Ovarian torsion (without associated tumor)	16	15.1
Corpus luteal cyst	15	14.2
Simple or follicular cyst	11	10.4
Paraovarian cyst	2	1.9
Hemo/hydrosalpinx	2	1.9
Biopsy of normal ovary	2	1.9
Ovotestis	1	0.9
Neoplastic: benign	47	44.3
Mature teratoma	42	39.6
Cystadenoma	3	2.8
Cystic lymphangioma	1	0.9
Immature teratoma	1	0.9
Neoplastic: potentially malignant	10	9.4
Dysgerminoma	4	3.8
Immature teratoma with yolk sac tumor	2	1.9
Yolk sac tumor	1	0.9
Juvenile granulosa cell tumor	2	1.9
Sertoli–Leydig cell tumor	1	0.9

Source: Cass et al. (11), with permission from Mosby.

ovarian torsion and 1 of 9 was from a juvenile granulosa cell tumor (1). Simple cysts are unlikely to be malignant (16).

In the premenarchal period beyond the first few months of life, large ovarian cysts are much less common and likely reflect follicular responsiveness within the prepubertal ovary to a pulsatile release of gonadotropins by the developing pituitary (7,17). Cysts in this premenarchal age group are known to present with precocious pseudopuberty, presumably from estrogen secretion by the cyst lining (18,19). These symptomatic cysts may resolve spontaneously. When cysts in this age group do occur, they are more likely to be malignant, particularly if complex or hemorrhagic (7,20). As with cysts in other age groups, torsion is also a concern, particularly with larger cysts (18).

In girls entering menarche, corpus luteum cysts become more predominant. These cysts may appear to be hemorrhagic and complex, but may resolve spontaneously if <7 cm (13). Cysts in this age group are less likely to be malignant than in the premenarchal age group (7). Ectopic pregnancy and pelvic inflammatory disease should also be considered in the differential diagnosis of ovarian cysts in adolescent girls (7). Less commonly, para-ovarian lesions such as mesenteric or intestinal duplication cysts can mimic ovarian cystic lesions.

2.2. Solid Ovarian Lesions

Most solid ovarian masses are benign, with mature teratoma being the most common of this group (11,21,22) (Table 21.1). Roughly 10% of solid masses, or 5% of all ovarian lesions, are potentially malignant (11). Characteristics of potentially malignant tumors are given in the table (includes tumor markers, prognosis, treatment). Unfortunately, the differentiation between benign and malignant ovarian masses is not easy. Solid masses are more common in the premenarchal and perimenarchal age groups, and can present with torsion (11,12).

Of the benign solid ovarian masses, most are mature cystic teratomas, comprised of both cystic and solid components, lined with well differentiated ecto- and endo-dermal derived cyst walls such as sebaceous cysts (11,23). Fewer than 20% of pediatric ovarian teratomas in most series are malignant. Cystadenomas, cystic lymphangiomas, and torsed ovaries (in the absence of tumor) make up most of the remainder (11,23).

Malignant ovarian masses comprise $\sim$10% of all ovarian lesions, and $\sim$20% of all solid ovarian masses (11, Table 21.1). Various classification schemes have been proposed, but most malignant ovarian tumors are believed to be derived from one of three types: germ cell, sex cord/stromal, or epithelial. Two-thirds of malignant ovarian tumors are germ cell tumors, which in turn have five subtypes (24).

Dysgerminomas are the most common germ cell tumor. They are bilateral in 10–15% of cases, and usually do not produce tumor markers (4). Treatment with resection and chemotherapy usually results in cure. While radiation may result in a complete response, the patient will be rendered infertile.

Endodermal sinus tumor, also known as yolk sac tumor, is less common but is extremely aggressive. αFP correlates with tumor burden, but may not be produced with recurrent tumor (4). Survival even with surgery and multi-agent chemotherapy is 15%.

Embryonal carcinoma is less well differentiated than the other germ cell tumor subtypes, and often exists as part of a mixed germ cell tumor. β-HCG is usually elevated, and αFP may be elevated if the tumor contains elements of endodermal sinus tumor. Precocious puberty is a frequent manifestation of this tumor. Prognosis of this tumor, while poor, is better than that of endodermal sinus tumor, and treatment consists of surgery and multi-agent chemotherapy (4,11,19).

Choriocarcinoma is uncommon in children, and may present with precocious puberty. β-HCG levels are elevated with this tumor. After treatment with surgery, additional adjuvant platinum-based chemotherapy is required (4,11,19).

Teratoma is the most common germ cell tumor overall, but only the second most common malignant germ cell tumor (4,11,25). Most are benign and are cured by extirpation. αFP may be elevated, but β-HCG is usually normal. Survival is related to histologic grade, ranging from 30% to 90%. For low-grade, malignant teratomas, surgery alone may be curative, but for higher-grade tumors, multiagent chemotherapy is required.

Malignant germ cell tumors may also present with characteristics of more than one germ cell subtype. In general, tumor markers, treatment, and survival are dictated by the most aggressive element. The overall prognosis is poor.

While germ cell tumors are the most common malignant tumor type, sex cord/stromal tumors are less so, accounting for 5–15% of pediatric ovarian malignancies. These are in turn subdivided into granulosa–theca tumors and Sertoli–Leydig tumors (arrhenoblastomas). Granulosa–theca tumors often present with early pubertal changes because of high estrogen secretion. Prognosis is good with resection alone for early stage tumors and surgery with chemotherapy for advanced disease. Sertoli–Leydig cell tumors, or arrhenoblastomas, are rare, and behave unpredictably. They are characterized by the production of large amounts of male hormones. While surgery is a mainstay of treatment, widespread agreement does not exist on the value or type of chemotherapeutic regimen (4,11,25).

Epithelial tumors, while much more common in older women, are unusual in girls, and extremely rare prior to menarche. This tumor may be bilateral, and in younger girls, widespread at the time of its discovery. CA-125 is usually elevated in these tumors (26,27). Treatment consists of resection when possible and multi-agent chemotherapy. Prognosis is fair in adolescent girls to poor in premenarchal girls, but long-term survivors have been reported (26,27).

3. A DIAGNOSTIC APPROACH TO OVARIAN MASSES

In addition to an understanding of the natural history of ovarian masses, a principles-based laparoscopic approach must include a thorough diagnostic workup of these masses. A detailed history and physical examination, guided by the child's age, will provide important information. A history of pain, gastrointestinal complaints, and (when appropriate) menstrual patterns and sexual activity are particularly relevant. Physical examination should focus on both general and pubertal development as well as characterization of any abdominal masses. Bimanual rectoabdominal examination and, in some adolescents, pelvic examination will provide important information regarding the size, texture, and fixation of an adnexal mass.

Laboratory tests should be ordered during the evaluation of any suspicious ovarian mass. αFT, β-HCG, and LDH should be sent. While CA-125 levels are useful in the evaluation of ovarian pathology in older women, they are unlikely to prove helpful in the pediatric patient.

The abdominal and pelvic ultrasound remains the most important radiologic diagnostic test in the evaluation of adnexal masses. Size, septation, fluid content, calcifications, and location can all be evaluated. Sonographic findings that may be suggestive of malignancy include heterogeneity, a significant solid component, and the presence of excrescences (28–30). Color-flow doppler can identify blood flow and aid in the diagnosis of

torsion (7). Low impedence to flow may also be suggestive of an increased risk of malignancy.

CT is indicated when malignancy is suggested by history, laboratory values, and ultrasound findings. We do not routinely employ CT when evaluating simple cystic lesions where malignancy is not suggested by history, physical findings, or laboratory values. CT is well suited for evaluating regional lymph nodes, assessing metastatic spread, and further characterizing complex solid lesions. Findings suggestive of malignancy include large size, presence of calcifications, and predominantly solid character with occasional areas (19).

Although MRI is not routinely indicated during the evaluation of ovarian cysts, it is highly sensitive and specific for evaluating pelvic pathology in adults (31). It is well suited for differentiating between masses of uterine and ovarian origin. Ovarian torsion with infarction has a characteristic MRI pattern with a T1-enhancing rim (19). MRI may have some role in the evaluation of Muellerian duct remnants and endometriosis. However, more liberal use of MRI in children is limited by the need for sedation or anesthesia to complete the examination in younger patients.

The role of percutaneous or incisional biopsy is limited for obvious reasons of tumour spill, and remains controversial.

4. THE ROLE OF LAPAROSCOPY FOR DIAGNOSIS AND TREATMENT

An understanding of the natural history and diagnostic workup of both cystic and solid ovarian masses in children serves as the basis for a laparoscopic–based approach to the treatment of these lesions. Considerable controversy continues to exist as to the proper role of laparoscopy in the treatment of certain ovarian lesions. With continuing advances in the detection of ovarian lesions, new questions are being raised as to which lesions should be treated at all. Furthermore, as advances in both laparoscopic technology and technique are being made, previously unforeseen treatment options become available, if not always advisable. Thus, two questions have to be addressed: (1) Is surgery indicated, and if so, (2) what is the proper role for laparoscopy in the surgical approach?

4.1. Fetal and Neonatal Cysts

In fetuses and neonates, most ovarian masses are benign follicular cysts which will resolve over time. The principle goal of treatment if undertaken should be to prevent potential torsion, which may occur prior to regression. Some authors recommend observation for all neonatal cysts, with surgical intervention reserved for those with symptoms or those failing to regress (1,3,7). Some authors feel that such cysts should be at least aspirated if over 5 cm in neonates to decrease the risk of torsion (7). Van der Zee et al. (32) advocate laparoscopically guided aspiration for neonatal ovarian cysts followed by mini-laparotomy. Given the very low likelihood of malignancy in this age group, particularly with a simple cystic mass, a laparotomy for the resection of an intact cyst may be excessive. The literature in this area is scant and mainly based on case reports (8,32). Laparoscopic drainage and/or resection of ovarian cysts in neonates are feasible. Although there is no evidence to support that laparoscopic-guided aspiration is superior to ultrasound-guided aspiration, for large simple cysts over 5 cm in diameter, aspiration and complete cyst resection done laparoscopically is a reasonable approach, provided that a maximal amount of ovarian tissue can be spared. For more complex ovarian

cysts in neonates, attempted laparoscopic resection may be reasonable; low likelihood of malignancy makes tumor spill unlikely. If at any time, preservation of ovarian tissue is in doubt, formal laparotomy should be undertaken.

4.2. Premenarchal Ovarian Cysts

After the first few months of life, the risk of malignancy in an ovarian mass becomes greater. Simple unilocular cysts in premenarchal girls can resolve spontaneously, and should be followed for several weeks before recommending surgical resection (7,20). Laparoscopy for simple unilocular cysts failing to resolve may be undertaken, but conversion to laparotomy is indicated if complex features are found at laparoscopy (7). Similarly, laparotomy should be undertaken from the outset for more complex ovarian cysts due to increased risk of malignancy and potential tumor spillage. Again, evidence supporting this suggestion remains anecdotal and mainly based on case reports (1,3,7,20).

4.3. Perimenarchal Cysts

With the onset of menarche, the characteristics and treatment of both simple and complex ovarian cysts change significantly. For asymptomatic unilocular cysts in adolescent girls, observation and follow-up over 6–8 weeks is appropriate (33). The use of oral contraceptives over a couple of cycles may help differentiate functional cysts from pathologic ones and also helps to prevent the development of functional cysts during the observation period (33). For simple cysts failing to resolve, laparoscopic fenestration offers the advantages of avoiding a laparotomy, sparing a maximal ovarian tissue and providing tissue for pathologic examination (34). Ultrasound or laparoscopy-guided cyst aspiration (as opposed to fenestration or removal) has a disadvantage of unacceptably high recurrence rate (35–37). Complex or hemorrhagic ovarian cysts should be observed for 2–4 weeks for resolution. If at any time, the ovarian mass is not simple or highly suspicious based on size, organic functional appearance, laparoscopy is indicated as the initial evaluation (33). Laparoscopy for initial evaluation of persistent complex cysts is appropriate, but we favor conversion to laparotomy if the complex appearance is confirmed and cyst resection is planned.

4.4. Solid Ovarian Lesions

There is relatively little controversy as to whether surgery is indicated for solid ovarian lesions in any pediatric age group. Following a thorough diagnostic workup, resection of any solid ovarian mass is indicated. If the lesion appears to be a benign teratoma by appearance and frozen section histology, an attempt at resection of the mass with preservation of remaining ovarian tissue may be considered (38). Otherwise, resection of the involved ovary and, if necessary, the involved adnexa should be undertaken (1,33).

The role of laparoscopy in the evaluation of solid or complex ovarian lesions is more controversial (12,39). Many authors recommend diagnostic laparoscopy followed by attempted laparoscopic resection for ovarian lesions with serologic, radiologic, and clinical features suggesting benign lesion (33). Most studies in the adult literature based on surveys of practice patterns and case reports have not demonstrated an increased risk to the patient following laparoscopic resection of solid ovarian masses but the literature in children are scant (12,33,40,41). While laparoscopic resection with an endo-bag may in fact be safe, most authors conclude that decreased risk of tumor spillage and dissemination and a better ability to preserve as much viable normal ovarian tissue as possible in young

patients make the open approach outweigh the potential benefits of laparoscopic resection (33,42,43). For highly suspicious lesions, few authors advocate attempted laparoscopic resection in children at present, although diagnostic laparoscopy at the beginning of the procedure may provide better visualization of relatively inaccessible areas. As additional experience is gained with laparoscopy for ovarian pathology in children, the results of follow-up studies may well expand the accepted indications for resection in children as they have in adult patients.

4.5. Ovarian Torsion

The issue of laparoscopic or open oophoropexy as treatment or prophylaxis for ovarian torsion is also controversial in any age group. The normal tube and ovary are extremely mobile and are capable of a rotation of 90° without giving rise to symptoms (44). Of all reported cases of torsion in childhood, 70% of the adnexal torsion contained pathologic lesions in the adnexal tissues (44). However, many authors feel, based on case reports, that the risk of fertility from adhesions related to oophoropexy outweighs the possible decreased risk of ovarian torsion, and do not recommend oophoropexy, either as treatment or prophylaxis (11). Depending on the patient's age group, open or laparoscopic aspiration, resection, or fenestration of a cystic lesion associated with torsed ovary is reasonable. Solid masses associated with ovarian torsion should be resected, preferably by laparotomy (33,43). If the so-called "torsion of normal ovary" is found, the ovary can be detorsed laparoscopically; if the ovary appears viable, it should be left in place (45). Whether ipsilateral or contralateral fixation following ovarian torsion should be done remains controversial without any strong evidence to support either way. (11,44–47). Laparoscopic oophoropexy may have additional use, other than the circumstances of torsion. For example, pexying the ovary(s) away from planned field of radiation in the pelvis for ovarian preservation has been effectively used by the authors as well.

5. SUMMARY

Knowledge of the natural history of ovarian lesions in children as well as the appropriate indications for laparoscopiy continue to expand. Today, most cystic lesions are amenable to laparoscopic treatment following a period of observation. For suspicious complex and solid lesions, the role of laparoscopy in children remains limited but can be expected to expand.

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22
Intestinal Rotation Abnormalities

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1. INTRODUCTION

> "During its development the abdominal portion of the alimentary canal may suffer a large variety of perversions . . . The condition therefore requires especial significance in regard to the surgery of infancy." Norman M. Dott (1)

Embryologic errors that lead to abnormalities of rotation and fixation of the intestine constitute a unique group of surgical anomalies that are managed by the pediatric surgeon. Intestinal malrotation and faulty fixation of the bowel, which may lead to the development of bowel obstruction or midgut volvulus and have life-threatening consequences, must be differentiated from

nonrotation, an abnormality that does not require surgical correction and is not associated with the risk of volvulus. The incidence of intestinal malrotation has been reported with a frequency of 1 in 500 to 1 in 6000 (2,3). Intestinal rotation or fixation abnormalities are associated with a high incidence of other associated congenital anomalies (3). Most patients diagnosed with malrotation will present with symptoms in the neonatal period, but problems may not occur until late in childhood or as adults (3). The presence of such an anomaly requires the prompt attention of a pediatric surgeon due to the risk of midgut volvulus.

Early investigation in the field focused on the embryology of the intestine. In 1898, Mall described the process of rotation and fixation of the bowel (4). In 1923, Dott elegantly published the first clinical correlation relating the embryology of intestinal malrotation to surgical anatomy (1). Ladd, in 1936, reported the successful treatment of intestinal rotation abnormalities in the infant (5). In regards to surgical management, he emphasized the lysis of peritoneal bands, the evisceration and derotation of the bowel, and the placement of the bowel in a state of nonrotation (5). These remain the principles of surgical treatment today.

Recent advances in minimal access technology have led to the development of a laparoscopic approach to managing patients with malrotation. The first report of laparoscopic management of malrotation was published in 1995 (6). This has been followed by many case reports and small case series describing the successful management of rotation abnormalities using the laparoscopic approach in neonates and infants (6–13), older children (8,12–16), and adults (13,17). As a collection, the available literature proposes many benefits for laparoscopic surgery over traditional measures in the management of intestinal rotation abnormalities. This chapter will evaluate the existing evidence supporting the application of minimal access techniques in intestinal rotation and fixation abnormalities.

2. EMBRYOLOGY

During its development, the midgut goes through four physiologic stages: (1) herniation, (2) rotation, (3) retraction of the herniated loops, and (4) fixation. Disruption of these critical steps will lead to midgut rotational and fixation abnormalities. The primary intestinal loop consists of a cephalic (duodenojejunal) limb and a caudal (cecocolic) limb and its associated blood supply (18). Between the 4th and 6th weeks of development, the primary intestinal loop enters into a state of rapid growth that leads to physiological herniation into the extraembryonic coelom. Each intestinal limb makes three separate 90° turns in a counterclockwise direction with the superior mesentery artery (SMA) serving as the axis of rotation, while outside of the abdominal cavity, the limbs of the primary intestinal loop undergo the initial 90° rotation. The second 90° rotation occurs as the bowel returns to the abdominal cavity and the final 90° turn occurs in the abdominal cavity. After a total turn of 270° around the axis of the SMA, the duodenojejunal limb is positioned to the left of the SMA and the cecocolic limb to the right of the SMA. The colonic mesentery fuses with the parietal peritoneum and the colon develops peritoneal bands that anchor the ascending and descending portions of the colon to the posterior abdominal wall (18). Failure of the cecum to rotate to the right side will lead to abnormally developed peritoneal bands known as Ladd's bands. These bands that anchor the malpositioned midline colon to the right postero-lateral aspect of the abdominal wall may obstruct the duodenum or proximal small intestine that lies to the right of the cecum (19).

3. CLASSIFICATION

A wide spectrum of anatomic abnormalities may arise during embryogenesis. The intestine may undergo no rotation at all or any combination of incomplete rotation as outlined

in the steps described above. The most common abnormalities include nonrotation, "classic" malrotation, duodenojejunal malrotation, and cecocolic malrotation. Abnormal fixation may also occur, but this tends to occur in the cecum.

Nonrotation is the failure of rotation of both limbs of the primary intestinal loop. In a state of nonrotation the small bowel lies on the right side of the abdomen and the colon on the left. The mesenteric base is usually wide enough to prevent midgut volvulus; however, this can be variable. This is the position of the intestine following a Ladd's procedure.

In "classic" malrotation there is incomplete rotation of both the duodenojejunal and cecocolic limbs. The ligament of Treitz lies to the right of the midline and the cecum is usually found in the upper abdomen close to the duodenum. The mesenteric base is narrowed and the midgut is at risk for volvulus. Duodenal obstruction may occur due to Ladd's bands or kinking of the duodenum on itself.

Duodenojejunal malrotation occurs with the failure of the duodenojejunal limb to rotate in the presence of normal cecal rotation and fixation. Duodenal obstruction may occur due to peritoneal bands that kink the duodenum. The base of the midgut mesentery is usually broad, making volvulus unlikely.

Incomplete rotation of the cecocolic limb may lead to problems with fixation of the colon. Peritoneal bands may obstruct the duodenum. Although rare, the lack of fixation also places the cecum at risk for volvulus. Nonrotation of the cecocolic limb in the presence of normal rotation of the duodenojejunal limb creates a narrow mesenteric base between the ligament of Treitz and the cecum; therefore, the midgut is at increased risk for the development of volvulus.

4. ASSOCIATED ANOMALIES

Rotational abnormalities of the midgut are often associated with other congenital anomalies. Associated anomalies are found in 30–59% of patients with the diagnosis of malrotation (3,19–23). Defects that may be encountered include Meckel's diverticulum, Hirschsprung's disease, imperforate anus, esophageal atresia and tracheo-esophageal fistula, cardiac anomalies, asplenia/polysplenia syndromes with cardiac anomalies, and situs inversus. Malrotation should be suspected in children with these anomalies. Conditions where the primary intestinal loop does not return to the abdominal cavity, such as congenital diaphragmatic hernia and abdominal wall defects, are universally associated with rotational abnormalities. The rotational defect usually does not need surgical correction beyond that of the repair of the primary disease.

5. CLINICAL PRESENTATION

The clinical presentation of rotational and fixation defects are determined by the type of abnormality. Children may present with midgut volvulus or an isolated duodenal obstruction in an acute, recurrent, or chronic setting. The majority of patients will present with symptoms in the newborn period. Reports show that up to 65% of patients will present within the first thirty days of life (3,22); however, rotational anomalies may remain silent until adulthood.

The most common symptom, particularly in young patients, is bilious emesis. This may be due to duodenal obstruction from peritoneal bands or due to midgut volvulus (3,22,23). Others symptoms include nonbilious emesis, blood per rectum, intermittent diarrhea, early satiety, weight loss, failure to thrive, or vague abdominal cramping.

Constipation is also very common. Some patients are asymptomatic. Physical findings range from a totally benign examination to an acute abdomen with peritonitis.

5.1. Midgut Volvulus

Midgut volvulus is a surgical emergency. The narrow mesenteric base present in malrotation places the midgut at increased risk for volvulus. When the bowel twists around its mesentery the vascular supply becomes compromised. Most patients with midgut volvulus present within the first months of life (24). Sudden onset of bilious emesis is the most common sign of acute midgut volvulus. Other signs include abdominal distension, dehydration, and irritability. As intestinal ischemia develops the symptoms may progress rapidly to lethargy, a firm abdomen, hypovolemia, and shock. Bilious emesis and other signs should raise concern for midgut volvulus, and a definitive diagnosis must be established before intestinal viability is compromised.

Intermittent and partial midgut volvulus occurs less frequently and is more commonly encountered in patients >2 years of age (25,26). Patients typically complain of chronic abdominal pain, intermittent nonbilious emesis, early satiety, weight loss, failure to thrive, and diarrhea. These patients may have poor nutrient absorption due to obstruction of mesenteric lymphatics and veins. Failure to suspect the diagnosis has led to repeated dietary manipulations and psychiatric consults in some patients (19).

5.2. Duodenal Obstruction

Duodenal obstruction may be partial or complete and chronic or acute. Acute duodenal obstruction is caused by extrinsic compression of peritoneal bands or by the kinking of incompletely rotated or nonrotated duodenum. Acute duodenal obstruction is most common in the neonate, but it may occur later in life. Bilious emesis is the primary sign; however, nonbilious emesis may be seen. The presence of abdominal distension is variable. Chronic or recurrent obstruction of the duodenum results from the kink of the duodenum that results from improper rotation. Key findings are bilious or nonbilious emesis, poor weight gain, and cramping abdominal pain. Contrast studies may underestimate the degree of duodenal kinking.

5.3. Vague Nonspecific Symptoms

Rotational anomalies may be diagnosed during a workup in children with vague and nonspecific symptoms such as intermittent abdominal pain, vomiting, poor weight gain, diarrhea, or constipation. It is difficult to know whether these symptoms, which are very common in children, are caused by the rotational abnormality.

5.4. Asymptomatic

Many patients with rotational abnormalities remain asymptomatic. It is important, but often difficult or impossible, to differentiate nonrotation from rotational defects that require surgery. Rotational anomalies are often discovered as incidental findings during operations for unrelated reasons or after a gastrointestinal tract contrast study. The primary concern with asymptomatic patients is trying to predict which patients are at risk of midgut volvulus and the potentially life-threatening consequences related to its development. There is no evidence in the literature that clearly differentiates nonrotation from malrotation or identifies factors that will effectively predict when patients with

malrotation will develop midgut volvulus. Therefore, it is common practice to operate on children with rotational abnormalities and surgically correct malrotation upon discovery. Nevertheless, this practice remains controversial because there is inadequate evidence in the medical literature to show a benefit in the application of surgical therapy over expectant management in asymptomatic patients.

6. DIAGNOSIS

Radiologic studies play a critical role in establishing the diagnosis of malrotation. Any infant with bilious emesis should have immediate imaging studies performed to confirm or rule out the diagnosis. The diagnostic evaluation should include plain abdominal radiographs and contrast imaging of the gastrointestinal tract.

Plain abdominal radiographs are usually nondiagnostic, but may show patterns and signs that may be helpful, such as the "double-bubble" indicative of duodenal obstruction. A "gasless" abdomen often characterizes volvulus; however, patients with volvulus may have a nonspecific or normal bowel gas pattern on plain abdominal radiographs. Bowel thickening and edema, indicating vascular compromise in the bowel, can often be seen with midgut volvulus. Since plain abdominal radiographs are often nondiagnostic, rapid steps must be taken to perform contrast radiologic studies, or the patient should be taken to the operative theatre for exploration to establish the diagnosis.

Contrast studies of the intestinal tract are needed to establish the diagnosis of abnormalities of intestinal rotation and fixation. The upper gastrointestinal contrast series is performed as the initial study of choice at most centers. The duodenum should be seen coursing across the spine in a cephalad direction to the ligament of Treitz. The diagnosis is established when the ligament of Treitz does not cross to the left of the patients' spine. There is controversy as to the significance of a low-positioned ligament of Treitz that crosses the spine. Other findings include obstruction of the duodenum and the jejunum positioned on the right side of the abdomen (27,28). Hallmark signs for volvulus include the "bird's beak" sign and a corkscrew appearance of the small intestine. Delayed films or small bowel follow through into the colon may reveal abnormalities of colonic rotation or fixation. Barium enema can also be used to evaluate the colon in patients suspected of having malrotation; however, there are many limitations of the barium enema. For instance, the barium enema will miss the diagnosis of duodenal obstruction that occurs due to malrotation with a normally positioned cecum (19). Other limitations include missed diagnosis in patients with a mobile cecum and nondiagnostic imaging due to redundant bowel (3). In their report, Simpson et al. showed that upper gastrointestinal contrast imaging made the diagnosis of 100% in comparison to a 50% rate for barium enema (28). Because of its limitations, the barium enema should be used to define the location of the cecum or to rule out other pathology such as intussusception, and not as the sole radiologic test for determining the presence of malrotation.

With improvements in technology, ultrasound has been used in some centers to aid in the diagnostic evaluation of malrotation. Ultrasound can be used to rule out other causes of obstruction such as pyloric stenosis, to establish the orientation of the superior mesenteric vessels, and to define the presence of flow through the superior mesenteric vessels in patients with suspected volvulus. While the use of ultrasound is becoming increasingly widespread, it is extremely operator dependent and should only be used to provide adjunctive information in the diagnosis of intestinal rotation abnormalities. Ultrasound cannot, however, differentiate malrotation from nonrotation.

7. PREOPERATIVE MANAGEMENT

An acutely ill patient with suspected midgut volvulus or bowel obstruction requires prompt surgical attention. Resuscitation with appropriate intravenous fluids should begin immediately. Other adjunctive measures include the placement of a nasogastric tube to decompress the stomach, the placement of a urethral catheter to measure urinary output, and the administration of intravenous antibiotics. In patients with suspected malrotation without acute symptoms, corrective surgery can be delayed to allow a thorough work-up and preoperative planning.

8. SURGICAL TREATMENT

Exploratory laparotomy and the Ladd procedure is the "gold standard" for treating malrotation with or without volvulus. The key steps of the procedure have not changed since it was first described by Ladd and Gross (5). The surgical steps are as follows:

1. evisceration of the midgut and inspection of the mesenteric root;
2. counter-clockwise de-rotation of the midgut volvulus (if present);
3. lysis of peritoneal bands with mobilization of the right colon so that it moves to the left side of the abdomen and straightening of the duodenum along the right abdominal gutter;
4. widening of the mesentery so that the small bowel sits to the right side of the abdomen and the cecum in the left lower quadrant (a state of nonrotation); and
5. appendectomy.

The principles of the open surgical technique also apply to the laparoscopic correction of abnormalities of intestinal rotation and fixation. This will be discussed in following sections.

8.1. Laparoscopic Treatment

Recent advances and experience in minimal access surgery have led to the development of techniques for the successful correction of abnormalities of intestinal rotation and fixation (29–34). The first report of the laparoscopic management of malrotation was by van der Zee and Bax (6) in 1995. They reported on the successful treatment of acute midgut volvulus with the use of minimally invasive techniques.

Since 1995, there have been 12 additional reports that have described the successful use of minimal access techniques in treating malrotation with or without volvulus (see Table 22.1). This group of publications includes five case reports (6,14–17), six case series (7–11,13), and a retrospective study comparing patients treated by open technique vs. laparoscopic technique (12). As a collection the studies account for the care of 58 patients who have been treated with minimal access surgery for malrotation. The patients span across all age groups and can be divided into neonates and infants (6–13), older children (8,12–16), and adults (13,17). Furthermore, the studies include patients treated for malrotation with volvulus and without volvulus, and the reports address the role of minimal access techniques in both the diagnostic evaluation of malrotation and in corrective therapy. The case reports and case series all praise the benefits of a short hospital stay, rapid return of bowel function, minimal postoperative pain, and improved cosmesis that were achieved with the use of minimal access surgery. However, only one study compares outcome data of laparoscopic surgery to the traditional open technique (12).

Table 22.1 Publications of Minimal Access Surgery for the Treatment of Rotation Abnormalities

Authors	Type of study (# of patients)	Ages of patients
Malrotation without Volvulus		
Lessin and Luks (8)	Case series (2)	6 years, 17 years
Bass et al. (9)	Case series (12)	Mean: 2 weeks (5 days–4 months)
Cheikhelard et al. (10)	Case series (3)	4, 11, 3 days
Gross et al. (11)	Case series (2)	6 weeks, 4 months
Frantzides et al. (17)	Case report	38 years
Waldhausen and Sawin (16)	Case report	4 years
Yahata et al. (15)	Case report	17 years
Mazziotti et al. (13)	Case series (7)	Median 7 years (4 days–23 years)
Chen et al. (12)	Retrospective comparison of laparoscopic to open surgery (LS, 18; OS, 20)	LS, median 8.5 months (3 days–17 years) OS, median 5.3 months (1 day–7 years)
Bax and van der Zee (7)	Case series (9)	Range 7 days–28 months
Malrotation with volvulus		
Bax and van der Zee (7)	Case series (9)	Range 7 days–28 months
Van der Zee and Bax (6)	Case report	7 days
Yamashita et al. (14)	Case report	13 years

8.2. Technique

The technical aspects of the laparoscopic treatment of abnormalities of malrotation without volvulus are based upon the principles established by Ladd as discussed above. The four major steps consistent in the reports from all of the authors are:

1. inspection of the mesenteric base and detorsion of the bowel;
2. lysis of peritoneal bands;
3. placement of the bowel in a state of nonrotation with broadening of the mesenteric base;
4. appendectomy (7–13,15–17). Port placement differed among authors, but we believe that the port placement configuration used in the case series by Mazziotti et al. provides the surgeon with flexibility and comfort to perform the procedure; furthermore, the technique described by Mazziotti et al. has been safe and effective at our institution and is detailed below (12,13).

A 10-mm port is placed in the infraumbilical fold with the use of the open technique. Two 5-mm ports are inserted in the right and left lower quadrants respectively, and a 10-mm port is positioned in the midline above the umbilicus (13). The abdomen is insufflated according to the age of the patient with a pressure range of 8–15 mmHg. After an adequate pneumoperitoneum and visualization are established, the base of the mesentery is inspected by identifying the duodenojejunal junction and the ileocecal junction. Since there are no guidelines established to predict whether a volvulus will occur, a mesenteric base extending greater than half the diameter of the abdomen was arbitrarily selected as being broad enough base to prevent volvulus. If the length of the mesenteric base is less than half of the transverse diameter of the peritoneal cavity, a Ladd procedure is performed.

The first step of the Ladd procedure is complete mobilization of the right colon. It is reflected to the left. Next, the entire duodenum is mobilized with straightening of the C-loop of the duodenum and peritoneal bands are lysed. Any remaining peritoneal bands associated with the mesentery should be lysed, and the base of the mesentery broadened. An appendectomy is performed with the use of laparoscopic techniques. The last step is to leave the bowel in a state of nonrotation with the cecum in the left upper quadrant and the small intestine to the right of the spine (13).

Cecopexy was used in the care of an adult patient in the report by Frantzides (17), but there are no descriptions in the pediatric surgical literature. There is no existing evidence to show that a benefit is derived from the use of cecal fixation in performing a Ladd procedure. In addition, cecal fixation creates a potential axis for rotation around the point of fixation and predispose patients to the development of a volvulus.

8.3. Results in Patients Without Volvulus

Since 1995, ten reports have shown that minimal access surgery can be applied safely and effectively to the evaluation and treatment of abnormalities of intestinal rotation and fixation (7–13,15–17). The studies are a collection of case reports, case series, and one retrospective analysis of open vs. laparoscopic technique. The studies address management in neonates, children, and adults. Symptomatic and asymptomatic patients were treated with laparoscopic techniques. Symptoms at presentation included bilious and nonbilious emesis, recurrent abdominal pain, and peritoneal signs (Table 22.2). Radiographic images were used consistently to evaluate the patients, and upper gastrointestinal contrast studies were used most commonly.

Many detractors of minimally invasive techniques for complex surgical problems cite technical feasibility, increased operative time, and potential for intraoperative complications as contraindications for minimal access surgery in infants. However, together the reports describe a reasonable level of operative times ranging from 35 min to 3 h and 25 min (Table 22.2). No intraoperative complications or incidents of conversion to open technique were reported. The outcome data generated by the reports were consistent. Time to the resumption of a regular diet ranged from one postoperative day to five postoperative days (Table 22.2). The time to discharge ranged from 1 day to 6 days postoperatively (Table 22.2). All patients detailed in the reports had complete resolution of symptoms at follow-up.

The data provided by the reports, supports the application of minimal access techniques to malrotation without volvulus. Many institutions have successfully applied minimal access surgery to malrotation without volvulus. The weaknesses of this body of literature are typical of case reports and small case series. First, there is tremendous potential for reporter bias in the data presented. It is difficult to interpret the lack of negative outcomes reported. While this may indeed be true, it is not verifiable. The second major weakness is the lack of concurrent controls or comparison data for the outcomes generated by these reports.

Recently, Chen, Minkes, and Langer reported a study comparing the outcomes of the laparoscopic and open approaches in children with malrotation without volvulus. The retrospective study evaluates the outcomes of 18 patients treated with minimal access techniques and 20 patients treated by an open Ladd procedure over the same time period. The ages of the patients were similar with the median age of 8.5 months in the laparoscopic surgery group (LS) and 5.25 months in the open surgery group (OS) (Table 22.3). Twenty-nine percent of the patients in the study were asymptomatic, and all rotational abnormalities were confirmed by contrast studies. The outcome data of the study shows

Table 22.2 Outcome Data from Publications for Laparoscopic Technique for Malrotation

Authors	Operative time	Time for return to regular diet	Time to discharge	Resolution of symptoms	Length of follow-up	Complications
Malrotation without volvulus						
Lessin and Luks (8)	NR	POD 1	POD 2	Yes	NR	None
Bass et al. (9)	Ave. 58 min (35–120 min)	POD 2 (10 cases) POD 3 (2 cases)	Ave. 2.2 days (2–4 days)	Yes- for all pts	NR	None
Cheikhelard et al. (10)	NR	Pt #1, POD 5 #2, NR #3, complicated by duodenal web	#1, POD 6 #2, NR #3, NR	Yes	#1, 5 months #2, 5 months #3, 2 months	None
Gross et al. (11)	#1, NR #2, 76 min	#1, POD 4 #2, POD 2	#1, POD 5 #2, POD 4	Yes	NR	None
Frantzides et al. (17)	1 h and 45 min	POD 1	POD 1	Yes	3 months	None
Waldhausen and Sawin (16)	3 h	POD 3	POD 3	Yes	5 months	None
Yahata et al. (15)	2 h and 20 min	POD 1	POD 6	Yes	6 months	None
Mazziotti et al. (13)	Median 2.0 h (1.25–3.25)	Median POD 2	Median POD 2	Symptoms Resolved in 5/7 pts	Median 15 months (10–16)	None
Bax and van der Zee (7)	3 h first few pts 1 h for later pts	NR	POD 4–11 days	Yes	NR	None
Malrotation with volvulus						
Bax and van der Zee (7)	3 h first few pts 1 h for later pts	NR	POD 4–11 days	Yes	NR	None
Van der Zee and Bax (6)	90 min	POD 1	NR	Yes	NR	None
Yamashita et al. (14)	NR	POD 1	NR	Yes	6 months	None

Note: NR, not reported.

Table 22.3 Group Characteristics From Chen et al. (12) Comparison of Laparoscopic to Open Technique

	Laparoscopic ($n = 18$)	Open ($n = 20$)
Gender		
Male	8	10
Female	10	10
Median age (months)	8.5 months (3 days–17 years)	5.25 months (1 day–7 years)
Broad-based mesentery	6	8
Operatvie time (min)	70 (35–194)	68 (25–105)
IV narcotics (h)*	24	46
Time to clear feeds (h)***	18	68
Time to full feeds (h)***	32	111
Hospital stay (days)**	2.6	5.3
Cost (×$1000)	4.3	4.5
Late complications		
Small bowel obstruction	0	2

Note: $^{*}p = 0.05$; $^{**}p = 0.001$; $^{***}p < 0.001$.

that laparoscopic surgery is as safe and effective as traditional open technique in the pediatric patient population. It was associated with a faster recovery time, which is exemplified in the faster return to clear liquids (LS = 18 h vs. OS = 68 h, $p < 0.001$), faster tolerance of full feeds (LS = 32 h vs. OS = 111 h, $p < 0.001$), and the shorter hospital stay (LS = 2.6 days vs. OS = 5.3 days, $p = 0.001$). The LS group had less postoperative pain determined by a shorter time span for the use of intravenous narcotics (LS = 24 h vs. OS = 46 h, $p = 0.05$). Operative time was similar between the groups (LS = 70 min vs. OS = 68 min, p = NS), and there were no intraoperative or perioperative complications in either group. Hospital charges did not differ significantly between groups. The symptoms resolved in all patients in the study. The mean follow-up was 22.1 months for all of the patients in the study. There were no long-term complications reported in the LS group; however, in the OS group two patients developed adhesive small bowel obstructions at 13 days and 21 months postoperatively. The patients both required laparotomy for treatment.

This study supports the conclusions of the authors from the case reports and series. The use of laparoscopic surgery is as safe and effective as the open Ladd procedure and is also associated with faster recovery, less postoperative pain, and a potentially lower risk of future adhesive bowel obstruction. While these data support the use of minimally invasive techniques, a multi-institutional prospective randomized control study comparing the laparoscopic approach to the traditional open technique would be needed to determine the true benefit of this approach.

While most of the reports advocated a full Ladd's procedure despite the width of the mesenteric base (7–11,15–17), Chen et al. (12) differed in their management by not performing the entire Ladd's procedure on patients with a wide mesenteric base in the LS treatment group. If a patient in the LS group had a broad based mesentery defined by a mesentery greater than half the transverse diameter of the abdomen, then the patient underwent appendectomy alone. Six (33%) patients of the LS group were found to have a broad-based mesentery and managed with appendectomy alone. All patients in the open group underwent a full Ladd procedure even when mesentery was broad enough to prevent volvulus. After a mean follow-up of 27.1 months, there were no reports of late volvulus in any

patient in the LS group of the study. Thus, the laparoscopic approach appears to be ideal in patients with "soft" radiologic findings of a rotation anomaly. In the report by Chen et al. (12) more than one-third of all patients were determined intraoperatively to have a broad-based mesentery and were felt not be at risk for volvulus. Thus, a full Ladd's procedure may be over treatment in these patients. Late follow-up in this patient population will be critical to establish the safety of this approach.

8.4. Results in Patients with Volvulus

Malrotation with volvulus presents different challenges for the laparoscopic surgeon. In malrotation with volvulus the bowel is often very friable and edematous; therefore, great care must be taken to avoid injury with the laparoscopic instruments. Expeditious detorsion of the bowel becomes a difficult task in the laparoscopic setting due to the small amount of operative space and the limitations of the laparoscopic instruments. In addition, speed may be the most critical element in preserving the viability of the bowel. This may be hindered by the laparoscopic approach. Many authors have discouraged the use of minimal access techniques in the presence of malrotation with volvulus (12,13). The laparoscopic management of patients with known midgut volvulus is controversial. There have been three reports of the use of laparoscopic techniques in the management of malrotation with established midgut volvulus (6,7,14). The studies describe the care of four patients. Three patients were infants, and the fourth patient was a 13-year-old girl. The infants presented with the acute onset of bilious emesis, and upper gastrointestinal contrast studies confirmed the presence of obstruction and volvulus. The 13-year-old girl presented with severe abdominal pain after 1 year of multiple hospitalization for intermittent abdominal pain.

The patients all underwent laparoscopic Ladd's procedure with successful outcomes. All of the patients tolerated a regular diet on postoperative day one. There were no intraoperative or perioperative complications, and the patients were asymptomatic at long-term follow-up.

Bax and van der Zee described the surgical technique employed by all three authors (6,7,14). The laparoscopic treatment of malrotation with volvulus relies upon the same principles outlined for treatment of malrotation without volvulus. In the care of patients with volvulus the surgeon must pay greater attention to the detorsion of the bowel. To detorse the bowel, Bax and van der Zee emphasize that the surgeon must first focus on the identification of the second portion of the duodenum. After it is identified, peritoneal bands are lysed, and the duodenum and jejunum are completely mobilized. According to Bax and van der Zee (7), pulling the duodenum and jejunum to the right side of the patient in a position underneath the liver will reduce the volvulus and place the bowel in a state of nonrotation. All other steps of the procedure should be conducted as described for patients with malrotation without volvulus.

While three reports have demonstrated success in four patients, most authors discourage the use of laparoscopic techniques in patients with confirmed midgut volvulus (9,11–13,16). Detractors believe that complications will arise from the greater technical difficulty (11) and the handling of ischemic bowel with laparoscopic instruments (9,12). In addition, expeditious detorsion of bowel may not be feasible with laparoscopic techniques. For the above reasons and the lack of sufficient evidence to support its safety, minimal access surgery should not be used routinely in the management of patients with malrotation with volvulus. If it is attempted, the surgeon must carefully choose patients that are in an early state of volvulus. In addition, the surgeon must be very experienced and adept at laparoscopic surgical techniques in pediatric patients.

9. SUMMARY

The data presented in this chapter concerning the laparoscopic treatment of malrotation without volvulus show that it is both safe and effective in pediatric patients. The one available study to compare laparoscopic surgical technique to open surgical technique showed favorable outcomes for the laparoscopic surgery treatment group. In addition, laparoscopy may be most useful in assessing patients "soft" radiologic findings of rotational abnormalities. Collectively the available data show support of minimal access surgical techniques for the management of malrotation without volvulus; however, prospective studies with larger sample sizes, long-term follow-up, and possibly randomization should be conducted to provide confirmation of the conclusions from the present literature. Patients with malrotation who have confirmed midgut volvulus should not undergo laparoscopic treatment, except in highly selected circumstances, due to the great risk for intraoperative complications.

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23
Varicocele

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1. INTRODUCTION

Varicoceles are considered to be the most identifiable cause of male infertility. The incidence of varicocele in prepuberal age varies from 10% to 15%, according to different series; the importance of early treatment, in childhood, to prevent testicular damage is widely accepted (1). Treatment options include spermatic vein sclerotherapy or embolisation, classical open surgical treatment via a scrotal, high retroperitoneal or inguinal approach, microsurgical bypass, and more recently, laparoscopy (2–4).

The main problem for the adolescent patient is establishing objective criteria for intervention. The decision for varicocele surgery in men usually is based on documented infertility and abnormal semen analysis; in adolescents these criteria are not applicable. In general, the intervention is indicated only when a persistent and measurable asymmetric decrease in testicular size is documented, along with a readily apparent varicocele and scrotal pain or discomfort (5,6).

2. DIAGNOSTIC EVALUATION

Most varicoceles are detected during adolescence by routine physical examination. The clinical classification of varicoceles is based on Horner's (7) definition: grade I, palpable but not visible; grade II, palpable and visible; and grade III, large varicocele. To confirm the presence of a varicocele detected by physical examination, a scrotal ultrasound with color-flow Doppler scanning is recommended. Other tests include thermography and venography; the latter can be used in associated with a transvenous ablation procedure.

3. ANATOMY

The knowledge of the vascular anatomy of the varicocele is fundamental to guide the surgical strategy. The venous drainage of the scrotum occurs via a superficial system (the anterior and posterior scrotal veins, which drain into the saphenous vein) and a deep system (which drains via the pampiniform plexus, the internal and external spermatic veins, and the ductus deferens veins). These two systems anastomose with each other (8). The left internal spermatic veins drain into the left renal vein, whereas the external spermatic vein drains—via the inguinal canal—into the inferior epigastric vein near its termination at the femoral vein (9). The ductus deferens vein drains into the internal iliac vein.

Three forms of varicocele can be differentiated: type I, where the etiologic factor is the internal spermatic vein itself (absence of competent valves in the proximal portion of the internal spermatic vein); type II, with the presence of the proximal nutcracker phenomenon (the compression of the left renal vein between the aorta, from the posterior side, and the superior mesenteric artery, from the anterior side); and type III, the presence of the distal nutcracker phenomenon (due to the obstruction of the left common iliac vein often because of compression from the left common iliac artery) (8,9). Two or three types of varicocele may be present at the same time.

4. PRINCIPLES OF TECHNIQUE

Types I and II are the most frequent, comprising $\sim$90% of cases (1–5). In these cases, the surgical procedure for the correction of the varicocele consists of the ligation of the inner spermatic veins alone (Ivanissevich procedure), or the ligation of the spermatic veins and artery (Palomo procedure) in the suprainguinal region (10,11). In the case of a type III varicocele, it is necessary to ligate both the internal spermatic veins and the deferential veins (10,11). Both procedures can be performed via laparoscopy or retroperitoneoscopy.

5. OPERATIVE SET-UP

In case of the laparoscopic approach, the patient is placed in the supine position, in a Trendelemburg position of at least 15–30°, with the entire abdomen, genitalia, and upper legs included in the operative field. For retroperitoneoscopy, the patient is positioned in lateral decubitus. In laparoscopy, the surgeon stands on the patient's side contralateral to the pathology, the assistant stands on the other side facing the surgeon, and the scrub nurse on the same side as the surgeon (5). In retroperitoneoscopy, the surgeon stands in front of the patient with the assistant on his/her right and the scrub nurse in front. For laparoscopy,

the screen is positioned at the patient's feet and the laparoscope and video units on the patient's side contralateral to the pathology. For retroperitoneoscopy, the screen is positioned in front of the surgeon. Laparoscopy is performed using one umbilical trocar for the telescope and two trocars in triangulation.

For retroperitoneoscopy, only one 10 mm trocar is necessary. This trocar is positioned half way between the costal margin and the superior iliac spine. An operative telescope is used with a 5 mm operative channel. A 450 mm grasper, a hook, and scissors are used to dissect and then coagulate the inner spermatic vessels (12,13).

6. OPERATIVE TECHNIQUE

6.1. Laparoscopy

The first step consists of the inspection of the abdominal cavity to identify the internal inguinal ring and evaluate the course of the cord (5–12). The peritoneum above and lateral to the inner spermatic vessels is opened with scissors at 5 cm above the inguinal ring (2–14). Sometimes, it may be necessary to mobilize the sigmoid colon to clearly expose the spermatic cord. The testicular vascular pedicle is fully exposed using scissors and atraumatic forceps. At this time, the surgeon must decide whether to perform an Ivanissevich procedure and spare the spermatic artery or to perform a Palomo procedure and ligate both spermatic veins and testicular artery (5–15,16).

Usually the spermatic vessels are sectioned between clips; alternatively, the vessels can be ligated and then sectioned. It is also possible to use bipolar or tripolar energy, laser energy, or ultrasonic scalpel. Any other vein around the spermatic bundle must be clipped and sectioned to reduce the risk of recurrence. It is very important to examine the veins around the vas deferens to assess whether they are macroscopically dilated. The incision on the posterior peritoneum can be closed at the end of the procedure with one or two stitches (5).

6.2. Retroperitoneoscopy

The retroperitoneal operative chamber can be created by Gaur's balloon technique or with the simple dissection of the retroperitoneal space using the tip of the telescope and with the aid of the pneumodissection, as described by Valla (5–13).

In retroperitoneoscopy, because of the small operative chamber, it is preferable to adopt one trocar surgery.

The inner spermatic vessels are identified, bluntly dissected for $\sim$3–4 cm of their length, coagulated using monopolar or bipolar energy, and then sectioned.

7. COMPLICATIONS

Data from a large multicenter series (13) demonstrated that most authors (85.8%) prefer the laparoscopic technique described by Palomo. This general tendency seems to be related to the lower number of recurrences with this technique (1.6% in this series) as compared with other procedures. As to the cause of recurrences in this series, postoperative venography demonstrated that they were due to reflux through the deferential veins related to an obstruction of the left common iliac vein. This extremely rare event, occurring in $\sim$5–10% of children with varicocele, can be prevented by the routine use of venography in the preoperative period or by also sectioning the deferential veins—should they

appear varicose when compared with the contralateral side—during the laparoscopic examination (13–15). This is not as easily visualized by retroperitoneoscopy. The 3.8% incidence of intraoperative complications was due to bleeding from the inner spermatic vessels during dissection, or instrument/light source/video-camera malfunction.

In the case of retroperitoneoscopic varicocelectomy, the main problem is linked to the smaller operative field and the risk of entering the peritoneal cavity during dissection, which would require conversion to the laparoscopic or to the open approach. Postoperative complications include a high rate of hydrocele using Palomo's procedure, (6.6%), related to the ligation of the lymphatic vessels when the artery is ligated (17–19).

The prevention of hydroceles seems to be most successful with Ivanissevich's procedure, because it spares the lymphatic vessels adherent to the spermatic artery and thus reduces the rate of this complication. Unfortunately, the Ivanissevich procedure is correlated with a higher rate of recurrence than that of Palomo's (5–13). In this study, which is in agreement with most reports in the literature, there were no cases of testicular hypoplasia or atrophy using Palomo's technique. This advantage is because of the collateral blood supply to the testis from the gubernaculum, the anterior and posterior scrotal vessels, the intra-scrotal anastomosis, and the deferential vessels (13). However, this study cannot demonstrate any improvement in spermatogenesis, because $<5\%$ of these patients have undergone semen analysis because of their young age at the time of surgery (20,21).

8. DISCUSSION AND CONCLUSIONS

Several other procedures can be used to treat pediatric varicoceles: venous embolization by interventional radiologists, open surgical technique, and microsurgical venous bypass (22–25). However, a review of the medical literature of the last 5 years shows that an increasingly frequent approach for both adults and adolescents is laparoscopy

Table 23.1 Results of Different Procedures of Varicocelectomy in Pediatric Patients

References	Year	Number of patients	Level of evidence	Type of procedure	Success (%)
Luque Mialdea et al. (22)	1995	22	II	Microsurgical anastomoses	100
Ardela Diaz et al. (23)	1996	15	II	Operative radiology	74
Lima et al. (4)	1997	207	II	Microsurgical anastomoses	97
Campobasso (25)	1997	172	II	"Blue venography" open surgery	100
Minevich et al. (26)	1998	32	II	Inguinal microsurgical varicocelectomy	100
Mazzoni et al. (24)	1999	277	I	Interventional radiology	79.4
Mazzoni et al. (24)	1999	124	I	High ligation via open surgery	88.7
Cayan et al. (3)	2000	236	II	MHIV	97.9
Cayan et al. (3)	2000	232	II	High ligation via open surgery	84.5
Esposito et al. (5)	2000	28	II	Laparoscopy–Ivanissevich	96.5
Esposito et al. (5)	2000	133	II	Laparoscopy–Palomo	97.8
Poddoubnyi (2)	2000	180	II	Laparoscopy–Ivanissevich	99.4
Cohen (12)	2001	40	II	Laparoscopy–Ivanissevich	83

Note: I, prospective study; II, retrospective study, review, or anecdotal report.

(Table 23.1) (5–16). The most striking piece of evidence is a common trend among several teams to adopt the laparoscopic Palomo's technique (13–16). This general tendency is probably related to its low recurrence rate (2–3%) as compared with other procedures (5–14). The 5–10% recurrence rate can be prevented by a systematic use of preoperative venography (23–25).

Critics of the laparoscopic method cite the high costs as a drawback (14–28). This problem is virtually eliminated when reusable trocars and instruments are employed: the only nondisposable instrument used in laparoscopy is the clip applier, and this is sometimes replaced by traditional low-cost ligatures.

The literature (5–13,28,29) demonstrates that the laparoscopic approach, in terms of recurrence and complications rate, is comparable to, if not better than, those achieved with the open surgical or radiological approach (28,29).

In addition, laparoscopy can assess and easily treat varicosed deferential veins which would be a cause for recurrence if only spermatic vein ligation is performed.

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24
Nonpalpable Undescended Testis

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1. INTRODUCTION

The incidence of cryptorchidism is 1–3% in male infants, 20% of whom have a nonpalpable testis (NPT) at presentation (1). The treatment of a palpable cryptorchid testis is not controversial and consists of a transinguinal orchiopexy. Laparoscopy has replaced ultrasound and magnetic resonance imaging for the localization of a NPT (2). Controversy exists concerning the operative technique in the cases of an intra-abdominal testis (IAT). The laparoscopy-assisted orchidopexy (LAO) without sectioning the spermatic vessels and the two-stage Fowler–Stephens (FS) procedure seem to be the techniques with the highest success rate (3,4) and have been rapidly gaining popularity.

2. HISTORY

The use of laparoscopy has brought about a remarkable improvement in the study and management of the NPT. The laparoscopic procedure was first described by Cortesi

et al. (5) in 1977, who showed that an IAT could be located in the pelvic region close to the bladder; in that position, the testis is practically unidentifiable with a traditional open approach performed via an inguinal incision. At the end of the 1980s, Bloom (6) and Elder (7) proposed the use of a laparoscopic FS orchidopexy for the management of the NPT, with the first step via laparoscopy and the second via open surgery. In 1994, Jordan and Caldamone reported the possibility of mobilizing laparoscopically an IAT and of accomplishing also the second phase of the two-step FS procedure laparoscopically (8–10). More recently, Esposito and Baker proposed a laparoscopy-assisted orchidopexy without sectioning the spermatic vessels (4–11).

3. ANATOMY

The knowledge of the anatomy and the embryology of the testis is fundamental to the accurate interpretation of the laparoscopic findings. The development of the testis may be divided into three phases: an intra-abdominal (1–7 months), a canalicular (7–8 months), and a scrotal phase (8–9 months). The failure of the testis to migrate in any one of these three phases causes cryptorchidism, the position of the cryptorchid gonad depending on the phase in which the migration stops. Concerning vascularization, blood flow occurs through the inner spermatic artery and inner spermatic veins. The inner spermatic artery derives directly from the aorta, and at 3–4 cm proximally to the testis, it anastomoses with the deferential arteries; these anastomoses are very important for the success of the FS intervention (6–8).

In the case of a high IAT, the testicular vessels are short, but the vas deferens and its companion vessels are longer than normal. The secondary vascular loop develops from the vessels of the vas deferens, the collaterals arise from the deep epigastric vessels, and the myriad branches enter the posterior wall of the processus vaginalis from the area of the gubernaculum testis. This vascular network results in a rich collateral circulation. The inner spermatic veins go directly into the vena cava on the right side and into the renal vein on the left.

4. PRINCIPLES OF THE TECHNIQUE

In all cases of NPT, the clinical examination under general anesthesia is essential and should be followed by laparoscopy. On the basis of the laparoscopic findings, the surgeon can easily select the most appropriate surgical strategy (Fig. 24.1). When there are blind ending cord structures, no inguinal surgical exploration is necessary. In the case of cord structures entering the internal inguinal ring (IIR), an open inguinal exploration is necessary to identify, and remove if necessary, an atrophic intracanalicular or ectopic testis (2). Interestingly, malignancy developing in a remaining atrophic testis has only been reported once in the international literature (11).

Concerning the surgical treatment of IAT, there is no general agreement over the procedure to adopt—the choices are an open groin exploration with abdominal extension for all cases, an FS procedure, microvascular transplantation, and laparoscopy (12,13). Presently, the best outcomes are reported with the two-step FS procedure (with both steps laparoscopically) and the LAO with the spermatic vessels intact (4–14). These will be described in the subsequent paragraphs.

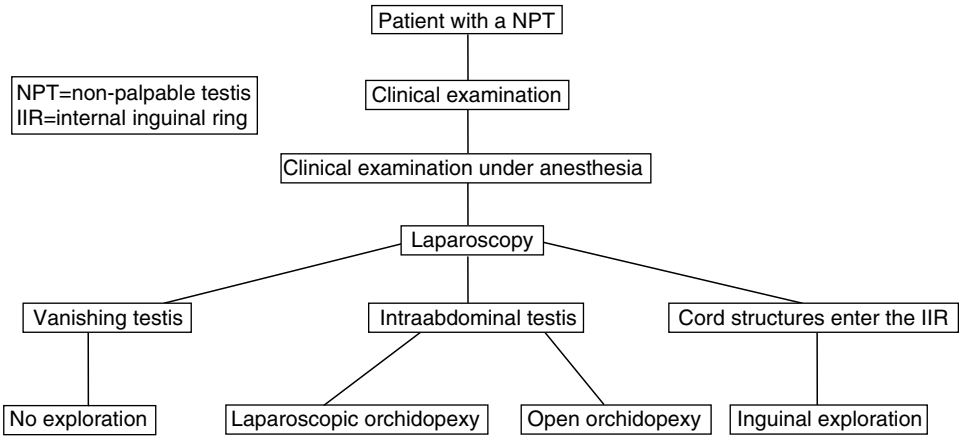


Figure 24.1 Algorithm for the workup of patient with a NPT.

5. OPERATIVE TECHNIQUE

The patient is placed supine with slight Trendelenburg. The entire abdomen, genitalia, and upper legs are included in the operative field (7). The bladder is drained. The surgeon stands opposite the side of the NPT. The assistant stands on the other side, facing the surgeon. The video monitor is positioned at the patient's feet. The telescope is placed transumbilically and two 3 or 5 mm trocars are placed in the right and left iliac fossae, at $\sim$3–4 cm below the umbilicus. The exact position of the trocars always depends upon the patient's age and size. If an IAT is found, a fourth trocar is placed in the ipsilateral hemiscrotum (11).

5.1. Diagnostic Laparoscopy

There are three principle laparoscopic findings: IAT (37%), abdominal blind-ending cord structures (14%), and cord structures entering the IIR (49%) (2). In cases of blind-ending cord structures (also called vanishing testis), there is no need for an open inguinal exploration. When cord structures enter the IIR, an inguinal exploration is necessary to look for an ectopic or an intracanalicular atrophic testis (1,2) or a nonpalpable but normal testis.

5.2. Laparoscopic Two-Step FS Procedure for IAT

The first step of the FS is very simple: two or more clips are positioned on the intra-abdominal spermatic vessels 3–4 cm away proximally to the testis. Collateral vascularization is derived from the vas deferens which provides blood to the last 2 cm of the intra-abdominal spermatic vessels (6,7). The second step of the FS procedure is performed 6–12 months later. This interval is necessary for the development of the collateral circulation. Laparoscopically, a wide peritoneal pedicle flap is created using scissors, and a blunt dissection is performed laterally to the internal spermatic vessels, near the clips placed closest to the testis during the first operation. The dissection is continued distally to the clips, around the internal ring; it is then extended medially to the umbilical ligament as far as 1 cm from the vas deferens (14,15). At this point, the spermatic vessels are sectioned near the clips and the dissection then extended medially, as far as 1 cm from the other side

Table 24.1 Results of FS Orchidopexy

No. of references	Year	Testes	Only one intrevention	Two-step procedure	Success (%)
Elder (7)	1992	12	—	12 (1st lap. 2nd open)	92
King (16)	1996	22	22 (open)	—	100
Law et al. (15)	1997	20	—	20 (1st lap. 2nd open)	95
Esposito and Garipoli (14)	1997	33	—	33 (both laparocopy)	100
Kirsch et al. (12)	1998	38	33 (open)	5 (1st lap. 2nd open)	74
Baker et al. (4)	2001	50	—	50 (both laparoscopy)	87.9
Chang et al. (3)	2001	29	20 (laparoscopy)	9 (both laparoscopy)	85
Baker et al. (4)	2001	20	20 (laparoscopy)	—	74.1

of the vas deferens. The testis is thus supported on a peritoneal flap attached to the peri-deferential peritoneum (15,16).

A blunt grasper is introduced deep into the hemiscrotum via the internal inguinal ring (14). A dartos pouch is then created through an open scrotal incision; a 5 mm trocar is inserted at the level of the bottom of the corresponding hemiscrotum and pushed into the abdominal cavity along the blunt grasper. With the use of a fenestrated forceps introduced through the scrotal trocar, the testis is grasped and brought down into the scrotum, carefully avoiding any torsion of the new vascular pedicle (14). The intra-abdominal portion of the procedure is completed by transperitoneal closure of the IIR. This technique consists of bringing the conjoined tendon closer to the crural arch with a nonabsorbable suture (6–14). The results of the FS procedure are presented in Table 24.1.

5.3. Laparoscopy-Assisted Orchidopexy Without Division of the Spermatic Vessels

This technique consists of sectioning the gubernacular attachment, opening the posterior peritoneum laterally to the spermatic vessels, and in mobilizing the testicular vessels and the vas deferens in a retroperitoneal position for ~8–10 cm (11–19). The vessels are preserved by performing a blunt dissection using a peanut or grasping forceps, without using any type of thermal energy. At the end of the dissection, the testis is free from adhesions to the posterior abdominal wall and pedicled onto the inner spermatic vessels and the vas (12–19). At this point, if the inner inguinal ring is open, a grasping forceps is introduced from inside the abdomen through the IIR into the scrotum. After creating a dartos pouch, a 5 mm trocar is introduced from the scrotum into the abdomen using grasping forceps (11). If the inner inguinal ring is closed, a neo inguinal ring is created medially to the obliterated umbilical artery and between the bladder, using the same procedure as described earlier; a 5 mm trocar is then introduced from the scrotum into the abdomen (17,18). At the end of the procedure, the testis is then brought down into the scrotum through either an already open IIR or a newly created inguinal ring. The results of the LAO are reported in Table 24.2.

6. COMPLICATIONS

In both the FS and the LAO procedures, a delicate dissection avoids injury to the iliac vessels, the ureter, and bladder. Concerning the FS procedure, atrophy of the testis after

Table 24.2 Results of Orchidopexy Performed Without Sectioning the Spermatic Vessels

No. of references	Year	Testis	Technique	Success (%)
Poppas and Lemack (18)	1996	10	LAO	100
Lindgren et al. (17)	1998	31	LAO	93
Kirsch et al. (12)	1998	33	Open inguinal	97
Esposito et al. (11)	2000	20	LAO	95
Baker et al. (4)	2001	140	LAO	97
Chang et al. (3)	2001	72	LAO[a]	92

[a]LAO, laparoscopy-assisted orchidopexy.

the first step of the procedure is an extremely rare event, whereas its incidence is 20–30% after the second step (7–12). Concerning the LAO procedure, once the spermatic vessels are isolated from the posterior peritoneum, they are very susceptible to injury when the testis is pulled down (11). A clinical assessment of all patients in the postoperative period is mandatory to evaluate for hypoplasia and atrophy, which when present, occurs several months later.

7. DISCUSSION AND CONCLUSIONS

To date, the pediatric surgery literature is devoid of sound evidence on this topic, as there are no meta-analyses, or randomized or prospective studies. On the contrary, there are several retrospective studies of single center experiences (2–4). The literature demonstrates that most authors consider laparoscopy the best type of diagnostic exploration in case of a boy with a NPT (2–11–20,21). Concerning the operative approach, laparoscopic orchidopexy seems to be better than the open procedure in correctly placing the abdominal testis in the scrotum, whereas it is difficult to find evidence regarding the best operative technique to adopt in case of IAT (3–22). On the basis of several publications reviewed, we believe that the inner spermatic vessels should be spared whenever possible to guarantee a good vascularization of the testis, and the laparoscopic mobilization of the spermatic vessels seems a good method to achieve this (21–23). In cases of very high IAT, with a distance between the testis and the IIR $>3-5$ cm, a two-stage laparoscopic FS procedure is the procedure of choice. If the inner spermatic vessels are very short, a high retroperitoneal dissection (as in the LAO procedure) will cause critical tension when pulling the testis down into the scrotum (11–25).

An important criticism of all the reports on NPT published in the international literature is that most have a short or inaccurate follow-up period, with the patients sometimes followed only by means of clinical examinations. Also, for each procedure there are sometimes similar results published using different procedures, via laparoscopy, via open surgery, or via microsurgery; this suggests that each author seems to prefer the technique adopted by his/her team. This point is very important as, except for the rare intraoperative complications related to the laparoscopic procedure, problems are rarely reported. We would like to stress the importance of a proper follow-up to verify the status and position of the testis and to identify hypoplasia or atrophy related to the procedure. The authors believe that a well-designed multicenter prospective study with long-term follow-up is necessary.

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25
Lung Biopsy, Lung Resection, and Pneumothorax

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1. INTRODUCTION

While minimal access surgery (MAS) in infants and children is a relatively new phenomenon, thoracoscopy has been performed in adults since the early 1900s (1). Jacobeus first described its use in 1910 when he reported on a series of thoracoscopic pleural adhenolysis in patients with tuberculosis using a rigid trocar and a cystoscope. Over the next 50 years there was limited experience with thoracoscopy and few advances (2). The technique remained limited to use in adults and consisted only of small biopsies of pleural-based lesions and limited explorations (3). The complexity of the procedures performed was restricted because of the simplistic nature of the optic system, light sources, and instrumentation. Procedures were limited to a single-port system with a working forceps inserted through or around the telescope sheath.

The first physician to make significant advances with the use of thoracoscopy in the pediatric population was Rodgers, who wrote of his initial experience in the mid-1970s (4). He used slightly modified cystoscopy equipment and newly developed instruments to perform small biopsies, evaluate intra-thoracic lesions, and perform limited pleural debridement in the cases of empyema (5,6). While his reports were landmark in nature, there was very little acceptance in the pediatric surgical community for these techniques. It was not until the early 1990s with the revolution in surgery that was spawned by the first successful reports of laparoscopic cholecystectomy and the technological advances in MAS technology that thoracoscopy in children gained more interest (7,8).

Numerous technological advances facilitated the advancements in pediatric MAS. These included improved camera systems, light sources, and optics. Also specific pediatric instrumentation was developed that was shorter (18–20 cm) and of smaller diameter (3 mm). However, a few specifically designed instruments greatly improved the ability to perform more complex thoracic procedures. The instrument with probably the greatest impact was the development of an endoscopic linear stapler. This instrument had an immediate impact on the management of patients with interstitial lung disease and, to a lesser extent, those with pulmonary metastatic lesions (9). The stapler allowed the surgeon to obtain large wedges of lung tissue without significant risk of air-leak or bleeding. This instrument along with later development of 5 mm endoscopic clips and various hemostatic energy devices set the stage for successful completion of even the most complicated thoracic procedures.

2. LUNG BIOPSY

The first successful thoracoscopic lung biopsies were performed in adults and one of the first large reports was by Bensard et al. in 1993 (9). They compared thoracoscopic vs. open lung biopsy in a series of patients with interstitial lung disease (ILD). They showed that the procedure was relatively easy to perform, was associated with minimal morbidity, and had a high rate of diagnosis. The morbidity, complication rate, and hospital stay were all less in the thoracoscopic group.

Unfortunately the linear stapler was too large (12 mm) for many pediatric patients and made universal application of this technique in small children difficult. A method using pre-tied ligatures to loop a tongue of lung tissue proved to be equally effective and required only a 5 mm port. Rothenberg et al. (10) reported the first large series of thoracoscopic lung biopsy (TLB) in infants and children in 1996. Adequate tissue for histology and diagnosis was obtained in 97% of cases and therapy was altered based on the results of the biopsy in 83% of cases. There were no complications and the average length of stay was 1.5 days.

Fan et al. (11), in a comparison of transbronchial, open, and thoracoscopic lung biopsy, further supported the use of thoracoscopic lung biopsy as a diagnostic tool in children with ILD. They showed that the highest diagnostic yield and lowest morbidity was in the group undergoing TLB. Similar results have been demonstrated in the use of TLB for metastatic pulmonary nodules. There is a large volume of evidence in the adult thoracic literature supporting thoracoscopic resection of single pulmonary metastasis. Swanson et al. (12) reported a large series of patients with suspect pulmonary nodules. They found that the sensitivity and specificity of TLB in this situation approached 100%. The morbidity was low in this group and the length of stay was less then three days. Similarly, Ginsberg et al. (13) from Memorial Sloan-Kettering had similar results in 426 patients. While their conversion rate was relatively high (25%), the operative mortality was very low (0.25%) and there was only one port site recurrence. There is larger controversy about this approach when there is more than one nodule, but most authors and series are relatively supportive of using this approach if there are only two to four nodules. There is still little long-term data to document outcomes in these patients.

Similar results have been documented in the pediatric population in dealing with pulmonary metastasis. In a series reported by Holcomb et al. (14) for the Children's Cancer Study Group, diagnostic material was obtained in 97% of cases. However, this was an early series from multiple institutions and the complication rate and conversion rate were $\sim$10%. Rothenberg reported similar accuracy of 98% with no complications

and a conversion rate of only 3% (15). These series, as well as others, report success rates for lesions approaching 1 cm in diameter and which are relatively peripheral as ~100%. Smaller lesions, especially those <0.5 cm or those deep in the parenchyma, are more difficult to identify thoracoscopically. This problem has been addressed to some degree by the use of CT-guided preoperative localization with either a pleural patch (blood or dye) or needle localization. A report by Smith et al. (16) documented success rates of >90% using these techniques.

The real disadvantage of TLB in metastatic disease is the inability to palpate the lung to detect unsuspected lesions. As yet neither intraoperative ultrasound nor any other imaging modality is sensitive enough to identify small parenchymal lesions, which are not pleural based and easily visualized. Significant controversy exists over whether undetected lesions are being left behind when only a thoracoscopic approach is being used, especially in cases of osteogenic sarcoma. The question is whether or not this will affect long-term survival. Most reports are anecdotal and there are no good studies to decide the controversy at this time.

Smith et al. (16) reported on an 8 year experience of thoracoscopy in pediatric oncology patients including resection of isolated lesions in patients with osteosarcoma. In this relatively large series from one institution there is no difference in recurrence or survival in this group of patients. However, further follow-up and review will be required before any meaningful conclusion can be reached. What is clear in all of these studies is that the procedure has an excellent diagnostic yield and much lower morbidity than the traditional open lung biopsy.

3. LOBECTOMY

There are relatively few reports in the pediatric surgical literature concerning more extensive resections such as lobectomy or segmentectomy. There are more reports in the adult thoracic literature and the results are generally favorable. These procedures are more technically difficult and require that the surgeon have very advanced dissecting and suturing skills. Therefore the results may differ considerably, based on the experience of the surgeon.

Many of these procedures have been performed using a combination of thoracoscopic ports and a mini-thoracotomy. However, with the development of improved instrumentation and energy sources for vessel ligation and tissue sealing, many can now be performed completely endoscopically.

The early reports, primarily in adult lung cancer patients, showed significantly longer operative times and conversion rates of 15–25% (17,18). However, the patients had a lower postoperative morbidity and a quicker recovery. Kirby et al. (19) compared video-assisted (VATS) lobectomy with a muscle-sparing thoracotomy and found significant advantages to the VATS approach. There was a significantly less postoperative pain and hospital stays where decreased on the average by 20%. McKenna et al. (20) showed similar results in patients undergoing lobectomy and mediastinal lymph node dissection. More recent studies have also suggested VATS lobectomy is also associated with a significantly reduced postoperative release of cytokines compared to a traditional open approach (21). This maybe a factor in the decreased morbidity associated with this group of patients. These studies all suggest that VATS lobectomy is preferable to open thoracotomy in selected adult patients with primary lung cancer.

The majority of pediatric patients requiring lobectomy are not oncology patients but instead have congenital lung lesions or infectious complications, such as severe

bronchiectasis. The issues in pediatric patients are more variable because of the varied etiology of the pathology and the size of the patients. Many of the instruments that were critical in the successful development of VATS lung resections in adults, such as the endoscopic stapler, have no role in smaller infants and children, since the stapler's cartridge often spans the entire width of the infant's hemithorax. This has required the development of alternate techniques, as well as instruments, to provide hemostasis and tissue sealing. The Ligasure® (Valleylab; Boulder, CO) is a 5 mm curved bipolar sealing device that can seal vessels up to 7 mm in size and can also seal lung parenchyma, which is quite useful when there is an incomplete fissure.

The first reported large series ($n = 113$) of thoracoscopic pulmonary procedures in infants and children was by Rothenberg in 2000 (15). The vast majority of these were pulmonary biopsies and wedge resections. Lobectomies were performed with a hybrid approach of thoracoscopy and mini-thoracotomy. This was before the advent of the Ligasure®. In 2003, Rothenberg (22) and Albanese (23) independently reported relatively large series of completely thoracoscopic lobectomies, mainly for congenital lung lesions. The majority of these are lower lobe resections, which are technically less difficult. In Rothenberg's series of 45 patients, the mean operating time for the three-port procedure was 125 min. There was one conversion to an open procedure due to an intraoperative complication related to application of the endoGIA stapler. The mean hospital stay was 2.4 days. Albanese's series comprised 14 patients, all of whom had a prenatal diagnosis of cystic adenomatoid malformation (12) or sequestration (2), with a mean age of 6 months at operation. There were no conversions to the open procedure and no intraoperative complications. Follow-up ranged from 4 to 35 months. The results of these studies compare quite favorably with standard open technique. This does not take into account the long-term benefits of avoiding a thoracotomy early in life, which is known to be associated with a higher incidence of shoulder girdle weakness and scoliosis (24).

4. PNEUMOTHORAX

Pneumothorax is a relatively common problem in adults and children occurring in 8 per 100,000, and is one that is well suited to treatment by thoracoscopy. The etiology in children and adolescents is generally a small subpleural bleb that ruptures and causes the air leak. This may or may not be associated with more significant parenchymal disease such as cystic fibrosis, or in older patients, emphysema. Standard therapy has consisted of chest tube placement with some type of surgical intervention if the air leak persists for more than 5–7 days, or earlier intervention in cases of recurrence.

Procedures routinely employed have included various types of chemical or talc pleurodesis, mechanical abrasion, pleurectomy, and bleb resection. These types of interventions are well suited to a thoracoscopic approach. The endoscopic linear stapler allows for easy resection of apical blebs and in smaller patients pre-tied suture ligatures (Endo-loops) can be used to snare and ligate the bleb. Electrocautery and laser have also been used to obliterate smaller blebs. Pleural irritants are easily inserted via the endoscopic ports and apical pleurectomy can be performed without significant difficulty.

One of the first reports on the thoracoscopic treatment of spontaneous pneumothorax in adults was by Hazelrigg et al. (25). They performed stapled bleb resections and pleural abrasion with minimal morbidity and recurrence. There have since been numerous reports in adults with recurrence rates of 0–12% (26,27). There has been no mortality and the procedure is associated with the same decreased morbidity and recovery as other thoracoscopic procedures. The only randomized study between thoracotomy and thoracoscopy

was reported by Waller et al. (28). This study showed a VATS approach to be superior in patients with primary spontaneous pneumothoracies, but was associated with a higher recurrence in patients with secondary pneumothorax.

There are only a few reports dealing with this subject in the pediatric literature (29). The largest is by Rodgers et al. (30) and involves 27 cases over 12 years. There were no operative complications and there were two recurrences (7.4%), which were treated by repeat thoracoscopy. These studies suggest that thoracoscopic management of spontaneous pneumothorax is the preferable method of treatment.

The one question which has not been adequately answered is the timing of intervention. Most standard approaches involve the initial placement of a chest tube with surgical intervention if an air leak persists after a few days. However, chest tube placement in a child often involves significant sedation at times with general anesthesia. The question then arises, since thoracoscopy adds little morbidity, whether or not VATS should be used at the time of initial presentation in order to expedite the patient's care, shorten hospital stay, and decrease the need for a second surgical intervention.

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26
Minimal Access Surgery in the Management of Empyema

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1. INTRODUCTION

The principle of adequate closed drainage for pleural empyema has been a surgical axiom since the early 1900s. Sir William Osler identified the importance of pleural drainage for empyema, but ironically succumbed to inadequately drained pleural sepsis (1).

Many surgeons are now using video-assisted thoracoscopy to treat empyema. There are a number of studies documenting its safety and effectiveness in children, and evidence has accumulated that thoracoscopy can replace thoracotomy when empyema debridement is indicated in adults (2). Although it is being used more liberally, thoracoscopy remains controversial as the initial treatment for all children with empyema. This chapter will delineate the principles behind the management of children with empyema and review current evidence for the use of minimal access surgery in this disease.

2. PRINCIPLES OF EMPYEMA MANAGEMENT

Most empyemas in children arise as a complication of a parapneumonic pleural effusion, usually secondary to streptococcal or other bacterial forms of pneumonia. Others may

follow surgery or trauma or may be associated with underlying lung or gastrointestinal pathology.

Pneumonia with parapneumonic effusion is typically managed with appropriate intravenous antibiotics, physiotherapy, and oxygen as needed. Thoracentesis is indicated for a large, symptomatic or loculated effusion and can help to determine whether the fluid is a transudate or an exudate. An exudative effusion, with or without identifiable bacteria, is equivalent to an empyema, "pus in the pleural space." An exudative effusion has low glucose (<2 mmol/L), low pH (<7.2), and high LDH (>1000 IU/L), and the presence of leukocytes (3). Bacteria, usually gram-positive cocci, may be identified on Gram stain or by culture.

The natural history of undrained empyema can be classified into three stages: 1—exudative, 2—fibrinopurulent, and 3—organizing (4). The transition from exudative to fibrinopurulent empyema may occur within a few days, whereas the organizing phase may persist for weeks. When undrained, the empyema either resolves or can lead to chronic complications such as sepsis, bronchopleural fistula, and erosion into neighbouring vital structures or through the chest wall. Death from empyema is rare in the antibiotic era.

Early and adequate drainage of empyema allows lung re-expansion, promotes normal pulmonary function, restores normal mobility of the chest wall and diaphragm, reduces the length of illness and hospital stay, and minimizes the associated morbidity of treatment (5). Traditionally, this has been done with thoracentesis or thoracostomy tube.

The presence of loculations predicts a poorer prognosis with chest tube drainage alone (6). Ultrasound and CT scan may be useful in identifying loculations and pleural thickening (Figs. 26.1 and 26.2), although the CT scan does not reliably differentiate empyema from transudative effusion (7).

When fever and loculated empyema persist in spite of chest tube drainage, the current options for further treatment are (a) intrapleural instillation of fibrinolytics, (b) thoracotomy with pleural debridement or decortication, or (c) video-assisted pleural debridement.



Figure 26.1 Ultrasound of loculated pleural effusion.



Figure 26.2 CT scan confirming loculated empyema.

3. FIBRINOLYTIC THERAPY

Pleural instillation of fibrinolytics, such as urokinase and streptokinase, has been used to either forestall or prevent the need for decortication, or as an adjunct to surgical debridement or decortication in children (8–10). A Cochrane review found three randomized controlled studies of intrapleural fibrinolytics for empyema in adults (11). The pooled data found small benefits with fibrinolytics, but insufficient evidence to support their routine use. The risks of allergic reactions may outweigh the potential benefits of streptokinase, and urokinase is currently unavailable because of concerns about its safety. In some centers, TPA has now become the fibrinolytic of choice (JC. Langer, personal communication).

4. THE EVOLUTION OF SURGICAL APPROACHES TO EMPYEMA DRAINAGE

Prior to the advent of antibiotics, the major innovation in empyema treatment was the introduction of closed pleural drainage, which decreased the mortality rate from 60% to 10% (12). Since then, chest tubes connected to closed drainage systems have become the standard of care. Open drainage is still preferred by some physicians for longstanding chronic empyema when the lung is no longer at risk of collapse and may allow outpatient follow-up.

Empyema patients with chest tubes who have persistent fever, toxicity, and inaccessible undrained or loculated empyema over many weeks have been traditionally treated with surgical decortication and pleurectomy to evacuate the empyema and remove its "peel" (5,13,14). This is usually accomplished through a standard posterolateral thoracotomy and may require rib resection for adequate exposure. Open decortication is bloody, and transfusion is frequently required. Because of the high accompanying morbidity, the operation may be delayed and prolong hospitalization hoping that the empyema will resolve without surgery. One review of 70 children with stage 2 or 3

empyema found that 70% required surgical intervention and that a delay in surgery was associated with more procedures, more radiographs, and an increased length of stay in hospital (15).

Proponents of early aggressive surgical drainage have decreased the postoperative morbidity of thoracotomy by using a muscle-sparing thoracotomy or limited mini-thoracotomy for pleural debridement. This approach depends on accurate localization of the empyema and the blind educated digit to clean out the cavity (16). The success and lower morbidity of early mini-thoracotomy led to its wider acceptance. There is evidence from two retrospective studies using historical controls that this approach has decreased the overall morbidity by allowing earlier recovery and averting subsequent full decortications (17,18).

5. VIDEO-ASSISTED THORACOSCOPIC DEBRIDEMENT FOR EMPYEMA

Pleuroscopy was invented early in the 20th century, but had to await improvements in optical telescopes to become more widely utilized. Dr. Stephen Gans led the development of the Hopkins rod-lens telescope in the early 1970s and was the first to apply this technology to visualization of intrathoracic pathology in children (19). The addition of video-enhancement technology in the 1980s made thoracoscopy more practical and it is now widely available.

The procedure is done in the operating room with the patient intubated under general anesthetic. The child is placed in the lateral decubitus position with the affected hemithorax up. Single lung ventilation will often not be tolerated or necessary, and positive pressure pleural insufflation is generally not required. The initial port is placed at the anterior axillary line through the fifth or sixth interspace, using blunt technique. If a chest tube has previously been placed, the chest tube site can be used for the initial camera port. It is useful to make the initial opening into the pleural space large enough to accommodate the surgeon's finger to facilitate exposure and debridement. A flexible port may be used, through which the angled telescope is inserted. The consolidated lung may not collapse, and visualization can be awkward. Once some initial adhesions are cleared, one or two more 5 mm ports can be inserted under direct vision. The process of breaking down loculations and debridement of fibrinous exudate is facilitated by blunt thoracoscopic graspers, suction/irrigator, and ovum forceps (Figs. 26.3–26.6). This avoids some of the practical difficulties of using only 5 mm instruments. After removing as much of the empyema as possible, the pleural space should be examined especially laterally and inferiorly to ensure that all loculated fluid pockets are opened and drained and the pleural space is irrigated generously with saline. One or two chest tubes are placed dependently, and the lung is re-expanded. The procedure usually takes at least an hour or more depending on the amount of pleural disease. The chest tube is removed usually within 4–5 days (Fig. 26.7).

The potential complications of thoracoscopy are the same as for thoracotomy, with bleeding and bronchopleural fistula reported complicating pediatric video-assisted thoracoscopic debridement (VATD) (20).

5.1. Safety and Effectiveness of VATD

Anecdotal reports of the use of thoracoscopy to treat empyema were published in the discussion of Raffensperger's 1992 paper (16). The first pediatric case series published in 1992 by Kern and Rogers reviewed a 10-year successful experience of treating nine children with



Figure 26.3 Video-thoracoscopy of pleural loculations.

empyema using thoracoscopy (21). Other pediatric centers quickly followed with reports of small case series demonstrating the safety and effectiveness of VATD for empyema (22–27).

One dissenting report documented an unsuccessful experience using VATD for empyema in seven children, five of who required a thoracotomy (28). The reasons for VATD failure included thick loculations, difficulty aspirating through a 5 mm suction,

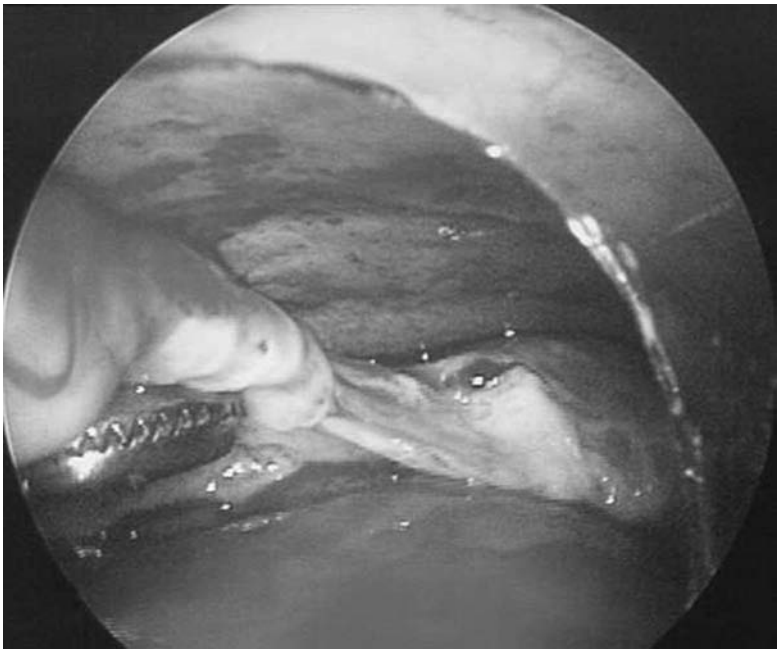


Figure 26.4 Fibrinopurulent stage of empyema.

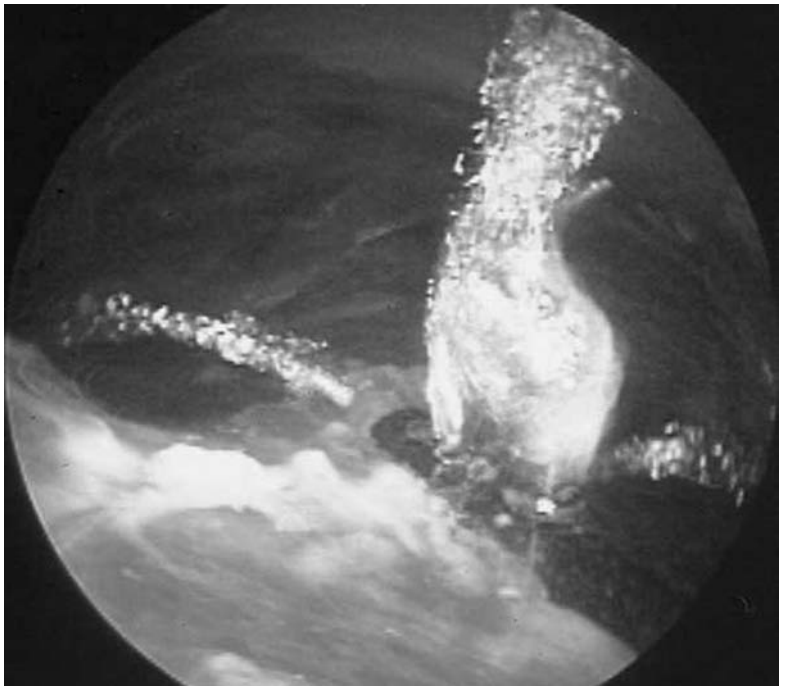


Figure 26.5 Pleural irrigation under video-thoracoscopic guidance.

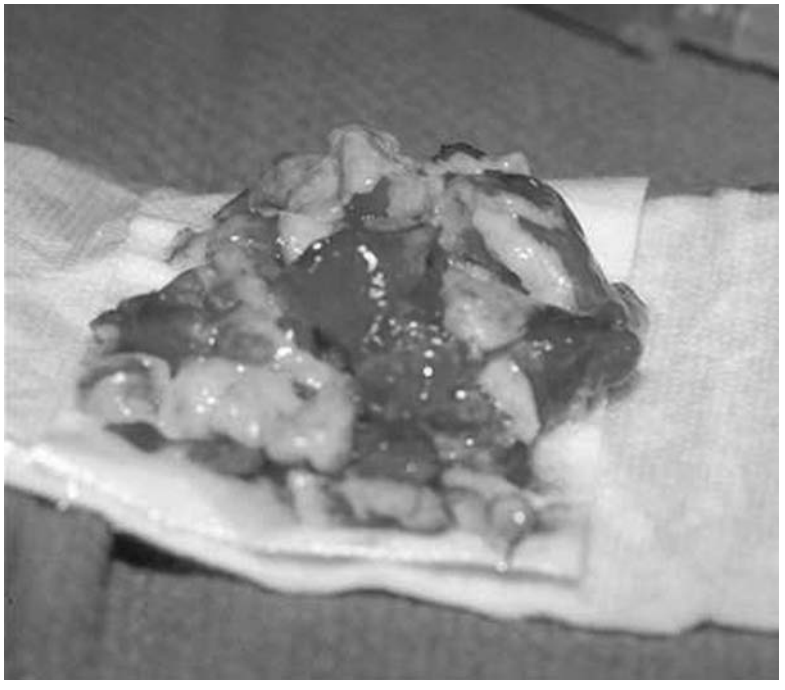


Figure 26.6 Gelatinous fibrinopurulent pleural debris removed by VATD.



Figure 26.7 Minimal scarring 1 month after VATD.

bleeding, stiff pneumonic lung preventing access, and thick hard pleura. Some of these difficulties may have been because of advanced disease and the learning curve of the procedure.

Variations in the reported success rates with VATD can be attributed to inconsistency in definitions and indications for surgery. Some reports refer to "decortication" (removal of the visceral pleura with the empyema peel of an organizing empyema) when the described procedure was in fact pleural debridement of the empyema at an earlier fibrinopurulent stage. Few studies record the duration of symptoms and effusion prior to intervention, but most agree that surgery is technically easier during the fibrinopurulent stage and that VATD may not even be indicated for the organized stage.

5.2. Comparison of VATD with Thoracotomy for Empyema

Prior to the advent of thoracoscopy, the high failure rate of chest tube drainage for stage 2 fibrinopurulent empyema prompted some surgeons to advocate earlier thoracotomy for nonresolving empyema. Video-assisted thoracoscopy allowing direct inspection of the pleural space, and visualization of surgical maneuvers has quickly supplanted thoracotomy for empyema without the evidence of randomized prospective trials. A few studies have used historical controls to compare VATD with limited thoracotomy for patients with fibrinopurulent empyema.

One report compared 22 children with empyema treated by VATD with 17 historical controls treated by thoracotomy (20). The VATD group was also treated at an earlier stage of disease. The VATD group had significantly fewer blood transfusions, shorter duration of postoperative fever, shorter duration of postoperative chest tube drainage, lower amount of postoperative analgesia, and shorter postoperative length of hospital stay. Two

VATD patients were converted to open thoracotomy to allow lung resection for parenchymal necrosis and bronchopleural fistula.

5.3. VATD as First-Line Treatment for Empyema

The overall rate of operative treatment is obviously greater when VATD is used liberally, which has raised concern that some of the operations might not be necessary since many empyemas ultimately resolve. With the establishment of its safety and effectiveness, VATD has evolved from being a delayed second-line treatment used only after traditional chest tube drainage has failed to being a primary first-line treatment used at the time the empyema is diagnosed.

Because the stage of empyema and presence of loculations may be difficult to determine at the outset, some centers now advocate early VATD at the time of the initial chest tube placement (29–31). Placement of the initial chest tube under general anesthetic is better tolerated in an anxious child and allows VATD to be done at the same time.

One prospective randomized controlled study of stage 2 fibrinopurulent empyema in 20 adults compared primary VATD with chest tube drainage and intrapleural fibrinolytics (32). Primary VATD was associated with significantly greater efficacy and shorter hospital duration. However, all fibrinolytic treatment failures were salvaged with VATD.

Others have found that not all empyemas that fail chest tube and fibrinolytic treatment can be cured with VATD. A large series compared 41 children with parapneumonic effusions treated by primary VATD with 98 historical controls treated with primary chest tube drainage (29). No child in the primary VATD group required a thoracotomy, and the duration of chest tube drainage and hospital stay were significantly shorter in the VATD group than in the historical controls. However, 38/98 children whose empyema did not resolve with chest tube drainage required delayed VATD and three of these ultimately needed a thoracotomy.

If VATD is used to salvage chest tube failures, it is important to intervene early. One series of 21 children with empyema treated by VATD concluded that VATD was most likely to succeed when undertaken within a week of diagnosis of an exudative effusion (33). There is no convincing evidence that undertaking VATD at the time of the initial chest tube placement for a parapneumonic effusion has any advantage over waiting several days to see whether the chest tube alone leads to rapid clinical resolution.

Potential disadvantages of undertaking routine early VATD are the extra-anesthetic morbidity and operating room procedure costs. One series looked at the costs incurred with VATD and found that the overall hospital charges with primary VATD were less than the charges when VATD was reserved for failures of conventional chest tube drainage (34). The initial costs of a protocol in which VATD is used liberally for early empyema are greater, but are offset by shorter hospital stays and fewer thoracotomies.

The accumulating evidence has led one institution to introduce a critical pathway that selects children for VATD based on either early identification of loculated pleural fluid by ultrasound or failure of chest tube drainage. This protocol has led to an average hospital stay of only 4–5 days and decreased cost when compared with historical controls and a national children's hospital database (35).

6. CONCLUSIONS

VATD is a safe and effective modality to treat fibrinopurulent empyema. The advantages over thoracotomy include smaller incisions, less pain, and less interference with

pulmonary mechanics. It is most likely to be successful when undertaken early in the disease process within a week of the diagnosis. Early VATD can decrease the likelihood of open decortication and its attendant morbidity of thoracotomy, transfusion, and prolonged hospitalization. Although early VATD appears to be more effective than intrapleural fibrinolytics in treating fibrinopurulent empyema, there is some evidence that routine VATD at the time of initial chest tube insertion for loculated parapneumonic effusion shortens hospital stay and is cost effective. Controlled studies looking at the role of newer fibrinolytics and the morbidity and cost-benefit of routine primary use of VATD for pediatric empyema are needed.

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27
Mediastinum, Esophagus, and Diaphragm

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The application of thoracoscopic techniques in the treatment of various lung diseases has already been discussed. The same advances which have allowed surgeons to approach the most complex lung pathology using minimal access surgery (MAS) techniques has also expanded the indications in other intrathoracic lesions involving the mediastinum and esophagus. Standard approaches to the mediastinum and esophagus have included mediastinoscopy, anterior thoracotomy, sternotomy, and posterolateral thoracotomy. Each technique has its advantages and limitations but all can be associated with significant morbidity and recovery. The least invasive approach to the anterior mediastinum, mediastinoscopy, is severely limited by the degree of access and control it allows the surgeon, and the visual field is extremely confined. This procedure allows the surgeon to perform only limited evaluations and biopsies of the paratracheal space and is a difficult technique to master. Anterior thoracotomy provides greater access but the visual field is still significantly limited. The greatest advantage of these approaches is that they allow the surgeon to stay extrapleural. However, if bleeding or other problems are encountered the surgeon has little choice but to hope the bleeding tamponades or to perform a sternotomy. Posterolateral thoracotomy and sternotomy greatly improve the surgeons' access and visualization but both have significant morbidity and recovery.

With minimal morbidity (1), thoracoscopy provides much better visualization and access to the anterior and posterior mediastinum than the aforementioned open approaches. Through three or four small ports, the surgeon can access anywhere from the thoracic inlet to the diaphragm with excellent visualization. This allows for critical evaluation of mediastinal structure as well as diagnostic or therapeutic procedures. The theoretical disadvantage of a transpleural approach seems to far outway the disadvantages of the limited access provided by other techniques.

There are now numerous reports in the adult and pediatric literature that support the use of thoracoscopy in the evaluation and treatment of anterior mediastinal structures. Early reports in the adult literature (2–4) documented the safety and efficacy of this approach for diagnosis and treatment of solid and cystic lesions. The sensitivity in diagnosing lymphoma, teratoma, and other malignancies approached 100% with little associated morbidity. The greater visual field also aided in the staging of primary lung malignancies because the entire hilum as well as the paratracheal area can be accessed. Gosset et al. (5) compared thoracoscopy with mediastinoscopy for biopsy of mediastinal tumors and found thoracoscopy to

be more accurate with no greater morbidity. Later reports documented success with more aggressive therapeutic procedures such as resection of thymic cysts and tumors, thyroid lesions, teratomas, and other solid tumors (6). The conversion rate to sternotomy has been documented at 0% to 20% and appears to be associated in some degree to the experience of the surgeon. There have been no documented deaths, and the same benefits of decreased recovery and pain as seen with other MAS procedures. A similar experience has been documented with posterior mediastinal tumors, primarily neurogenic, in adults. Reardon et al. (7) and Liu et al. (8) have reported large series with successful resection of ganglioneuromas with few complications and no recurrences reported to date. In one of the few comparative studies of open vs. a thoracoscopic approach for mediastinal masses, Bousamra et al. (9) reviewed the management of benign neurogenic tumors. He found that operative times were moderately longer in the thoracoscopy group (171 vs. 112 min) but hospital stay (2.6 vs. 4.5 days) and return to work (4.3 vs. 7.7 weeks) were significantly shorter. Often the biggest challenge in dealing with these lesions is their size and extracting them after resection. Generally, these lesions can be placed in a sac and brought out piecemeal through an enlarged trocar site without compromising the integrity of the procedure or the pathologist's ability to accurately analyze them.

A similar experience has been documented in the pediatric population. Lesions most appropriate for primary resection in the anterior mediastinum include teratomas, thymic masses, and aberrant thyroid tissue. In the posterior mediastinum foregut duplications, ganglioneuromas and neuroblastomas are the masses most commonly encountered. These are generally amenable to complete resection or biopsy, when indicated. The first report of thoracoscopic evaluation of these lesions was by Rodgers et al. (10) in the early 1980s. Even with very rudimentary equipment he was able to approach 100% specificity and sensitivity in obtaining adequate tissue for diagnosis. He also noted that more extensive disease than expected was found in 45% of patients, supporting the use of thoracoscopy over mediastinoscopy or mini-thoracotomy. After this there were only a few case reports and limited series until the late 1990s and 2000. Partrick and Rothenberg (11) have reported the single largest series of mediastinal masses treated by thoracoscopy in children. This study included 39 anterior and posterior mediastinal masses. Ninety-seven percent of the procedures were completed successfully and diagnosis was obtained in 100%. Postoperative morbidity and recovery were favorable when compared with open thoracotomy. Sandoval and Stringel (12) and Moffat et al. (13) documented similar results in a smaller series of primarily diagnostic thoracoscopies. The sensitivity was 100% and there were two conversions to an open procedure

There are two large studies (14,15) documenting the use of thoracoscopy in pediatric oncology patients. Both report 100% diagnostic accuracy with thoracoscopic biopsy of anterior and posterior mediastinal tumors with no operative complications. Posterior tumors, especially ganglioneuromas, were easily excised with little morbidity and short hospital stays and recovery periods. These reports suggest that thoracoscopy should be the procedure of choice in children with mediastinal disease. However, because thoracoscopy requires at least partial collapse of the lung on the ipsilateral side, attempted resection of a giant anterior tumor with evidence of respiratory compromise, is a contraindication to this approach.

Case reports and small series over the last decade suggest that bronchogenic cysts and esophageal duplications are particularly suited for thoracoscopic resection. In general, these lesions may share a common wall with the native esophagus or trachea, but rarely there is a communication between the two structures. Resections can be achieved bloodlessly without the need for complicated reconstruction. Several reports (11,16,17) demonstrated that these lesions can be resected with minimal morbidity

(<5%), reasonable operative times (<2 h), and shorter hospital stays (range 1–2 days). The only significant complication reported was the recurrence of a bronchogenic cyst that was incompletely excised (17).

The other disease worth separate mention is total thymectomy for myasthenia gravis. Thoracoscopy provides superior access and visualization compared with the open cervical approach, and avoids the morbidity of a sternotomy in a patient with weakened respiratory muscles. One of the initial reports in adults by Mack et al. (18) confirmed the benefits of this approach exhibiting minimal complications. Thirty-three consecutive thoracoscopic thymectomies were performed with only one conversion to open. The mean hospital stay was 3 days and there was clinical improvement in 88%. In children, Kogut et al. (19) reported the largest series of thoracoscopic thymectomies for myasthenia gravis. In 10 patients, there were no intra- or postoperative complications, and the mean operating time was 114 min. All patients were extubated at the end of the procedure and discharged on the first postoperative day. After an average of 10 months follow-up, all patients were off their medications and symptom-free. The one serious complication of this operation is injury to the phrenic nerve. It is easily visualized on the side ipsilateral to the thoracoscope (left) but not on the contralateral side. The surgeon must take care to avoid either direct or indirect injury to this structure during the dissection. Phrenic nerve injury has also been reported following the open transcervical and median-sternotomy techniques, so the same care must be exerted in all approaches.

In addition to foregut duplications, there are two other esophageal lesions in children that have been dealt with using MAS. This includes achalasia and esophageal atresia. The debate as to whether an abdominal approach or a thoracic approach for achalasia is a long standing one and the application of MAS has not changed that. Both laparoscopy and thoracoscopy provide excellent access to the lower third of the esophagus and the myotomy can be performed without significant difficulty. The most important technical point is to extend the myotomy adequately through the gastroesophageal junction in order to prevent an incomplete myotomy. The real point of controversy is whether an antireflux procedure needs to be performed at the same time as the myotomy. Reports by Patti et al. (20) suggest that the incidence of reflux postmyotomy reaches 80%. Albanese and coworkers (21) and Rothenberg et al. (22) have reported similar findings in the pediatric population. The specifics of this are discussed in another chapter.

The last esophageal lesion common to pediatric surgery is the treatment of esophageal atresia. The application of MAS techniques to this disease process is still in its infancy and the number of patients treated and their outcomes are too small to derive definitive conclusions. The first report of a successful repair was of a pure esophageal atresia by Lobe et al. (23) in 2000. This child had a long gap, which had been dilated antegrade and retrograde for ~3 months. The procedure took 4 h and the patient had a stricture that eventually resolved with dilatation. The first tracheoesophageal fistula repair was reported the next year (24). Rothenberg (25) reported the first series of eight patients in 2001. The mean operating time was 90 min and there were no operative complications. One patient developed a leak that resolved spontaneously on postoperative day 8. Seven patients were started on feeds on postoperative day 5. Three patients required at least one anastomotic dilatation. In 2002, Bax and van der Zee (26) published a series of eight patients, all of whom had a tracheoesophageal fistula, and had an uncomplicated thoracoscopic operation in a mean of 198 min. There was one leak that closed without reoperation and four anastomotic strictures that required dilation. Feeds were begun a median of eight days after surgery. No long-term follow-up was reported. That same year, Martinez-Ferro et al. (27) reported their first consecutive nine patients who underwent primary thoracoscopic repair of esophageal atresia with tracheoesophageal

fistula. There were no intraoperative complications and no conversion to an open procedure; the mean operating time was 105 min. There were two leaks and three strictures at the anastomosis, all of which were successfully treated nonoperatively. Although the four reports do not provide firm conclusions, the results are promising. The greatest advantage is the avoidance of a posterolateral thoracotomy incision in infancy with its associated incidence of scoliosis, chest wall asymmetry, and shoulder girdle weakness (28). Clearly, further evaluation and follow-up will be necessary before definitive conclusions can be made.

The other structure amenable to intervention by thoracoscopy is the diaphragm. Evaluation of traumatic injury to the diaphragm using thoracoscopy was documented as early as the 1940s (29). In the early 1980s, Jones et al. (30) reported the first large series of 36 patients. The sensitivity of this approach is >95% with little morbidity. More interventional procedures to the diaphragm were documented in the mid-1990s. There were case reports of both congenital and traumatic diaphragmatic hernia repair using a thoracoscopic approach in children and adults (31,32). Using the laparoscope, Hendrickson et al. (33) reported 11 patients with congenital diaphragmatic hernia. Nine were infants and two were older children. There were eight Bochdalek and three Morgagni hernias. The mean operating time was 90 min and there were no conversions to an open procedure. Of note is that all 11 patients tolerated intra-abdominal insufflation and there were no complications referable to the insufflated gas being in both the hemithorax and the abdomen simultaneously. There was one recurrence in a neonate at 9 months of age. A European multicenter retrospective study (34) analyzed the outcome of 22 patients in whom a laparoscopic Morgagni hernia was repaired. In 18 of the 22 patients, a prosthetic patch was necessary. One iatrogenic small bowel injury was repaired during the procedure. There were no conversions to an open technique and the mean operating time was 94 min. The mean duration of follow-up was 13.5 months and in this time, one patient developed a recurrence and underwent successful laparoscopic reoperation. These initial results suggest that this approach is acceptable in patients who are not in severe respiratory distress.

MAS has been used for diaphragmatic plication. There are single case reports of a thoracoscopic approach with good results (35–37). Again the operative times are slightly prolonged but the recovery and hospitalization are shorter. Partrick and Rothenberg (38) reported laparoscopic placation of three ventilator dependant children. The average operative time was 40 min and there were no complications. The respiratory status improved in all three patients allowing extubation and follow-up chest radiograph at 1 month showed the repairs to be intact. The real debate is not so much whether an MAS approach is beneficial in the treatment of diseases of the diaphragm but which approach, laparoscopic or thoracoscopic, should be used. Most of the reports and the author's own experience would favor a laparoscopic approach especially when suturing is necessary. The rib cage provides a fixed point for the trocars and instruments, and can make fine motions, especially suturing, difficult. Also the angles can often be awkward. Abdominal insufflation provides a self-retaining retractor and gives excellent visualization of the entire diaphragm, even over the dome of the liver. The pliability of the abdominal wall also allows more flexibility for trocar placement and instrument movement. Whichever route the surgeon chooses, it would appear that the MAS approach to the diaphragm is beneficial to the patient.

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28
Bariatric Surgery

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1. INTRODUCTION

Adult bariatric surgical procedures on morbidly obese patients are on the verge of becoming the most commonly performed operations by general surgeons. An estimated 90,000 adults in the United States will undergo bariatric operations in 2003, which is twice as many those performed in 2002 and almost three times the number done in 2000 (1). Part of this increase in the volume of bariatric procedures is supply related in that obesity has become an epidemic affecting over 300 million adults across the world (2). Furthermore, bariatric surgery is currently the only successful long-term treatment for the severely obese patient. It has been well established that improved outcomes and favorable results in patients undergoing these procedures are documented by the resolution of multiple medical comorbidities of obesity following surgically induced weight loss. The introduction of minimally invasive surgical techniques to obesity surgery with reduction of wound complications and operative morbidity has been a major contributor to the logarithmic growth of bariatric surgery in the treatment of morbid obesity.

Since almost 50% of adults in the United States are considered overweight and 20% clinically obese (3), it is not surprising to learn that >15% of children and adolescents are overweight (4). The objectives of this section will be to review the general principles of bariatric surgery as they have been applied to morbidly obese adults and determine how these principles may apply to obese children or adolescents, as well as review the evidence for the efficacy of the application of bariatric procedures in adolescents at the present time.

2. PRINCIPLES OF BARIATRIC SURGERY

An extensive review of bariatric surgery is beyond the scope of this chapter; however, a review of the various operations and outcomes is necessary to understand the rationale for the application of these techniques in younger patients, specifically the adolescent patient.

Bariatric surgery has been validated as an effective strategy for long-term management of morbid obesity since the publication of the proceedings of the National Institute of Health Consensus Development Conference on gastrointestinal surgery for severe obesity in 1991 (5). In this statement, both gastric bypass and gastric restrictive procedures are included as viable options for motivated patients who have reasonable operative risk. Since that time, the medical literature has been replete with various reports from multiple institutions in an effort to provide evidence that one approach is superior to the other. The goal of bariatric surgery is to help patients lose their excess body weight and to prevent, control, or resolve any obesity associated comorbid conditions. The most common comorbidities associated with severe obesity are fairly well recognized and include type 2 diabetes mellitus (6), hypertension (3), obstructive sleep apnea (7), degenerative joint disease (8), and an overall decrease in life expectancy (9). The following paragraphs outline the recent available literature with respect to the effectiveness of both gastric by pass procedures and gastric restrictive procedures in providing sustained weight loss, correction of comorbid conditions, and overall success and complication rates.

2.1. Bypass Procedures

There are several gastric bypass procedures that can be performed that provide substantial weight loss; however, the most commonly performed procedure in the United States is the Roux-en-Y gastric bypass (10). All bypass procedures, including the biliary-pancreatic diversion with or without duodenal switch and the distal gastric bypass, stimulate weight loss, to some degree, by including malabsorption. Procedures other than the Roux-en-Y gastric bypass have a high rate of adverse nutritional sequelae and thus would not be applicable to the pediatric or adolescent population (11,12). Still, there are groups who contend that malabsorptive procedures can be performed safetly without long-term nutritional complications; however, these studies are in adult patients (13,14). Until this controversy is resolved, the conservative approach for adolescents would be the Roux-en-Y gastric bypass and thus the remainder of the discussion of bypass procedures will focus on this technique.

The Roux-en-Y gastric bypass has been established as a safe and effective means to achieve significant and sustained weight loss in the adult population (15). In 2003, two large series documented that laparoscopic Roux-en-Y gastric bypass results in approximately a 60–66% loss of excess body weight, 80–66% resolution of type 2 diabetes mellitus, and control of hypertension in nearly two-thirds of patients (16,17). The outcome are equally encouraging for the treatment of other comorbidities. The group at

Emory University reported their experience with both laparoscopic and open procedures in patients over 50 years of age and found that Roux-en-Y gastric bypass resulted in a marked reduction in the incidence of degenerative joint disease, gastro-esophageal reflux disease, and CPAP-dependent obstructive sleep apnea, in addition to a 66% loss of excess body weight and control of hypertension and diabetes (18).

2.2. Restrictive Procedures

Restrictive operations such as vertical-banded gastroplasty and laparoscopic adjustable gastric banding ("Lap-band") work by creating a very small gastric reservoir, without any malabsorptive component. Proponents of restrictive obesity procedures cite excellent results in the management of the complications of obesity. Groups from Australia (19,20) and Italy (21) have reported durable weight loss with resolution of hypertension and diabetes similar to that seen with Roux-en-Y gastric bypass using laparoscopic adjustable gastric banding.

Similarly, Israeli surgeons have reported that laparoscopic adjustable gastric banding can produce a 100% reduction in the incidence of obstructive sleep apnea (22). Whichever bariatric surgical option is undertaken, it is clear that comorbid conditions and overall quality of life can be markedly improved with substantial and sustained weight loss.

3. MALABSORPTIVE VS. RESTRICTIVE PROCEDURES

The debate regarding which surgical approach is preferable mostly focuses on geography. The majority of centers that advocate the adjustable gastric band are from overseas while the mainstay of bariatric surgery in the United States remains the Roux-en-Y gastric bypass (10). There are relatively by few studies that directly compare restrictive and bypass procedures. Sugerman et al. (23) published the landmark paper comparing Roux-en-Y gastric bypass with vertical-banded gastroplasty in 1987. This randomized prospective study was stopped early because patients with Roux-en-Y gastric bypass lost significantly more weight than those with gastroplasty. "Sweet-eaters" had the poorest response to gastroplasty and some have used these results to suggest that gastroplasty may not be useful in adolescents (11). Vertical-banded gastroplasty has basically been replaced by the adjustable gastric band, especially overseas. This technique has gained widespread popularity in Europe (24) and Australia (25) with favorable results. Until recently, the adjustable gastric band was not approved for use in the United States. Since its availability, most of the results in the U.S are not as encouraging as the foreign data and much of the enthusiasm for this technique has dampened (26,27). In fact, one center has converted many patients with the adjustable band to Roux-en-Y gastric bypass due to unfavorable results (28). Another comparison of laparoscopic adjustable gastric banding and laparoscopic gastric bypass revealed that patients after bypass had a higher excess weight loss but more early postoperative complications than patients after gastric banding (29). However, other centers have found that they could duplicate the foreign experience with the laparoscopic adjustable gastric band in the United States (30,31). There are some theoretical advantages of the adjustable gastric band over Roux-en-Y gastric bypass for adolescents with morbid obesity. The weight loss after adjustable gastric banding is somewhat more gradual when compared to Roux-en-Y gastric bypass (17,20), and the band could be loosened during times when nutritional needs are increased, such as pregnancy (32). Furthermore, placement of the adjustable

band would not preclude conversion to bypass (28). However, a prospective randomized study comparing the two techniques with long-term follow-up would be needed to definitively address the issue.

Both the Roux-en-Y gastric bypass and the adjustable band techniques may be associated with significant complications. The complication most commonly associated with the open Roux-en-Y gastric bypass had been wound infections that occur in up to 15% of patients (33). However, most bariatric surgeons now perform this procedure laparoscopically, which may decrease major complication rates. A meta-analysis of over 3400 laparoscopic cases compared to over 2700 open cases revealed that the laparoscopic procedure was associated with a decreased incidence in not only wound infections, but also iatrogenic splenectomy, incisional hernias, and mortality when compared to the open technique (34). However, there was an increased incidence of early and late bowel obstruction, gastrointestinal hemorrhage, and stomal stenosis in procedures performed laparoscopically. There was no difference in anastomotic leak rates that typically occur in about 2% of patients following either approach. The major complication associated with the laparoscopic approach appears to be internal hernia, which occurs in approximately 3% of patients (34,35). The presenting symptoms may be vague and the diagnosis must be suspected in all patients with abdominal pain after laparoscopic Roux-en-Y gastric bypass (35). It has been suggested that surgeon experience, sleep apnea, and hypertension may be predictors of complications after laparoscopic Roux-en-Y gastric bypass, and that a history of diabetes mellitus may be associated with poorer weight loss (36), although a much larger series would be needed to confirm these results.

In general, laparoscopic adjustable gastric banding has been associated with lower morbidity and mortality rates than gastric bypass procedures (25,37). The complications that occur are most often related to migration, erosion, or mechanical failure, which occur in 12.5%, 2.8%, and 3.6% of patients, respectively, in one prospective study (20). Pouch dilation can also occur (38). Most problems can be dealt with using a laparoscopic approach by either re-banding, removal of the band, conversion to bypass operation, or revision of the access port (37,39,40). The morbidity associated with reoperation was $\sim$5%. Some authors report that the placement of a larger band (11 cm) may prevent the majority of complications (39,41).

4. PEDIATRIC OBESITY DEFINED

Body mass index (BMI), designated as kilograms per meters squared (kg per m^2), is a relatively simple means to define overweight in adults who have attained full growth. An adult with a BMI $\geq 30\ kg/m^2$ is considered obese. In children and adolescents who are still growing, there is no definitive BMI that can be used as a marker for obesity. Thus, the application of growth charts and multiple percentiles are necessary to determine overweight and obesity for age and sex in this group (42). Pediatric obesity has been defined as a BMI greater than the 95th percentile for age and sex. Currently, this constitutes approximately 10% of all children and adolescents with another 10% falling into the category of overweight, or at risk for overweight, with a BMI $>$ 85th percentile (43,44).

The health implications of obesity in adults are well known to be increased risk of cardiovascular disease (especially hypertension), dyslipidemia, diabetes mellitus, gall bladder disease, increased prevalence's and mortality ratios of selected types of cancer, and socioeconomic and psychosocial impairment (45). This leads to the question of what are the medical consequences of obesity in children as well as the persistence of obesity into adulthood. In fact, obese children have an overwhelming chance of carrying

their obesity into adulthood. Studies report that on average 50–77% of obese children become obese adults, with this percentage increasing to >80% if only one parent is obese (46–49).

The magnitude of the adverse health consequences of obesity in adults is underscored by multiple studies that demonstrate an increased incidence of morbidity, mortality, and specifically early death in obese adults (50–52). Similar evidence is reported in a longitudinal study in adolescents by Must et al. who found that overweight increased all-cause mortality by 1.8-fold (53). Furthermore, deaths from coronary artery disease, cerebrovascular disease, and colon cancer were all increased in adults who were obese adolescents. The incidence of premature disease in obese adolescents is shown to be increased over the baseline population with its effects spread over many organ systems (54). Risk factors for atherosclerosis and coronary artery disease coexist in obese adolescents with hyperlipidemia also being more common in this group (55,56). Almost 60% of obese children in the Bogalusa Heart Study had one risk factor for cardiovascular disease, with 20% having two or more risk factors (57).

Glucose introlerance is a frequent consequence of adult obesity as manifested by noninsulin-dependent diabetes mellitus (NIDDM). In one pediatric center, an increasing percentage of individuals with newly diagnosed NIDDM were obese (58). Certainly, the adverse health consequences of glucose intolerance and diabetes are well known and this trend may signify the effects of the increased prevalence of pediatric obesity in general.

Other health problems associated with obesity include sleep disorders and obstructive sleep apnea syndrome. There are data to suggest that children with obstructive sleep apnea exhibit adverse cardiac profiles such as left ventricular hypertrophy and abnormal ventricular dimension related to the syndrome (59). Sleep deprivation and excessive daytime sleepiness have been noted to be more common in obese children, and learning disabilities may be associated with disordered sleep patterns in these children (60,61). It may follow that correction of these problems may improve school performance.

Skeletal disorders related to obesity are related to the inability of the bone and cartilaginous structures to withstand the pressures of excess weight. In Blount's disease, which is characterized by abnormal bowing of the tibia and the resultant overgrowth of the medial aspect of the proximal tibial metaphysis in children, over two-thirds of afflicted children are obese (62). Slipped capital femoral epiphysis is also seen in obese children related to the effects of increased body weight on the cartilaginous growth plate of the hip. Up to 50% of children with slipped capital femoral epiphysis are overweight and risk of recurrence is common if weight loss is not achieved (63).

Hypertension, which is less frequently found in children overall, occurs at ninefold increased rate in the obese (64). Pseudotumor cerebri is a rare childhood disorder associated with increased intracranial pressure and presents with headaches. As may as 50% of children with this disorder are obese; however, the relationship between obesity and symptom onset is unclear (65). Up to 30% of women with hyperandrogenism and polycystic ovary disease are obese (66). Nonalcoholic fatty liver disease and steatohepatitis are seen to occur more frequently in obese children and adolescents (67). Finally, the risks of certain cancers, in particular gynecologic malignancies, have been associated with obesity in adolescents (68–70).

Psychosocial and quality of life issues are among the most prevalent in obese adolescents. The patterns of discrimination against obese children are established early in life and become ingrained in a culture in which thinness is admired (54,71). Although young children do not exhibit negative self-esteem or low self-image (72), adolescents develop a negative self-concept that may persist into adulthood (73). Moreover, obese individuals report that their weight has a negative impact upon several aspects of their daily lives

including physical functions, self-esteem, sexual function, and work life (74). Wang and Dietz (75) determined that over the past decade (1979–1999), the cost of healthcare for children between 6 and 17 years of age with obesity-related diagnoses had tripled from $35 million to $127 million. They attributed increased incidences of diabetes, complications from gall bladder disease, and obstructive sleep apnea as responsible for the overall increase.

5. HISTORY OF BARIATRIC SURGERY IN ADOLESCENTS

The initiative of bariatric surgery in adolescents is a relatively new one, and thus one must extrapolate results from the adult practice in an effort to determine its appropriate use. However, experience with bariatric surgery is starting to mount in the pediatric population. Breaux (76) published his results with bariatric surgery in 22 children aged 8–18 in 1995. He performed open vertical-banded gastroplasty in 5 children, open Roux-en-Y gastric bypass in 14 children, and open biliary-pancreatic diversion in 4 children. Substantial weight loss was found in each group. He concluded that bariatric surgery was safe and effective in the pediatric population, although he did report two late deaths. Sugerman in Sugerman et al. (77) reviewed his 20-years experience with bariatric surgery in adolescents, which comprises the largest study. Standard Roux-en-Y gastric bypass and long-limb gastric bypass were the most commonly performed procedures; few of these procedures were approached via the laparoscopic technique. Sugerman found that results in the adolescent population were similar to those in adults, including sustained weight loss, correction of comorbidities, and improved self-image. Garcia et al. (11) currently have the largest recent series of adolescents who have undergone laparoscopic Roux-en-Y gastric bypass. Their results have also been favorable, but long-term follow-up is pending.

Recently, two groups from overseas have reported their results with laparoscopic adjustable gastric banding. Dolan et al. (19) from Australia reported 17 adolescent patients who lost a median of 59% of their excess body weight. Only two complications were encountered: a slipped band and a leaking port after 2 years of follow-up. Similarly, Abu-Abeid et al. (22) from Israel reported their experience with 11 adolescent patients and laparoscopic adjustable gastric banding. They reported a drop in mean body mass index of 14 kg/m^2 and no complications with a mean follow-up of 2 years.

The evidence suggests that bariatric surgery is safe and effective adults, and the modest data available in the adolescent population concurs. Most of the experience in the United States is with Roux-en-Y gastric bypass, either via open or laparoscopic approaches. Thus, it must still be considered the gold standard for bariatric surgery in adolescents. However, foreign data suggests that the laparoscopic adjustable gastric band may be applicable for pediatric patients. The theoretical advantages of more gradual weight loss and the ability to ease gastric restriction during periods of increased nutritional needs are attractive for adolescent bariatric surgery. Unfortunately, the device is not currently approved for use in patients less than 18 year of age. Thus, no prospective trial can be conducted to compare gastric banding and bypass to evaluate which technique is preferable in the pediatric population. It would be an important trial to pursue if gastric band use approval is secured.

6. RATIONALE FOR BARIATRIC SURGERY IN ADOLESCENTS

The rationale for performing bariatric procedures on adolescents is to prevent or alter the pattern of adverse health consequences and early death shown in this group who become

Table 28.1 Indications and Contraindications for Bariatric Surgery in Adolescence

Indications
Failure of at least six months of organized, medically supervised weight loss attempts; and
Attained or nearly attained physiologic maturity (unless comorbidity is extreme); and
Severe obesity (BMI $\geq$ 40) with significant obesity-related comorbidity or BMI $\geq$ 50 with less severe comorbidities; and
Exhibit commitment to comprehensive medical and psychological evaluation, both before and after surgery, and agree to avoid pregnancy for at least one year postop; and
Be capable and compliant with postoperative nutritional guidelines; and
Provide informed assent to surgical management.
Contraindications
Substance abuse within the preceding year; or
Psychiatric diagnosis that would impair ability to adhere to postoperative dietary or medication regimen (e.g., Psychosis); or
Medically correctable cause of obesity; or
Inability or unwillingness of patient or parent to fully comprehend the surgical procedure and its medical consequences; or
Inability or refusal to participate in lifelong medical surveillance.

obese adults. If the observed health benefits of bariatric surgery for adults, with respect to the elimination of obesity-related comorbidities and maintenance of excess weight loss, can be realized in the adolescent population, then the application of bariatric surgery for select adolescents would seem reasonable. Guidelines and recommendations for offering bariatric surgery to adolescents have been proposed by Inge in conjunction with experts in the field of pediatric obesity and pediatric surgery (78). Adolescents being considered for bariatric surgery would ideally meet the inclusion or exclusion criteria depicted in Table 28.1.

7. RECOMMENDED COMPONENTS OF A MULTIDISCIPLINARY TEAM

As part of a comprehensive pediatric weight management program, with one component being dedicated to bariatric surgery, several other specialists are required to meet the unique needs of adolescents. Physicians in adolescent medicine having experience with obesity evaluation and management, as well as experts in the areas of adolescent psychology, nutrition, and exercise physiology should be involved with patient assessment. Other specialists would be utilized depending upon an individuals needs, but may include pediatric experts in endocrinology, pulmonology, gastroenterology, cardiology, anesthesiology, and orthopedics.

Since adolescence represents a significant period of substantial growth and maturation, both physically and emotionally, special attention to developmental issues in adolescents is critical when considering bariatric procedures that will have marked impact upon future growth and development. Attainment of $>$95% of linear growth (or adult stature) may be a reasonable goal at which bariatric surgery may be considered depending upon individual characteristics. Based upon peak height velocity measurements in normal weight girls (8–9 cm/year) and boys (9–10 cm/year), girls should achieve $>$95% linear growth by 13 years of age, and boys by 15 years of age (79). The onset of menarche has also been a useful marker for the completion of linear growth in girls. If any doubt exists as

to whether linear growth is achieved, determination of bone age by plain X-ray of the wrist is recommended to assess physiologic maturation. The importance of a committed pediatric psychologist and continued postprocedural follow-up cannot be overstated. As a corollary to physiologic growth, the adolescent has rapidly developing psychological profile with varying ability to understand health information and behavior patterns related to them.

Historically, this age group has demonstrated poor compliance (<50%) to medical treatment regimens and follow-up when groups with chronic illnesses were studied (80,81). Rand and Macgregor (82) reported poor compliance by adolescents following gastric bypass surgery with <15% of adolescents demonstrating compliance to postoperative dietary multivitamin and nutrient supplementation. However, several studies suggest that adolescent adherence to strict medical and dietary regimens can be improved with the application of behavioral therapy (83–85). Therefore, the continued support of the behavioral psychologist is essential for success in dietary compliance and long-term follow-up after bariatric procedures in adolescents.

8. SUMMARY

Surgical approaches for morbidly obese adolescents may be appropriate for individuals who have serious obesity-related health risks and have been unsuccessful in achieving sustained weight loss following multiple, medically supervised attempts. Individuals should be considered based upon the severity of comorbid conditions, physiological and emotional maturity level, and other relevant supportive data. The laparoscopic Roux-en-Y gastric bypass is currently considered the procedure of choice for morbidly obese individuals; however, further study is required to determine which procedure may be the most efficacious when applied to the adolescent population. There may be a significant role for laparoscopic gastric banding or gastric restrictive procedures in younger patients. The importance of family support and individual commitment to permanent lifestyle change following bariatric surgical procedure cannot be overemphasized.

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29

A Miniature Access Approach to Pectus Excavatum

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1. INTRODUCTION

Pectus excavatum (PE), also known as funnel chest or trichterbrust, is by far the most common disorder of chest wall formation. Approximately 90% of patients with chest wall disorders have PEA the incidence is 1 in 300 live births (1). Pectus carinatum (PC), the next most common chest wall disorder, is seen in only 7% of patients with chest wall deformities. A great deal of controversy exists as to the indications, timing, and method of repair of PE. Recently, Nuss et al. (2) have introduced a novel and minimally invasive method of repair. Their technique has intrigued pediatric surgeons and provoked further discussion amongst the pediatric surgical community as to the optimal method of repair. This chapter reviews the available literature regarding the efficacy and safety of this miniature access approach compared to more conventional open repairs, that is, the Ravitch technique.

2. EMBRYOLOGY/ETIOLOGY

Multiple theories exist as to the cause of PE; however, the etiology remains obscure. The development of PE may be a result of overgrowth of costal cartilage, displacing the sternum posteriorly. Abnormalities of the diaphragm, rickets, or elevated intrauterine pressure have also been theorized to cause posterior displacement of the sternum (3–8). This is supported by reports of the coexistence of PE with diaphragmatic agenesis and congenital diaphragmatic hernia (5,6,9). The coexistence of PE with other musculoskeletal disorders, such as Marfan's syndrome and scoliosis(15% of patients have scoliosis and 11% have a family history of scoliosis), suggests some abnormality of connective tissue may be involved in the development of PE. Further support for a genetic predisposition comes from the fact that 37% of patients have a family history of PE (10).

3. CLINICAL PRESENTATION AND EVALUATION

PE can range in appearance from mild, shallow defects, to defects in which the sternum almost touches the vertebral bodies (Fig. 29.1). The appearance of the defect is the result of two factors. The first is the degree of posterior angulation of the sternum and the second is the posterior angulation of the costal cartilages as they meet the sternum. Additional sternal or cartilaginous asymmetry adds to these defects that become quite challenging for the pediatric surgeon.

PE is generally present at birth or arises shortly thereafter. It is often progressive, with the depth increasing as the patient grows (10). It is much more common in males (3:1, males to females). As mentioned above it can be associated with other congenital abnormalities, including diaphragmatic abnormalities and in 2% of cases with congenital



Figure 29.1 An infant and adolescent with pectus excavatum.

cardiac anomalies (10). In some instances, the repair of the PE is mandated before the repair of the cardiac anomaly (11).

Perhaps the most important association of PE is with Marfan's syndrome. Approximately 2% of PE patients have Marfan's syndrome and these patients typically have the most severe PE (10). After the diagnosis of Marfan's syndrome, these patients should have genetic evaluation, ophthalmologic screening for subluxation of the lens, and have a cardiac echo performed to evaluate for dilation of the aortic root and mitral valve prolapse. A simple office screen for Marfan's syndrome is the thumb sign, which is positive if the proximal phalanx of the thumb extends beyond the ulner border of the palm when the thumb is maximally opposed (Fig. 29.2).

Several methods have been developed to quantitate the severity of PE. These usually involve measuring the distance from the sternum to the spine. Perhaps the most commonly used method is that of Haller et al. (11), who use a ratio of the transverse distance to the anterior–posterior distance derived from chest CT scans (T/AP) (Fig. 29.3). In their system, a score of 3.25 or higher was associated with a severe defect requiring surgery.

PE generally has no discernable physiological effect on infants or children. Some children have reported pain in the area of the sternum or costal cartilage especially after vigorous exercise. Other children have noted palpitations that may or may not be related to the presence of mitral valve prolapse that commonly occurs with PE. A flow murmur may also be detectible in some patients. This is related to the close proximity of the sternum to the pulmonary artery resulting in transmission of a systolic ejection murmur (12).

Asthma is sometimes noted in children with both PE and PC and an association has been proposed. In a review of a large series of patients, however, asthma was no more frequent than in the general population (10). PE also did not seem to affect the clinical course of the patient's asthma. It does not appear that any real association exists between the two.

The impact of PE on the pulmonary or cardiovascular system continues to be debated. Most studies of pulmonary function fail to show any clinically significant



Figure 29.2 Patient with Marfan's syndrome demonstrating the thumb sign.

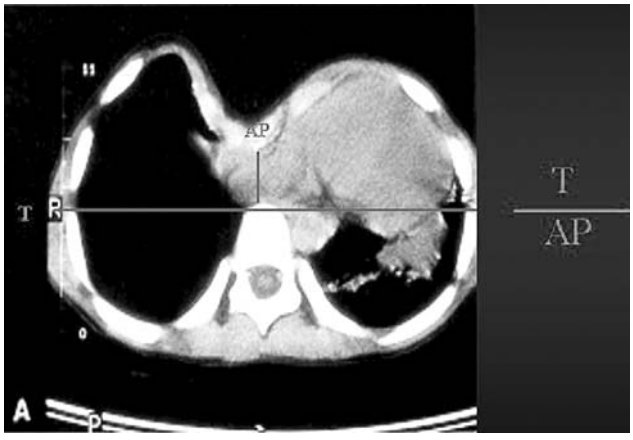


Figure 29.3 CT scan of a patient with severe PE.

difference or show only mild restriction in children at rest or at exercise. In older children and adolescents, a severe pectus deformity has been associated with significantly lower lung volumes with exercise, but no significant difference has been shown at rest (13). Whether lower lung volumes at exercise has any significant physiological effect is unknown. In small children it is difficult to obtain reliable pulmonary function tests, and, therefore, it is unclear what effect, if any, PE has in this age group.

The impact of PE on cardiac function is similarly unclear. Studies using angiography have shown deformities of the heart as a result of the PE (14). Exercise studies have shown decreased cardiac output, increased heart rate and decreased stroke volume in the upright or sitting position in patients with PE (15). In the supine position, these parameters improved, suggesting that PE limits cardiac filling. Improvement in cardiac function could be demonstrated in many of these patients after surgical repair. Despite this, no consistent effect on exercise tolerance has been demonstrated in patients with PE.

4. INDICATIONS

Several indications exist for repair of PE. Some pediatric surgeons believe that the pectus deformity has little physiological consequence. An exception, perhaps, is the competitive athlete, in whom a slight decrease in exercise tolerance may result in poorer performance. The major indication is probably psychological. The sunken appearance of the chest wall has been associated with poor self-image in children with PE, especially as they approach adolescence (16). Improvement in the appearance of the chest wall after repair can lead to improved socialization in these children/adolescents.

Patients with PE can require cardiac or aortic surgery as a result of congenital heart disease or Marfan's syndrome, and sometimes repair of PE is indicated prior to surgery. Shamberger et al. (17) noted a 1.5% incidence of cardiac anomalies in patients undergoing surgery for anterior chest wall deformities. In patients requiring extra cardiac conduits, they recommended repair of the PE prior to cardiac surgery to avoid the possibility of external compression on the conduit. One patient in their series underwent repair of the PE at the time of cardiac surgery. This patient subsequently expired from intrathoracic bleeding. Shamberger et al. (17), therefore, recommended not performing simultaneous

cardiac and PE repair. Similarly, repair of PE prior to aortic surgery in Marfan's patients may be required to avoid the possibility of extrinsic compression.

5. OPEN SURGICAL REPAIR

The first surgical repair of Pectus Excavatum occurred in 1911 by Meyer and, subsequently by Meyer and Sauerbruch (18,19) in 1913. In 1949, Ravitch (20) reported a technique that until recently formed the basis of modern Pectus surgery (Figs. 29.4 and 29.5). His technique included excision of all deformed cartilage from the perichondrium, division of the xiphoid from the sternum, division of the intercostals bundles from the sternum, and finally transverse sternal osteotomy. The sternum was then displaced anteriorly and held into position by wires.

Welch (21), in 1958, altered the procedure by preserving the perichondrial sheaths of the costal cartilage. He preserved the upper cartilage intercostal bundles and fixed the sternum anteriorly with silk suture. Other workers further modified the repair by adding a metal strut to ensure stability of the sternum (22,23). An interesting recent modification has been introduced by workers in Japan (24). They use an endoscope to assist in resection of the costal cartilage. Their reported advantages include smaller operative incisions and the ability to dissect the pleura under endoscopically magnified visualization. Their report details results in only six patients, all of whom appeared to have satisfactory outcomes

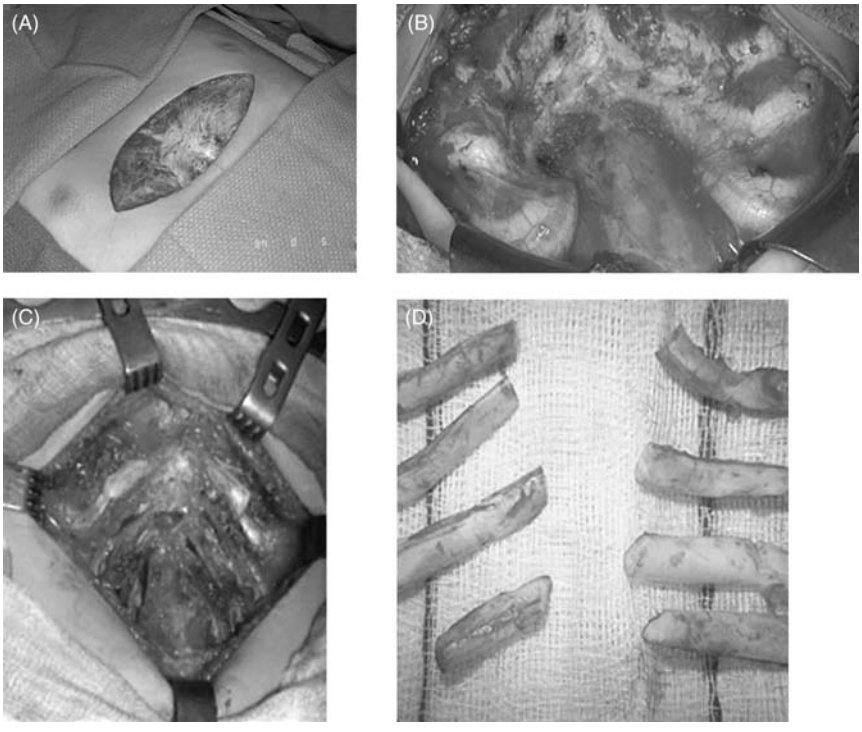


Figure 29.4 Ravitch procedure: (A) incision is made in chest wall; (B) exposure of cartilage after elevation of muscle flaps; (C) sternum after resection of costal cartilage; (D) resected cartilage specimens.

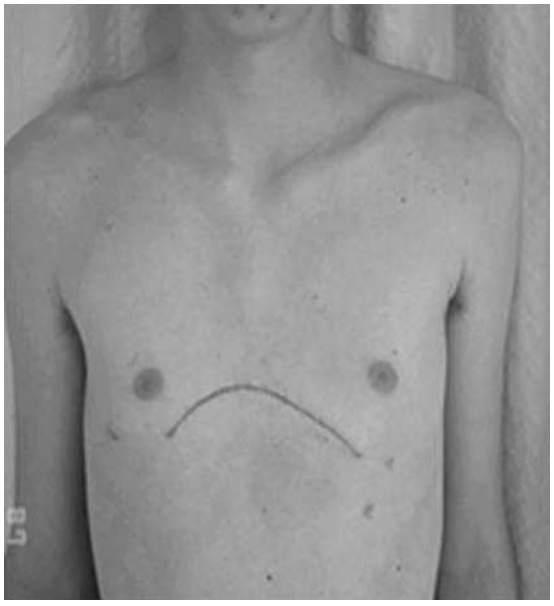


Figure 29.5 Patient post-op after Ravitch repair.

cosmetically. They did report one pneumothorax. No other significant complications were noted.

Haller et al. (25) have described a technique of tripod fixation of the sternum. This method uses a posterior sternal osteotomy, subperichondrial resection of the lower deformed cartilage, and oblique division of normal upper second or third cartilage. The obliquely divided cartilages are positioned to override themselves and are secured in an anterior position to the sternum.

A technique developed in France and used primarily in Japan divides the sternum from the cartilage (26). The sternum is then flipped 180° and reattached to the cartilage as a free graft. This technique has a high complication rate compared to more conventional methods. Finally, a procedure has been described to correct the deformity by introducing silastic implant into the subcutaneous space above the sternum (27). This improves the appearance of the defect but does not increase the size of the thoracic cavity.

6. OUTCOMES AND COMPLICATION OF STANDARD REPAIRS

In general, reports of outcomes and patient satisfaction following the Ravitch (open) repair and variants thereof have been excellent. Several large series of patients exist both with a metal strut in the repair and without, and report >90% satisfactory results, respectively (28,29). Presently, no direct prospective comparison exists between the use of metal struts and no struts in open repairs. Haller et al. (25) showed 100% satisfactory results in a series of 45 patients who underwent their tripod fixation method.

Complications of open repair of PE are unusual. The most common complication is a pneumothorax. These are usually small and can be observed. Large pneumothoraces usually need only aspiration of air. Recurrence is seen in up to 20–30% of patients in long-term follow-up. Approximately half of these are major recurrences requiring a second operation (28,29).

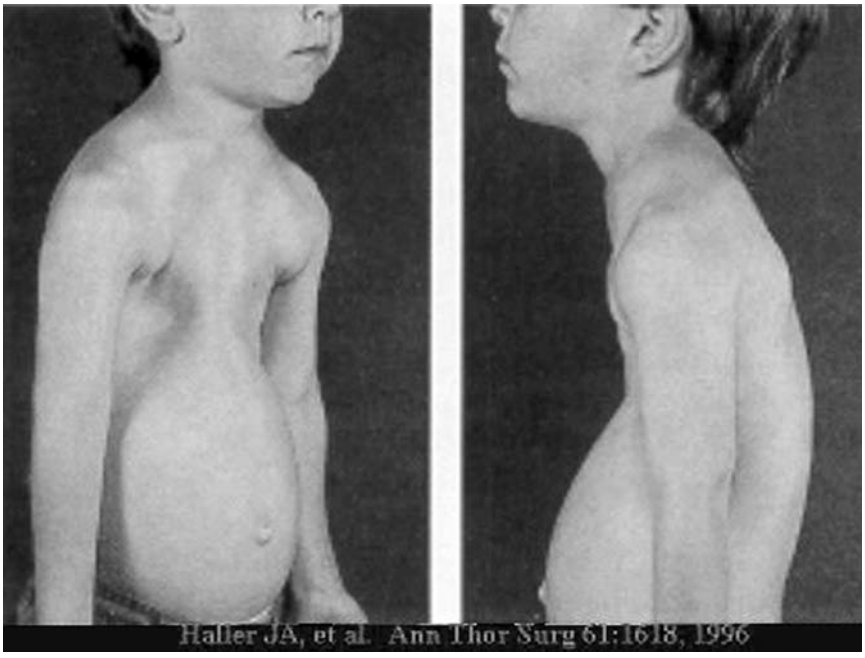


Figure 29.6 Two patients with thoracic dystrophy after open PE repair.

The most devastating complication of the open repair is impaired development of the thoracic cavity and thoracic dystrophy (Fig. 29.6). This occurs most commonly when the surgery is performed in preschool age children. Haller (30) reviewed 12 patients who underwent PE repair and developed thoracic dystrophy. In each instance, repair had been performed under the age of 4 and more than 5 ribs had been resected. These patients presented with severe exercise intolerance and significant reduction in pulmonary function tests. Based on their report, Haller (30) recommended delaying operative repair until at least age six and limiting the number of resected cartilage. This author recommends delaying surgery until a significant portion of adolescence has occurred, that is, age 12–16 years. Theoretically, at this age a significant portion of chest wall growth has already been completed and dystrophy at this point would be inconsequential.

7. MINIMALLY INVASIVE REPAIR OF PE (THE NUSS PROCEDURE)

In 1998, Donald Nuss et al. (2) presented their 10–year results of a new and minimally invasive approach to repairing PE (2). Their repair is based on the observation that even the chest wall of adults can be remodeled, as seen in adults with barrel chest from emphysema, without the need for resecting ribs or cartilage. Moreover, remodeling in adults occurs long after the chest wall has "matured". Therefore, it should be possible to remodel the chest wall in children without the need for cartilage resection or sternal osteotomies. The other key observation is based on the management of orthopedic conditions such as scoliosis and club foot. These conditions can be corrected conservatively by placing splints and leaving them in place for long periods of time.

The Nuss procedure involves inserting a custom bent, curved metal bar underneath the sternum through lateral chest incisions. The bar is then turned placing the convexity of

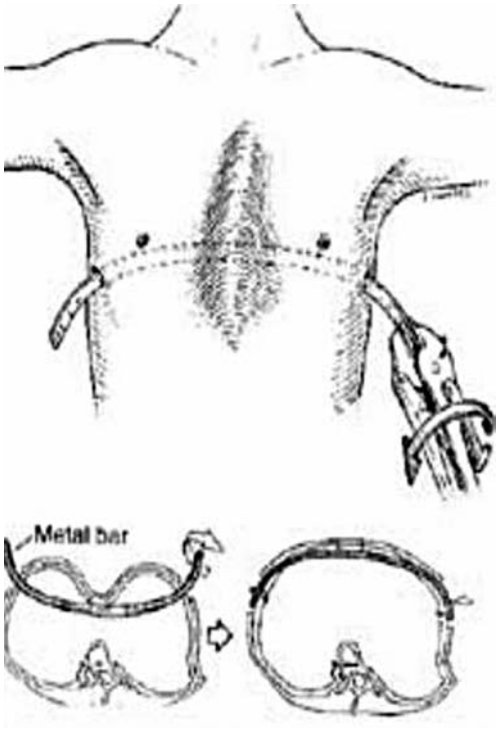


Figure 29.7 Schematic of the Nuss repair.

the bar upward and instantly correcting the pectus depression. The bar is then secured to the lateral chest wall/ribs and left in place for 2–3 years, after which it is removed in an outpatient procedure. The details of the procedure are outlined in Figs. 29.7 and 29.8.

8. EVIDENCE-BASED OUTCOME OF THE NUSS PROCEDURE

In the original published description by Nuss et al. (2), 127 patients with PE were evaluated over a 10-year period (2). Fifty of these patients underwent operative repair (8 by open methods and 42 by the Nuss method). The age range of patients undergoing the Nuss procedure was 15 months–15 years, with most patients in the 3–5 year range. The ratio of male to female was 4:1. The twelve most recent patients had chest CT scans and the CT index in almost all cases was >4. The most common clinical complaints were exercise intolerance in 16, recurrent upper respiratory tract infections in 15, asthma in 9, and chest pain with exercise in 6 of the patients.

At the time of that publication, the Nuss procedure called for the bar to remain in position for two years. Thirty patients had undergone bar removal by the end of the 10-year study period. Follow-up after bar removal ranged from 6 months to 7 years (mean 2.8 years). Good to excellent results were reported in 26 of the 30 (87%) (Fig. 29.9). In three of the four patients, with poor to fair results, there was recurrence of the pectus deformity because the bar was too soft. Subsequent repairs used in the series were performed with stiffer metal bars. One of the four patients with a poor outcome also had Marfan's syndrome.

In the 42 patients undergoing the minimally invasive procedure, complications were reported in 12 (29%). Complications were as follows: pneumothorax in four patients (one of which required a chest tube), skin irritation in four patients, two bar displacements requiring repositioning, one wound infection, and one viral pneumonia. No serious complications related to the surgery were noted in their series despite long follow-up.

In 2001, the group in Indianapolis published their series of 38 patients undergoing the Nuss procedure during a 5-year period compared to 68 patients who underwent an



Figure 29.8 Operative steps of the Nuss repair: (A) placement of epidura catheter; (B) model of Nuss bar is bent specifically for each patient; (C) bending of the Nuss bar to the shape of the model; (D, see p. 340) (right) thoracoscopic ports are placed to monitor bar placement (left) bar is passed under sternum; (E, see p. 340) (right) preparing to flip the Nuss bar (left) Bar in position after being flipped.

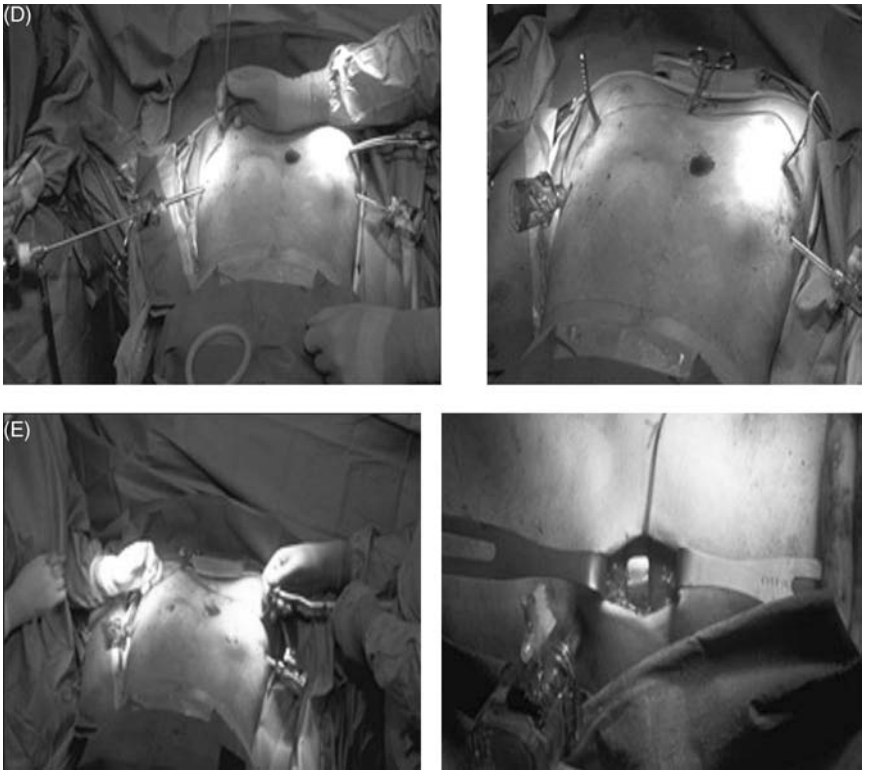


Figure 29.8 *Continued.*

open repair during the same period (31). The Nuss group tended to be younger (avg. 9.5 years vs. 12.6 years) and had slightly fewer associated anomalies (6% vs. 8.8%). Complications were seen in 43% of the Nuss patients compared to 20% in the open group over the same period of time. These complications were typically minor and

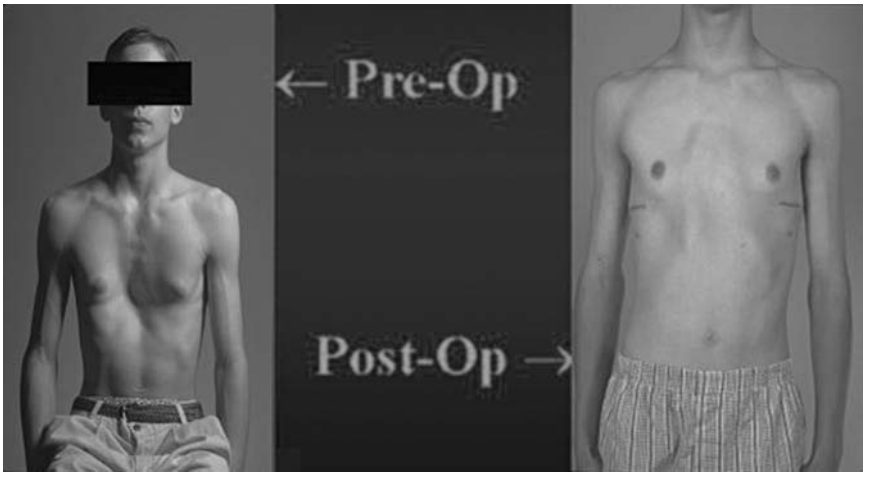


Figure 29.9 Patient before and after Nuss repair.

included pneumothorax, pleural effusion, bar migration, contact dermatitis, and ileus. One serious complication was noted: mediastinitis requiring bar removal. In this series, only two poor cosmetic outcomes were noted. In one child the PE converted to a PC, while another developed a mild chest asymmetry. Therefore 33 of 35 patients achieved at least a good cosmetic outcome (94%).

A third small series of patients undergoing minimally invasive PE repair has been reported from the University of Connecticut (32). They compared 36 patients undergoing the Nuss repair to six patients undergoing the Ravitch repair. Mean age and weight as well as severity of the PE deformity were equivalent in the two groups. Operative time, blood loss, and ICU admissions were significantly reduced in the Nuss group. These results are similar to that seen in the Indianapolis series. However, the length of stay increased and analgesic requirement was also higher. Again these results were consistent with those seen in the Indianapolis series.

The most common complication following Nuss repair seen by Wu et al. was pneumothorax (6 of 36 patients, 17%) (32). Only one, though, required tube thoracostomy (2.8%). Other complications included pleural effusion (8.3%) and epidural catheter-related problems (2.8%). Interestingly, bar displacement was seen in only one patient (2.8%). This is lower than rates published in the two other series and may be related to the use of a stabilizing bar in all patients in this series.

The fourth and most recently reported series is from the group at Kansas City (33). They reported 80 patients undergoing Nuss procedure compared to 32 previous patients undergoing a Ravitch procedure. This is the largest single institutional experience reported so far. The patient characteristics are similar to the other series. The mean age is 11.5 years compared with 9.4 years in their open cohort. Operative time was 53 min, significantly shorter than the open procedure (143 min). Blood loss was very low and hospital stay was short. The results are similar to the other series. The complication rate was 11% with pneumothorax and bar displacement being most common.

A recent survey of pediatric surgeons in the American Pediatric Surgical Association (APSA) membership was conducted by Hebra and coworkers in an attempt to determine outcomes and complications of the Nuss procedure in a large volume of patients (34). Of 74 responders, 42% used the Nuss procedure as their primary method of repair. The overall experience of the responders was 251 cases. A complication rate of 21% was reported. This rate is somewhat lower than that reported by Molik et al. (20%) and Nuss et al. (28%). But this number is quite high compared to complication rates noted in several large series of open pectus repairs ($<10\%$). The most common complication was bar displacement (9.2%). A complication seen in 4% of patients in Nuss' series, 11% of patients in the Indianapolis series, and 4.7% of patients in the Kansas City series. The next most common complication was pneumothorax requiring chest tubes (4.8%). Pneumothorax is a very common complication seen in all series. In general, however, these are very small and resolve spontaneously. In Nuss' series only one patient required a chest tube for pneumothorax.

The APSA survey also found a significant re-operative rate for the Nuss procedure. As mentioned earlier, the most common reason for reoperation was bar displacement. In the Indianapolis series, reoperation was required in eight patients (29%) and bar displacement was the most common reason. Only two patients in Nuss' series required reoperation, both for bar displacement. A recent introduction of a lateral stabilizing device may decrease the incidence of bar movement.

A feature of all reported series is a wide age range of patients undergoing minimally invasive repair. The impression of most surgeons polled by Hebra et al. was that older children experience more complications than the younger cohort. Specifically, older children

were thought to have a higher incidence of bar displacement. Perhaps this is related to larger forces generated by the larger rib cage of teenager and engagement in higher risk activities such as contact sports. Interestingly, Molik et al. were not able to appreciate an increased complication risk in teenagers. One thing that is clear is that teenagers have significantly more pain from the Nuss procedure and some of the studies have documented a longer period of analgesia in these patients.

So far there have been very few serious complications reported with the minimally invasive pectus repair (34,35). One case of cardiac perforation was reported. This occured in an 8-year-old child with a moderately deep and asymmetric deformity. The child required a median sternotomy, emergency heart–lung bypass, and repair of the right atrium, tricuspid valve and left ventricle. At two years follow-up, the child was doing well. One case of bilateral empyema with pericarditis has also been seen. In this case bilateral chest debridement and open pericardial debridement was performed. Three cases of thoracic outlet syndrome have also been reported. At the time of the report one of these patients required bar removal for severe symptoms referable to the brachial plexus and arm cyanosis relieved with arm elevation. The other patients had gradual improvement of symptoms and still had the bar in place.

It is noteworthy that to date no reports of thoracic dystrophy after the Nuss procedure exist. Haller (30) found this complication primarily in younger patients. In the series of patients presented by Nuss et al., 19 were 5 years or younger when they underwent PE repair. None were reported to have developed thoracic dystrophy. In general, no conclusions can be made yet from the other series. Molik et al. operated primarily on older patients and no patient under the age of 5 underwent Nuss repair in their series. Thoracic dystrophy was not reported as a complication by surgeons polled by Hebra et al., presumably since the growth plates are not injured or removed in the Nuss procedure compared to the classic Ravitch procedure. However, the age of these patients is unknown as well as the length of follow-up. Therefore, while preliminary indications suggest that thoracic dystrophy may not be a complication of the Nuss procedure, the jury is still out, long-term follow-up of the youngest patients is required.

Determining acceptable cosmetic outcomes is quite subjective. As mentioned previously, Nuss reported good to excellent outcomes in 26 of 30 patients after bar removal. Molik et al. reported two poor outcomes (6%), but most patients in their series had not reached the point of bar removal, so that it remains to be seen what the cosmetic outcome in that series will be. Cosmetic outcomes are also difficult to determine in the University of Connecticut series. The authors state that they achieved a 100% satisfactory cosmetic outcome. However, at the time of their publication, only five patients had reached the point of bar removal. The Kansas City series reported good to excellent results in 76 of 80 patients (95%) and in 15 of 15 patients after bar removal (100%). Most surgeons polled by Hebra et al. considered their cosmetic outcomes to be good to excellent in 96.5% of patients. It remains unknown in that poll what the length of follow-up was and how many patients had achieved bar removal. Since PE can recur even 5–10 years after open repair, much longer follow-up than currently exists with the Nuss procedure will be needed before determining how cosmetically acceptable and durable this method of repair will be. Also, it remains unclear if teenagers will derive the same cosmetic benefit as younger children. Few of the patients in Nuss' series (the series with by far the longest follow-up) were teenagers. In contrast, three other series of patients are comprised of much older children (mean age 6.8 years vs. 12.3 years, 9.5 years and 11.5 years).

At the present time no information exists as to the physiological benefit of the minimally invasive approach to PE repair. As discussed previously it is difficult to reliably

demonstrate any cardiopulmonary deficit in patients with PE. In open repairs, several studies have been performed comparing cardiopulmonary effects before and after repair. In his review of the subject, Shamberger (36) found no consistent evidence that open PE repair provided any cardiopulmonary benefit. In fact, several studies have demonstrated worsening of pulmonary function after repair, perhaps as a result of increased chest wall rigidity from the surgery. In contrast, studies assessing workload and exercise tolerance often show improvement after PE repair. Therefore, the cardiopulmonary benefit of open PE repair is, at best, uncertain. It will be interesting to see what affect, if any, the minimally invasive repair will have in the long-term. Without the need for cartilage resection and sternal osteotomy, one might expect less postoperative chest wall rigidity and a greater likelihood of improvement in pulmonary function. Borowitz et al. (37) have compared preoperative and postoperative pulmonary function in ten patients undergoing the Nuss procedure (37). They find no significant difference between preoperative function and that of 1-year postoperatively with the bar in. Therefore, having the bar in place has no detrimental effect on pulmonary function. It remains to be seen what effect there is after bar removal.

9. MODIFICATIONS TO THE NUSS PROCEDURE

In response to complications seen so far, several modifications to the Nuss procedure have been proposed. Implementation of a lateral stabilizing bar was perhaps the first major modification and was a response to a significant number of bar displacements (Figs. 29.10 and 29.11). In the Indianapolis series they documented bar migration in 35% of their first fifteen patients and none in 21 subsequent patients using the stabilizing bar. Wu et al. had only a single episode of bar displacement in 38 patients using the stabilizing bar. Other surgeons have used somewhat different modifications to combat bar displacement. A recent report proposes the use of a three-point fixation method in conjunction with the stabilizing bar (38). In this technique, a stitch is placed around a rib and bar adjacent to the sternum with the aid of thoracoscopy. In 20 patients undergoing the Nuss procedure with the three-point fixation modification, only one bar displacement was noted in 1-year follow-up (5%). In that patient, an absorbable suture was inadvertently used.

Another recent modification is the use of thoracoscopy to monitor safe passage of the clamp across the mediastinum. Thoracoscopy came into widespread use after the report of the cardiac injury discussed earlier. A survey of surgeons performing the Nuss found that 61% now use thoracoscopy on a routine basis (34). A separate variation is described by Miller et al. (33), in which a subxiphoid incision is through which a subxiphoid tunnel is created into the anterior mediastinum. This allows direct visualization of the bar as it passes underneath the sternum.

Recommendations for the length of time before bar removal have also been changed. In the original description of the operation, the bar was left in for 2 years. Recently, some workers have recommended increasing that time to 3 years in an attempt to decrease the incidence of PE recurrence. Time will tell if this will increase the number of positive outcomes.

Because of the variety of modifications, it can be difficult to compare results from one series to the next. A prospective study comparing the Nuss procedure to open repair is currently underway. This will be a prospective, nonrandomized observation study. Patients will undergo preoperative assessment by CT scan and by a pediatric pulmonalogist. Multiple variables will be measured pre and postprocedure The main outcomes from the study, however, will result from comparison of pre and postoperative CT

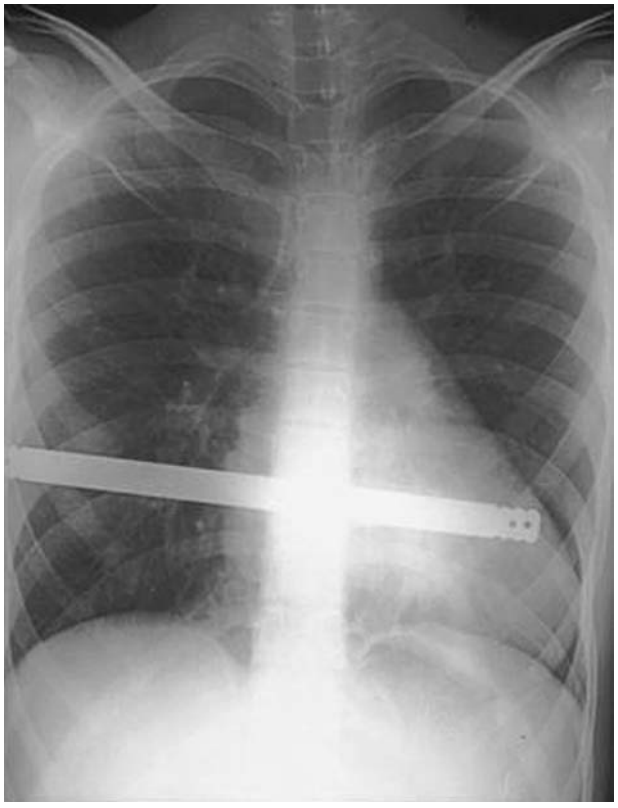


Figure 29.10 Chest X-ray of a patient with displacement of the Nuss bar.

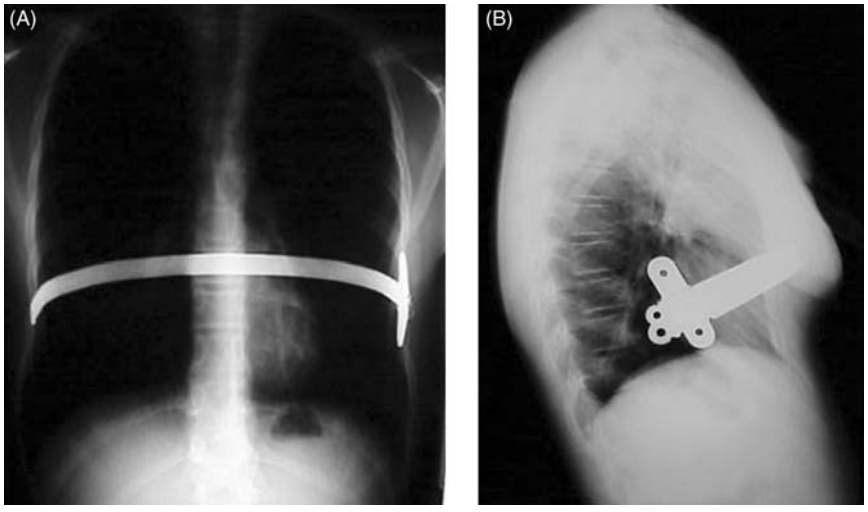


Figure 29.11 Chest X-ray of a patient with a lateral stabilizing bar in place: (A) anterior–posterior view and (B) lateral view.

scan and pulmonary function tests. This study should determine differences, if any, in cosmetic and function outcomes between minimally invasive and open repair. Unfortunately, it will not be feasible to randomize this study as most patients have preconceived ideas about the type of operation they want. This will somewhat limit the utility of this study.

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Minimal Access Surgery in Other Pediatric Surgical Specialities

30
Minimal Access Surgery in Pediatric Urology

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1. INTRODUCTION

Despite the expanding application of laparoscopy in general surgery, laparoscopy is not yet widely performed by pediatric urologists. This is mainly due to a lack of laparoscopic training. In the early experience, the indications for laparoscopy in pediatric urology were unclear and unproven compared to the advantages of open procedures. Gradually, indications are expanding from ablative surgery to major reconstructive procedures. Historically, management of nonpalpable testis is the most common laparoscopic procedure performed by pediatric urologists, and this is covered in Chapter 24 (Nonpalpable Undescended Testis). In the current chapter, we will deal with specific procedures for the upper and lower urinary tract in pediatric urology. As experience in children is still limited, we have also included the adult urology experience in this review.

2. UPPER URINARY TRACT

Upper urinary tract surgery in children includes two main categories: ablative and reconstructive. Except for rare exceptions, both are limited to benign diseases. The most common ablative procedures are stone removal and nephrectomy, and the most common reconstructive procedure is pyeloplasty.

Extracorporal shock wave lithotripsy or percutaneous and endoscopic techniques can be used to manage most pediatric kidney stones. Open surgery may be required in rare cases when these techniques are not applicable, such as in young children with bulky cystine lithiasis. For these exceptions, laparoscopic pyelolithotomy is a suitable alternative to open surgery, and may avoid parietal scarring in these children who usually need repeat surgery later in life (1–3).

The first laparoscopic nephrectomy in adults was reported in 1991 by Clayman et al. (4). One year later Erlich et al. (5) reported the first series of pediatric cases. Since then many authors have reported successful results for nephrectomy and nephroureterectomy in children, all advocating the transperitoneal approach (6,7). Roberts suggested a retroperitoneal approach to the kidney in 1976 (8), reporting his experience using retroperitoneal endoscopy with gas insufflation in animals. Retroperitoneal operative laparoscopy was described for the first time by Gaur in 1992 (9) and then by others in adult and pediatric urology (10–12). Despite the expanding application of retroperitoneal laparoscopic renal surgery in adults (13,14), this technique is still only performed by a limited number of pediatric urologists (2,12,15–17). We previously reported that the retroperitoneal approach is a well-adapted laparoscopic technique for renal surgery in children and is comparable to that of conventional renal surgery (2,18).

Guilloneau et al. (11) reported in a retrospective study of adults and children that retroperitoneal and transperitoneal approaches were equivalent in terms of morbidity and postoperative stay, but that the operating time was shorter with the retroperitoneal approach. There have not been any randomized studies done thus far comparing the transperitoneal and retroperitoneal approaches to nephrectomy in either adults or children.

Effects of retroperitoneal CO_2 insufflation have been studied in both animals and children (18,19). These studies demonstrated a significant increase in systolic blood pressure and end-tidal carbon dioxide, while there was no modification of the other hemodynamic or ventilatory parameters. These changes do not need any special modification of the ventilatory parameters, whereas special care with hypertensive patients is required.

2.1. Renal Access

The upper urinary tract can be accessed by retroperitoneal or transperitoneal approaches.

2.1.1. Lateral Retroperitoneal Approach

The patient is placed lateral, with sufficient flexion of the operating table so as to expose the area of trocars placement, between the last rib and the ileac crest. Retroperitoneal access is achieved through the first incision, 10–15 mm in length, and one finger width from the lower border of the tip of the 12th rib (Fig. 30.1). The use of narrow retractors with long blades allows deep dissection with a short incision. Gerota's fascia is approached by a muscle-splitting blunt dissection, then it is opened under direct vision and the first blunt trocar (5 mm or 10 mm) is introduced directly inside the opened Gerota's fascia [Fig. 30.2(A)]. A working space is created by gas insufflation dissection, and the first trocar is fixed with a purse-string suture that is applied around the deep fascia to ensure an airtight seal and to allow traction on the main trocar if needed to increase the working space. The second trocar (3 mm or 5 mm) is inserted posteriorly in front of the lumbosacral muscle, at the costovertebral angle. The third trocar (3 mm or 5 mm) is inserted in the anterior axillary line, a finger width from the top of the iliac crest. To avoid transperitoneal insertion of this trocar, the working space is fully developed and the deep surface of the anterior wall muscles is identified before the trocar insertion. Insufflation pressure does not exceed 12 mm, and the CO_2 flow rate is progressively increased from 1 L to 3 L/min. Access to the retroperitoneum and creation of the working space are the keys to success in the retroperitoneal renal surgery. Age is not a limiting factor for this

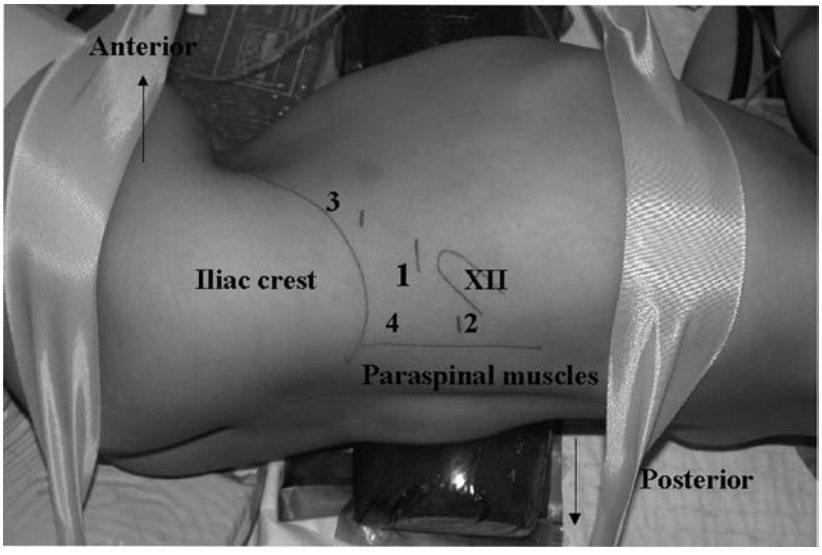


Figure 30.1 Ports placement and landmarks for left retroperitoneal laparoscopic pyeloplasty. The child is positioned lateral. (1) Retroperitoneal access is achieved via the first incision, at the tip of the 12th rib, and is used for the laparoscope. (2) Second port, placed in the costovertebral angle, and is used for the needle holder, scissors, and to place the double J stent. (3) Third port is placed close to the iliac crest at the anterior axillary line, and is used for the grasping forceps. (4) Fourth port, is eventually placed if needed, is used for traction on the UPJ to stabilize the suture line during anastomosis. The first port is 5 mm diameter and the other ports are 3 mm.

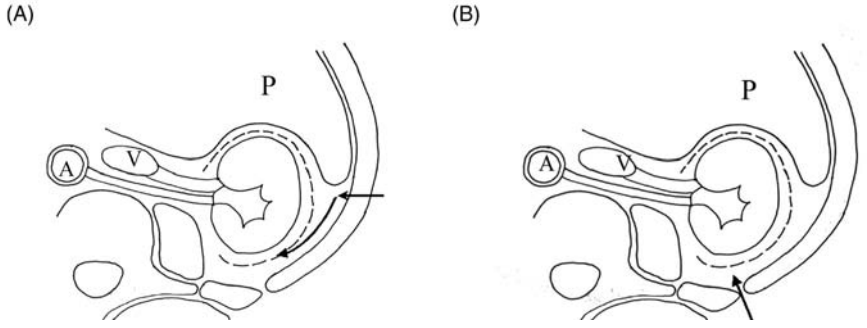


Figure 30.2 Retroperitoneal access (right side). The retroperioneal space can be approached either by the lateral or by the posterior approach. (A) Lateral approach: the child is positioned lateral. The Gerota fascia is exposed laterally but retracted anteriorly to be opened posteriorly, the direction of the approach is shown by the arrow. By the posterior approach of the Gerota the peritoniuem is far and the insufflation starts behind the kidney, thus the kidney is pushed anteriorly with the peritonieum without the need to retract the kidney. (B) Posterior approach: the child is positioned prone, the Gerota fascia is opened posteriorly and the renal pedicle is directly approached. P: peritonieum; V, inferior Vena Cava; A, aorta.

approach. Young children have less fat and the access is easier. Our youngest patient using this technique was 6 weeks of age.

2.1.2. Prone Posterior Retroperitoneal Approach

Access begins with an incision in the costovertebral angle at the edge of the paraspinous muscles [Fig. 30.2(B)]. The secondary trocars are placed just above the iliac crest, one medially at the edge of the parasinous muscles, and one laterally at the posterior clavicular line (20). Borzi (21) compared the lateral and posterior retroperitoneal approaches in a randomized prospective study on 36 complete and 19 partial nephrectomies in children, and found no significant difference in operative time or outcomes. He observed that the posterior approach gave easier access to the renal pedicle, while the lateral approach gave better access to ectopic kidneys and allowed complete ureterectomy in all cases.

2.1.3. Other Retroperitoneal Approaches

Since the description by Gaur (9), balloon dissection has been the method applied by most urologists to develop the retroperitoneal space. Disadvantages include the cost of the disposable material and possible complications from rupture of the balloon (22). Balloon dissection permits creation of a working space without opening Gerota's fascia, a consideration which is important for radical nephrectomy of malignant tumors in adults, but which may be less relevant in children. Micali et al. (3) reported the use of the Visiport visual trocar to access the retroperitoneal space directly. The advantage of this method is the possibility to use a small incision for the first trocar, which is interesting in reconstructive surgery but not in ablative surgery as the first incision is needed for organ retrieval.

2.1.4. Transperitoneal Access

There are several options for patient positioning. The most frequently described is the flank position described by Peters (20). The pneumoperitonium is created through an open umbilical approach. The child is positioned with the surgeon standing in front of

the abdomen (opposite side of nephrectomy). The most frequent configuration has been with the umbilical port and two ipsilateral ports in the mid-clavicular line above and below the umbilicus. A fourth trocar may be placed in the mid-axillary line for exposure to retract the liver or spleen if needed. The kidney is exposed by medial mobilization of the colon.

2.2. Nephrectomy

2.2.1. Indications

In adults, the current feeling is that laparoscopic radical nephrectomy is safe and appropriate for renal cancers measuring <5 cm in size (13). The most common renal tumor in children is nephroblastoma (Wilms tumor). These tumors are almost always very large, frequently extend outside the kidney, and may be prone to rupture during dissection, with a significant impact on staging if this happens. For these reasons, most authors feel that the laparoscopic approach is inappropriate for renal tumors in children.

Most nephrectomies in children are done for nonfunctioning kidneys secondary to obstructive uropathy or reflux, and the laparoscopic approach is excellent for most of these cases. Although laparoscopic nephrectomy for multicystic dysplastic kidney is an easy and safe procedure, we agree with other pediatric urologists who do not feel that these kidneys require routine resection because of the high rate of spontaneous involution (23). Generally acceptable indications for nephrectomy are increase in size of the cysts or patients who develop hypertension or infection.

Nephrectomy may be indicated in children with end-stage renal disease before transplantation when the primary renal disease is associated with hypertension, severe nephrotic syndrome, or severe hemolytic uremic syndrome (18). During open surgery in these children, a large incision is necessary to control the renal pedicle and to extract a large kidney, and therefore the laparoscopic approach is particularly advantageous for this population. Bilateral laparoscopic nephrectomy has been performed in adults (24). We have performed simultaneous bilateral nephrectomy in 10 children. The procedure was performed using the lateral retroperitoneal approach, and the position and draping were changed between the first and the second side (25). This procedure can also be performed through a posterior retroperitoneal approach (26).

2.2.2. Technique of Nephrectomy Using the Lateral Retroperitoneal Approach

After accessing the retroperitoneal space, the landmarks are identified to maintain orientation. The psoas muscle is the posterior landmark and should remain at the bottom of the monitor. The kidney is attached anteriorly to the peritoneum and should remain at the top of the screen.

The renal pedicle is identified and approached posteriorly (Fig. 30.3). To avoid multiple ligation of branches of the renal vessels, the vessels are dissected close to their junction with the aorta and the vena cava. On the left side, the vein is ligated distal to the genital and adrenal branches. On the right side, the vein is short and careful dissection at its junction with the vena cava will avoid the confusion of dissecting the vena cava. The renal artery is usually dealt with first, followed by the vein. The vessels can be clipped, ligated, or coagulated. The choice of method depends on the vessel diameter and the surgeon's experience. Small arteries such as those associated with multicystic dysplastic kidneys can be coagulated using bipolar cautery, harmonic scalpel, or ligasure. The method most commonly used for larger vessels is titanium clips. If the diameter of the vessel is bigger than the length of the clip, resorbable intracorporeal knots can be used

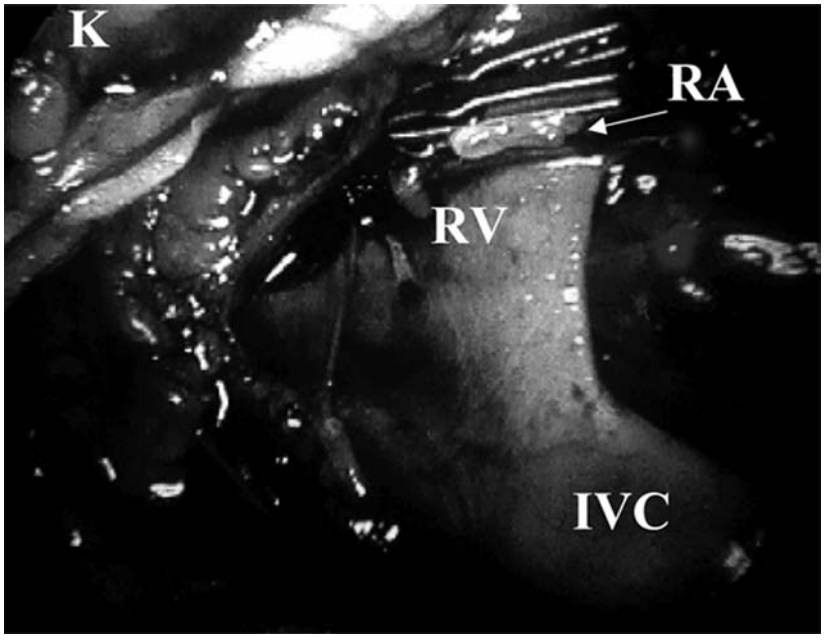


Figure 30.3 Exposure of the renal pedicle via retroperitoneal approach. Right pretransplant nephrectomy for congenital nephrotic syndrome in a child aged 8 months. The kidney (K) is held anteriorly by the peritoneum and stays on the top of the screen. The renal artery (RA) is clipped and sectioned. Note the good exposure of the renal vein (RV) and the inferior vena cava (IVC). In this case, the size of the vein is bigger than the 5-mm clips, indicating the need for endocorporeal ligation of the renal vein.

either with or without clips. Staples have been described for large vessels during nephrectomy in adults, but this technique requires a 12-mm port and is not needed in most pediatric patients.

The ureter is then identified and dissected as far as necessary. In the absence of reflux, the ureter is coagulated and sectioned at the lumbar level below the pyeloureteric junction. This is particularly important in patients undergoing pre-transplant nephrectomy, since the native ureter might be used for the transplanted kidney. In the presence of reflux, the ureter is ligated as close as possible to the ureterovesical junction, taking care to avoid injury to the vas deferens in males. During this distal dissection, the surgeon moves towards the head of the child, and the screen goes towards the feet. In the beginning of our experience, we were using a fourth trocar to dissect and ligate the distal ureter (2). Currently, we use an endoloop or, if the ureter is large, a transparietal suture to fix the ureter to the abdominal wall to facilitate its distal dissection and ligation. As the peritoneum is very close to the ureter in this distal part, its dissection is done at the end of the procedure to avoid tearing the peritoneum.

The last part of the dissection is the anterior surface of the kidney. The kidney is dissected from the peritoneum very close to its capsule in the cleavage plane of the areolar tissue. Usually no hemostasis is necessary in this plane, although sharp dissection with bipolar coagulation may be necessary for inflammatory adherent kidneys. In rare cases of xanthogranulamatous pyelonephritis, we do the dissection of the adherent kidney through the subcapsular plane to avoid injury to the intraperitoneal structures (27).

2.2.3. *Kidney Retrieval*

The kidney is usually retrieved through the main incision. A 5 mm telescope is inserted through the accessory port and a toothed grasping forceps is introduced through the 10 mm port to extract the kidney. The kidney is grasped at one of the poles and pulled in this axis. In most cases, the kidney can be divided under vision during extraction through the muscle wall. In cases of severe pyelocaliceal dilatation or a multicystic dysplastic kidney, direct evacuation by puncture helps in organ retrieval. An extraction bag is used for infected or large kidneys, and the kidney is morcellated inside the bag. If nephrectomy is being done in conjuction with an open lower urinary tract procedure, the nephrectomy is done first and the kidney is placed near the bladder without transecting the ureter. Retrieval is then done through the Pfannenstiel incision.

2.2.4. *Laparoscopic vs. Open Nephrectomy*

No prospective randomized study has been done to compare the laparoscopic and open approaches to nephrectomy in adults or children. We have retrospectively studied a comparable group of children who underwent pretransplant nephrectomy before beginning our experience with retroperitoneal laparoscopic nephrectomy (18). In this specific group of patients with end-stage renal disease, the hospital stay was significantly shorter after laparoscopic vs. open nephrectomy (5.2 vs. 8.4 days). Even though the operating time for laparoscopic nephrectomy was longer (120 vs. 104 min), the difference was not statistically significant (18). Hamilton et al. (28) found comparable results on transperitoneal laparoscopic nephrectomy with a significant decrease in hospital stay after laparoscopic compared to open nephrectomy (22.5 vs. 41.3 h). Operative time was significantly longer in the laparoscopic group (175.6 vs. 120.2 min).

Abbou et al. (13) retrospectively compared retroperitoneal laparoscopic radical nephrectomy to open radical nephrectomy for renal cancer in adults. Operative time was longer in the laparoscopic group (121 vs. 145 min). In the laparoscopic group there was significantly less blood loss, less postoperative pain medication, and a shorter hospital stay.

2.3. Partial Nephrectomy

2.3.1. *Indications*

Partial nephrectomy is usually done in children to remove a non-functioning upper or lower pole secondary to complicated duplex anomalies of the kidney. The usual pathology of the upper pole is obstruction associated with a ureterocele or incontinence secondary to an ectopic ureter. The usual pathology in the lower pole is reflux. Laparoscopic partial nephrectomy is technically more demanding than total nephrectomy. Jordan and Winslow (29) described the first case of upper pole partial nephrectomy in a child by transperitoneal laparoscopy. Janestschek et al. (30) reported their series of 14 cases of successful partial nephrectomies in children. These procedures can be performed using a retroperitoneal or transperitoneal approach. Laparoscopic techniques are well suited to this procedure, with the benefits of perfect global exposure to the anatomy of the full kidney and its vascularization without the need to mobilize the remaining part of the kidney. Another advantage is that there is no need for a second inguinal incision to excise the distal ureter. Our preference is the retroperitoneal approach as it provides a posterior access to the kidney without dissecting the main renal pedicle, an unavoidable step during the transperitoneal anterior approach.

We have shown, in a retrospective comparative study, that retroperitoneal laparoscopic partial nephrectomy can be performed in a comparable operative time to that of an open procedure (31). The mean operative time was 146 min (50–180) and 152 min (75–240) for the open surgery and the laparoscopy groups, respectively. The main advantage to the laparoscopic approach is that the hospital stay is shortened significantly compared to that of an open procedure. The mean hospital stay was 1.4 days (1–3) and 3.9 days (3–5) for both the laparoscopy and the open surgery groups, respectively ($p < 0.0001$). Eight of 13 children were discharged on the day following that of the laparoscopic procedure.

2.3.2. Technique of Retroperitoneal Partial Nephrectomy

The upper pole ureter is identified at the lower pole of the kidney and dissected very close to its wall to avoid injury to the vascular supply of the lower pole ureter. We find it helpful to ligate the proximal ureter before cutting it, so the proximal ureter remains dilated facilitating the dissection of the upper pole. Because the exposure is posterior, contrary to the transperitoneal anterior approach, the upper pole ureter is lifted off the vessels by blunt dissection superiorly. The upper pole ureter is used as a handle to facilitate this part of dissection. The plane between the dilated upper pole pelvis and the lower pole parenchyma is easily identifiable by blunt dissection until the edges of the thin parenchyma of the upper moiety are recognized. At this step, the upper pole vessels are identified running from the aorta or the renal vessels to the upper pole parenchyma. They are either clipped or coagulated depending on their size. The upper pole is identified by color changes after vessel ligation and by the difference in appearance between the normal lower pole and the dilated dysplastic upper pole. The duplex system anomalies have a well-defined vascular line of demarcation between the upper and lower poles of the kidney. However, sometimes it is difficult to individualize upper pole vessels, and the parenchymal transection is started before vascular control of the upper pole. Transection can be done by electrocautery, but we prefer the Harmonic scalpel with the curved jaws, as it provides a clean cut at the junction between the upper and lower poles.

In partial nephrectomy where there is more substantial parenchyma, other means may be required for specific control of bleeding. These may include bipolar electrocautery, electrosurgical snare electrode, microwave coagulator, Gill renal tourniquet, or argon beam, Nd:YAG or holmium:YAG laser (32–38).

To minimize mobilization of the lower pole and consequently the risk of indirect vascular trauma of the renal pedicle, the lower pole remains attached to the peritoneum during all the steps of the procedure. The upper pole is completely freed from peritoneal attachment before transecting the parenchyma to avoid transperitoneal bowel injury.

If upper pole partial nephrectomy is done using a transperitoneal approach, the proximal ureter must be passed behind the renal hilum and delivered under the vessels from above after mobilization of the upper pole ureter. This step is technically the most difficult part of the procedure and requires dissection of the lower pole vessels (32). Identification of the upper pole vessels and parenchymal transection are identical to the retroperitoneal approach.

2.3.3. Retroperitoneal Lower Pole Partial Nephrectomy

Access is the same as for the upper pole. The lower pole ureter is identified and followed to the lower pole pelvis to be sure of its identity. Contrary to the upper pole nephrectomy, full dissection of the lower pole vessels is necessary before transecting the parenchyma. As the main pathology is reflux with repeated infections, the lower pole is usually retracted and easily identified from the healthy upper pole parenchyma. The ureter should be ligated close to the bladder to avoid postoperative reflux into a long ureteral stump.

2.4. Pyeloplasty

2.4.1. Indications

Open pyeloplasty remained the standard treatment for both adult and pediatric patients until the mid-1980s, when the morbidity associated with the flank incision led urologists to explore less invasive alternatives. In the 1990s, the endourologic management of uretero-pelvic junction (UPJ) obstruction became the treatment of choice in the adult population. Although the postoperative morbidity is significantly less after the endoscopic procedures, their success rate does not exceed 80% (39).

Laparoscopic pyeloplasty was introduced in adults in 1993 (40,41). In the initial report of five cases, the operative time ranged from 3 h to 7 h. The procedure has gained in popularity, and recent series have reported a success rate of >95% (42–44). The techniques reported were either a dismembered or non-dismembered pyeloplasty.

Soulie et al. (45) have compared retroperitoneal laparoscopic vs. open pyeloplasty with a minimal incision in 53 consecutive nonrandomized adults. The mean operating time (165 vs. 145 min) was similar in both groups. Incidence of complications, hospital stay, and functional results were equivalent for both groups, but the return to painless activity was more rapid with laparoscopy in younger patients. Bauer et al. (46) did a similar study, with no difference in the postoperative outcome between laparoscopic and open pyeloplasty. In children, the experience is still limited to a few centers with small numbers of patients. Tan (47) reported his experience with transperitoneal laparoscopic dismembered pyeloplasty in 18 children aged 3 months–15 years old (mean 17 months). Mean operative time was 89 min. There was no conversion to open surgery. Two patients with persistent obstruction underwent repeat laparoscopic pyeloplasty.

Retroperitoneal laparoscopic dismembered pyeloplasty is also feasible in children. Yeung et al. (48) have reported their initial experience with retroperitoneal dismembered pyeloplasty in 13 patients, one or whom required open conversion. The mean operative duration was 143 (range 103–235) min. All patients had a rapid and uneventful recovery. Drainage was satisfactory in all 12 patients on a follow-up scan. The longer time needed for the retroperitoneal approach is probably related to the limit of the working space that renders suturing more difficult. Although postoperative urine leak has not yet been reported, this complication would be better tolerated in the retroperitoneal space than in the peritoneal cavity. For this reason we prefer the retroperitoneal approach. In our experience, we attempted the retroperitoneal approach in 21 children, four of whom required conversion to open surgery (49). Conversion was decided at the time of suturing, one for a large redundant renal pelvis of 2 L capacity, one for kidney rotation, and two exceeded the time permitted within our teaching hospital. Operative time was longer than the previously reported series, ranging from 3 h to 5 h. Mean hospital stay was 2 days, and return to full activities was 5 days after surgery. After a mean follow up of 18 months (range 6 months–3 years), none of our patients demonstrated persistent obstruction on follow-up.

The gold standard in children remains the open retroperitoneal dismembered pyeloplasty. The advantages of open pyeloplasty include mucosa-to-mucosa anastomosis and excision of redundant renal pelvis and diseased ureter. The retroperitoneal laparoscopic dismembered pyeloplasty represents an attractive alternative to conventional open pyeloplasty. It is technically challenging but with practice and technical adaptations (50) to improve suturing, it may be completed in the same time as conventional open pyeloplasty. In the future, surgical robotics may facilitate suturing in such a limited working space. Recently, Peters (51) has presented his first preliminary results of robotic-assisted laparoscopic transperitoneal pyeloplasty in children. The first impression is that the robotic-assisted technique

makes suturing easier and may allow to expand advanced laparoscopic reconstructive surgery to bigger number of surgeons without expertise in laparoscopic surgery.

2.4.2. Technique of Laparoscopic Retroperitoneal Pyeloplasty

The same access is used as described for nephrectomy. Yeung et al. (48) used different positioning according to the side of the kidney; semi-prone for the right side and semi-lateral for the left side. The kidney is approached posteriorly and the renal pelvis is first identified. The pyeloureteral junction is identified and a minimal dissection is done to free the junction from connective tissue. Small vessels are divided using bipolar electrocoagulation. Care is taken not to section ureteral blood vessels. A fourth trocar (3 mm) is inserted at the anterior axillary line, anterior to the first trocar. A stay stitch is placed at the junction. Aberrant crossing vessels are identified. The renal pelvis is partially divided using scissors at the most dependant part and gentle traction on the stay suture helps to define this point. Keeping the traction, the ureter is partially divided and incised vertically for spatulation. The traction suture helps to mobilize the ureter so the scissors can be in the axis of the ureter, usually introduced through the last trocar. The anterior surface of the kidney is left adherent to the peritoneum so that the kidney is retracted medially without the need for individual kidney retraction. The ureteropelvic anastomosis begins using a 6-0 absorbable suture with a tapered 3/8 of circle needle, placed from the most dependant portion of the pelvis to the most inferior point or vertex of the ureteral spatulation. The suture is tied using the intracorporeal technique with the knots placed outside the lumen. The same stitch is used to run the anterior wall of the anastomosis. The UPJ is kept intact for traction and stabilization of the suture line, and removed just before tying the last suture on the pelvis. A double-pigtail stent is inserted through the last trocar and its position in the bladder is assured under fluoroscopy. The posterior ureteropelvic anastomosis is then done. The pelvis is trimmed if needed. In case of aberrant crossing vessels, the technique is slightly different. After placement of the stay suture, the ureter is completely divided and the UPJ and the pelvis are delivered anterior to the vessels with the help of the stay suture. Then the anastomosis is performed as described. A Foley catheter is left in the bladder for 24 h postoperatively.

3. LOWER URINARY TRACT

Contrary to upper tract surgery, experience with laparoscopic procedures for the lower urinary tract is still in the era of development and evaluation. Rare indications specific to pediatric patients, such as excision of a complicated prostatic utricle, can be done successfully by laparoscopy, offering a good surgical view and permitting easy dissection in a deep and narrow pelvic cavity (52). The most common indication for lower urinary tract surgery in children is the treatment of vesicoureteral reflux. Major reconstructive operations are less frequent in pediatric than in adult surgery.

3.1. Treatment of Vesicoureteral Reflux

The surgical treatment of vesicoureteral reflux has been well established for several decades. Open ureterovesical reimplantation procedures are highly successful with success rate of over 99% in children. Efforts to improve these procedures have been directed toward reducing the perioperative morbidity and shortening hospitalization while maintaining the success rate.

3.1.1. Laparoscopic Extravesical Reimplantation

This technique was first reported in animal models with satisfying results (53). Early clinical reports (54,55) did not recommend the procedure because the advantages of the laparoscopic technique were outweighed by the disadvantages, such as long operative time, technical difficulty, and uncertain long-term results. Later, Lakshmanan and Fung (55) modified the Lich–Gregoire laparoscopic extravesical approach and reported 100% success rate with 71 refluxing ureters in 47 children. They recommended patient selection and advised open surgery for children younger than 4 years and also in cases of megaureter requiring ureteral tapering.

3.1.2. Endoscopic Intravesical Reimplantation

Okamura et al. (57) reported their initial experience with endoscopic trigonoplasty in 12 patients. Using the urethral route, a pneumo-bladder, and two trocars on the abdomen, the ureteric orifices were approximated close to the midline. Initially, vesico-ureteric reflux was eradicated in all patients. In a subsequent report, the use of the technique in a larger number of children with longer follow-up revealed a lower cure rate and higher rate of recurrence (58). At 12 months, there was no reflux in 59% of children and in 74% of adults. Trigonal splitting caused recurrence of reflux, which was greater than grade II. Gatti (59) reported a similar experience, but their results were improved using the Cohen technique instead of the previously described Gil–Vernet technique; resolution of reflux was obtained in five of six treated children. Recently, Jeff Valla, Nice, France, has presented a successful experience with laparoscopic Cohen reimplantation in four children, reproducing exactly the open surgical technique (presented at the annual meeting of the French Society of Pediatric Surgery, 5–7 September, 2001, Paris, France). His current series is now 20 patients without recurrence of reflux (personal communication). CK Yeung, Hong Kong, has nearly the same experience using transvesical 3-ports technique, with an average operative time of 2 h for bilateral cases (personal communication).

A new surgical concept with modified instruments, easily usable inside the bladder, may replace conventional intravesical surgery in children with a comparable success rate.

3.1.3. Endoscopic Subureteral Injection

Endoscopic management of reflux is by far the least invasive with respect to postoperative discomfort, cosmetic results, and hospital stay (in the majority of patients subureteral injection is performed on an outpatient basis). The remaining problem is the choice of bulking agent, which must be safe, stable, and effective on long-term follow-up.

Endoscopic management of vesicoureteral reflux in children has become an accepted alternative to open ureteral reimplantation in many centers. Until recently the most common bulking agent used was polytetrafluoroethylene. Puri and Granata (60) reviewed a considerable multicenter survey of 12,251 refluxing ureters treated endoscopically with subureteral polytetrafluoroethylene injection, and followed for 1 to 13 years. Subureteral injection failed to correct reflux in 554 ureters (4.5%). In this review, no clinically untoward effects were reported in any patient due to the use of polytetrafluoroethylene as an injectable material. However, intense local granulomatous reaction and migration to lungs and brain have been reported and have discouraged its further application by most urologists (61). More recently, polymethylsiloxane (Macroplastique, Uroplasty Inc., Geleen, The Netherlands), a silicone elastomer, has been introduced as a more ideal agent because of bulky consistency, lack of migration, minimal local reaction. Dodat et al. (62) have reported a significant experience in reflux treatment by endoscopic

implantation of polymethylsiloxane in 297 children with 454 refluxing vesicorenal units. Regardless of the etiology and the grade, reflux resolved in 91.2% of children (93.3% of ureters). The experience with this agent in North America is limited to one recent publication by Herz et al. (63), documenting a similar success rate in low-grade reflux but a lower success rate in high-grade reflux. Other substances, as autologous chondrocytes, have been used and reported in smaller numbers of patients, but had $<70\%$ success rate (64).

3.2. Major Lower Urinary Tract Reconstruction

Urogenital cancer and neurogenic bladder are the main indications for bladder replacement and augmentation in adults, and the most common indication in children is a small, poorly compliant bladder secondary to neurogenic bladder, posterior urethral valves, or bladder exstrophy. The development of laparoscopic radical prostatectomy in adults by Guillannau et al. (65) has opened a new era in laparoscopic reconstructive urologic surgery. Several techniques were reported in animal models to adapt laparoscopic techniques to standard bladder augmentation surgery (66,67).

Docimo et al., were the first to report laparoscopic gastrocystoplasty in a 17-year-old girl with a poorly compliant bladder (68). Their experience extended later to involve eight patients who underwent laparoscopic-assisted enterocystoplasty using a variety of bowel segments. The enterovesical anastomosis was performed using an open technique through a Pfannensteil or midline incision (69).

Gill et al. (70) have reported an initial clinical experience with laparoscopic augmentation enterocystoplasty using the ileum, sigmoid, or right colon in three patients with functionally reduced bladder capacities due to neurogenic causes. Bowel reanastomosis was performed by exteriorizing the bowel loop outside the abdomen through a 2-cm extension of the umbilical port site. The enterovesical anastomosis was done using an intracorporeal laparoscopic suturing technique. Various bowel segments can be fashioned, as with open surgery, including creation of a continent, catheterizable stoma.

Although preliminary results are encouraging and indicate the technical potential for major reconstructive surgery, the efficiency of these procedures and the benefits over the standard open technique still need further experience with larger series and long-term results. In pediatric patients, indications are less frequent and validation of the efficiency of these complex procedures should come through the experience of the adult urologists.

4. COMPLICATIONS OF LAPAROSCOPIC UROLOGICAL PROCEDURES

Complications of abdominal laparoscopy for urologic procedures, such as bowel and great vessel injury, have been documented in the adult and pediatric populations (71–73). In a multicenter survey involving 5,400 pediatric urological laparoscopic procedures, Peters (71) showed that the clearest predictor of complications was laparoscopic experience. Soulie et al. (73) have reported a decrease in complication rate from 9% for the first 100 to 4% for the subsequent 250 procedures. Most intraoperative complications (2.6%) were vascular and visceral injuries, while postoperative complications (2.8%) were predominantly thromboembolism and wound infection at trocar sites. Complications of retroperitoneal renal surgery are rare and mainly include vascular or colonic injury. Kumar et al. (74) reported a major complication rate of 3.5% in 316 patients (aged 4–88 years) who underwent retroperitoneal laparoscoscopic urological surgery. Vascular

injuries occurred in seven, five of which required immediate conversion to open surgery. Four patients (1.2%) had other major complications including colonic injury, retroperitoneal collections, and incisional hernia.

In our experience with 65 retroperitoneal nephrectomies, we have had one vascular tear at the origin of a lumbar vein in a case of xanthogranulomatous pyelonephritis (27). A clip without the need to convert to open surgery successfully closed the tear. In retroperitoneal procedures, traction on the kidney towards the top of the screen stretches renal vessels, reducing bleeding while evaluating the feasibility of the hemostasis by laparoscopic measures. Our only postoperative complication was a hematoma after pretransplant bilateral nephrectomy, which was drained percutaneously.

Pneumoperitonium secondary to peritoneal tear is commonly seen postoperatively in patients undergoing the retroperitoneal approach (2,74). This occurred in ~30% of our cases in the early experience, and could be avoided by careful preparation of the retroperitoneal space for insertion of the anterior working ports. When pneumoperitoneum occurs at the beginning of the procedure, the retroperitoneal working space is reduced by the effect of the pneumoperitoneum. This can be managed either by laparoscopic suturing of the tear or, if not possible, by inserting a Veress needle in the peritoneal cavity to evacuate the gas during the procedure. If the tear occurs after the ligation of the renal vessels, during dissection of the anterior surface of the kidney or the ureter, the procedure can usually be accomplished without specific management of the pneumoperitoneum.

Additional complications seen after partial nephrectomy are injury of the main renal pedicle or injury of the duodenum during right upper pole nephrectomy (2).

5. CONCLUSIONS

Indications for laparoscopy in pediatric urology are expanding, with more centers being involved in the evolution of various procedures. To avoid a discouraging learning curve, we recommend that pediatric urologists acquire their experience in a progressive pattern (75). Nephrectomy for multicystic dysplastic kidney or hydronephrosis is a relatively safe and easy procedure, which acquaints the surgeon with laparoscopic exposure to the upper tract. When the surgeon is familiar with this exposure he/she can proceed to more difficult nephrectomies (pretransplant, partial nephrectomy). Reconstructive procedures as pyeloplasty should be restricted to surgeons with advanced laparoscopic expertise.

Time can only be limited by training. Today, training is easily available in many centers of adult and pediatric surgery. Experienced peers are also available to accompany the surgeon during the initial experience, especially in the era of telerobotic surgery (76). This might improve the results during the initial experience with laparoscopy and encourage its development among a larger number of pediatric urologists.

Minimal access procedures emphasize our goals of improving patient comfort and safety while adapting the laparoscopic procedures as closely as possible to conventional surgical techniques with respect to the operative time, cost, and surgical principles.

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31
Minimally Invasive Pediatric Neurosurgery

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1. INTRODUCTION: HISTORICAL PERSPECTIVE

Minimally invasive surgical techniques have had a major impact on pediatric neurosurgery. The history of this development is actually reasonably long. The earliest reported neurosurgical endoscopic procedure was by Lespinasse (1) who was a Chicago urologist.

He fulgurated the choroid plexus bilaterally in two infants with hydroceplus using a small cystoscope: one died immediately after the operation. A few years later, in 1918, Dandy (2) devised a new treatment for hydrocephalus. He used a hand-held cystoscope illuminated by room light and inserted it into the lateral ventricles to drain in cerebro-spinal fluid and avulsed the choroid plexus. Three of the four patients died. In 1922, he reported using a cystoscope to aid coagulating the plexus and coined the term Ventriculoscope (3).

The field was relatively static, due to this initial failure rate, until Harold Hopkins, a British physicist, designed the rod lens system and flexible fiberoptic lenses in the 1960s. These systems tremendously improved the quality of images and illumination. In the 1980s, video endoscopy was developed using the charged coupled device (CCD) technology allowing multiple observers and enhancing teaching and documentation.

Neurosurgeons typically work with a single portal and scope with minimal diameter and inside the brain, normally under water (Fig. 31.1). Larger rigid scopes—under a cm in diameter—with multiple working channels and an irrigation system are typically used. Common working instruments include coagulation probes, scissors, and dissectors. Smaller rigid scopes as well as flexible fiberoptic endoscopes are used for inspection (Fig. 31.2).

2. HYDROCEPHALUS

Hydrocephalus results from a pathological accumulation of cerebrospinal fluid (CSF) within the ventricles of the brain at increased pressure (Fig. 31.3). Untreated it leads to progressive neurological injury and death. The commonest form of treatment, cerebrospinal fluid shunts have very high complication rates including shunt malfunction, shunt infections, altered CSF hemodynamics, and overdrainage causing slit-ventricle syndrome (4). The commonest cause of hydrocephalus amenable to endoscopic surgery is obstruction of the cerebral aqueduct (connecting the third to fourth cerebral ventricles) from pathologies such as tumours or congenital stenosis of the aqueduct (5) (Fig. 31.4). Third ventriculostomy, or creating a hole in the floor of the third ventricle, bypasses this block and more importantly avoids the use of a shunt. The pediatric group of patients suffering from hydrocephalus benefits most from this procedure since they suffer the

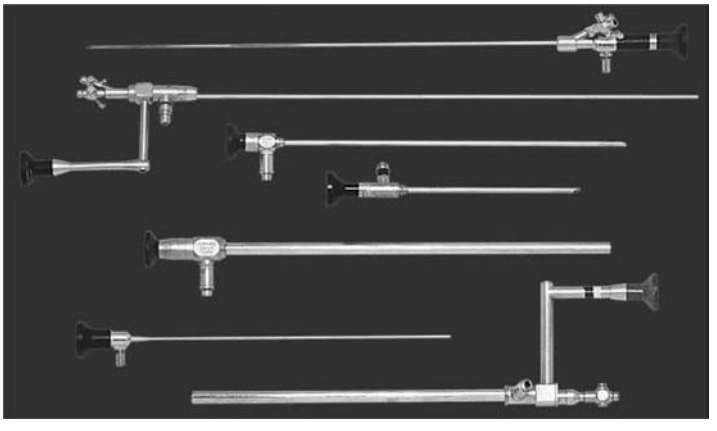


Figure 31.1 Neuroendoscopes are available in different sizes and lengths.

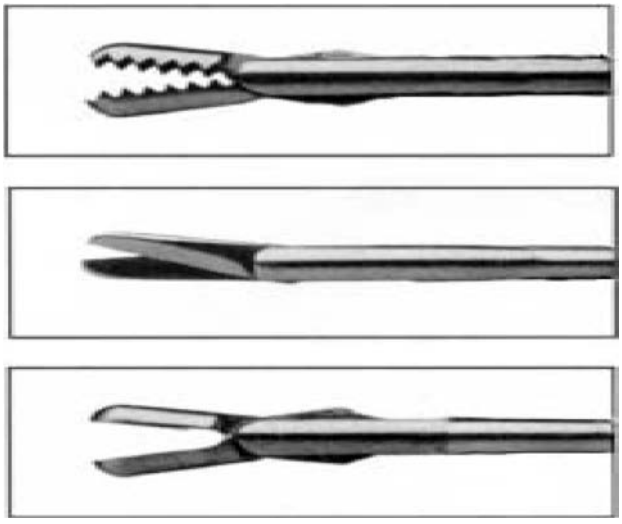


Figure 31.2 Micro-forceps and scissors are used through the operative channels of the endoscope.

longest with the disease and are most prone to shunt complications and repeated revisions (6).

Aqueductal stenosis is usually present in the second decade of life. The common presenting symptoms are headache, gait disturbance, ataxia, cognitive dysfunction, enlarged head size and endocrine dysfunction. The possible etiologies of aqueductal



Figure 31.3 CT of obstructive hydrocephalus.



Figure 31.4 Obstructive hydrocephalus by 4th ventricular tumor. Arrow showing the trajectory of the endoscope and the place of ventriculostomy.

stenosis include neonatal or infantile infection (7), x-chromosome transmitted (8), and vitamin deficiency (9). However in the majority of cases, a known cause is absent.

The diagnosis of aqueductal stenosis is by demonstrating enlargement of the lateral and third ventricles and a small fourth ventricle, also known as tri-ventricular hydrocephalus. Further investigation by an MRI scan may demonstrate the site of blockage and lack of flow. Also, the MRI should show a downwardly bulged floor of the third ventricle (10).

2.1. Third Ventriculostomy

Dandy (11) was the first to conceive of an internal shunt for obstructive hydrocephalus. Third ventriculostomies were initially performed as open operations and then percutaneously (12) using stereotactic guidance (discussed subsequently) under radiographic control. However, modern neuro-endoscopy has made this technique safer by using direct visualization. For the technique, the patient is put in a supine position. A frontal burr hole is placed about 3 cm lateral from the midline on the coronal suture. After opening the dura, the endoscope is inserted either according to surface landmarks under computer-assisted navigation (discussed subsequently) into the lateral ventricle. Neuronavigation is particularly useful in cases where the lateral ventricles are not dilated, in order to choose the optimal trajectory aiming at the floor of the 3rd ventricle. In most instances, a rigid endoscope with working channels is used.

The next step is to identify the foramen of Monroe, assisted by the identification of the normal venous anatomy and choroid plexus (Fig. 31.5). The endoscope is passed through the foramen of Monro into the 3rd ventricle under direct vision. The floor of the 3rd ventricle is inspected (Fig. 31.6). The site of ventriculostomy is between the infundibular recess of the pituitary stalk and the mammillary bodies to enter into the prepontine cistern. The floor is usually thinned out into a translucent membrane from chronic hydrocephalus, and the Basilar artery which sits in front of the mammillary bodies can be

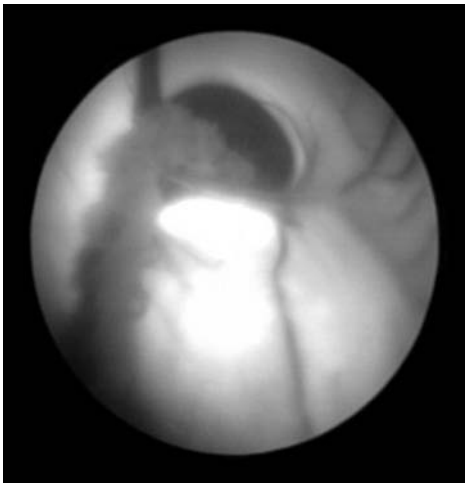


Figure 31.5 Endoscopic view of foramen of Monroe.

visualized and avoided. In cases of thickened floor, because of previous infection or fibrosis, there is a potential risk of injuring the Basilar artery.

Many techniques have been described to perforate the floor of the 3rd ventricle, such as using the endoscope itself (13), blunt probe (14), balloon catheter, monopolar or bipolar coagulation (15), and laser (16). Heat producing methods such as coagulation or laser may be associated with higher risk and are generally not recommended. Traumatic Basilar artery aneurysm after 3rd ventriculostomy has occurred (17). The most acceptable and safe method is to perforate with the blunt probe. After perforation, a Fogarty balloon catheter of 3–5F is gently repeated inflated to enlarge the opening. The endoscope is then passed through the stoma to confirm free flow of CSF into the prepontine cistern. Coexistent tumors may also be biopsied. Tumors situated in the posterior part of the third ventricle, around the corner from the foramen of Monro, may require a flexible scope for access, to avoid injury to the venous structures and choroid plexus.

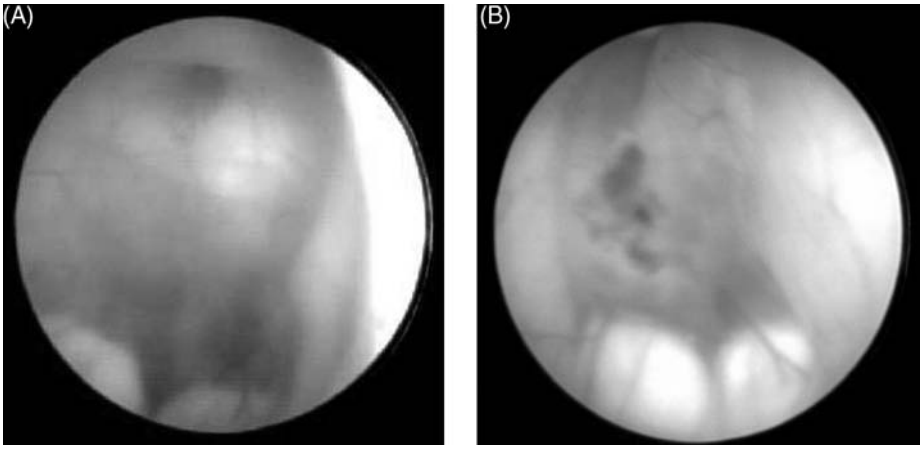


Figure 31.6 (A) Floor of 3rd ventricle and (B) opening created at floor of 3rd ventricle with balloon catheter.

The success of 3rd ventriculostomy is assessed both radiologically and clinically. Radiographically, the patency of the opening is assessed by MR flow studies, which show flow void signal through the floor of the 3rd ventricle. Lev et al. (18) described a method of determining the functional status of 3rd ventriculostomies by using phase-contrast MR velocity measurements, finding the ratio between the high pontine cistern and the space anterior to the spinal cord.

The ventricular size may or may not change significantly compared to preoperative images, especially in patients with long-standing hydrocephalus (19). But radiographic evidence of raised intracranial pressure, such as peri-ventricular signal changes, is absent. Schwartz looked at the change in ventricular size after 3rd ventriculostomy and found that a decrease in the 3rd ventricle size is a more reliable indicator of a successful outcome.

Recently Hopf et al. (20) looked at the outcome of a heterogenous group of 100 consecutive patients who underwent 3rd ventriculostomy, including both adult and pediatric group of patients. The overall clinical improvement was 76%. Best results were in the group of patients with benign space-occupying lesions causing obstruction in CSF flow, with a success rate of over 90%. In contrary, patients with progressive tumorous lesions have a success rate of only 64%.

A majority of patients suffering from spinal dysraphism also develop hydrocephalus. Jones et al. (21) reported 25 patients in which he treated 11 patients <6 months of age but with one success only. The group also reported 69 patients with myelomeningocele and hydrocephalus. Fifty patients were shunted previously. The overall success with 3rd ventriculostomy was 72%.

The optimal timing of 3rd ventriculostomy is unknown. Most operators think that the procedure is significantly more effective in pediatric patients who are older than 2 years of age. Teo and Jones (22) reported a poor success rate for patients with spinal dysraphism and hydrocephalus operated at <6 months of age.

Performing third ventriculostomy in place of a shunt revision is an attractive alternative in patients plagued by repeated shunt failure or infection. However, long-term shunting in patients previously having normal CSF absorptive ability may lead to atrophy or involution of the resorption sites, leading to failure of third ventriculostomy. Kehler et al. (23) illustrated this point in two patients who experienced symptoms of raised intracranial pressure in the first few days after ventriculostomy. Both patients drastically improved after the initial period. The author postulated that the initial high pressure was required to "reopen" the CSF absorptive channels during this adaptation period. Cinalli et al. (24) reported a series of patients with shunt malfunction treated with 3rd ventriculostomy, with an overall success rate of 76% at median follow-up of 8.7 years. Eight out of 13 patients with shunt infection had their shunt removed successfully after ventriculostomy.

Third ventriculostomy is not without risk. Although most complications have been few and relatively mild, serious complications including cardiac arrest, hypothalamic damage, and bleeding and laceration of the basilar artery leading to the formation of basilar tip aneurysm, and neurological disability have been reported.

3. ENDOSCOPIC TREATMENT OF INTRACRANIAL CYSTS

Benign intracranial cystic lesions include arachnoid cysts, intraventricular cysts, porencephalic cysts, and colloid cysts. These cysts normally have a very slow rate of growth. The indications for treatment of these cystic lesions include hydrocephalus from obstruction of

the CSF circulation pathway (25), focal neurological deficit, seizure disorder, and nonspecific symptoms such as headache.

Conventional treatment options for symptomatic intracranial cysts include resection or marsupialization of the cyst, fenestration of the cyst into the basal cisterns to establish a communication with the normal CSF circulation. Both methods have previously required a formal craniotomy. Another option is shunting of the cyst to an extracranial compartment; most commonly to the peritoneum. However, the shunt apparatus, like any implanted hardware is prone to malfunction and infective complications. With the recent advance of neuroendoscopy, cyst fenestration may be achieved through a less invasive approach.

Hopf and Perneczky (26) have classified endoscopic use in the treatment of intracranial cysts according to the optical device used; whether the endoscope was used alone or in conjunction with the microscope, and whether the surgical manipulation is done outside or inside the endoscope. The procedures were categorized into pure endoscopic neurosurgery (EN), endoscope-assisted microneurosurgery (EAM) and endoscope-controlled microneurosurgery (ECM).

Endoscopic neurosurgery (EN) is best for deep-seated lesions such as intraventricular cysts where there is no extension to accessible regions of the skull. All the manipulations are performed with the endoscope. For cysts with the extensive contact with the accessible part of the skull, such as in the case of large middle fossa cyst, Hopf recommended the use of EAM. This combines the strength of both microscopy and endoscopy. Difficult and hazardous dissections are done by conventional microsurgical techniques under the microscope. The endoscope is used to aid inspection of areas not accessible by the microscope.

For ECM, microsurgical manipulation is done solely under endoscopic guidance. It is used in lesions where there is moderate contact to accessible regions of the skull, requiring difficult dissection. The endoscope provides an excellent view and the surgeon can use a wide variety of familiar microsurgical tools.

3.1. Arachnoid Cysts

Arachnoid cysts are developmental in origin and were described by Bright (27) more than 170 years ago. An arachnoid cyst is a collection of CSF-like fluid in between the split layer of the arachnoid membrane under pressure. They are lined by a single layer of flattened arachnoid cells. The pathogenesis is still unknown. Several mechanisms have been proposed for their origin: the ball-valve mechanism (28) and the abnormal secretion of CSF into the cysts (29). Schroeder reported a case of suprasellar arachnoid cyst where they observed a slit-valve-like mechanism that opened and closed synchronously with arterial pulsation.

They have a special affinity for the middle cranial fossa. Wester (30) found that 65% of 126 patients had arachnoid cysts located in the middle cranial fossa. The second most common location is the posterior fossa, followed by suprasellar location and in the quadrigeminal cistern. Approximately 75% of patients present in the pediatric group. They present with signs and symptoms of raised intracranial pressure, headache, and midline shift on imaging studies. Others may have seizure disorders. Patients with posterior fossa lesions may present with cranial nerve deficits or hydrocephalus from obstruction of CSF flow. Some arachnoid cysts are small and are discovered incidentally. Galassi et al. (31) devised a classification for arachnoid cysts in the middle cranial fossa based on their appearance.

- Type 1 cysts are small lenticular-shaped cysts located proximally in the Sylvian fissure at the anterior pole of the middle cranial fossa immediately posterior to the sphenoidal ridge and not associated with any mass effect or midline shift.
- Type 2 cysts appear rectangular in shape and involve the proximal-to-mid aspect of the fissure outlining the insular cortex. Minimal midline shift may be present.
- Type 3 cysts are large lenticular-shaped lesions, often with significant midline shift.

3.1.1. Treatment

Arachnoid cysts that are small and are found incidentally are treated conservatively in view of their benign and slow-growing nature. Some cysts may become symptomatic by expanding and compressing the surrounding brain tissue (Fig. 31.7). For patients exhibiting evidence of raised intracranial pressure, focal neurological deficits, and intractable seizures (28,32) surgical intervention should be considered if these symptoms are referable to the cyst. Surgical options include open craniotomy and fenestration, with or without marsupialization of the cyst, endoscopic fenestration, and shunting.

3.1.2. Cystocisternostomies/Ventriculocystostomies

In fenestration surgery, part of the cyst wall is excised to enter into the cyst. The medial aspect of the cyst wall is then fenestrated into the basal cistern (cysto-cisternostomies), or the ventricular system (ventriculo-cystostomies), creating a communication between the cyst and the CSF circulating space (Fig. 31.8). The relatively avascular nature of arachnoid cysts makes these lesions particularly well suited for endoscopic fenestration. Substitution of the endoscope for the microscope permits the surgeon to work in a very small space and eliminates the need for brain retraction.

The procedure is performed under general anesthesia. A burr hole is made over the area where the arachnoid cyst is situated. After opening the cyst, a rigid endoscope is introduced into the cyst under constant gentle low-pressure irrigation. This is important

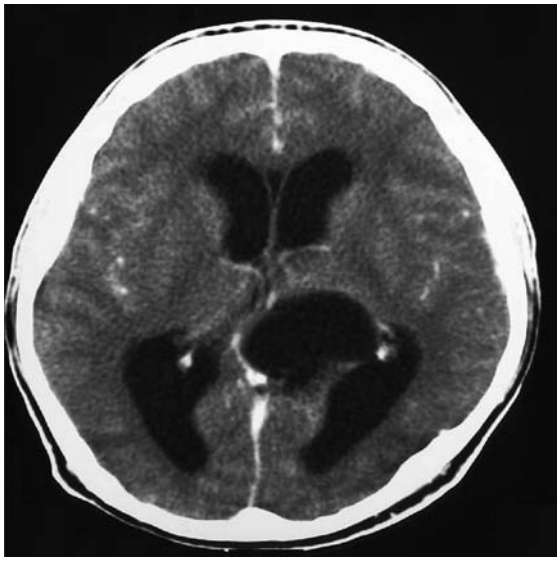


Figure 31.7 Arachnoid cyst obliterating posterior part of 3rd ventricle and aqueduct.

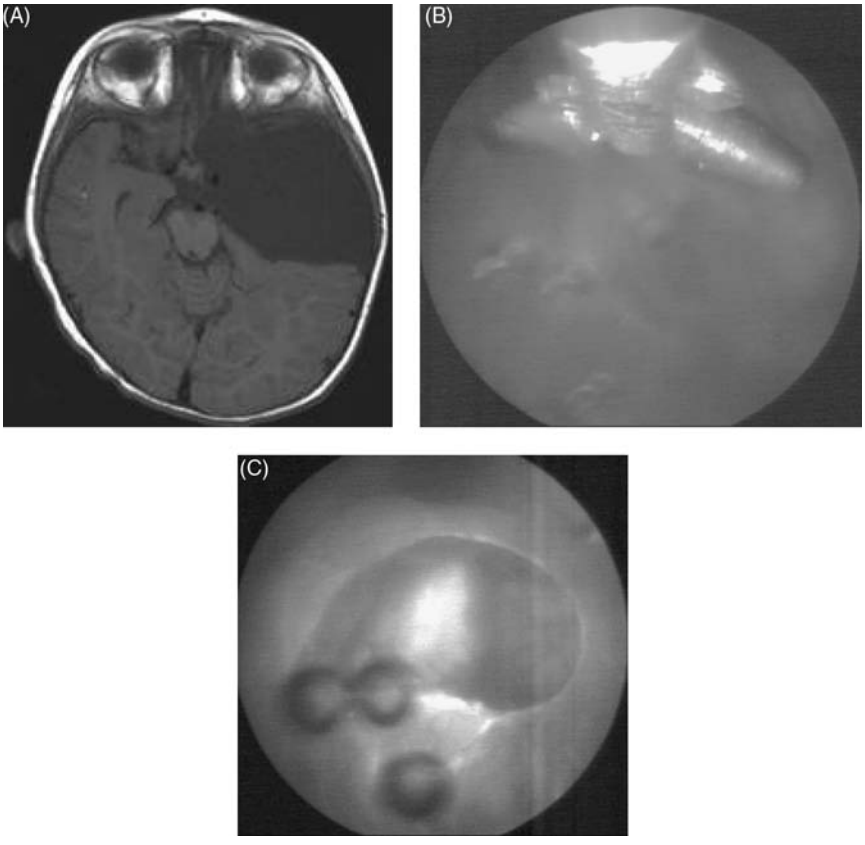


Figure 31.8 (A) Large left temporal arachnoid cyst; (B) fenestration of arahnoid cyst with microforcep and (C) opening created between arachnoid cyst and basal cistern.

for all neuroendoscopic procedures in order to maintain optimal visual feedback. After inspection of the cyst wall, the thin, often translucent arachnoid membrane on the medial aspect is opened into the basal cistern or the ventricle. This may be achieved by micro-grasping forceps or blunt hook. The opening is then enlarged with the help of a balloon catheter. The endoscope is then advanced further through the opening to ensure the two compartments are freely communicating.

McComb reported 36 children with middle fossa arachnoid cysts. The majority were symptomatic. Those that were symptomatic showed a progressive increase in size on imaging studies. Thirty-five patients were successfully fenestrated with one patient requiring a shunt operation. Schroeder reported 11 patients with arachnoid cysts who underwent endoscopic fenestration. The follow-up was up to 45 months. Ten patients had decreased cyst size and nine patients had resolution of preoperative symptoms. Others have achieved less dramatic results. Hopf reported only half of their patients with arachnoid cyst were successfully fenestrated on long-term follow-up. Neuroendoscopy, however is not a risk-free procedure. Small veins or perforating vessels may be lacerated during the procedure. Any small amount of bleeding will severely affect the quality of the visual image, making hemostasis more difficult. Overall the success rates of endoscopic fenestration of middle fossa arachnoid cysts have been reported to be $\sim$50–70%. Patients who failed the procedure continue to have symptoms and signs related to the lesion, requiring shunting operation.

3.1.3. Cystoperitoneal Shunt

Another treatment option for arachnoid cysts is to divert the cyst fluid to an extracranial space; in most cases, the peritoneum. Some authors reserve shunting for cases where open or endoscopic fenestration have failed. Others use shunting as their first line treatment believing that up to 50% of fenestrated cases eventually require placement of shunt. The advantage of shunting has been stated to be a minor operation with less risk and fewer complications (25,33). Arai et al. (34) reported 77 patients with middle fossa cysts that were treated with cystoperitoneal shunting. A total of 12 revisions were required for infection and shunt malfunctions, concluding that shunting is a safe and viable option to cyst fenestration.

The controversy continues regarding the best surgical treatment. In experienced hands, endoscopic fenestration is a safe and effective procedure. A proportion of the patients can be treated without implantation of hardware. Should the endoscopic procedure fail, microsurgical fenestration or cystoperitoneal shunting can then be performed.

3.2. Colloid Cyst

Colloid cysts are located in the roof of the third cerebral ventricle at the foramen of Monroe and represent ~1% of all intracranial neoplasms (35) (Fig. 31.9). They can either intermittently or continuously obstruct the foramen of Munroe, producing enlargement of the lateral ventricles. Although they are histologically benign, the natural history is not clearly known. Some cysts are asymptomatic and remain static in size radiologically for many years (36). On the other hand, colloid cysts are known to be associated with sudden death as a result of acute obstruction of the CSF pathways (37). The current consensus is that symptomatic cysts such as in patients with headache, nausea, gait disturbance, coma, and ventricular dilatation should be treated aggressively. However the treatment of small, asymptomatic cysts remains controversial. Treatment modalities include simple shunting of the ventricles to relieve the hydrocephalus, stereotactic cyst aspiration, craniotomy, and microsurgical excision of the cyst and neuroendoscopic fenestration of the cysts (Fig. 31.10).

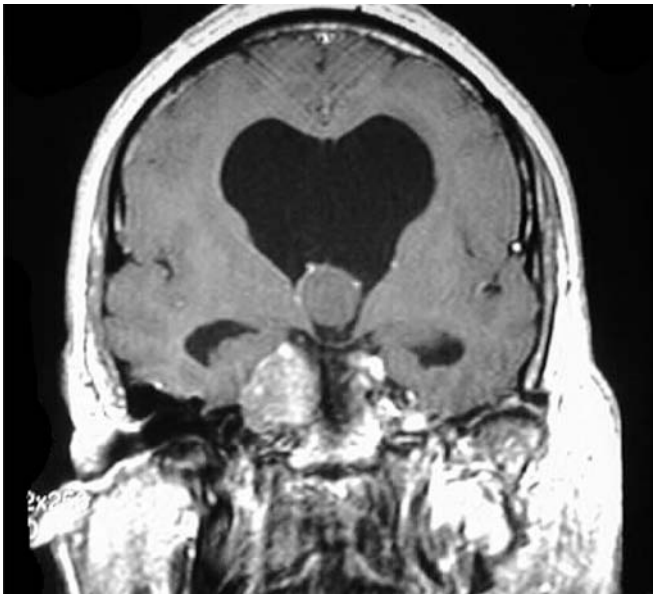


Figure 31.9 Colloid cyst obstructing foramen of Monroe causing obstructive hydrocephalus.

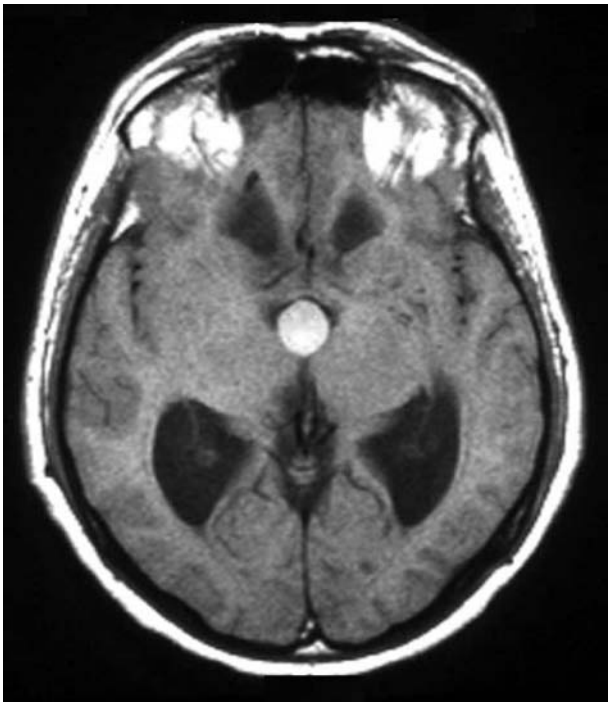


Figure 31.10 Colloid cyst: axial MRI scan.

3.2.1. Shunting

Colloid cysts cause symptoms by producing hydrocephalus. The simplest treatment to alleviate the hydrocephalus is to insert a ventriculoperitoneal shunt. However, shunting alone is not an ideal treatment. The colloid cyst often obstructs both foramen of Monroe, requiring shunting of both lateral ventricles separately. Also shunts are prone to blockage, which precipitate the patient to sudden obstructive hydrocephalus. Therefore, simple shunting is not longer considered acceptable.

3.2.2. Cyst Aspiration

Another option is stereotactic aspiration of the colloid cyst. It was first described in 1975 (38). This is probably the least invasive method. Successful aspiration depends on the viscosity of the cyst content and the size of the cyst. Some colloid cysts have very viscous content rendering needle aspiration difficult or impossible. Cyst size is also important in the success of aspiration. It is more difficult to puncture a small firm cyst than a big cyst with the needle. Kondziolka and Lumsford (39) reported a series of 22 patients with colloid cysts. Stereotactic aspiration alone was successful in 11 patients. However simple aspiration, leaving the cyst wall behind, may run the risk of recurrence. Mathiesen et al. (40) looked at the long-term outcome of a series of 16 patients with colloid cysts treated by stereotactic-guided aspiration. Thirteen of these patients required reoperation due to an acute comatose state, development of hydrocephalus or regrowth of the cysts.

3.2.3. Craniotomy

Open surgical removal is the gold standard for colloid cysts of the third ventricle. This is the only method that can remove the cyst totally in most instances to ensure a definite cure

for this condition. Dandy (41) in 1930 first described the open surgery for colloid cysts using the transcortical–transventricular approach. Advocates of craniotomy and microsurgical approach argue that a stereotactic-guided transcallosal approach with total removal, utilizing a 1 cm opening in the corpus callosum, is as minimally invasive as a 0.5 cm endoscopic tunnel through the cortex. Total removal offers a better long-term result.

3.2.4. Endoscopic Removal

Guiot et al. (42) in 1963 first reported direct visualization of a colloid cyst using a rigid endoscope. Today, the procedure is performed with the patient under general anesthesia. A skin incision is made about 3 cm lateral from the midline at the level of the coronal suture. A burr hole is made, the dura is opened, and the neuroendoscope is inserted into the lateral ventricle either freehanded or under image-guided navigation if the lateral ventricles are small. The colloid cyst is first aspirated, which may be more difficult in cysts when the colloid material is very viscous. The cyst wall is then opened widely with microscissors and forceps, facilitating further aspiration of its contents. The cyst wall is excised carefully in a piecemeal fashion. Any residual cyst wall is then coagulated to decrease the chance of cyst recurrence.

Compared to open craniotomy and cyst excision, endoscopic approach cannot usually accomplish complete removal. Attempt to remove the cysts completely by endoscopic route may be hazardous and may cause damage to surrounding structures such as the fornices. The advantage of endoscopy is that this technique is a less invasive technique with postoperative care being as simple as that required after a stereotactic puncture. Longer follow-up is needed to assess the efficacy of endoscopic excision before this technique is considered as the treatment of choice for colloid cysts of the third ventricle.

4. ARTERIO-VENOUS MALFORMATION (AVM)

An anteriovenous malformation (AVM) is an abnormal collection of high-flow vascular channels supplied by arterial feeders with outflow into draining veins. In the pediatric population, it accounts for 30–50% of all intracerebral hemorrhages in children (43,44). A large proportion of patients present with hemorrhage; some pediatric series reported up to 85% with hemorrhage as their initial presentation (45,46), compared to the adult incidence of ~60% (47). Other presentations include seizure and focal neurological deficit either from mass effect or from vascular steal phenomenon by the AVM channelling the blood away from surrounding functioning brain.

The best treatment for AVM in children remains controversial and the management of AVMs have changed tremendously over the last few decades due to the development of stereotactic radiosurgery and microvascular embolization. The options for treatment of cerebral AVMs in general include a combination of microvascular resection, endovascular embolization, stereotactic radiosurgery, and conservative treatment in patients where the risk of treatment is determined to be too high. However, because of the high rate of bleeding during the child's lifetime, conservative treatment is seldom recommended. The most commonly quoted annual risk of hemorrhage from a cerebral AVM is 2–4% (48,49). For an annual risk of 2%, the projected risk of hemorrhage over 50 years is 65%.

5. GENERAL GOAL OF TREATMENT OF AVM

In the treatment of cerebral AVMs, the goal is complete obliteration of the nidus with preservation of neurological function.

5.1. Microsurgical Excision of AVM

Many authors have recommended surgical removal as the first line treatment for small and accessible AVMs. Complete surgical excision offers an immediate cure and the patient is free from any future risk of hemorrhage from the lesion (Fig. 31.11). However, in high-risk lesions, surgical treatment may be associated with significant morbidity and mortality (45). Spetzler and Martin have devised a widely used classification of cerebral AVM based on the size, venous drainage, and proximity to eloquent areas of the brain with regard to the risk of surgery. Low-grade AVMs are treated with surgical resection, with or without preoperative embolization. When a lesion is considered high grade and too

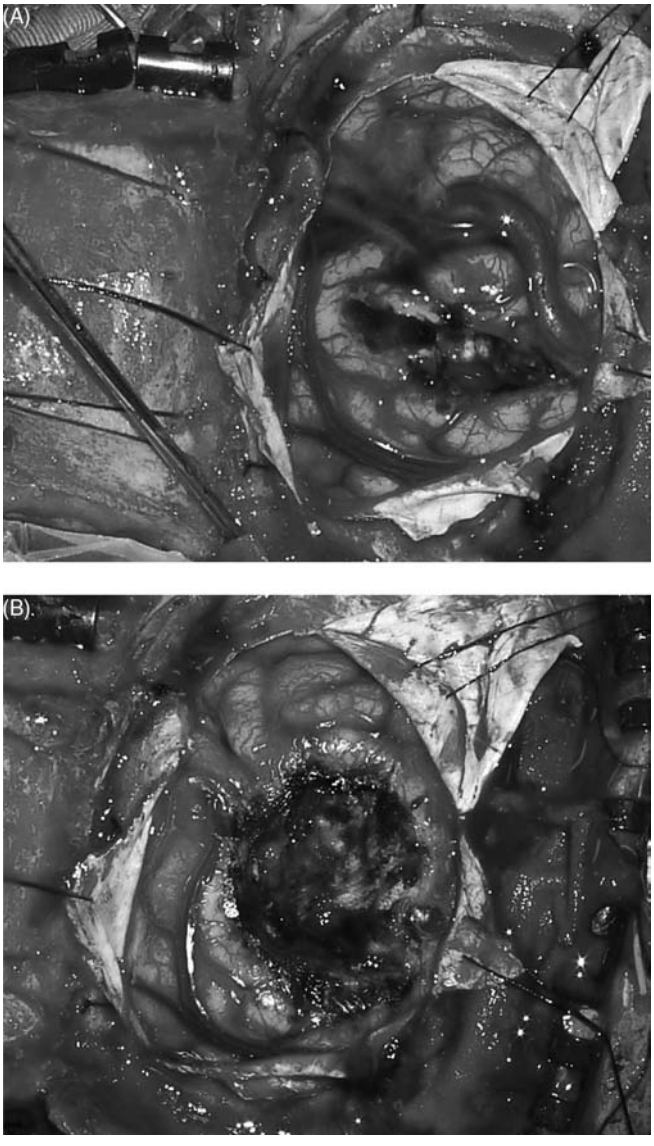


Figure 31.11 (A) Operative view of a large AVM demonstrating the large draining vein and (B) after total excision.

dangerous to be tackled surgically, other modalities like embolization and radiosurgery are considered.

Before surgery, the patient's cerebral angiogram and MRI scans are studied in detail. From the angiogram, the number and position of the arterial feeders, the nidus, and the draining veins are identified. The details of surgical approach is further planned from the anatomical information obtained from the MRI. For AVMs that are deep-seated and small, image-guided neuronavigation system can be very helpful in guiding the surgeon to the AVM with minimal disturbance to surrounding tissues. Once the AVM is identified, the nidus is carefully dissected from the surrounding brain. The arterial feeders are identified, coagulated, and sectioned. As the feeders are divided, the pressure and flow inside the nidus decreases and become less engorged. The draining vein(s), which may be enormous in size, must be kept patent and not disturbed until the very end. Otherwise the back pressure will cause catastrophic rupture of the AVM.

5.2. Embolization of AVM

Embolization of AVM in the adult population is done with the patient awake, so that the patient can be assessed clinically throughout the course of the procedure. Some unique issues pertained to the pediatric group. In infants or small children, vascular access required for endovascular procedure may be difficult or impossible. Embolization of pediatric AVMs is usually performed via the transfemoral route under general anesthesia (50) (Fig. 31.12). A femoral sheath is placed in one of the femoral arteries. Then flow-directed or steerable microcatheters of various sizes and configurations are passed through cerebral vessels into individual feeding arteries. Contrast material is injected to obtain a superselective angiography, which is used to plan the area of embolization and determine the rate of flow through the nidus. The angiogram can also be brought up during embolization as a superimposed image providing a road map to aid negotiation of catheters through tortuous feeding vessels. Special glue, n-butylcyanoacrylate (NBCA) is the agent most commonly used for embolization of AVM. Once set, it permanently occludes an area of the nidus. It has been shown from experience that solidified NBCA is relatively soft and does not cause difficulties when the surgeon resects the AVM. Other particulate materials, although are safer and easier to use, tend to recanalize with time and are not used for AVM embolization. NBCA is mixed into different concentrations, which have different solidifying time. The goal is to let the glue set inside the nidus without getting it into the draining vein or the feeding artery. Blocking the draining vein will cause outflow obstruction and increase in back pressure, exposing unprotected areas of the AVM to risk of rupture. On the other hand, blocking the feeding artery will preclude further access into that segment of the AVM. Embolization may be done in a single session or as a staged procedure, especially for patients with large AVMs, blocking off a number of feeding arteries at a time. This stepwise reduction in nidus flow and size would decrease the risk of postprocedure perfusion breakthrough bleeding as the residual AVM and the surrounding brain can adapt to the change in hemodynamic gradually.

In the pediatric population, endovascular embolization is seldom used alone as the treatment of AVMs. It is used as an adjunct with either surgery or radiosurgery. With microsurgery, embolization is used to reduce flow and to occlude deep-feeding arteries that are difficult to control during the course of dissection. For preradiosurgery embolization (51), the goal is to permanently reduce the size of the nidus since a smaller nidus has a higher chance of obliteration and lower morbidity (52).

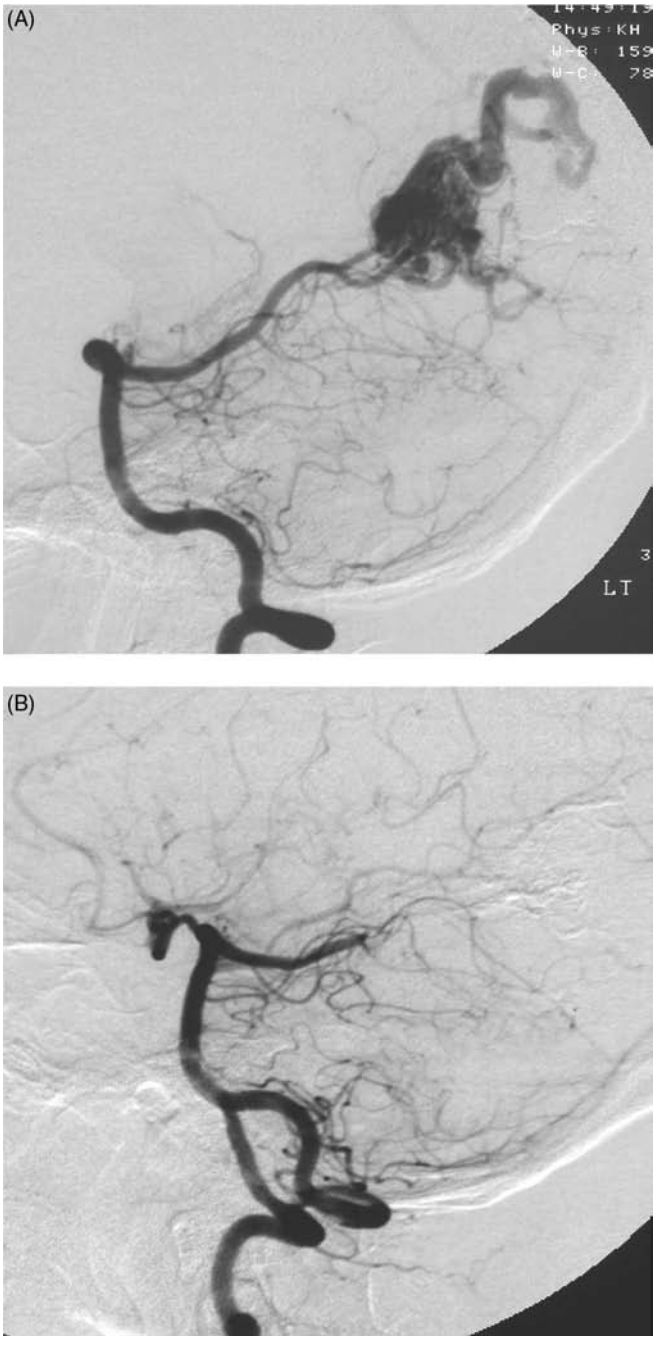


Figure 31.12 (A) Vertebral artery angiogram: occipital lobe AVM and (B) after embolization.

5.3. Radiosurgery

Radiosurgery is a single-fraction radiation treatment. It causes obliteration of AVMs by inducing a gradual sclerosing process of intimal hyperplasia of the abnormal vessels, ultimately leading to thrombosis. This is a slow process, which takes place over a course of

few months to few years. The basic principle of stereotactic radiosurgery is beam convergence. Irradiation is delivered from different directions with the lesion placed in the centre focal point. Steiner et al. (53), in 1972, first reported treating a patient with cerebral AVM by Gamma Knife Radiosurgery. Today the most widely used systems are the linear accelerator (LINAC)-based system and the gamma knife (GK) system; other new systems are emerging and are gaining clinical popularity. The LINAC system uses a mechanical modification of the existing linear accelerator apparatus for conventional radiotherapy. Beam convergence is achieved through adjustments of the table and gantry angles. The lesion is irradiated while the gantry rotates through a pre-set range. The GK system utilizes multiple (106 in total) cobalt radiation source directed at the lesion. Because of the precision required, mechanical accuracy and calibration must be achieved down to the millimeter level. Another important principle of radiosurgery is a steep gradient of irradiation at the treatment field edges. This has the effect of marked reduction of irradiation dose to the surrounding normal structures. The average distance achieved from 90% to 50% isodose line is 2 mm and from 90% to 20% is 4 mm (54,55). As a result, only a small volume embracing the lesion receives a significant irradiation. In general, the irradiation profile and accuracy of LINAC and GK do not differ significantly.

Stereotactic radiosurgery had been widely utilized in the adult population in different fields of neurosurgery. Indications include growth of tumors, both primary and secondary, arteriovenous malformation, and in functional neurosurgery. In the pediatric population, the use of radiosurgery had been more limited in view of the yet uncertain effect of radiation on the developing brain. The only established indication is in the treatment of cerebral AVMs. The success of radiosurgery is significantly influenced by AVM volume and the radiation dose delivered. The idea lesion for radiosurgery is a small, deep-seated AVM.

Radiosurgery for AVM is a one-session treatment. A typical stereotactic radiosurgical treatment is divided into three stages: image acquisition, planning, and treatment. It begins with fixation of a stereotactic head frame to the head of the patient. This is fixed rigidly by applying threaded pins to the outer table of the skull for the entire procedure. The frame may be MRI compatible if MRI images are to be obtained on the day of treatment. Alternatively, MRI images taken sometime before the treatment can be incorporated or fused into the treatment plan by specially designed software. Children $<13-15$ years old are less cooperative and less tolerant to long procedures; radiosurgery is usually done under anesthesia. For patients over this age, the treatment is done under sedation.

With the head frame positioned, a contrast enhanced MRI scan or CT scan is obtained to define the nidus of the AVM. MRI studies obtained before the day of treatment can be incorporated into the CT scan using sophisticated fusion software. The patient is then transported to the angiogram suite for lateral and anterioposterior angiography to further define the anatomy of the nidus. Intra-arterial angiography is the gold standard for imaging of AVMs. However it only provides two-dimensional information, whereas CT and MRI are helpful in demonstrating the lesion in three-dimensions. For lesions situated in eloquent areas of the brain such as the motor and speech centers, useful images from functional MRI can also be incorporated for planning. All the images are transferred to the computer planning workstation where the radiosurgical team consisting of neurosurgeons, neuro-oncologist, and neuroradiologist will formulate a treatment plan. The nidus is outlined in the different imaging modalities for optimal target localization. Critical structures like the optic apparatus and brainstem are also marked to minimize the radiation dose received. Since the AVM nidus is usually irregular rather than spherical in shape, a conformal plan needs multiple isocenters of various sizes. The neuro-oncologist will then calculate and decide on the radiation dose delivered by each isocenter. The marginal dose

is determined, which is the amount of radiation delivered to the margin of the lesion. This marginal dose is designed that it is situated at the steep drop-off region of the radiation curve. Therefore the centre of the nidus receives higher and the surrounding normal brain tissue is subjected to a lower radiation dose. Treatments are prescribed to the 80% isodose. The actual dosage delivered depends on the localizations, size of the nidus, and the radiosensitivity of critical structures. A smaller dose is prescribed for a larger lesion. The usual marginal dose is $\sim$18–20 Gy.

The patient is then transported to the radiosurgery couch for treatment. After treatment, the stereotactic frame is removed and the patient discharged the following morning. Patients are maintained on anti-convulsion medications if they have history of seizures. For larger AVMs after radiosurgery, a short course of steroid may be given to decrease the amount of surrounding edema.

The end point of treatment of AVM is complete obliteration of the nidus. For radiosurgery, there is a latent period of $\sim$2–3 years. Cerebral angiogram is the gold standard to confirm the absence of abnormal AVM vessels and normalization of cerebral vasculature. The angiographic finding would be one of progressive obliteration of the nidus and gradual decrease of caliber of the dilated feeding arteries. However, because of the invasiveness of the procedure, angiography cannot be repeated frequently to assess the degree of obliteration. Therefore, after radiosurgery, patients are followed by serial cranial MRI studies, which is less expensive and more acceptable to patients, every 6 months. MRI has a predictive value of 91% for nidus obliteration (56). If imaging indicates that the nidus is obliterated and there is no abnormal flow-void on the MRI scan, a set of cerebral angiograms are obtained for confirmation.

Complications of radiosurgery for AVMs is directly associated with the dose delivered and volume treated. These include an area of alopecia for AVMs close to the brain surface, evidence of edema (57) surrounding the AVM, increased seizure activity, and neurological deficits. The greatest risk after radiosurgery is the persistent risk of hemorrhage. Although studies have shown that radiosurgery does not increase the risk of hemorrhage (58,59), it takes 2–3 years after treatment to achieve vessels obliteration; during this period, the patient is unprotected from further bleeding (60). Engenhart-Cabillic and Debus (61) reported a series of 145 patients with AVMs treated by radiosurgery over a 10-year period; eleven patients suffered from hemorrhage between 4 months and 7 years after radiosurgery.

5.4. Success of Radiosurgery for AVMs

The obliteration of AVMs with radiosurgery is related to the volume treated and the dose (62) delivered. The radiation dose delivered is adjusted to provide the greatest chance of obliteration with an acceptable risk of complications. Current AVM radiosurgical series have reported obliteration rates of 64–95% for AVMs smaller than 3 cm in diameter (63–65). For larger AVMs, the chance of success is lower. Mathis et al. (66) reported an obliteration rate of 50% in 24 patients with AVMs with volumes of over 14 mL. The Pittsburgh group (67) using the gamma knife, reported an 81% obliteration rate for lesions $<$20 mm in diameter and 45% rate for lesions between 20 mm and 44 mm diameter. Pollock et al. (65) analyzed 220 patients treated with radiosurgery and found that the success of AVM obliteration is associated with a smaller AVM, fewer draining veins, younger age, and hemispheric location. Pre-radiosurgical embolization was found to be a negative predictor of success.

It was generally assumed that once the abnormal AVM vessels are obliterated, the patient is cured and free from further risk of hemorrhage. However, in the pediatric

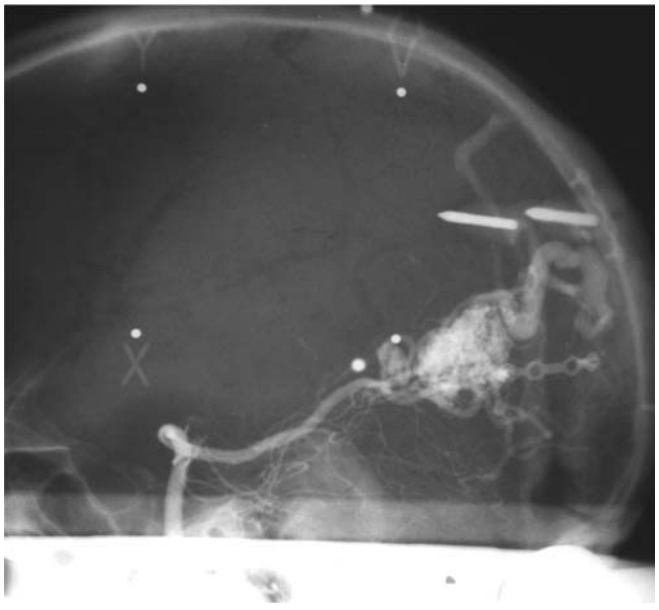


Figure 31.13 Angiogram for localization of AVM.

population, there is a rare but well-reported phenomenon of recurrent AVMs (68). Kader et al. (69) reported that among 141 patients with complete surgical resection of AVMs proven by angiograms (Fig. 31.13), five children had recurrence hemorrhage. One recurrence occurred 9 years after treatment. In over 40 years of pediatric experience, two recurrent AVMs were reported from the Hospital for Sick Children after complete microsurgical resection (70). Lindqvist et al. (71) studied the long-term effect of gamma knife radiosurgery in 48 patients, who were followed by angiogram with a median time of 9 years. There were totally four hemorrhaging episodes, which their AVMs were previously documented to be completely obliterated. Three of them were in the pediatric group. Therefore, in the pediatric group of patients, a prolonged follow-up with serial imaging is necessary even after the AVM is obliterated.

5.5. Conclusions

In the treatment of patients suffering from AVMs, a multidisciplinary team approach is necessary. The strength of each modality is utilized to maximize the efficacy and minimize the morbidity. Although surgery carries higher risk and morbidity, it offers immediate and complete removal of the AVMs. On the other hand, radiosurgery avoids the surgical risk and invasiveness, especially for high-grade lesions and is effective in obliteration; however, the delay between treatment and cure makes it a less attractive option for lesions which present with bleeding. As an adjunct, endovascular embolization makes surgery and radiosurgery easier or feasible in selective cases.

6. NEURONAVIGATION

Since the brain parenchyma is generally devoid of any unique landmarks, and vital areas are susceptible to injury, it is a challenge to stay oriented within its substance. Before the

era of CT scan, most intraparenchymal surgery was based on free-hand techniques with the help of conventional radiographs and clinical signs. The surgical exposures often were much larger than necessary.

The advent of CT and MRI scans (Fig. 31.14) has revolutionized neurosurgery, eliminating much of the guess work. Surgeons now can see the pathology inside the skull in different image planes. This allows making precise diagnosis and formulation of treatment plan. Also surgeons can better discuss with the patients and their relatives the risk of the operation in terms of neurological deficits. These images are also of great use to surgeons in the operating room in planning resections and trajectory of approaches. However, the surgeon has to study these images carefully, taking in all the anatomy and reconstruct in



Figure 31.14 (A) Patient in CT scan and (B) patient in angiogram suite.

his or her own mind. The incision and approach is transferred to the patient's head by known references such as facial features and other bony landmarks.

Further advancement of technology came with neuro-navigation. The principle of image-guided neurosurgery is to transfer the preoperative imaging data to a computer workstation inside the operating room (Fig. 31.15), allowing the surgeon to manipulate the images to provide a real-time accurate three-dimensional localization of surgical targets and trajectories (Fig. 31.16). The first apparatus for stereotactic neurosurgery was developed by Zernov (72) in 1889 in Moscow where aluminum rings were attached to the patient's head to predict surface anatomy of the brain. Thereafter, various different stereotactic systems had been developed. The basic components included a frame rigidly fixed to the patient's head, a system of data analysis for determining the coordinates, a mechanism for directing the instruments, and a probe directed at the target point.

Stereotactic neurosurgical procedures were most popular in 1950s for thalamotomy and pallidotomy in the treatment of parkinsonian tremor. However, with the emergence of L-dopa, these operations had significantly decreased in number. There was a resurge of frame-based stereotactic procedure when the CT scan came into existence in 1970. The scan provided a detailed anatomical representation of the lesion where biopsies and accurate resections can be carried out with the use of the stereotactic system. However, the major drawback was the need to fixate the frame on the patient's head and the overall complexity of the system.

The next development in neuronavigation was the articulated mechanical arm using position sensors to accurately locate the surgical tools in three-dimensional space on preoperative images during operation. At least six sensors are required; three for translation and three for rotation. The preoperative CT or MRI images are fed into the workstation. The patient's position is mapped in three-dimensional space by reference to fiducial markers applied on the patient's scalp or by anatomical landmarks. Theoretically at least three fiducials are needed; in practice, four to six are applied. Doshi et al. (73) has found that increasing numbers and wide distribution of fiducial points would increase the accuracy. These systems were easier to use and less cumbersome compared to the stereotactic frame system. Many of these systems are made light enough for easy handling and the weights are much reduced by balancing with counterweights at each articulation.

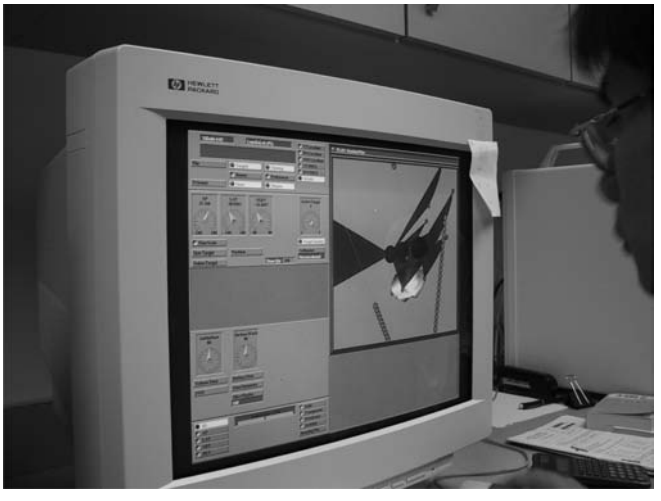


Figure 31.15 Treatment planning at the computer workstation.

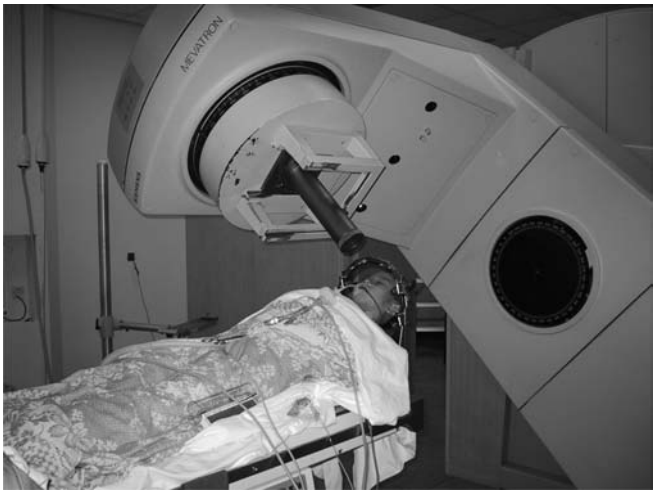


Figure 31.16 Patient undergoing Linac Knife radiosurgery treatment.

6.1. Armless Neuronavigation

Further development of "armless" systems relied on optical, acoustic, or electromagnetic tracking systems. The optical tracking system consist of a series of light-emitting diodes (LED) attached to the pointer and an array of three charge coupled detector (CCD) cameras that can accurately map out the positions of the LEDs. This is based on the triangulation technique for three-dimensional acquisitions of position coordinates. Some system uses active infrared LEDs, while other systems use passive plastic spheres of highly reflective surfaces for CCD cameras detection.

Today these navigation systems are quick to set up and easy to use. Indications for use of frameless stereotactic system include planning of skin incision and site of craniotomy, especially for lesions that are small and close to the surface where an inaccurate bony opening may miss the lesion completely. Also the system can help in determining the boundary of resection, especially for infiltrative tumors where the margin between the tumor and normal brain is not discrete. It enables the surgeon to perform a more complete resection, which is closely related to the patient's prognosis.

6.2. Accuracy

Accuracy in neuronavigation is particularly important in order to achieve its goals (Fig. 31.17). Galloway and Maciunas (74) defined accuracy in stereotactic frames. They define "accuracy" as the ability of a device to correctly find a point in space. Therefore a smaller mean distance between the desired and actual points denotes greater accuracy. Accuracy is further divided into mechanical and application accuracy. Mechanical accuracy is the ability of the system to bring the tip of an instrument to a given coordinate within its range. This is a measure of the precision of the joints in an articular arm system and the optical resolution in an armless optical system. This accuracy is usually in terms of a fraction of a millimeter. However this value does not take into account the inexactness of imaging and on patient registration. In order to maintain a high degree of mechanical accuracy, regular calibration of the system is needed.

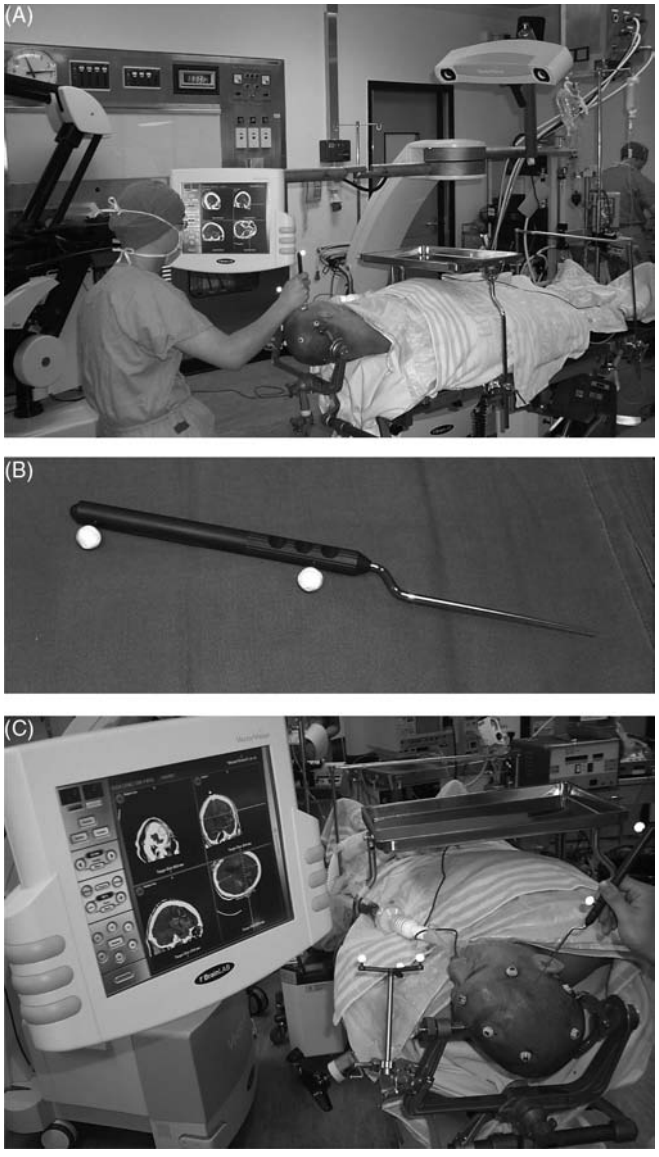


Figure 31.17 (A) Registration and preoperative planning using neuronavigation system; (B) navigation probe for real-time localization and (C) defining the tumor and planning of the surgical incision.

Application accuracy is the accuracy that concerns the neurosurgeon and is the accuracy one gets when using the machine on patients. It includes the mechanical error and the steps of surgical localization. The latter comprises imaging techniques, such as slice thickness, patient moving at the time of scanning, and errors in the steps of fiducial point selection and patient registration. This accuracy is much lower than the mechanical accuracy and may be up to few millimeters.

The accuracy however deteriorates over the course of the operation. During removal of the spinal fluid, tumor tissue, and distortion of the brain with retractors, the brain

parenchyma shifts. Therefore the navigation based on the preoperative images become less reliable. Nauta (75) showed in a series of 200 stereotactic operations that neither the application nor the mechanical accuracy were the limiting factors in the navigation system. Once the dura is opened letting out some CSF, the accuracy quickly dropped to 5 mm. Black (76) reported their documentation of brain shift from 1.8 mm to up to 30 mm. Various methods, like intraoperative ultrasound, intraoperative CT scan, and MRI scans are available to overcome this problem. These imaging modalities provide real-time image update thus permitting instant assessment of the extent of surgical resection.

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Minimal Access for Surgery in Pediatric Spinal Surgery

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1. INTRODUCTION

Video-assisted thoracoscopic (VAT) surgery has recently gained increasing popularity in the management of spinal disorders. Thoracoscopic spinal applications represent a new technique, not a new operation, and true it is minimal access surgery but it is definitely not minimally invasive. The endoscopic approach to the spine has involved an evolutionary approach. What began as an isolated drainage of a vertebral abscess has continued as a method of single diskectomy, release of the annulus fibrosis with or without ligation of segmental vessels, rib resection for thoracoplasty, rib harvesting for intervertebral fusion, and most recently, insertion of correctional implants and fusion. The body of knowledge currently available began with a single report in 1993 (1) and progressed to about 15 articles in the year 2000 (2–16).

In December 1993, at Children's Hospital Medical Center in Cincinnati, we performed our first VATS procedure for anterior release of severe spinal deformities in children and adolescents. The potential benefits of this procedure would include diminished postoperative pain and ventilatory compromise, decreased hospitalization time, improved wound care, a faster recovery time with minimal shoulder dysfunction and reduced infection risks to patients and OR personnel due to the decrease in operative time and exposure. Optimistically all of the above should reduce overall health care costs.

2. HISTORY OF THORACOSCOPY AND THORACOSCOPIC SPINAL SURGERY

Surgeons have long attempted to maximize the therapeutic benefit of interventional procedures for their patients while minimizing the disruption of the normal healthy tissue. Thoracoscopy has been utilized for lung lesions over the past 75–85 years (17). In 1910, Hans Christian Jacobeus introduced thoracoscopy to medical practice for the diagnosis and treatment of pulmonary tuberculosis (17,18). Jacobeus popularized thoracoscopy, as a bedside procedure under local anesthesia, where he first used a cystoscope. The thoracoscopic procedure was performed to lyse tuberculous pleural adhesions (intrapleural pneumolysis), which became the principal therapeutic tool in the treatment of tuberculosis from the 1920s through the 1940s. In 1931, Unverricht (19) reported 1500 thoracoscopies over a 16-year period with no injuries. Following the availability of antituberculous drugs, intrapleural Pneumolysis was less frequently used. In 1951, Stejnzajg (20) used thoracoscopy for the treatment of tuberculous empyema, hemopneumothorax, foreign bodies of the chest, and malignant pleural lesions. Interest in thoracoscopy waned and was followed by an era of neglect. Explosive growth of endoscopic procedures in other medical and surgical specialties occurred between the 1960s and 1980s.

The first report of thoracoscopy used in children for mediastinal masses, cysts, lung anomalies, spontaneous pneumothorax, and empyema was made in 1971 by Klimkovich et al. (21). This was followed by a report by Rodgers and Talbert (22). Interest was regained in thoracoscopy in the 1990s; by then considerable technical improvements in the video monitors, optical lens systems, and optic illumination systems had been developed. Video monitors removed the awkward and potentially sterile contamination necessity for the surgeon's head to be at the end of a telescope close to the chest wall and allowed the use of assistants. By 1992, Mack et al. (23) was able to report a series with no mortality, morbidity, or emergency thoracotomy, as did Lewis et al. (24). Excellent visualization of the spine during thoracoscopic surgical procedures led to the development of thoracoscopic spine surgery. In the early 1990s, thoracoscopy for the treatment of spinal

pathology was developed independently by Michael Mack, John Regan, and coworkers (1), Blackman and Neal (25) with animal, cadaveric, and clinical studies in the United States, and by Daniel Rosenthal and colleagues (26) in Germany. Riley et al. (27) presented on thoracoscopic corpectomy in a canine model. In the late 1990s, thoracoscopic techniques for spinal surgery gained considerable clinical momentum as an alternative to thoracotomy. Both surgical techniques and instrumentation have become fairly sophisticated, and the field of VATS has evolved rapidly in the last several years. Blackman et al., in 1998 (28) and Picetti et al., in 1999 (29) reported on the endoscopic instrumentation, correction, and fusion of thoracic and thoracolumbar scoliosis.

3. ENDOSCOPIC ANATOMY

The thoracic spine is divided into three regions because of anatomic differences among these areas. The upper field includes T1 to T5, the middle field includes T6 to T9, and the lower field includes T10 through L1 and is often obscured by the diaphragm, which may necessitate retracting it manually.

1. Upper field: On the right side, the veins to the 2nd, 3rd, and 4th intercostal spaces join to form the superior intercostal vein and then empty in the azygos vein. The 1st intercostal vein empties into the brachiocephalic vein and the 5th intercostal vein empties directly into the azygos vein. The junction of the azygos and the superior vena cava can also be visualized. The 1st and 2nd intercostal arteries arise from the supreme intercostal artery, a branch from the costocervical trunk of the subclavian artery, the remaining intercostal arteries arise directly from the aorta. The 1st rib may be difficult to visualize because of surrounding fat; palpation is usually used to help identify it followed by counting and marking of the lower ribs. The head of the 1st rib articulates with the 1st thoracic vertebra, whereas the heads of the 2nd, 3rd, 4th, and 5th articulate with the bodies above and below that particular disc space (e.g., the head of the 3rd rib spans the disc between the second and third vertebrae). The sympathetic chain can be seen lying over the rib heads. The esophagus can be seen between the trachea and the spine. On the left side, the aorta lies on the vertebral column making exposure of the discs more challenging. The intercostal arteries arise from the aorta and the intercostal veins arise from the hemiazygos vein in the majority of cases. The left superior intercostal vein is the termination of the hemiazygos and can be seen crossing the aortic arch and empties into the brachiocephalic vein.
2. Middle field: In this field the costovertebral articulation lies directly over the disc space. The intercostal veins arising from the azygos vein on the right are accompanied by their segmental arteries. The intervertebral discs are identified by the mounds observed on the spinal column and the vertebral bodies by the valleys. The segmental vessels are nested in the valleys directly overlying the bodies. The esophagus is anterior to the azygos vein. The greater splanchnic nerve is seen along the anterolateral vertebral body. On the left side, the aorta overlies the vertebral column otherwise the anatomy is the same as on the right side.
3. Lower field: The diaphragm attaches to the 12th rib, the transverse process of L1, and the anterolateral aspect of the upper three lumbar vertebrae. The T12–L1 disc can be visualized with retraction of the diaphragm.

The 10th, 11th, and 12th ribs do not cover a disc space, they articulate only with their respective vertebral body. There are no significant differences between the right and left sides.

The lumbar spine can be visualized through a retroperitoneal endoscopic approach. This allows exposure from L2 to L5. The retroperitoneal structures are visualized after divisions of the mesosigmoid, allowing direct access to the posterior abdominal wall and spine. The intervertebral discs bulge outward between the concave vertebral bodies. The segmental vessels lie within the concavity of the vertebral bodies. The anterior longitudinal ligament covers the anterior surface of the vertebral bodies. The psoas major lies on either side of the spine, it originates from the anterior surface of the transverse processes and the lateral portions of the vertebral bodies and discs from T12 to L5. The lumbar plexus lies within the substance of the psoas major muscle. The combined retroperitoneal and thoracoscopic approach can allow endoscopic exposure of the thoracolumbar junction.

4. INDICATIONS

The indications for thoracoscopy in pediatric spinal surgery are the same as were previously determined for thoracotomy. These indications are as follows:

1. rigid idiopathic scoliosis deformities at or about 75° in magnitude with correction to $<50^\circ$ on side-bending radiographs;
2. preventing crank shaft phenomena in the skeletally immature child with $>50^\circ$ curvature;
3. kyphotic deformities of $>70^\circ$ which do not correct to $<50^\circ$ on a hyperextension film over a bolster;
4. progressive congenital deformities within the thorax requiring anterior epiphysiodesis;
5. patients with neuromuscular deformities with at-risk pulmonary status;
6. patients with progressive spinal deformity and metabolic disease;
7. severe rib hump deformity not corrected by spinal instrumentation;
8. patients with neurofibromatosis who have intrathoracic tumors in addition to a significant spinal deformity;
9. pseudarthrosis following anterior intervertebral fusion;
10. rib and intercostal nerve tumors, and most recent;
11. instrumentation of thoracic spinal deformities above the diaphragm.

We have now extended our indications to include all procedures to the thoracic spine previously approached by thoracotomy.

5. CONTRAINDICATIONS

1. Inability to tolerate single lung ventilation;
2. severe or acute respiratory insufficiency;
3. high airway pressures with positive pressure ventilation;
4. pleural symphysis;
5. empyema; and
6. previous thoracotomy or thoracostomy are relative contraindications only.

Many children with progressive scoliosis underwent cardiothoracic surgical procedures during infancy. We have now successfully performed VATS in patients following previous thoracotomy, although resection of adhesions are time consuming and there is a higher risk of infection.

6. PREOPERATIVE PLANNING

A thorough and meticulous preoperative workup is mandated prior to the surgical intervention. Our preoperative protocol includes a thorough neurological examination, body mass index evaluation, and, if the patient is morbidly obese, a weight reduction program is started, Standing PA and lateral scoliosis views and supine-bending films are obtained on all scoliosis patients. A hyperextension film over a bolster is obtained for patients with kyphosis. A whole spine MRI is ordered on patients with spinal deformities secondary to neurofibromatosis, congenital spinal deformities, infantile scoliosis, patients with idiopathic scoliosis who have asymmetric abdominal reflexes, a difference in foot size or limb girth and patients with a documented rapid progression of the deformities. Left-sided curves with neurological findings, and patients with marked truncal asymmetry also are further investigated by magnetic resonance imaging of the spine. Lab work includes a CBC with a differential, renal and electrolyte panel, a coagulation profile, UA examination, and a pregnancy test in female adolescent patients.

Iron loading by elemental iron intake is started at least 6 weeks prior to surgery. Patients who meet the criteria for autologous blood transfusion undergo timely harvesting of up to four units. Patients with chronic illness undergo a routine preoperative pulmonary function testing. All patients undergo a preoperative anesthesia evaluation to determine if any further work-up is required. As a part of preoperative education, our patients are required to take a walk through the OR and the floor in order to obtain first hand information regarding the surgical process and to meet their future caregivers. The patient is reevaluated a week prior to surgery by the physician to answer any further questions and informed consent is obtained.

7. TECHNIQUE

7.1. Room Set-Up

Two surgeons usually perform the procedure, one is responsible for visualization or access, and the other is the spinal surgeon. Some surgeons prefer to work on the same side of the table usually facing the patient with the patient in the lateral decubitus position, whereas others would prefer to work opposite each other and to view the intrathoracic contents by placing monitors directly across from them usually looking over their associates' shoulder; we prefer the latter.

7.2. Endoscopic Instruments

A 30°-angled scope is best for anterior spinal release and is the most commonly used scope. Specialized thoracospinal instruments include long handle rongeurs, curettes, periosteal elevators, extended insulated tip electrocautery, both monopolar and bipolar, harmonic scalpel, and suction devices. Rigid and flexible portals, spinal cord monitoring leads, video recorder, and image processor are required as well.

7.3. Anesthesia

The anesthesiologist must be capable of using the fiber-optiscope and selectively deflating one lung. This can be done best with the use of a double lumen tube, although bronchial blockers may be necessary in smaller children. Twenty minutes is required to get complete resorption atelectasis.

7.4. Positioning

The patient is placed convex side up onto the lateral decubitus position with kidney rest support. Although not draped out, the arm is hyperflexed at the shoulder to allow the placement of portals higher into the axilla. An axillary roll is placed under the downside axilla. Lower extremities are wrapped with Ace wraps and the greater trochanter on the downside is especially well padded. Protective padding of the downside peroneal nerve adjacent to the fibular head is carried out.

7.5. Procedure

The first portal, that is, the visual panorama portal is most frequently placed at or about the T6 or T7 interspace (to avoid the diaphragm) in the mid-axillary line. The incision is made over the top of the rib to avoid the intercostal vessels. There tends to be less bleeding when the electrocautery is used to dissect the intercostal muscles into the chest cavity. After establishing the visual portal, further entry into the chest can be observed through the scope. Prevention of bleeding around the scope is achieved by coagulation of the vessels and is an important part of this technique. A 15-mm trocar is used through which a 10-mm 30°-angled rigid telescope is placed. The thoracoscope is introduced and the lung is observed as it deflates. A panoramic assessment and evaluation should be carried out of the intrathoracic space in an effort to determine the topographical anatomy. The portals should be as far apart as possible. The superior thoracic spine is well visualized without retraction once the lung is completely deflated, but retraction is necessary below T9–T10 because the diaphragm gets in the way. By percussing the chest and visualizing the percussions from within, other working portal sites are selected. A rigid thoracoportal is necessary for the thoracoscope because it can be damaged as one attempts to navigate between the resistant rib cage fulcrum. At this stage the ribs are counted. If there is a question of the specific level a spinal needle is inserted into an intervertebral disc and an X-ray taken. We prefer to open the parietal pleura in a longitudinal fashion very similar to what one would do when performing a thoracotomy. The intervertebral discs are identified by the mounds observed on the spinal column and the vertebral bodies by the valleys. The segmental vessels are in the valleys overlying the vertebral bodies. We consider transligation of the segmental vessels to be fairly safe in our practice. The segmental vessels are coagulated with the harmonic scalpel, incised, and retracted with the pleura. The pleura is further elevated and retracted using the harmonic scalpel, thoracoscopic periosteal elevators, and blunt dissectors. A Raytec sponge is packed under the pleural sleeve to assist in retraction and protecting the vessels, as well as other soft tissues. We then proceed directly to excising the annulus at the level of intervertebral disc. A transverse cut is made across the vertebral endplate parallel to the disc both rostral and caudal to it. An elevator is then used to elevate the vertebral endplate to isolate the disc. A transverse cut is made across the annulus fibrosis continuing down to the level of the nucleus pulposus. Rongeurs, curettes, and periosteal elevators are then used to assure complete removal of the disc material and the end plates. In the young child, it

is often possible to elevate the vertebral end plate apophysis and to completely excise the intervertebral contents back to the posterior longitudinal ligament. With experience, one is able to excise the annulus and disc space contents in an $\sim 250^{\circ}$ arc or from the rib head to opposite posterolateral body. The spinal column segment should be stressed with moderate external force by rotating a periosteal elevator in the disc space following each release to see if spinal segment mobility has been achieved. Our initial routine consisted of harvesting bone by removing a rib and performing an intervertebral fusion; this is still the case for anterior instrumentation cases, but for intervertebral release cases, we started using fibular allograft rings. We no longer attempt pleural closure. A chest tube is then placed through the inferior-most portal. Chest tube placement is observed through the scope as it is placed alongside the vertebral column. The patient is usually then re-intubated and turned prone for a posterior spinal fusion. It is important that the inflated lung be suctioned to prevent mucous plugging.

7.6. Procedure for VATS-Assisted Anterior Spinal Instrumentation

The patient is positioned in a lateral decubitus position. An image intensifier is used to assess the sagittal profile of the patient and a radio opaque marker is used to identify the rib heads at each level; 1.5 in. oblique portals are established at every other interspace. The oblique incision allows one to dissect above and below the rib for portal access. Pleural dissection is carried out in the same fashion as described above. Endplates are identified with the help of diathermy. Dissection is stopped at the anterior longitudinal ligament. The endplates are elevated in the same manner as described above. Image is used to confirm the dissection to the downside annulus. A rasp or curette may be used to thoroughly remove the remnants of the end plates without compromising the vertebral body. One may select a single rib or segments of several ribs to morcelize for bone graft. The use of emulsified allograft is discouraged. The morcelized bone graft is packed into the intervertebral disc space. After this one can proceed with the instrumentation.

7.7. Postoperative Care

We perform a wake-up test prior to extubation. The patient is moved to a horizontal frame and subsequently transferred to the ICU for 24 h. Chest X-rays are taken in the ICU to document chest expansion and chest tube placement. The majority of our patients are placed on 1 mg/mL morphine PCA. The antibiotics which were initiated at the time of incision are continued until 24 h following chest tube removal. Dressing is changed on POD2, at which time the patient is measured for a Boston type TLSO and fitted with one the next day.

7.8. New Advances in Technique

7.8.1. Allograft Fibular Rings

Autogenous rib grafting of the intervertebral space has been considered to be superior to allograft (30). Harvesting of the rib segments by VATS allows adequate material; however, the inability to secure the periosteum to prevent weeping leading to persistent pleural effusion has not been resolved. We therefore undertook the use of fibular allograft rings in those patients who would subsequently undergo posterior instrumented spinal fusion. Currently, 11 patients have had allograft fibular rings, 6 had Scheuermann's Kyphosis, 3 had idiopathic scoliosis, and 2 had neuromuscular scoliosis. Of these patients,

5 had over 8 months follow-up; all of these cases showed evidence of good anterior fusion mass. There is a notable decrease in persistent effusion since incorporating this technique. This technique has the potential to decrease operative time, blood loss, postoperative chest pain, and postoperative effusion.

7.8.2. Prone Positioning

A new modification on endoscopic transthoracic release involves prone positioning of the patient as opposed to the lateral position described earlier. The release is performed on the concave side for scoliosis and on either side for kyphosis patients. This technique permits simultaneous posterior exposure, instrumentation, and correction of the deformity. Papin et al. (31) performed thoracoscopic anterior release and fusion in the prone position on five adolescent patients whose mean age at the time of surgery was 12 years. The anterior release was followed by posterior instrumentation and fusion in all patients. Lieberman et al. (9) reported on prone position endoscopic transthoracic release with simultaneous posterior instrumentation for spinal deformity. The procedure was performed on 15 patients and no complications related to the endoscopic components of the procedure were reported. The major advantage of this technique is the elimination of the need for double lumen intubation and the amount of time required to extubate and reposition the patient. The prone position presents the opportunity for another team to simultaneously expose and instrument the posterior spine.

8. EXPERIENCE AT THE CHILDREN'S HOSPITAL MEDICAL CENTER, CINCINNATI

The first VATS procedure for anterior release of severe spinal deformity was first performed in December of 1993. By the end of May 2001, we had performed over a 112 cases. Seventy cases were available at 2 years. For deformity, patients ages varied from 5.1 years to 32 years. Thoracoscopic anterior release with discectomy and fusion were carried out on patients who had the following diagnoses: idiopathic scoliosis ($n = 45$), Scheuermann's kyphosis ($n = 19$), neuromuscular spinal deformity ($n = 15$), neurofibromatosis ($n = 7$), myelomeningocele ($n = 3$), congenital scoliosis ($n = 2$), infantile scoliosis ($n = 1$), dysplasia ($n = 1$), Marfan's ($n = 1$), and postradiation scoliosis ($n = 1$). Eight patients underwent anterior instrumentation in addition to thoracoscopic anterior release with discectomy and fusion. Repair of pseudoarthrosis of the spine was performed on one neuromuscular patient. Four patients had excision of the first rib for the treatment of thoracic outlet syndrome. One patient had excision of intrathoracic neurofibroma, and another had a benign rib tumor removed. One patient had anterior fusion following thoracic spine fracture dislocation and another had anterior fusion following vertebral osteomyelitis (Fig. 32.1).

Data analysis of the first 100 patients showed a mean preoperative scoliosis of 72° (range, 42°–120°) and a mean preoperative kyphosis of 83° (range, 65°–110°). The average operative time for the thoracoscopic anterior release with discectomy and fusion procedure was 250 min (range, 150–405 min). The average number of discs excised was 8 (range, 4–11), and there was no change in the number of discs excised as the series progressed. The time spent for discectomy varied from 8 min to 18 min, but when considering the total procedure including rib harvesting, end plate ablation, and intervertebral fusion, the average operative time per disc was 33 min in the first 45 deformity patients compared with 31 min in the last 45 deformity patients, which was

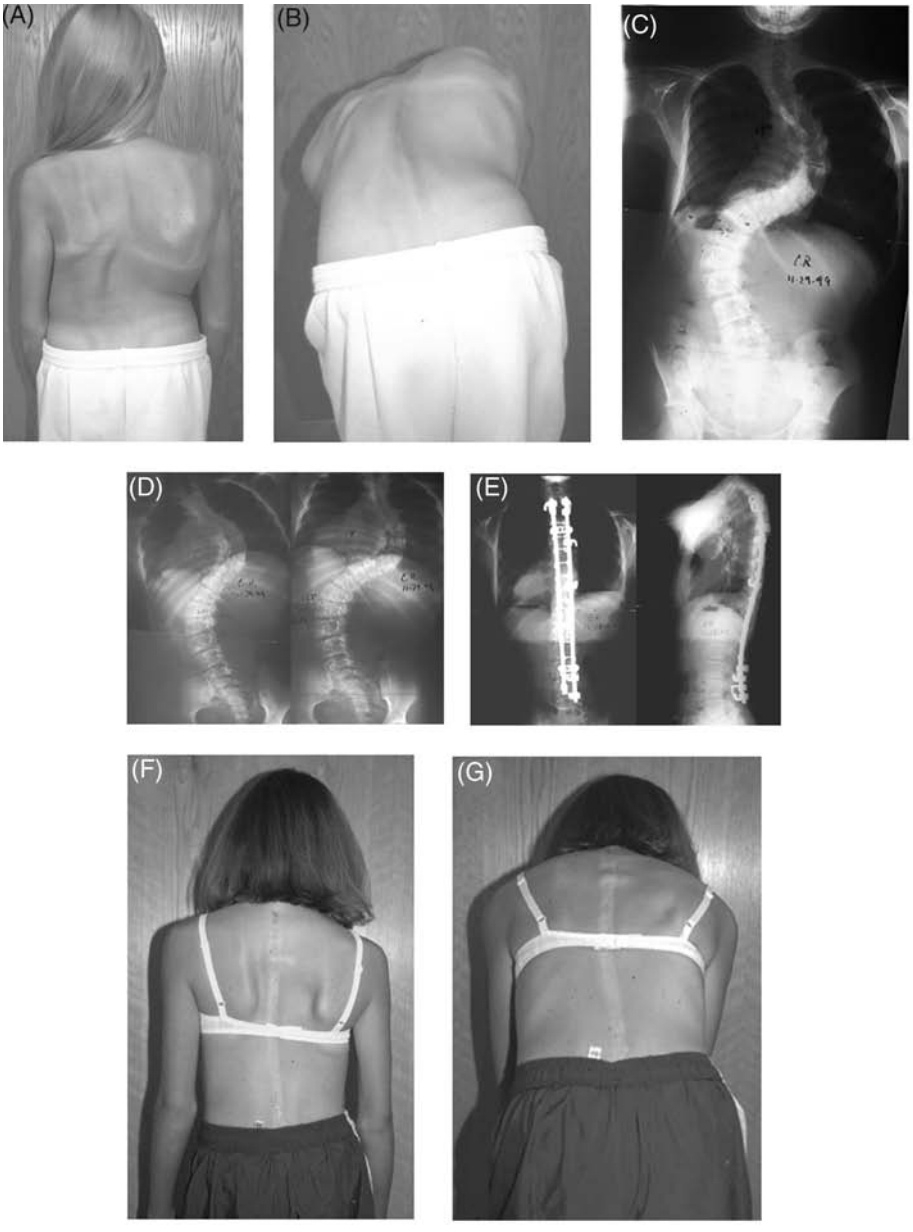


Figure 32.1 A 14-year-old female with a severe double primary scoliosis. Her thoracic curve measured 120° and the lumbar curve measured 73°. Both curves were extremely rigid and showed little correction on side-bending views. We elected to perform anterior release and fusion by VAT, posterior release followed by halo-femoral traction and subsequent posterior instrumented spinal fusion. She obtained an excellent result. (A) Clinical standing photograph taken from the posterior, illustrating the severe prominence of the right rib cage and left lumbar flank; (B) bending clinical photograph revealing rib hump of the right hemithorax and significant rotation of the left thoracolumbar spine; (C) standing PA view illustrating severe thoracic and lumbar curves. She is Risser 0; (D) right and left-bending films showing minimal flexibility of the curves; (E) postoperative PA and Lat views showing excellent correction in the coronal and sagittal plane; (F) postoperative clinical photograph illustrating clinical improvement and (G) bending clinical photograph illustrating correction of the rib hump and thoracolumbar flank rotation.

not statistically significant. The average blood loss during the thoracoscopic anterior release with discectomy and fusion was 290 mL (range, 50–1300 mL). The hospital stay averaged 7 days (range, 5–15 days); days in the ICU averaged 1.5 ± 1.5 days. Chest tube days averaged 3.1 ± 1.4 days with chest tube output averaging 999 ± 543 mL. Final postoperative scoliosis and kyphosis correction were 68% and 90%, respectively.

Complications occurred in 18 patients, five of whom were patients with myelomeningocele. Complications included a single case of intraoperative tension pneumothorax in an anterior instrumentation case (32), postoperative pneumonia, pulmonary embolus, chylothorax, mucus plug, fluid overload requiring reintubation in a patient with tetralogy of Fallot, atelectasis, a retained iliac crest drain, and two cases of superior mesenteric artery syndrome. Three patients had large pleural effusions that required intervention. One patient required ultrasound-guided thoracentesis and the other two had ultrasound-guided pigtail catheter thoracostomy. Complications in the myelomeningocele patients included one iliac crest wound infection, one decubitus ulcer, one patient developed a posterior spine wound infection, and two patients had left upper lobe atelectasis. These complications were evenly distributed throughout the series. Clearly, there is an extensive learning curve for VATS, but this learning curve is not prohibitive. VATS provides a safe and effective alternative to open thoracotomy in the treatment of pediatric spinal deformities in addition to its utility in the treatment of intrathoracic tumors, benign rib tumors, and vertebral osteomyelitis.

9. VATS VS. THORACOTOMY

Newton et al. (33) compared the efficacy of VATS vs. thoracotomy in achieving an adequate spinal release in a goat model. Anterior spinal release was performed in six mid-thoracic motion segments in five mature goats. VATS was used for three levels on one side and thoracotomy was used for alternating three levels on the contralateral side. Motion segments were then individually subjected to nondestructive biomechanical testing using sagittal, coronal and torsional bending torques, and the resultant angular displacement was measured. Duration of surgery per disc level decreased with increasing experience, while the intraoperative blood loss was comparable between the two groups. Both techniques resulted in a significant increase in the flexibility when compared to the intact levels, but there was no difference in the mobility achieved between the two techniques.

Cunningham et al. (34) utilized a sheep model to compare the efficacy of VATS to a traditional thoracotomy in promoting interbody spinal arthrodesis. The spinal column was destabilized in 14 sheep by resecting the anterior and middle columns at T5–T6, T7–T8, and T9–T10, followed by reconstruction using iliac autograft, the Bagby and Kuslich device packed with iliac autograft, and Z plate stabilization with iliac autograft. In half the sheep, the entire procedure was performed using VATS, while in the other half it was done through a traditional thoracotomy. Histomorphometric and biomechanical evaluation performed on the disarticulated spines of these models showed comparable bone formation and biomechanical properties. However, the VATS group showed increased incidence of interoperative complications, longer operative times, higher blood loss, and increased animal morbidity. These results were attributed to a substantial learning curve associated with the technique.

Wall et al. (29) compared the spinal flexibility achieved after thoracotomy and VATS in an animal model. The intervertebral disc between vertebrae T8 and T9 was resected from 30 live, anesthetized, adolescent pigs. In 15 pigs, the chest was opened

via thoracotomy of the eighth rib, and the disc was excised. In the other 15 pigs, the disc was removed endoscopically. These motion segments and six intact controls were tested mechanically in side bending, flexion-extension, and axial rotation. No statistically significant differences in flexibility were found between open and endoscopic groups in any loading direction. The statistical power to detect a 20% difference between surgical groups was $\geq$95%. They concluded that endoscopic and open techniques were equally effective in increasing spine flexibility.

Newton et al. (33) compared the results of anterior spinal release and fusion performed through a thoracoscopic approach in 14 patients to 18 performed via a thoracotomy. They reported a 56% and 88% postoperative correction of the scoliosis and kyphosis, respectively, in the thoracoscopic group compared to 60% and 94% in the thoracotomy group. A mean of 6.4 discs was excised in the thoracoscopic group compared to 6.1 in the thoracotomy group. Mean blood loss in the thoracoscopic group was 235 mL compared to 270 mL in the thoracotomy group. The mean operative time was 191 mL compared to 128 mL in the thoracotomy group. This difference was statistically significant. A learning curve was associated with the operative time as the first seven cases took 220 min compared to 162 min for the later seven thoracoscopic cases. Chest tube drainage was 1252 mL in the thoracoscopic group compared to 776 mL in the thoracotomy group. Chest tube drainage continued for 5 days in the thoracoscopic group compared to 3.1 days in the thoracotomy group. This difference in chest tube data was significant. The length of hospital stay was not reduced by the thoracoscopic procedure, while the cost of open procedure was 29% less than the thoracoscopic procedure. They concluded that the thoracoscopic technique was safe and effective for anterior spinal release and fusion in pediatric spinal deformity.

Vincent Arlet (2) reported a meta analysis of published papers dealing with the use of VATS for anterior spinal release in pediatric spinal deformities. Ten published articles comprising 151 procedures were selected for meta analysis. They reported that the number of excised discs varied between four and seven but the quality of disc excision was not uniformly reported. The procedure lasted between 2 h 30 min and 4 h depending on the experience of the physician. The mean curve measured 65° for scoliosis and 78° for kyphosis. The postoperative correction for scoliosis ranged between 25° and 37°, while that for kyphosis measured 44°. The average time spent in the hospital for a combined procedure was 9 days with a 28% increase in the cost for a VATS-assisted procedure. The total complications reported were 18%, most of them being pulmonary complications.

Durrani et al. (35) compared the adequacy of anterior spinal release and efficacy of anterior spinal fusion achieved by VATS and thoracotomy in reasonably matched pediatric patients. None of the patients had an anterior spinal release performed below L2, no anterior instrumentation was performed, and all patients underwent an instrumented posterior spinal fusion as well (Fig 32.2). Adequacy of release was judged using weighted correction, which was defined as the release times the stiffness of the curve. Efficacy of anterior spinal fusion was determined by comparing the immediate postoperative and the final Cobb angles of the deformity and by radiographic evidence of bridging across 50% of the width of the disc space at each level judged by a blinded reviewer on the final lateral spine radiograph. Twenty-eight patients formed the VATS group, while 29 patients formed the thoracotomy group. In the VATS group, the mean preoperative primary spinal deformity measured 65.18° (S.D. 19.67), while the immediate postoperative value measured 32.5 (S.D. 13.82). In the thoracotomy group, the same values measured as 68.52 (S.D. 17.08) preoperatively and 33.14 (S.D. 16.65) postoperatively. For thoracotomy, the difference in preoperative and postoperative primary Cobb angles was 35° ($\pm$13°) for primary curves, and 24° ($\pm$15°) for secondary curves. For VATS, these values were 33° ($\pm$17°) and 25° ($\pm$17°),



Figure 32.2 A 16-year-old girl with a 95° kyphotic deformity. She underwent a VATS anterior release and rib fusion followed immediately by a posterior instrumented spinal fusion. (A) Clinical photograph of a lateral bending position illustrating the severe kyphotic deformity; (B) composite photograph of pre and postoperative lateral radiographs illustrating correction of the kyphotic deformity and (C) postoperative clinical photograph taken in the lateral bending position 1 year later.

respectively, not significantly different from thoracotomy ($p > 0.5$). The mean weighted correction for spinal deformities in the thoracotomy group measured 24.55° for the primary deformity compared to 26.65° for the VATS group. The weighted corrections were not significantly different between the treatment groups ($p > 0.5$). In the thoracotomy group, the mean postoperative Cobb angle for the primary deformity measured 33.14° (S.D. 16.65), while the mean final follow-up value measured 41.24° (S.D. 17.5). In the VATS group, the mean postoperative primary deformity measured 32.5° (S.D. 13.82), while the mean final follow-up value measured 34.57°. For thoracotomy, the differences between immediate postoperative and final angles for primary curves was 8° (±9°), while for VATS, these values were 2° (±6°), a marginally significant ($p = 0.011$) difference. There was no difference in the number of radiographically fused levels between the two groups. The mean operative blood loss for the thoracotomy group was 804.5 mL compared to 412.2 mL in the VATS group. This difference was statistically significant ($p = 0.0025$). VATS, in their view, can achieve an adequate spinal release and an effective spinal fusion with decreased blood loss.

10. VATS-ASSISTED ANTERIOR INSTRUMENTED SPINAL FUSION

Anterior spinal fusion and instrumentation using VATS is gaining increased popularity. Sucato et al. (36) recently reported on the results of CT examinations on 12 patients

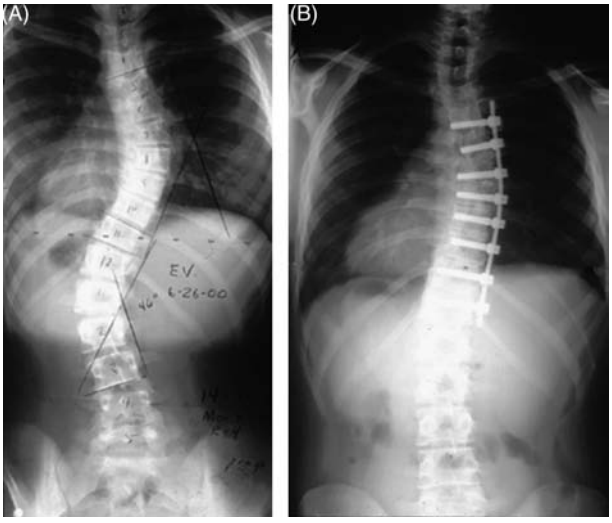


Figure 32.3 A 14-year-old girl with a double primary curve whose lumbar curve was extremely flexible. Selective fusion by VATS anterior instrumentation was carried out of the thoracic curve only. (A) PA radiograph view illustrating a double primary curve and (B) PA radiograph following VATS anterior instrumentation. Note almost complete resolution of the lumbar curve.

who underwent anterior spinal fusion and instrumentation for idiopathic scoliosis (Fig. 32.3). All patients had a single right thoracic curve measuring a mean of 55.9° preoperatively and 9.4° postoperatively. The average number of fused levels was 6.6. The average area of disc excision was 73.3%. Eighty-eight screws were used. The average distance of the screw tip from the aorta was 3.9 mm; however, in 27% the screws were just adjacent to the aorta. The average distance from the spinal canal to the posterior edge of the screw was 4.6 mm with three screws broaching the spinal canal. They concluded that spinal instrumentation using a VATS is a technically demanding procedure in which screw placement so close to the aorta and the spinal canal could become problematic.

11. COMPLICATIONS

11.1. Intraoperative Complications

Bleeding
Lung tissue trauma
Dural tear
Lymphatic injury
Tension Pneumothorax (32)
Spinal cord injury
Sympathectomy
Solid Organ Injury
Pressure Necrosis of the skin over the downside iliac crest and greater trochanter
Peroneal Nerve Palsy
Incorrect Fusion Levels

11.2. Postoperative Complications

1. Pleural effusion: Patients must have daily chest radiographs for the duration of the chest drain and one day after the removal of the chest drain. If the effusion is $>20\%$ of the hemithorax volume, we then recommend percutaneous aspiration under ultrasound guidance. If >400 cc of fluid is obtained then a pigtail catheter is left in place and removed when the drainage falls below 80 cc for a shift.
2. Pulmonary embolus: This is a very rare complication in children but must be suspected if the oxygen saturation is falling in the absence of a pleural effusion.
3. Chylothorax: This can be treated with parenteral hyper alimentation for 6 weeks during which time the chylothorax usually resolve.
4. Intercostal neuralgia: One can inadvertently damage the intercostal nerve while placing the portals, in addition to the pressure exerted on the nerve with a rigid port in place. There are less reported intercostal neuropathies following the use of flexible ports as apposed to rigid ones. The rigid port is only used for thoracoscope and is changed frequently to prevent intercostal neuralgia.

11.3. Delayed Complications

1. Pseudoarthrosis: Pseudoarthrosis is a legitimate concern with any spinal procedure that aims to achieve a spinal fusion and so is definitely a concern with VATS-assisted spinal fusion as well.
2. Early (unpublished personal communication) results of VATS instrumentation using only emulsified allograft for fusion show a trend to persistence of the disc space, pseudoarthrosis, and rod breakage.

Huang et al. (8) reported on the complications encountered in 90 consecutive patients who underwent anterior spinal lesions using VATS. The diagnosis were varied including deformity, infection, and tumors. The procedures included biopsy (3 patients), thoracic discectomy (3 patients), multiple level anterior discectomy and fusion (14 patients), corpectomy (6 patients), corpectomy and interbody fusion (32 patients), and internal instrumentation (28 patients). A total of 30 complications were noticed in 22 patients (24%). Two fatal complications resulting from massive blood transfusion and postoperative pneumonia were noted. Nonfatal complications included four cases of transient intercostal neuralgia, three superficial wound infections, three cases of pharyngeal pain, two cases of lung atelectasis, two cases of residual pneumothorax, two cases of subcutaneous emphysema, one inadvertent pericardial penetration, one chylothorax, one screw malposition and graft dislodgement. Four patients were converted to an open procedure: two due to adhesions and two due to poorly tolerated single-lung anesthesia.

12. CONCLUSION

VATS has provided the spine surgeon with a minimal incisional technique to achieve the same objectives that were previously being accomplished via thoracotomy. There appears to be immense patient satisfaction with the aesthetics of the incisions. The postoperative convalescence shows a trend to decreased pain support requirements, understandably because of preservation of para spinal and shoulder musculature. The surgical team concept is exemplified by all the associates observing the same exposure via the monitor. VATS also presents opportunities for surgical education unparalleled with previous exposures. For biopsies of benign lesions, anterior spinal release and fusion as

well as costoplasties, it may ultimately replace thoracotomy. [We now perform VATS for all procedures previously requiring a thoracotomy without instrumentation.] It is important to remember that a minimal incisional technique is by no means minimally invasive. There is a significant learning curve associated with this technique and only meticulous attention to fine detail can prevent potentially disastrous complications. Instrumentation via VATS is indeed another story. VATS-assisted anterior spinal instrumentation is on the horizon. It is an *extremely* complex and demanding undertaking. The thorax presents multiple opportunities for significant life-threatening complications. Needless to say the aorta, esophagus, heart, and lung are at risk and attention to complications are not as timely because of the constraints of the closed environment. The future of VATS instrumentation shall require a significant refinement of instrumentation and familiarity of spine surgeons to the environment before it reaches the same level of safety as posterior spinal instrumentation. Until this significant learning curve has been achieved, we feel that the gold standard for correction of scoliosis should remain a selective posterior instrumented spinal fusion.

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33
Minimally Invasive Surgery in Pediatric Cardiac Surgery

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1. INTRODUCTION

A recent and favourable trend, that is, a significant reduction in morbidity and mortality in the treatment of congenital heart disease, must be in part credited to recent technological advances. The introduction of routine pediatric cardiac surgery began in the mid-to-late 1970s; therefore, most innovations are relatively new. Many centers are now experiencing excellent immediate surgical results; the implications and thrust of this chapter therefore remains in; further improving long-term outcomes utilizing novel technologies. Obviously a superior cosmetic repair seems highly desirable especially in children where an obvious and unsightly incision(s) may have significant long-term social implications. In addition to the improved self-esteem with a reduction of an external scar, the concomitant reduction in tissue trauma allows the benefits of reduced pain, earlier discharge and decreased length of hospital stay. In this era of managed care, it remains rare to find a sound fiscal medical philosophy with a high degree of patient satisfaction.

The operating room continues to remain a highly conservative domain that has until recently remained unchanged in appearance and practices. Similarly, so too have the surgical techniques utilized for "open-heart" pediatric cardiac surgery. Advances in

videoscopic techniques were first evident with the introduction of arthroscopy in the mid-to-late 1970s. Gynecologists and soon general surgeons found scopes to have a beneficial role in the diagnosis and later treatment of their respective disciplines. The introduction of videoscopes in cardiac surgery occurred relatively late, in the 1990s. It seems that the pediatric cardiac specialists who finished their subspecializations in general surgery after the introduction of laparoscopy have spurned advances in the field of cardiac surgery.

Robotics too has only recently assumed a collaborative role assisting surgeons during delicate operations once only thought possible in science fictions of 20 years ago. An evolution from the once archaic, large and uni-task collection of metal and electronics to the newer 3D visually assisted highly mobile units has taken place only within the past decade. As such, neonates and infants and even adults born with congenital heart defects now have an alternate paradigm in which to undergo surgical repair.

Minimally invasive surgical techniques have allowed the development of novel intracardiac visualization techniques that may soon allow for the avoidance of the heart–lung machine as a routine component of pediatric cardiac surgery (see subsequently). The possible avoidance of the inherent and systemic inflammatory state found in contemporary "open-heart" practices may allow for improved neonatal/infant organ function including the brain. Neurological dysfunction (either congenital and/or related to perioperative period) remains a serious comorbidity that may significantly preclude successful long-term outcomes in children afflicted with congenital heart disease. The mechanisms of injury remain multifactorial with up to 25% of children having residual neurological sequelae postcardiac surgery. Newer techniques can now be utilized to allow for continuous cerebral perfusion avoiding both deep hypothermia and circulatory arrest (Fig. 33.1).

2. SURGICAL TECHNIQUES

2.1. Modifications to the Heart–Lung Machine

The limited size of the incisions has forced changes in the sometimes archaic heart–lung machine. The roller pump has only recently been modified to allow a downsizing of

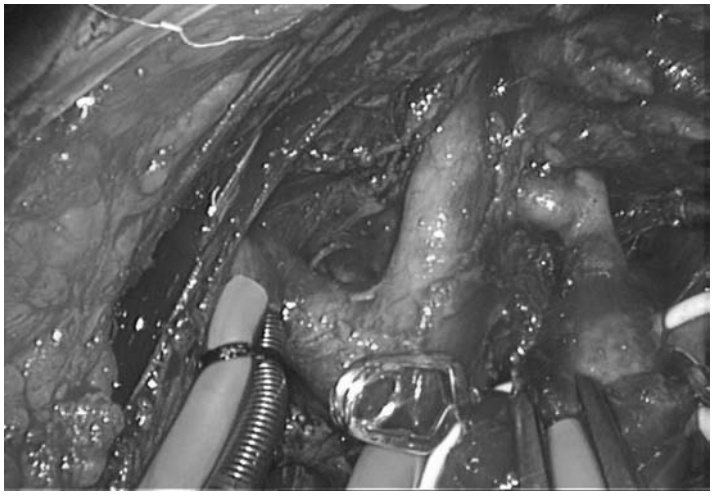


Figure 33.1 Aortic arch reconstruction in a neonate with avoidance of circulatory arrest. The cannula is providing continuous cerebral and coronary perfusion via the innominate artery.

cannulae needed for the miniature incisions of the 21st century. Both active venous and centrifugal suction can be applied to the circuit. Our modified circuit allows for continuous ultrafiltration (CUF) while delivering a low prime blood cardioplegia with rapid initiation of modified ultrafiltration (MUF) via the same cardioplegia system without elaborate modifications to the entire pump circuit. The use of the blood cardioplegia system provides both temperature control and macro filtration. The arteriovenous system is used to withdraw through the arterial line and return to the venous cannula. The entire cardiopulmonary bypass (CPB) is thus made available and the crystalloid chaser preserves the system integrity should the need arise for the reinitiation of CPB. MUF occurs in the range of 10–20 mL/kg per min with vacuum of 300 mmHg and temperature regulation according to the patient's temperature.

Venous-assisted drainage, both kinetic and vacuum, has allowed for a further shortening of lines and intracomponent lengths, reduced caliber, and alternative cannula for smaller incision. The application of as little as 40 mmHg has increased venous return by $\geq$50%. A cautionary note with respect to the potential embolization of air must be made when the latter techniques are utilized without prior experience (Fig. 33.2).

2.2. Surgical Incisions

Although several incisions are currently available to the pediatric cardiac surgeon, the hemi-median sternotomy remains a safe and comfortable incision. It lacks the pain and discomfort frequently found with the thoracotomy and avoids the risk of injury to the potentially developing breast tissue. Avoidance of groin cannulation and retrograde cerebral perfusion seem desirable. The excellent exposure of the mediastinum afforded by the hemi-sternotomy allows for the repair of a myriad of cardiac malformations distinct from atrial septal defects, that is, ventricular septal defects, fibromuscular obstruction to the right ventricular outflow tract, atrioventricular valvular abnormalities and endocardial cushion defects (Fig. 33.3).

During the past 3.5 years our surgical philosophy has continued to evolve; the cornerstone remaining the limited hemi-sternotomy. Infants, children, and adults have

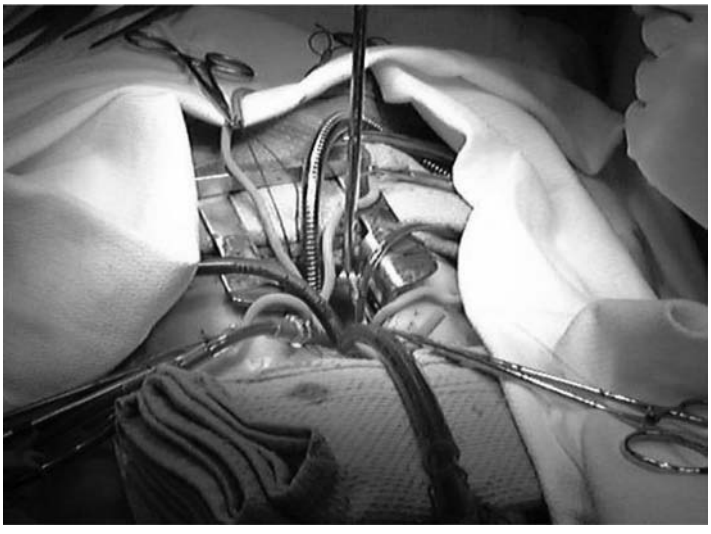


Figure 33.2 Smaller incisions demand innovative cannulaes, bypass circuits, surgical instruments, and better tools to visualize the intended targets.

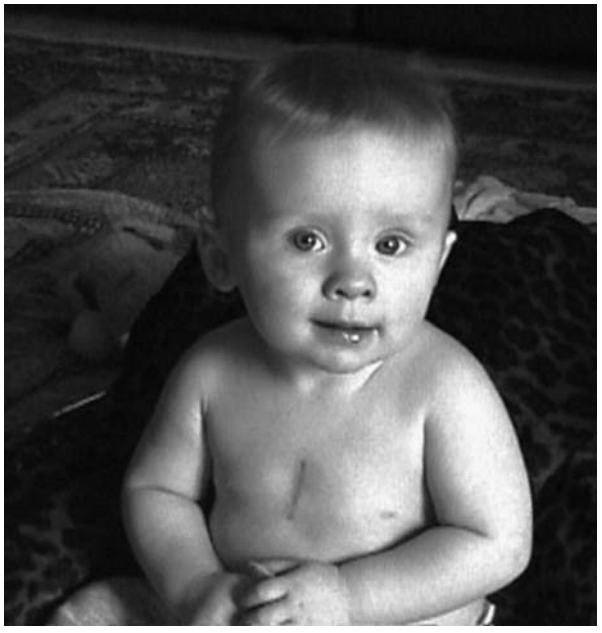


Figure 33.3 Complex cardiac repairs can be performed via the "hemi-median" sternotomy.

undergone operations that routinely include, epidural or spinal anesthesia, active venous suction, cardioscopy, and robotics.

Direct access to the mediastinum via the hemi-sternotomy allows for the correction of defects not previously appreciated, the ability to easily extend the incision if required, and superior de-airing of the cardiac chambers. The incision remains the least painful and stability is assured as the manubrium remains intact and the remaining divided sternum can be successfully stabilized.

2.3. Video-Assisted Cardiac Surgery (Cardioscopy)

Our preliminary experience utilizing the hemi-median sternotomy incision with "simple" defects [see atrial septal defect (ASD)] has enabled the repair of more complicated defects using a multitude of adjuvant techniques. Cardioscopy is one such example. With a marked reduction in the operating field and the inherent reduction of stereoscopic vision the addition of cardioscopy has assured a well illuminated, magnified field for all to view.

Allowing the surgeon to view each and every suture placed by a burgeoning trainee remains a key advantage of cardioscopy when deployed in a teaching or University environment. A multitude of rigid scopes with various diameters from 2.6 to 5 mm and various angled lenses $0-45^{\circ}$ are available and have made the correction of lesions involving shunts at various levels, obstructive outflow tracts lesions, and valvular pathologies routine. The recent addition of flexible cardioscopy has provided a safe and unique way to examine the mitral valve in a retrograde fashion, allowing careful examination of the subchordal apparatus frequently abnormal in children. Unfortunately even with the best optical and electronic filters the intraoperative videos and pictures obtained are frequently heavily pixelated and thus may require postimaging processing (Fig. 33.4).

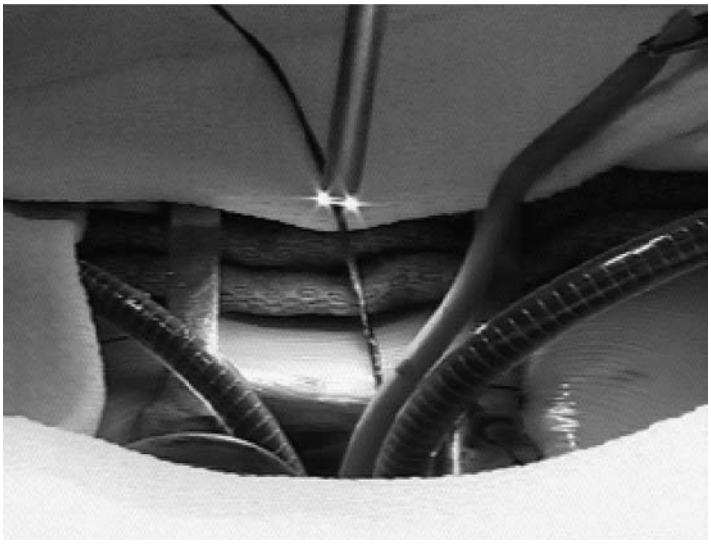


Figure 33.4 Minimally invasive cardiac surgery with a rigid cardioscope.

2.4. Robotic Video-Assisted Cardiac Surgery

Cardioscopy remains limited to the natural tremor amplification found when an assistant surgeon is asked to hold and direct the video picture. Even with best intentions, the assistant is sometimes unable to present the surgeon with the "key" anatomical features required to allow an effortless procedure, until he/she first learns the nuances of the technical exercise him/herself. Exposure to multiple previous procedures takes time and remains patient-limited especially when the pathology remains rare. Much interest therefore has recently been focused on "virtual training" as a method to reduce the "learning curve" frequently seen with newer techniques.

In order to reduce the inadequacies of the human assistant and to allow for improved rhythm and intraoperative performance, the Aesop 3000 (Computer Motion, Goleta, CA) robotic arm with the voice-activated Hermes system has allowed the surgeon to control his immediate external environment without asking others to anticipate his/her thoughts and/or movements. Voice-activated video robotics remains a valuable "adjuvant" for improving the needed visualization within the restricted and rigid confines of a reduced surgical field. The surgeon is now in control of his/her immediate environment. Legitimate concerns over children's safety, the length of the procedure, and the duration of hospitalization has been addressed in our initial and/or intermediate experience demonstrating no untoward mortality or significant morbidity. In fact the avoidance of the human finger, in part due to the confines of the access incisions, may allow for the avoidance of infections. Our experience over the past 3.5 years would verify such a hypothesis as our rates for serious postoperative infections remains negligible. Significant technological advancement continue to be required in order to enable improved micro-maneuvering and positioning of the cameras and instruments within the neonatal heart (Fig. 33.5).

One such advancement may be the use of novel cameras that may allow vision through the viscous medium of human blood! This latter device is currently being developed by our laboratory and should soon enable "open heart surgery" without the use of cardiopulmonary bypass—an advance for both children and adults. Using an array of ports with the aid of catheter-based technology, we are exploring collaborative methods

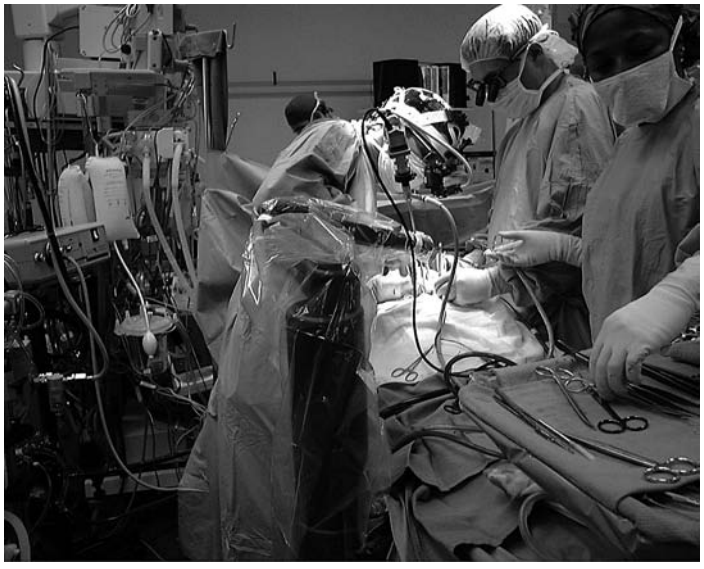


Figure 33.5 Cardiac surgery performed with the Aesop 3000.

of repairing intracardiac lesions without the use of cardiopulmonary bypass. It is likely that cardiac surgery may be performed on a same day of admission as many other ambulatory surgeries of today!

A second development developed in collaboration with a local Californian company, has enabled our first generation robot to be modified to correct both simple and complex types of congenital heart defects through incisions *no larger than 2 inches* (virtual port—avoiding thoracoscopic ports). The latter device reduces the multitude of potentially disfiguring thoracic ports regardless of age and body stature and thus should standardize the procedures allowing widespread acceptance into the medical community. In addition, the use of the above technologies has allowed a multitude of noncardiac disciplines to benefit from our endeavors, that is, prostatic or hepatobiliary surgery (Fig. 33.6).



Figure 33.6 The "virtual port" allows cardioscopy with/without the use of a static arm/robotic platform.

Some of the initial barriers to pediatric cardiac robotic deployment include:

- A relatively small distance between the thoracic cage and the mediastinum.
- The frequent association of "shunt" lesions and thus an enlarged heart (ideal focal point may be an internal position—maximizing robotic intra-cardiac repair).
- Small calibre femoral vessels (inability to successfully cannulate) do not allow the routine initiation of cardiopulmonary bypass.
- The potential for future maldevelopment of the osseous chest and breast tissue with multiple portal sites and nonsternotomy incisions.
- Children and young adults undergoing cardiac procedures typically have maldevelopment of their chest wall due to the abnormal size and positions of the underlying cardiac chambers.

2.5. Total Robotic Telemanipulation

Telemanipulation or the ability to repair surgical lesions from a distant location (from a few feet within the same operating room to over the ocean) has intrigued military surgeons for decades. The ability to repair lesions and pathologies via multiple port access incisions has allowed the correction of coronary arterial obstruction without the use of cardiopulmonary bypass. Since most congenital lesions are considered an "open-heart" procedure we applied our initial attempts at telemanipulation for the repair of extra-cardiac intrathoracic lesions (Fig. 33.7).

The repair of patent ductus arteriosus (PDA) and coarctation of the aorta are routinely performed via a thoracotomy. We have advanced the therapy of PDA using thorascopic techniques and would like to do the same for coarctation of the aorta (Fig. 33.8).

Due to the limitations of thoracscopic techniques (i.e., tremor amplifications, nonprecise micro-maneuvering, etc.), we pioneered robotic techniques to address the latter concerns. Due to FDA regulations most patients are being operated on abroad. However, the techniques are soon expected to be allowed in the United States. We anticipate early discharge from hospital [i.e., same day (short stay)], especially when encountered in the non-neonatal child or infant. Since economic concerns have become progressively significant as a variable in the modulation of the rate of medical change, procedures that potentially reduce hospital resources while maintaining patient safety should succeed and be further developed.

Although the correction of coarctation of the aorta is currently being performed on children and adults using a thoracotomy and in the case of PDA using thoracscopic techniques (limited to a few institutions), no one in North America currently has attempted or has successfully completed the repair of either of these lesions using telemanipulation. Robotic video-assisted and total robotic telmanipulation techniques will be used for ligation of a PDA by the time this chapter reaches publication. Vascular anastomosis can be accomplished via clips rather than sewing; however, the latter technique should be avoided when growth is anticipated and/or desired. Alternative anastomotic techniques will likely soon be developed including novel biologically degradable glues. Although somewhat oxymoronic surgeons may not be required to sew in the not too distant future.

2.6. Robotic Fetal Techniques

The diagnosis of fetal cardiac disease can be ascertained as early as the 16th week of gestation with current ultrasound technologies. Unfortunately, serial antenatal echocardiograms



Figure 33.7 Initial investigation of telerobotic manipulation were limited to animal studies.

are needed to follow the dynamic changes within the cardiovascular system. The development of the cardiovascular system occurs early, by the 12th week of gestation. It is believed that the primary defect (which may be initially a small morphological abnormality) can lead to more severe secondary changes due to the alterations of normal flow and pressures during the remaining gestational period. Alterations of the primary defects are believed to lessen the severity of the secondary lesions, provided the fetus has time during the third and possibly the second trimester to undergo alterations in anatomy due to normalization of blood flows and pressures.

The current philosophy of fetal removal from the confines of the protected uterine milieu, the institution of cardiopulmonary bypass, and the reimplantation of the fetus should be questioned. Since the triggers for spontaneous delivery of the human fetus are not fully known at this time, the potential for early and unwanted delivery is real. Fetal demise and lack of time for maturation of the fetal structures may occur, thus making all previous efforts futile. Placenta failure and fetal death are not uncommon complications of fetal cardiopulmonary bypass.

Alternatives to the latter philosophy have recently included fetoscopy. Micromanipulation with long instrumentation inherently amplifies human inadequacies, that is, tremor amplification. Catheter placement via the umbilical artery in fetal sheep has been successfully accomplished and recently the first successful human fetal cardiac manipulation in North America was reported on 02/23/02 in the New York Times.



Figure 33.8 Single post thorascopic surgery routinely can be used to repair extra-cardiac thoracic abnormalities.

The latter report too revealed the inadequacies of operating upon a fetus with long instruments and catheters.

We have successfully implemented the use of robotic video-assisted surgery as a potential viable option to the methods described above. Telemanipulation provides illumination and magnification as does fetoscopy but with no tremor amplification. Direct visualization and controlled robotic manipulation of the chordal vessels has allowed direct access of the cardiac chambers via catheters and the opportunity for intracardiac manipulation (Fig. 33.9).



Figure 33.9 First generation telemanipulations were used in fetal (sheep) studies.

3. CONCLUSIONS

The diagnosis and/or treatment of children born with congenital heart disease should routinely incorporate cardioscopy, active venous suction, epidural/spinal anesthesia, and robotic video assistance. Our initial experience with the repair of the most technically simple repairs has enabled us to perform more complicated repairs utilizing the hemisternotomy approach. We have since found cardioscopy with robotic video assistance to be an extremely valuable "adjuvant" for improving the needed visualization within the restricted confines of a reduced surgical field. Legitimate concerns over children's safety, the length of the procedure, and the duration of hospitalization has been addressed in our initial and/or intermediate experience demonstrating no untoward mortality or significant morbidity.

It continues to be our belief that minimizing surgical trauma may be of more benefit to higher risk children who would then require less narcotic analgesia in the postoperative period. Since respiratory complications are a significant cause of prolonged ICU and hospital stay, avoidance of narcotic analgesia should improve the clinical outcomes in patients undergoing minimally invasive cardiac surgery.

On-going developments should allow for the repair of selective intracardiac lesions using telemanipulation (full robotic assistance) without the use of cardiopulmonary bypass, hopefully in the near future, utilizing innovative visualization systems.

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The Interventional Radiologist's Role in Pediatric Minimally Invasive Surgery

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1. INTRODUCTION

Pediatric interventional radiology (PIR) services are a rapidly growing facet of many children's hospitals, given the emergence of fellowship trained pediatric interventional radiologists. Pediatric interventionalists perform diagnostic and minimal access therapeutic procedures using various medical imaging modalities to guide them. As in other areas, the minimally invasive nature of the procedures promises to result in less patient morbidity, shorter hospital stays, and more cost effective health care (1–3).

The role of interventional radiology has not yet been fully realized or determined in the pediatric population. The ongoing improvement in medical technology promises to result in evolution of this field with a need for continuing research to determine outcomes and help define the roles of both radiologists and surgeons. Future roles should be determined by evidence-based studies demonstrating whether surgeons or radiologists have the best outcomes, are least invasive, and most cost-effective for each particular procedure rather than making these decisions based on financial gain or "turf" issues. For some procedures a combined radiology–surgery approach may be beneficial.

As this specialty is in its infancy, large numbers of comparative, prospective and cost-analysis studies have not yet been performed making a detailed review of the literature quite limited. As a result, this chapter will outline the background behind

interventional procedures, review basic techniques, modalities and equipment, describe the basic workup of patients for interventional procedures and finally describe the types of procedures that are currently being performed. Results of evidence-based studies and other types of research that have been conducted, along with potential research directions, will be discussed where possible.

1.1. The Image-Guided Therapy Team and Multidisciplinary Collaboration

In addition to performing procedures, the image-guided therapy (IGT) service must take responsibility for pre- and postprocedure care, as well as monitoring outcomes and treating complications (4,5). A dedicated multidisciplinary team approach is fundamental to a successful PIR service (4–7). This approach includes 24 h availability of pediatric interventional radiologists, nurses and technologists, specifically trained in an interventional radiology environment and minimally invasive procedures. The addition of a pediatrician or nurse practitioner to the PIR team improves periprocedural care. Close collaboration with surgical and medical colleagues is essential when providing this type of care and in the development of new and combined minimally invasive procedures.

2. BASIC TECHNIQUE, IMAGING MODALITIES, AND EQUIPMENT

The first invasive radiologic procedure was performed in 1929. Dr. Werner Forssmann inserted a catheter through his own brachial artery and into his heart using a fluoroscopic screen for guidance (8). Modern interventional radiology techniques have their origins in the 1950s from work done by Seldinger and Cope (9). The number and variety of pediatric interventional procedures performed with image guidance has grown as a direct result of improvements in imaging technology, particularly ultrasound (US), fluoroscopy and computed tomography (CT) (9–17). Rapid advances in biotechnology with the development of new catheters, wires, balloons and other devices contribute substantially to sustaining this growth.

2.1. Seldinger Technique

Most PIR procedures are based on the Seldinger technique. Access is obtained with a needle to a structure either by direct visualization, palpation, ultrasound, or fluoroscopic guidance. Through the needle, a wire is passed, the needle is removed, and a catheter is introduced (over the wire) to drain, dilate, stent, or embolize. Through this small (<5 mm) incision, access is gained for diagnostic and therapeutic purposes to many anatomical structures and areas.

2.2. Imaging Modalities

There are a variety of imaging modalities used in interventional radiology. The majority of pediatric interventional procedures are performed using a combination of ultrasound with or without fluoroscopy. CT is used less often in pediatrics than in the adult population due to excellent visualization and lack of radiation that ultrasound offers.

2.2.1. *Ultrasound*

US uses the reflection of sound waves to form images. A US transducer uses piezoelectric crystals to send and receive sound wave pulses. The sound waves are reflected at tissue interfaces. The reflected pulses are then used to form real-time two-dimensional images. Colour, power, and pulsed-wave Doppler imaging are variations that allow colour or graphical visualization of motion. US is the most commonly used modality in PIR and is considered safe within the range used for most diagnostic procedures.

US guidance is ideally suited to the pediatric population secondary to good visibility related to the lack of body fat. It is used primarily for vascular and enterostomy access and biopsies. The advantages include lack of ionizing radiation, faster procedure times (18), multiplanar capability, and the ability to constantly visualize the needle during placement and sampling in real-time from a wide variety of angles and approaches. US is limited by the inability to penetrate air and intact cortical bone. Use of colour Doppler imaging allows vascular structures to be avoided during needle placement (19). Sonographic contrast agents and volumetric and harmonic imaging techniques are recent innovations that may further increase effectiveness of US-guided procedures (20–24).

2.2.2. *Fluoroscopy*

Modern pulsed, fluoroscopy uses intermittent low dose X-rays to provide "real-time" dynamic imaging. X-ray exposures on the other hand, use much higher doses of radiation (25). The fluoroscopic X-ray source is from a single plane or biplane piece of equipment called the C-arm. Biplane units allow simultaneous fluoroscopy or exposures from two directions and result in a lower radiation dose to the patient than using multiple runs with a monoplane unit (26). For anteroposterior (AP) imaging, the X-ray source is usually below the table and the image is captured above the table by the image intensifier.

Fluoroscopy is used primarily for enterostomy and vascular access, vascular procedures, and some biopsies (e.g., bone). Disadvantages include radiation exposure and little or no discrimination between soft tissue types.

2.2.3. *Computed Tomography*

CT units use a tightly collimated strip of radiation that circles around a patient to obtain a two-dimensional map of X-ray attenuation at that level. CT has undergone several major developments since its introduction. Most recent changes include the change from single static images obtained one at a time at specific levels to the introduction of helical and multislice acquisition techniques. CT fluoroscopy is a recently introduced modality that allows real time two-dimensional visualization of needle motion at the expense of increased radiation doses to both the patient and radiologist (27). With new imaging strategies and improved technology, studies are beginning to show decreased dose to patients and staff with CT fluoroscopy relative to conventional CT-guided biopsy (28).

CT offers excellent visualization with high resolution that is not limited by the presence of air or bone. It is used to biopsy lung, bone, and small abdominal and pelvic lesions that are not visible by US and occasionally for access to collections in deep locations. The drawbacks include radiation exposure, limited imaging planes, poor differentiation of some lesions related to lack of fat in babies and children, longer procedure times than sonographic biopsy (18) and, with conventional CT, lack of dynamic visualization of needle movement during placement and sample collection.

2.2.4. Magnetic Resonance

Magnetic resonance imaging (MRI) creates images by using variable magnetic gradients and radiofrequency pulses to cause hydrogen atoms to emit radio waves. Improvements in computer technology and pulse sequence strategies now allow images to be obtained at subsecond intervals. Near-real-time imaging allows the development of MRI-guided minimally invasive procedures.

At present, interventional MRI is in its infancy and is not yet being used on a widespread basis. However, uses of MRI for intervention and intraoperative assessment are being developed and have been increasing in scope in the last few years. Pediatric applications are only beginning to be discussed, with the only reports so far being in the fields of neurosurgery (29), cardiology (30), and orthopedics (31).

Initial interventional and intraoperative MRI units included a horizontal or vertical opening. The open design increases ease of access to the patient. Traditional surgical instruments can be used in relatively close proximity to units because of the low field strength of the open MRIs. Open MRI units are, by definition, low field strength (0.2–0.7 T) because the open access area disrupts the magnetic field and introduces significant field inhomogeneity. Currently, there is a move from open, low field strength MRI units to short-bore, high field strength (32) traditional "closed" MRI units. The desire for higher resolution, lower signal to noise ratios of high field strength magnets and the ability to perform high-end imaging techniques, such as spectroscopy, is behind the change in attitude. Higher field strength magnets are used at the expense of decreased patient accessibility and the necessity for the development of nonferromagnetic surgical instruments or significant changes in room design. While some procedures can be performed within the closed MRI bore by direct operator intervention (33), moving the patient in and out of the MRI bore may be necessary.

Several tools are being developed to improve MRI-guided procedures including patient registration software (32), robotics (34), and positionable MRI compatible operating room tables (35,36).

Current uses for intraoperative MRI include guiding tumor ablation, assessing margins following resection of masses (37) or determining the success of surgical intervention (38). Intraprocedural guidance during endoscopy and laparoscopy is also possible (39).

2.3. Equipment

Needles, wires, sheaths, catheters, balloons, and embolic materials are commonly used in interventional practice and will be briefly discussed below.

2.3.1. Needles

Needles are used for obtaining access and performing biopsies. Description of the multitude of needles and biopsy devices available are beyond the scope of this chapter. Needles are used to obtain access and perform biopsies.

Access Needles. When accessing a structure, the choice of needle size and type is based on patient size, depth of target, size/strength/thickness of guidewire, proximity to vital structures, and radiologist's preference. Angiocatheters are commonly used in the pediatric population as the soft-tipped catheter is easily introduced and minimizes damage during exchanges.

Biopsy Needles. There are two basic types of biopsy needles: aspiration and cutting. Aspiration needles are used to obtain samples for cytopathologic analysis. Aspiration

needles are small in gauge, ranging from ~20 to 24 Gauge. Examples of aspiration needles include spinal and Chiba types. Aspiration needles demonstrate increased yield with increased gauge and decreased bevel angle (40).

Cutting needles tend to be larger in gauge (14–19 Gauge) than aspiration needles and come in end-and side-cutting types. End-cutting needles are like aspiration needles with modification of tip design to increase yield. Examples include Franseen and Turner needles. Side-cutting needles shear the tissue sample. The most commonly used side-cutting needle type is the TruCut needle. The central portion, containing a recessed notch, is introduced into the lesion. A cutting outer cannula is subsequently introduced trapping the sample in the notch.

In the early 1980s, automated biopsy devices were introduced (41,42) in order to increase the ease of performing biopsies resulting in higher yields and shorter procedures while providing high quality core biopsy specimens.

In adults, fine needle aspiration biopsy is common as samples are primarily used for cytologic analysis (43) of adenocarcinomas. Accuracy in diagnosis of the common pediatric sarcomatous tumors is increased with core needle biopsy, which gives a larger sample for histopathologic study (44).

2.3.2. Wires

Wires are used to introduce, guide, and exchange catheters. There are two basic types of wires: solid and wrapped. Wrapped wires have a central solid core called the mandrel with an outer layer of wire wrapped around the mandrel. Mandrels have variable degrees of ability to torque and stiffness. The tips of wires may be angled, to ease introduction into distal branches, or have variable floppiness, to help protect vessel walls. Many have coatings to ease introduction, placement, and ease passage through stenotic areas. Common coatings include Teflon®, heparin, and hydrophilic polymer coatings. Infusion wires are specialized wires that are used to pump fibrinolytic agents directly into a thrombus.

2.3.3. Sheaths

Vascular sheaths are used to protect the arteriotomy site from trauma in cases where multiple catheter changes or prolonged examination times are expected. It is thought that the incidence of stenosis and thrombosis is increased with multiple catheter changes. The diaphragm at the proximal end has an air-tight seal that protects against hemorrhage and air embolism. Long sheaths are used for stabilization or specific purposes like snaring, angioplasty, and filter placement.

2.3.4. Catheters

The two basic types of catheters are angiographic and drainage.

Angiographic catheters are used to navigate to a specific area, perform diagnostic studies, and introduce therapeutic devices, agents, or other catheters. The ends of the catheters have various shapes, each designed to help access specific structures. Some are malleable to allow the operator to further vary the tip design as required for individual situations. Microcatheters—small coaxial catheters that are introduced through larger stabilizing catheters—can be negotiated far out into tiny vascular structures allowing the possibility of targeted, focal therapy with minimal surrounding effect.

Most radiologically placed drainage catheters are “Cope loop” catheters. These catheters have a self-retaining mechanism so that they do not have to be sutured in place. This type of catheter is used for all drainages including abscess and pleural.

2.3.5. *Balloons*

Balloons are used to perform controlled dilatations of both vascular and nonvascular structures. Balloon sizes range from 1.5 to 30 mm. Noncompliant balloon material increases the burst pressure and increases the radial force in the area of stenosis.

2.3.6. *Stents*

Stents are used to buttress open stenotic areas in vascular and nonvascular structures. Stents are usually metal but can be composed of other materials such as silicone. There are many variations in stent design including balloon expandable or self-expanding, covered or noncovered, and flexible or rigid versions.

2.3.7. *Embolic Material*

Embolic materials are used to stop blood flow. There are numerous uses for embolic materials including control of acute hemorrhage, embolizing a biopsy tract, preoperative tumor embolization to decrease intraoperative blood loss, treatment of vascular anomalies, and chemoembolization. Commonly used embolic materials include coils, particles, and glue.

Coils come in various lengths and shape designs. They may be covered with thrombogenic fibres. While coils are considered a permanent embolic agent, there are many reports of recannalization (45,46). Coils may interfere with subsequent procedures by blocking access.

Particles can be temporary, such as Gelfoam, or permanent, such as polyvinyl alcohol (PVA) or Embospheres®. Gelfoam, which is well known to surgeons, is a gelatin sponge that is cut into pledgets or made into a slurry. PVA is an irregular plastic material that is made by grinding down a block of PVA and separating the particles based on size. PVA has been used since the 1970s and, to our knowledge, there have been no reports of allergic reaction. While PVA is supposed to be permanent, recannalization has been reported (47). Embospheres are a relatively new product on market. They are spheres of acrylic polymer cross-linked with gelatin surrounding a saline core. Theoretically, the spherical shape means that a single particle can obstruct a single arteriole/vessel, therefore requiring less embolic material than other embolic agents.

Cyanoacrylate is an injectable polymer that results in thrombosis is used in multiple areas including embolization of Vein of Galen malformations (48), tracheoesophageal fistulae (49) and arteriovenous malformations (50,51), and fistulae, including carotid-cavernous (52).

3. PREPROCEDURE CARE

3.1. Preprocedure Workup

The role of the interventional radiologist begins with the consultation from the referring service. The referring physician must relate the reason for referral, the patient's pertinent history and physical findings, and the diagnostic workup that led to the referral. The patient's medical history and previous imaging are reviewed, allowing the interventionalist to determine if a procedure is feasible and advisable. Factors that help determine feasibility vary with the procedure to be performed. The only absolute contraindications for most procedures are absence of a safe access route and uncontrollable coagulopathy. Additional imaging, to further delineate or potentially diagnose an abnormality, may be requested prior to accepting a request. For example, specific contrast CT protocols can be used to diagnose a lung nodule as an arteriovenous malformation or a liver mass as a hemangioma, negating the need for a biopsy. Once the decision to proceed has been made, the most appropriate imaging modality to use for guidance is chosen based on lesion location, visibility and size, in addition to the radiologist's preference.

Consent and assent are obtained in accordance with regional laws and personal and departmental practice. Assent is the agreement of the child, who is unable to give consent according to regional law, age or inability to understand the implications of procedure performed. In general, the reasons for the procedure, alternative methods of treatment, technique, risks, and complications are discussed with the patient and/or their parents. The implications of not performing the procedure should also be discussed.

Preprocedure bloodwork should include prothrombin time (PT) and/or international normalized ratio (INR), partial thromboplastin time (PTT), platelets, and haemoglobin. The history should include determination of medications that can interfere with coagulation, such as aspirin and heparin, to allow adequate time for them to be stopped. Coagulopathies should be corrected prior to the procedure. While the preprocedure workup includes determination of clotting factors and correction of any coagulopathy, abnormal clotting factors are not a good predictor of haemorrhage (53).

Children fasting for a procedure should not become dehydrated, and orders for maintenance of intravenous fluids should be part of routine pre-procedure orders. Patients undergoing sedation or having a general anaesthetic, should have solid foods prohibited for 8 h before the procedure and clear fluids for 2 h before the procedure, although there is some disagreement in the literature (54–57).

3.2. Radiation Protection

In order to be able to operate imaging equipment, radiologists receive extensive training in radiation physics, radiobiology, and radiation protection (58,59). People who work in and around the interventional suite need to be aware of the invisible but real dangers of radiation exposure (60–64). Radiation dose to IGT staff is higher in procedures performed on children than on adults (65,66). Modern pulsed fluoroscopy with careful collimation minimizes the dose to both patients and staff (67,68). A lead apron with a thyroid shield must be worn during any fluoroscopic or CT procedure (69–74). An apron that wraps fully around the body is preferred if the person is going to be moving around the room so that he or she will still be protected if they turn their back to the beam. A properly fitted belt helps shift the apron's weight from the shoulders to the hips and reduces muscle fatigue. Hand protection should be used, when possible, when the hands are going to be close to the beam. Thin, compliant lead gloves can reduce the dose to the hands by 50% (69,75). Prescriptive and nonprescriptive lead glasses are available for additional eye protection. Because radiation dose is inversely proportional to the distance from the X-ray source, (dose α $1/\text{distance}^2$) every opportunity should be taken to increase the distance of those involved in a procedure (distance from the X-ray beam) when patient safety and stability permit. During exposures and angiographic runs, everyone except the person doing a hand injection should step out of the room or as far as possible (76). Power injectors should be used whenever possible. Studies to determine radiation exposure to patients and IGT staff are very limited in the pediatric population.

Studies looking at neurointerventional procedures and others have been performed. These demonstrate that doses to children and staff can be considerable, but are outweighed by the benefit of performing percutaneous therapy (63,77) and that appropriate techniques can decrease dose during these procedures (78).

3.3. Sedation/Analgesia/Anesthesia

The approach to the type of sedation or anesthesia depends largely on the procedure to be performed and the medical complexity of the patient undergoing the procedure. Most procedures are performed under sedation when feasible.

In general, most pediatric procedures that are painful or that require the patient to be completely immobilized will require deep sedation or general anesthesia. In both cases, the procedure should be done under the supervision of an anesthesiologist. General anesthesia may be indicated for lengthy procedures and for procedures where the area of interest is close to a vital structure. On the other hand, some neonates and patients with oropharyngeal abnormalities may be poor candidates for endotracheal intubation. In these patients (and in older patients undergoing less painful procedures), local anesthesia or light sedation may be all that is required.

Monitoring of sedated patients includes blood pressure checks every 5 min, ECG read out, and continuous pulse oximetry (79). Visual monitoring by physicians, nurses, and technologists is by far the most important assurance of safety and avoidance of complications. Regardless of whatever level of sedation is chosen, full resuscitation and anaesthetic equipment with suction should be immediately available.

A wide variety of medications are available for anesthesia and sedation (80–87). Radiologists tend to use a limited selection of drugs for sedation and analgesia. Infants weighing <5 kg may be given an oral dose of chloral hydrate (80 mg/kg) followed by either intravenous morphine or oral diphenhydramine. In young children (5–20 kg), intravenous pentobarbital (3 mg/kg per IV) followed by intravenous meperidine (1 mg/kg), repeated once if necessary. In older children and adolescents, sedation would include intravenous diazemuls (0.1 mg/kg) followed by intravenous meperidine, repeated once if necessary. Sedation for anxious patients undergoing short relatively painless procedures is usually achieved with oral midazolam. Ketamine infusions are used in some centers (88).

Liberal use of local anesthetics, even in sedated or anaesthetized patients, improves postprocedure recovery and decreases pain experienced at the operative site. Local anesthetics can be administered through a 27 G needle with minimal discomfort. Patient acceptance is improved if the site has been prepared with topical EMLA cream or Ametop gel (89). EMLA patches may also be used in neonates (90–92), but not where there is an open skin wound or mucosa. To decrease the pain of subcutaneous injection, the pH of lidocaine can be raised with a 1:9 mixture of injectable sodium bicarbonate (93). The maximum dose of 1% lidocaine is 0.5 cc/kg. For longer duration of analgesia, 0.25% bupivicaine (maximum dose 1 cc/kg) is used instead of lidocaine. Local anaesthetic with epinephrine (1:100,000) can be used to cause vasoconstriction. This results in prolonged anaesthetic effect and decreases bleeding. Epinephrine should not be used in an area with end artery blood supply (such as the digits, nose, and penis).

4. POSTPROCEDURE MANAGEMENT

After the procedure, a brief note is written in the patient's chart outlining the procedure, outcome, and any further interventions required. Once the procedure is completed, the patient is sent to the postanesthetic care unit (PACU) with orders for pain management. Patients are then followed by the interventional team as required.

5. BASIC/COMMON PROCEDURES

At the Centre for Image-Guided Therapy at the Hospital for Sick Children in Toronto, over 6000 interventional cases are performed each year (Fig. 34.1). The caseload has risen steadily over the last 7 years and is largely due to close collaboration with referring



Figure 34.1 Photograph of an image guided therapy suite from the Hospital for Sick Children in Toronto, Canada. The room operates at OR standards and includes integrated angiographic, ultrasound, and CT fluoroscopy equipment. The room was also designed to allow the integrated use of surgical equipment such as laparscopes, surgical lasers, and microscopes.

physicians, our surgical colleagues and other disciplines. The majority of procedures are vascular access, enterostomy access, biopsy, and drainage.

5.1. Vascular Access

In some centres, there has been a significant shift in the role of the radiologist in vascular access (94, 95). In the past, surgeons placed all central lines and radiologists diagnosed line malfunctions, repositioned misplaced lines, and retrieved fractured line fragments. The use of image-guided techniques for central venous access device placement have been shown to result in less infections, cost, and misplacements than surgical lines (94–98). The success of image-guided techniques has lead to the increasing placement of central venous lines (CVS) by radiologists in many centres. In addition, increasing use of lines and longer survival times has lead to an apparent increase in thrombosis making placement by traditional techniques difficult to impossible, and require placement of CVLs through collateral vessels (99).

All forms of vascular access devices are placed in the interventional radiology suite including PICCs (peripherally inserted central catheters), central lines, and subcutaneous ports (100).

5.1.1. Peripherally Inserted Central Catheter

A PICC is a long-term venous access device that is placed when the expected duration of therapy is 3 weeks to 1 year. It may be used for administration of fluids, drugs, total parenteral nutrition (TPN), and blood sampling (101–103). Preprocedural bloodwork is not usually required unless the patient is coagulopathic and/or unstable or there is a significant

chance that PICC insertion may not be successful and a tunneled central line may have to be placed. Unsuccessful PICC insertions most commonly occur in younger children (especially neonates) and those who have had multiple lines previously. Active sepsis is a relative contraindication unless the PICC is placed for antibiotic administration.

PICC's are placed under strict sterile conditions using visualization, palpation, ultrasound, or venography to obtain initial access (104–108). In adults, sonographic and venographic access techniques have similar complication rates but sonography has a higher initial success rate (107). Once venous access has been achieved, a guidewire is advanced into the vein, through the SVC into the right atrium. A dilator or peel away sheath is placed over the wire. The catheter is cut to length, placed through the sheath, sutured, and dressed. Determining the appropriate tip position can be difficult especially because the position of the tip varies with arm position (109). A centrally located PICC tip position is associated with a lower complication rate than a noncentral location (110). At our institution, we aim to leave the tip at the SVC—right atrial junction or lower SVC. In 92.5% of children, this is at the level of the sixth thoracic vertebral body (111).

The procedure may take anywhere from 20 min to 4 h depending on the status of the venous patency, size of patient, stenosis, and collaterals from numerous previous lines or thromboses and venospasm. Potential complications during insertion include vasospasm, causing difficulty or inability to advance the catheter, bruising at IV site(s), venous stripping, air embolism, and cardiac arrhythmias. The position of the wire in the right atrium can cause atrial or ventricular tachycardia and ectopic beats.

Later complications include infection, thrombosis, fracture, and tip perforation. Symptomatic thrombosis is seen in 1–4% of PICC lines (112,113). A combined adult–pediatric paper found high thrombosis rates with PICC lines (114). Thrombosis was found in 23.3% of patients with a single PICC insertion and 38% who had undergone more than one placement. Thrombosis was found in 57% of patients with cephalic PICC lines. Cystic fibrosis patients may be at increased risk of thrombosis (115). Other complications include PICC fractures with subsequent embolization of the fragment to the SVC, heart or lungs in 0.1 to 2.7% (116). The natural history of undetected fractures is unclear. In one case, a PICC line fragment was removed from an asymptomatic patient's pulmonary artery when discovered, 11 years after the embolization occurred (117).

PICCs can also be placed by specially trained nurses. If malpositioned, spontaneous correction of position can occur (118). Cost-effectiveness of radiologic vs. radiologic/RN placement depends on the cost of the interventional suite (119). A recent study, however, suggests that PICCS' may be less cost-effective than initially thought due to costs related to thrombosis and difficulty in placement (121). Follow-up studies to assess long-term effects in the pediatric population have not been performed.

5.1.2. Central Venous Line

Single and double lumen central venous lines are frequently inserted in the interventional radiology department for TPN, chemotherapy, and dialysis. For children and infants, CVL insertions usually require a general anesthetic. Many teenagers require anesthesia because of anxiety and poor cooperation. Intermittent positive pressure ventilation reduces the possibility of air embolism (122,123), a complication of both insertion (123–126) and removal (127–131). Paralysis is not necessary for CVL placement. Ideally the patient is placed in Trendelenburg position on a tilting table during insertion (132).

Radiographic CVL placement varies in technique from surgical insertion in that initial access is obtained using sonographic guidance and the catheter length/tip position is determined fluoroscopically. Sonographic access has been demonstrated to be safer than

traditional landmark techniques (104,105). Preferentially, the right jugular vein is punctured under ultrasound guidance with a 19 G needle, and a wire advanced into the SVC. The subclavian veins are almost never used, particularly in patients with renal disease (133,134). A dilator is passed over the wire followed by an appropriate peel away sheath. The catheter is tunneled from a small chest wall incision along the plane of the subcutaneous tissue and the end is externalized at the neck site. After shortening, the catheter is introduced through the sheath. After checking the position, the catheter is secured and dressed. The final fluoroscopic assessment allows misplaced catheters to be immediately repositioned. Potential complications include bleeding, cardiac arrhythmia, pneumothorax, and trauma to the great veins.

When central access by convention routes is not possible due to thrombus or other abnormality, placement of CVLs can be performed through collateral vessels (99) or utilizing transhepatic (135) or translumbar (136) routes.

There are no large, prospective series reporting CVL use and risk profile for pediatric population.

5.1.3. Port Insertion

Radiologic placement of ports has been shown to be cheaper than surgical placement in one study (96). Radiologic port placement has been demonstrated to be safe in children (137). Radiologic methods are similar to surgical and, like CVL placements, vary in image-guided access and fluoroscopic adjustment of the length and tip position.

5.1.4. Retrieval of Line Fragments

One complication of PICC and CVL lines, particularly medially placed subclavian lines (138), is line breakage. Fractured PICC and CVL fragments are retrieved from the cavae, heart, and pulmonary arteries (PA) by using a gooseneck snare to entrap the fragment. (Fig. 34.2) In the heart or PA's. directional catheters are used to introduce the snare to the desired area. Biplane fluoroscopy is sometimes helpful.

5.2. Enteral Access

5.2.1. Gastrostomy and Gastrojejunostomy

Percutaneous gastrostomy was first described in a child in 1980 (139). Since then several studies have demonstrated its safety and efficacy (140,141) (Fig. 34.3, 34.4). The majority of gastric (G) and gastrojejunal (GJ) tubes are placed in neurologically impaired children (140). G-tubes are direct gastric tubes and GJ tubes are tubes that pass through the stomach to feed into the jejunum. The indications for placement include neurologically impaired children with an absent gag reflex, inability to feed adequately by mouth, severe gastroesophageal reflux, oromotor incoordination, and excessively high metabolic needs. GJ tubes are placed as an alternative to fundoplication. A randomized, controlled trial comparing the two is being planned. As many of these patients have gastroesophageal reflux and an absent gag reflex; they are at risk of pulmonary aspiration. Contraindications to placement include coagulopathy and anatomic variants prohibiting safe access to the stomach.

Retrograde and antegrade insertion techniques have been described (140,142). A description of the retrograde technique follows. Prophylactic antibiotics are administered. To avoid injury to surrounding organs, the edges of the liver and spleen are mapped with ultrasound and barium is introduced to outline the colon. The stomach is distended with air

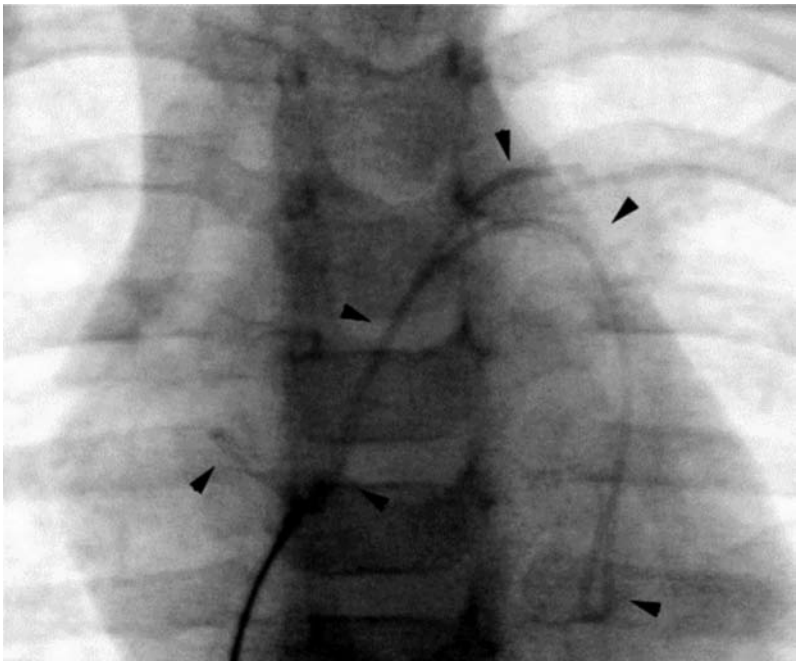


Figure 34.2 A gooseneck snare was used to entrap a PICC fragment (arrowheads) from the pulmonary artery.

through a nasogastric (NG) tube. Glucagon is administered to cause constriction of the pylorus and gastric atony (143,144). The side-effects of glucagon are usually not of major significance, but may include tachycardia (145), hypertension, and hyperglycemia (144). A significant degree of gastric distension required may be required to distend the

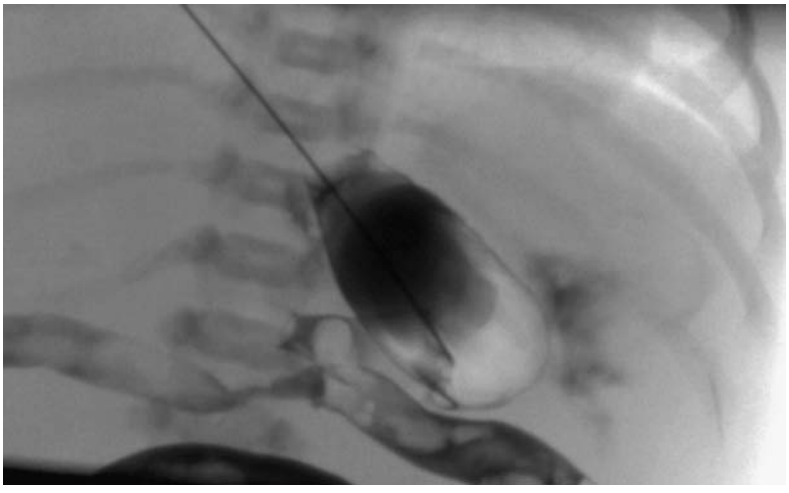


Figure 34.3 Gastrostomy in a newborn with esophageal atresia. A needle has been introduced into the stomach using sonographic guidance. The needle is used to inject contrast, to assure an intragastric location, and to insufflate the stomach prior to puncture.

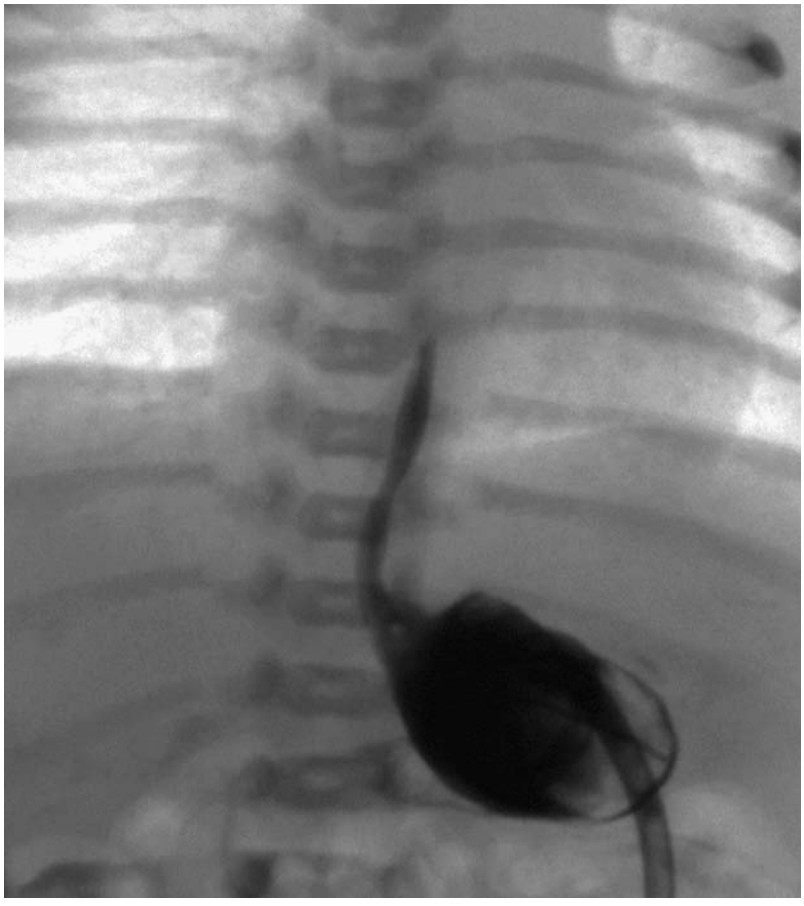


Figure 34.4 Frontal view showing the gastrostomy tube in place. Note the refluxed contrast demonstrating the level of the atresia.

stomach below the level of the lower margin of the liver, and to depress the colon away from the stomach. The distended stomach is punctured under fluoroscopic guidance avoiding the colon. Endoscopic assistance is occasionally used to chose a puncture location in patients with gastric varices (146). Contrast is instilled through the needle to confirm an intragastric location. Using the least amount of contrast possible minimizes the risk of aspiration. The G-tube is exchanged with the needle over a guidewire after dilation of the tract. GJ tubes are inserted over a longer wire that has been steered by a directional catheter from the pylorus into the jejunum. A small amount of pneumoperitoneum is almost the norm at the end of the procedure (147). Significant pneumoperitoneum that compromises respiratory excursions can be aspirated with a 27 Gauge needle.

VP shunts can become infected secondary to gastrostomy placement (148). Sane found that 2 of 23 patients with VP shunts became infected. During that study, antegrade gastrostomy (in 21 of 23 patients) was performed without prophylactic antibiotics. In a review at our institution 1 infection was found in 49 placements in patients undergoing retrograde placement with antibiotic prophylaxis.

5.2.2. Cecostomy

Cecostomy tubes are placed to administer antegrade enemas to treat fecal incontinence in children with spina bifida or congenital rectal anatomic abnormalities (149–152). (Fig. 34.5) Evacuating the colon from the cecum in an antegrade manner helps avoid intermittent soiling from fecal residue. Cecostomy insertion can allow the patient to become independent with the ability to perform their own enemas and avoid the need for rectal washouts (153). Percutaneous fluoroscopic cecostomy was developed as a minimally invasive alternative to the MACE procedure (154) where a surgical appendicostomy is created. Percutaneous cecostomy saves the appendix for a Mitrofanoff procedure. Endoscopic cecostomy insertion was subsequently described (155). The patient's anatomy may be distorted because of spina bifida and/or scoliosis. Sensory deficit often renders the procedure less painful for patients with a high level lesion. The majority of cecostomy tubes are placed under sedation. It should be noted that there is a higher incidence of latex allergy in patients with spina bifida (156). As such, latex precautions should be used routinely.

5.3. Biopsies

Image-guided percutaneous biopsy techniques have matured sufficiently to become the method of choice for obtaining diagnostic tissue specimens for pathologic analysis in

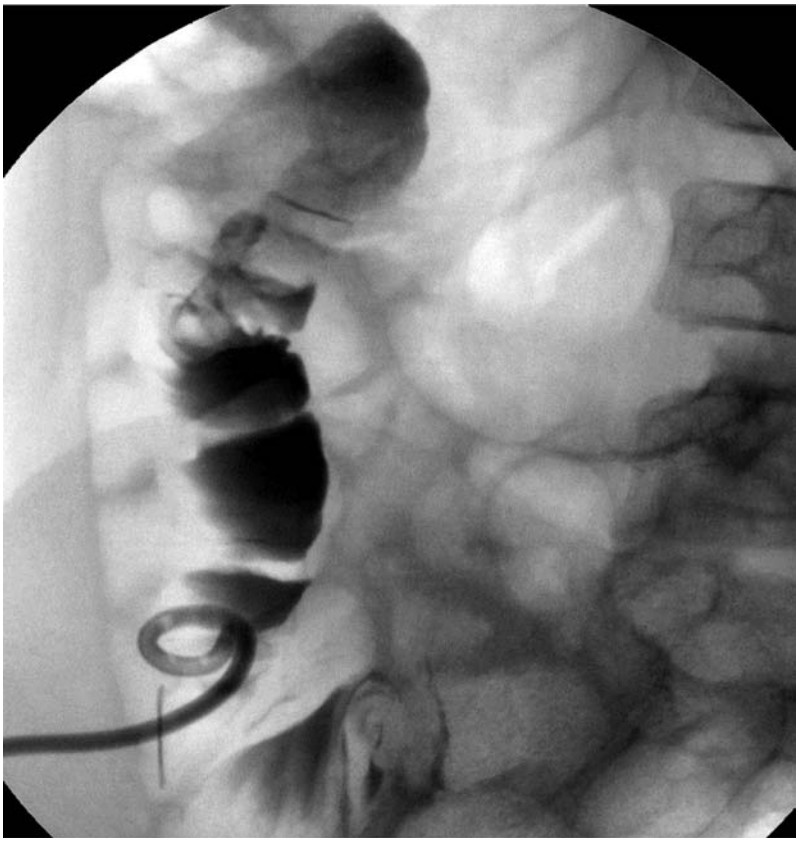


Figure 34.5 Cecostomy tube insertion in a 9-year-old male with fecal incontinence. This therapeutic intervention allows the administration of antegrade enemas to give patients a period without worry of incontinent episodes.

many areas and tumor types (157,158). In interventional radiology we perform most organ biopsies (i.e., kidney and liver) under sedation. The normal prebiopsy requirement is a normal coagulation profile.

When determining a biopsy route, large vascular structures and specific organs must be avoided. Organs that should not be traversed include lung and pleural space (in abdominal and mediastinal biopsies), gallbladder, pancreas, and bladder. Liver and bowel can be safely transgressed with small calibre needles (159). The likely etiology of the abnormality will also determine the route. For example, a retroperitoneal approach should be used in a suspected malignancy such as neuroblastoma and Wilms tumor to avoid peritoneal seeding. The bowel should not be transgressed in the face of infection because of the possibility of fistula formation. Peripheral liver lesions should be sampled through a parenchymal tract. Good visualization of the lesion and pathway is essential for the procedure to be performed safely and to ensure the sample is taken from the appropriate location. In certain oncological conditions, proposed surgical incisions and skin flaps, and so on must be avoided and should therefore be marked by the surgeon. Preferably, contamination (by bacteria, malignant cells, fungi, etc.) should be contained to its space of origin to avoid contamination of another space (e.g., pleural, peritoneal, retroperitoneum, skin dermatomes, etc.).

Biopsies of large lesions (within the liver, kidney, or spleen) can be performed during spontaneous respiration in a sedated patient. If the lesion is small and deep, poorly accessible or close to vital structures, then controlled respirations using general anesthesia with intubation and paralysis increases the safety of the procedure and likely the positive yield of the biopsy. End expiration is frequently required as a reproducible, easy to maintain state of respiration, allowing the lesion to be held in clear view and biopsied. The same holds true for lesions high in the liver, close to the diaphragm. In the spleen, lesions as small as 5 mm can be sampled with controlled respirations. Controlling respirations may also reduce the possibility of shearing or tearing of the spleen or the liver capsule theoretically reducing the potential complication rate.

Similar principles hold true for lung biopsies. Superficial peripheral pulmonary nodules ($\geq$3 mm) are sampled under ultrasound guidance. These require controlled respirations and close cooperation between the anaesthesiologist and the interventionalist. CT guidance may be required for lung biopsy as a lesion within the lung parenchyma will not be visible by ultrasound. A coaxial approach utilizes a guiding needle (e.g., 19 G) to puncture the pleura once and is advanced to the edge of the lesion. Through the outer needle, multiple passes can be made with a smaller biopsy needle (e.g., 21 G) through the lesion. Controlled respirations, usually at end expiration, enables the lesion to remain within the CT slice of interest with sufficient consistency to provide a high diagnostic yield ($\sim$85%) (160). Lesions that are too small for percutaneous biopsy can be localized using CT-guided placement of a hook wire (161–163) or India ink (164), prior to surgical biopsy.

When biopsy of the lung is performed, acute complications that may arise are pneumothorax and hemothorax (165). Other complications of biopsy include hemorrhage, infection, needle tract seeding (166), and sampling error.

5.3.1. Transvenous Biopsy

In patients with a bleeding diathesis or other contraindications to percutaneous biopsy, transvenous biopsy can be performed. The tenet is that if bleeding occurs, it does so into the venous system. The first description of the technique was published about dogs in 1976 (167). Transjugular liver (168–170) and renal biopsy (167,171) techniques

have been developed. In place of transjugular biopsy, some authors advocate for embolization of the tract of a percutaneous liver biopsy with Gelfoam in patients with a coagulopathy (172).

Transjugular Liver Biopsy. Transjugular liver biopsies are performed on children with severe coagulopathy that prohibits percutaneous needle biopsy (168). The biopsy system we employ is a 7 French (Fr.) Desilets–Hoffman catheter set with a Quick-core^{TM} cutting needle. Because of its size, this is suitable for infants who are really quite small (e.g., 5 kg). Access is obtained to the the right jugular vein and a directional catheter is used to cannulate the hepatic veins. A stabilizing catheter is then used to introduce the needle into the hepatic parenchyma through the hepatic vein wall. Firing the needle, advances the tip of the needle beyond the catheter, obtaining a 2 cm core. (Fig. 34.6) The biopsy is monitored using ultrasound and fluoroscopy to allow modification of the needle-tip trajectory to ensure adequate thickness or volume of liver tissue is sampled avoiding the capsule and major vascular structures. (Fig. 34.7) After a post-biopsy venogram to ensure no breach of the capsule, the inner and outer catheters are sequentially removed over a wire. Other complications include parenchymal hemorrhage and cardiac arrhythmias related to the wire or catheter manipulations.

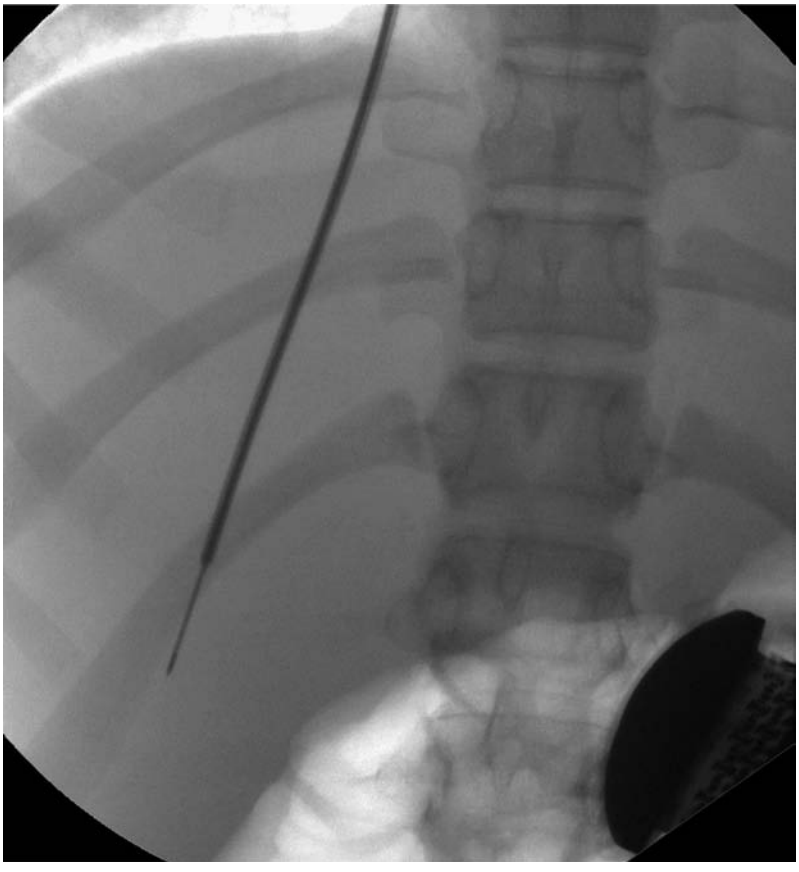


Figure 34.6 Fluoroscopic image of transjugular liver biopsy. The needle is seen extending from the guiding sheath within the liver parenchyma. Note the ultrasound transducer in the lower right corner of the image.

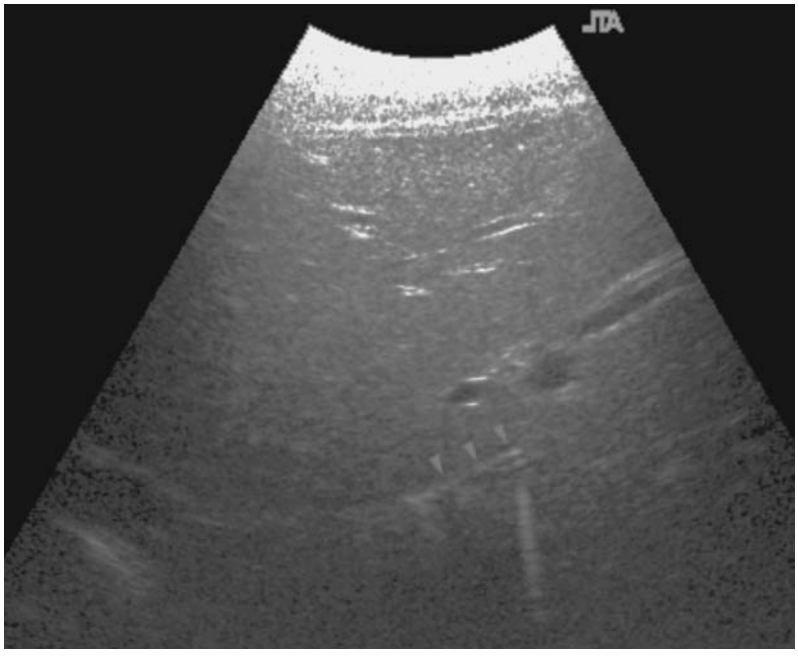


Figure 34.7 Oblique ultrasound image showing transjugular biopsy needle (arrowheads) within the hepatic parenchyma. Monitoring with ultrasound assures that the needle does not puncture the capsule or large adjacent vessels.

5.4. Drainages

Interventional radiology is frequently called upon to drain abscess and other fluid collections situated in a variety of locations. The majority can be approached using a percutaneous approach with a combination of ultrasound and fluoroscopy. CT guidance is rarely required for abscess drainage in pediatric patients. US is used to choose the optimum access point and then guide needle placement prior to fluid aspiration. Contrast is instilled to outline the cavity or space; a wire is advanced through the needle followed by a dilator and then a drainage catheter (4–22 Fr. appropriate to the size, viscosity, and location of the collection) is inserted under fluoroscopic guidance.

Complications of percutaneous drainage are infrequent and usually minor. Fever shortly after initial drainage may represent bacteremia as a result of manipulation or lavage of an abscess cavity. Severe hemorrhage occurs infrequently and is either related to unsuspected coagulopathy, laceration of a vessel, or pseudoaneurysm related to pancreatic inflammation. Disruption of the pleura during percutaneous drainage of subphrenic, hepatic, or splenic collections may result in pneumothorax, haemothorax, or pyothorax/empyema. It is possible to contaminate other spaces, by traversing them during drainage, such as the subphrenic and subhepatic spaces during drainage of a hepatic abscess. Similarly, generalized peritonitis may develop due to perforation of the wall of an abscess cavity. Rarely, bowel perforation may occur at the time of initial puncture or due to catheter erosion. Overall, morbidity associated with image-guided percutaneous drainage is significantly lower than with operative drainages, predominately related to the advantages of real-time ultrasound control. Mortality is extremely rare in children treated by percutaneous aspiration or drainage, and is more often related to underlying pathology and the clinical status of the patient than to the drainage procedure itself.

5.4.1. Peritoneal Abscess

The most common indication for abscess drainage in the pediatric population is following appendiceal rupture. Following acute appendicitis or ruptured appendiceal abscess, the high risk of recurrent abscesses and postoperative adhesions is a relative contraindication to immediate appendicectomy (173–178). In our institution, US-guided percutaneous drainage is the preferred treatment of localized collections presumed to be due to a ruptured appendix. These children may require several drains, repeated procedures, and a good deal of patience to achieve success. Aspiration with a 20 or 22 Gauge Chiba needle is an accepted treatment for collections that cannot be safely drained such as interloop abscesses or displacing the overlying loops of bowel with the ultrasound probe; this is more feasible in pediatric patients (178).

Pelvic abscesses are usually deep percutaneous options due to overlying bowel, vessels, urinary bladder, and bony pelvis. A transrectal approach has been our preferred technique in 67 cases where fluid collections have been located deeply within the pelvis (179). Under US control, a long trocar needle is advanced either along the index finger or protected within the enema tip. The needle tip is positioned adjacent to the abscess collection and then advanced into the abscess under real-time imaging. For lesions higher up in the pelvis, an enema catheter is used to protect the mucosa from the sharp tip during introduction of the needle. Similar techniques may be used for transvaginal (180) or transperineal (181) access to pelvic fluid collections, but these are seldom employed in the paediatric population. A transgluteal route is used by some centers, but this approach is associated with a higher incidence of pain, bleeding, nerve damage, and other complications (182–184).

Subphrenic abscesses are usually seen as a postoperative complication or in patients with a ruptured appendix. They are often difficult to access and may require a transhepatic subcostal approach or an intercostal approach. Combined US and CT guidance may be necessary to access these collections, using fluoroscopy for final tube placement (185,186).

5.4.2. Liver Abscess

Primary hepatic abscesses are uncommon in the paediatric population (187,188). Pyogenic abscesses are most commonly seen in patients with chronic granulomatous disease and other causes of impaired immunity and following abdominal trauma. Less common causes include appendicitis (septic emboli), PV thrombosis, or liver infarction (as in sickle cell disease or tumor necrosis). Depending upon their size, pyogenic abscesses are aspirated or drained (189–192). Small lesions are aspirated with a Chiba needle with US guidance. For larger collections, standard drainage technique is used with access under US guidance followed by contrast, wire placement, tract dilatation, and drainage catheter insertion under fluoroscopy. Just as for liver biopsy, a choice of access route that interposes normal liver tissue between the capsule and the collection will help prevent hemorrhage and, in this case, intraperitoneal contamination.

Percutaneous drainage plays an important role in the management of children with infected collections occurring after liver transplantation (193,194). Re-transplantation has been avoided in some children after successful drainage of infected bilomas, secondary to hepatic arterial thrombosis. In the immunocompromised patient, focal micro abscesses are often seen as a consequence of fungal infection. These are usually too small to allow for drainage but aspiration or biopsy can be performed for diagnostic purposes. The multiloculated cysts with satellite cysts characteristically seen on CT with Echinococcus granulosa infection are preferentially treated with puncture, aspiration, injection, re-aspiration technique (PAIR) using 95% alcohol as scolecide agent (195,196). Standard access

technique is used to catheterize the cysts, and portions of the cyst fluid are exchanged with a quantity roughly equivalent to one-third of the aspirated fluid. Insertion of a catheter is not necessary and the procedure can be performed with a needle alone. This replacement technique avoids intraperitoneal spillage of infected fluid during the procedure. Another series used a similar technique but hypertonic saline was injected instead of alcohol (197). Antimicrobial therapy alone has (albendazole) alone have been used on some series though (198). And in those, persistence of symptoms for greater than two weeks, while on appropriate medical therapy, is an indication for percutaneous management. Antibiotic therapy is continued for eight weeks or until the catheter is removed (199,200).

Patients who have immigrated from or traveled in underdeveloped areas, particularly in Africa, South America, and Asia are at increased risk for amebic hepatic abscesses. They present as single or lobulated masses, usually in the right lobe and are most often treated conservatively. Percutaneous drainage is indicated for failed antimicrobial therapy or imminent rupture.

Hepatic abscess is a known but rarely reported complication associated with umbilical venous catheter (UVC) placement (189). UVCs are used almost routinely in very low birthweight (<1000 g) infants. Percutaneous treatment of hepatic abscesses in very low birthweight infants can be performed safely, with gentle technique and small (5- or 6-Fr.) catheters, and may represent a desirable alternative to conservative treatment.

5.4.3. Cholecystic and Pericholecystic Collections

Drainage of pyogenic cholecystitis and infected pericholecystic collections is occasionally performed in the pediatric population using the transabdominal technique described above. Cholecystostomy can be performed using a transhepatic or peritoneal approach. The surmised advantage of transhepatic access is a decreased chance of intraperitoneal leakage of bile contents (201).

5.4.4. Splenic Abscess

The optimum approach to splenic abscesses is controversial. There are concerns about the safety of transsplenic percutaneous intervention. However, experience in both adult and pediatric patients suggest that percutaneous drainage of splenic abscesses can be performed safely and effectively (202,203). Percutaneous drainage offers the advantage of preserving splenic function, important for host immune response. Percutaneous aspiration can be performed in any size collection in the spleen. Drainage with a catheter should be considered in a significantly sized lesion that is not complex and is not close to any large vascular structures.

5.4.5. Pancreatic Collections

Percutaneous drainage of pancreatic fluid collections is indicated in the presence of infected, symptomatic, or persistently enlarging pseudocysts (204–208). Pancreatic phlegmon, peripancreatic necrosis, or the presence of pseudoaneurysms or varices are relative contraindications to percutaneous management. Pancreatic abscesses are rare, but have been seen in particularly immunocompromised children. Depending on their size and position, they may be aspirated with a thin needle or drained.

Pseudocysts usually develop as a result of acute pancreatitis from whatever cause. They may present as an upper abdominal mass, which appears hypoechoic on US, with or without debris. CT confirms the appearance of a low-density collection, which usually arises close to the pancreas in the region of the lesser sac. Clinically, most of

these pseudocysts will resolve with bowel rest and parenteral nutrition. However, persistent symptomatic collections require treatment, preferably by transgastric drainage. After defining the extent of the cyst on preprocedure CT, drainage is usually performed under combined US and fluoroscopic control.

6. CHEST INTERVENTIONS

The most common thoracic interventions are lung biopsy and placement of drainage catheters for pneumothorax and empyema. Angiography is most commonly performed for diagnosis and embolization of arteriovenous malformations, pulmonary embolus, and haemoptysis. Esophageal dilations and tracheal stenting will be discussed below.

6.1. Esophageal Dilatations

Since the advent of balloon dilatation of strictures (209,210), many esophageal strictures are now dilated in the interventional radiology suite under fluoroscopic guidance. The majority of dilatations are performed for anastomotic strictures secondary to tracheoesophageal fistulae but can also be required in epidermolysis bullosa, achalasia, gastroesophageal reflux, and following ingestion of a corrosive substance. The risks include esophageal perforation and bleeding. Complications following dilatation include pain, tracheal aspiration, and perforation with mediastinitis or hemorrhage (211). Patients with tight stenoses are at risk of aspiration from the retained contents within the esophagus.

The stricture is outlined with a small volume of water-soluble contrast. A soft guidewire is then advanced through the stricture to guide fluoroscopic placement of a balloon catheter. The balloon is inflated by hand or pressure device and the stricture is dilated. Narrowing of the balloon, seen as a waist, denotes the point of stricture. As the dilation progresses, the waist disappears. As marked distension of the esophagus with a balloon can cause pressure on the posterior wall of the trachea, balloon dilatation must be performed carefully in children with known airway compromise. At the end of the procedure, contrast is instilled into the esophagus to exclude an esophageal perforation.

In patients with strictures secondary to epidermolysis bullosa, this procedure is performed in a similar fashion but with extreme gentleness and avoidance of any tape to the face during the procedure. Similar principles of balloon dilatation are applied to colonic strictures secondary to necrotizing enterocolitis or anastomoses with variable results (212–214). Transcatheter balloon dilatation of strictures, related to necrotizing enterocolitis or other causes, is performed on occasion with variable results (212–214).

Placement of an esophageal stent has been performed in children with persistent strictures (215), using either a nitinol coil-type stent or a solid silicone stent (Fig. 34.8). The nitinol stent is easily placed but is frequently complicated by mucosal overgrowth. Recurrence of symptoms is commonly seen and is treated with serial dilatations. The persistence of a tight stricture following dilatation or the inability to physically dilate a stricture may ultimately necessitate surgical resection.

6.2. Tracheal Stenting

Tracheal stenting is uncommonly performed but is occasionally required in children with tracheal stenosis or tracheobronchial abnormalities (216,217). Water-soluble contrast is the agent usually employed to perform bronchography. Small volumes are injected

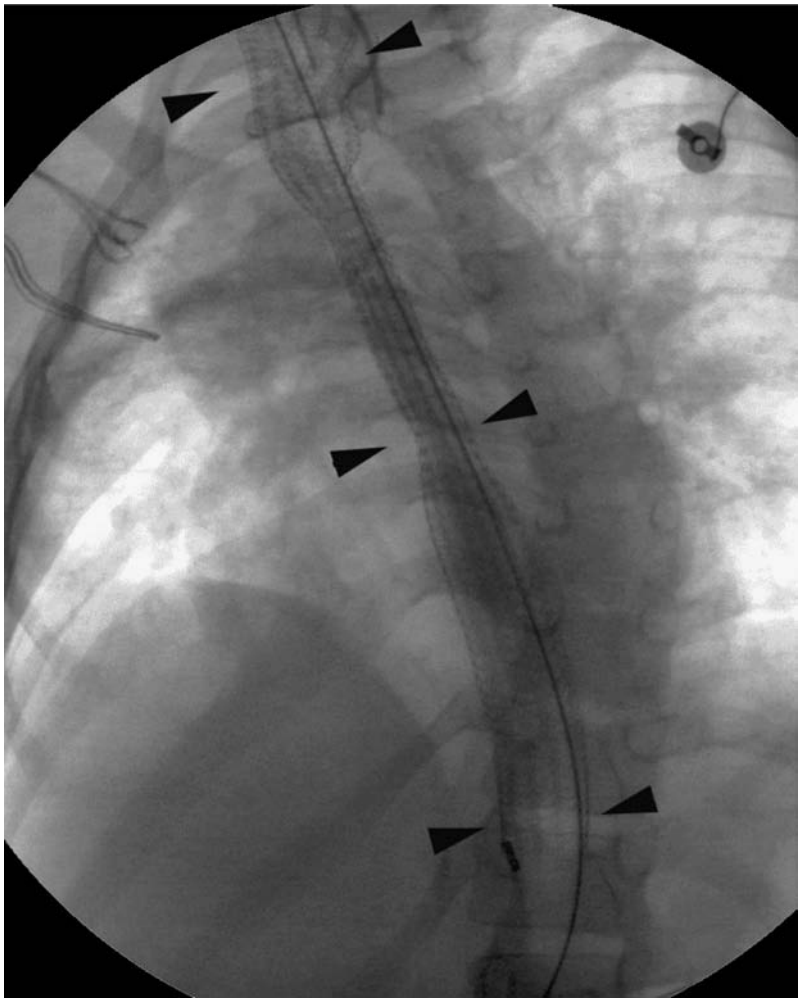


Figure 34.8 Nitinol stent (arrowheads) placed in the esophagus of a 9-year-old female with a long history of persistent esophageal stricture related following repair of a tracheoesophageal fistula as an infant. This procedure was a combined effort between the gastroenterology and the interventional radiology services.

through a small directional catheter selectively into the relevant bronchus. This is tolerated moderately well by the airway. Passage of catheters, balloons, wires and deployment of stents, requires full cooperation within the team, that is, anesthetists, radiologists, otolaryngologists, and respiratory physicians for preoxygenation prior to airway manipulation and for sharing of the endotracheal tube or bronchoscope.

7. BILIARY INTERVENTIONS

The most common pediatric biliary interventions are percutaneous transhepatic cholangiography (PTC), percutaneous transhepatic transcholecystic cholangiography (PTTC), and drainage and stenting in native and transplant livers. The necessity for these procedures has increased substantially with the increasing number of liver transplants performed in

the pediatric population (193,194,218–222). Transhepatic removal of biliary calculi and placement of transjugular intrahepatic portosytemic shunts (TIPS) to deal with sequellae of portal hypertension are uncommonly performed in children.

7.1. Percutaneous Transhepatic Cholangiography

PTC is performed to opacify the intra- and extrahepatic biliary tree with radio-opaque contrast in order to diagnose numerous conditions including obstruction, choledochal cysts, calculi, and sclerosing cholangitis and occasionally biliary atresia. PTC is commonly performed following transplant to assess for biliary injury or obstruction. While magnetic resonance cholangiopancreatography (MRCP) is becoming increasingly sophisticated, its use at this time is limited in the pediatric population, especially in those without marked biliary dilation, because of the inability to resolve tiny structures. PTC is also the initial step prior to therapeutic measures, for example, drainage, stenting, or dilating. Uncorrectable bleeding diathesis, a history of severe contrast reaction, vascular hepatic tumors, vascular malformations, and ascites are regarded as relative contraindications to PTC.

Biliary sepsis is a serious risk, especially in transplant patients and those with obstructed biliary tracts (223,224), making prophylactic antibiotic administration essential. Depressed immune function (225) and bacterial colonization with enteric organisms (223) is common in biliary obstruction, especially associated with liver transplantation (224) and bile duct reconstruction (226,227). Biliary sepsis from manipulation of infected ducts is the most common cause of serious complications during interventional procedures. Therefore, a low threshold should be maintained for the use of preprocedure antibiotics.

7.2. Transhepatic Transcholecystic Cholangiography

When PTC is unsuccessful, the biliary tree can be opacified through the gall bladder and cystic duct cholangiography (228,229). Ultrasound is used to introduce a small needle through the liver into the gall bladder. The transhepatic approach is used in an attempt to decrease bile leak following the procedure. Dilute contrast is injected to fill the biliary tree. Trendelenberg positioning and administration of morphine (to constrict the ampulla of Vater) may be used to aid in filling of the intrahepatic biliary ducts. At the end of the procedure, the gall bladder is emptied to minimize the chance of bile leak.

7.3. Biliary Drainage

Biliary drainage is commonly performed to relieve obstruction secondary to ischemic strictures, following hepatic artery thrombosis in children with liver transplants (193,222).

Biliary drainage is performed by accessing the biliary system with a combination of sonography and fluroscopy. Diagnostic cholangiography is performed, a wire is introduced through the biliary system (preferably into the duodenum) and a catheter is then passed over the wire following dilation of the tract. This is an internal–external drain that allows bile to drain through and around the tube into the duodenum. If initial attempts to reach the duodenum are unsuccessful in a patient with infective cholangitis, an intrahepatic drain will be left to drain externally and the patient brought back at a later date to advance the catheter into the duodenum. The risk of biliary sepsis in such a patient is high and precludes prolonging the procedure in an attempt to access the intestinal tract.

Percutaneous transhepatic drainage is particularly well suited to patients with large postoperative defects or with leaks associated with severe acute necrotizing pancreatitis (230). Percutaneous drainage may avoid the need for surgical re-exploration, surgical or endoscopic sphincterectomy, or even hepatic lobectomy. Although endoscopic drainage is an alternative, it is usually not possible in the presence of a hepaticojejunal anastomosis.

Patients are usually returned to the ward with the catheter left to gravity drainage. The catheter is flushed twice daily with 5–10 cc of sterile saline to keep the catheter and side-holes clear of thrombus or debris. To avoid protein and calorie malnutrition, fluid and electrolyte depletion, and to assist uptake of drugs (e.g., cyclosporine) that depend on enterohepatic circulation, early internalization of biliary drainage is desirable. Once the draining bile clears, a trial of internal drainage is attempted by capping the external drain. If this is successful and long-term drainage is anticipated, conversion to a completely internal drain or stent may be considered.

7.4. Biliary Dilatation and Stenting

Dilatation of biliary strictures is usually performed with balloon catheters over a wire (219,231–233). The size of the balloon used for dilatation should be gauged according the diameter of the duct proximal and distal to the area of stricture. Often, several dilatations are performed, each lasting no more than 15 s. Following dilatation, a biliary drain is usually left in position with side holes made above and below this area of stricture. Internal biliary drains are seldom used in the pediatric population. Occasionally, expandable Palmaz stents have been used for anastomotic strictures in transplants.

7.5. Percutaneous Endoluminal Bile Duct Biopsy

If a ductal or periductal obstructing mass is identified by imaging or suspected in relationship to a biliary stricture, the thin access needle may be exchanged for a suitable sheath. FNA biopsy may be performed through the sheath for cytopathologic analysis (234). A biopsy forceps (235) or transvascular needle (236), may be used to obtain histologic tissue specimens. In smaller children, a periductal tissue sample may be obtained through a second biopsy needle targeted at the region of obstruction under combined fluoroscopic guidance and US.

7.6. Portal Venous Interventions

The portal venous (PV) system can be accessed percutaneously via the transhepatic route quite easily under US guidance. This allows dilatation of posttransplant strictures, embolization of varices, and direct measurement of PV pressure when the PV and hepatic sinusoidal systems are discontinuous (e.g., extrahepatic PV obstruction, splenic vein obstruction, or presinusoidal portal hypertension) (237–239). Embolization of PV to hepatic venous malformations is possible in patients with hepatic haemangiomas (240–242).

7.6. Transjugular Intrahepatic Portosystemic Shunt

The TIPS procedure involves the creation of a parenchymal tract between the hepatic and portal veins with a metallic stent (243–248) (Fig. 34.9). Indications include treatment of GI bleeding, varices or intractable ascites due to portal hypertension. Other indications for this procedure include relief of hepatic outflow obstruction (i.e., Budd–Chiari syndrome)

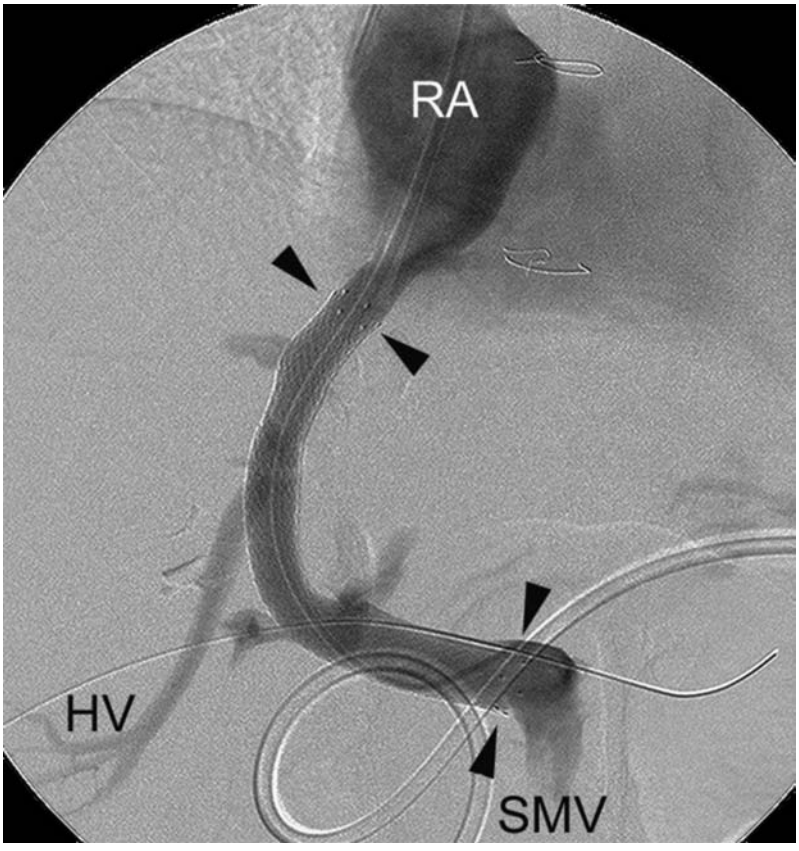


Figure 34.9 Six-year-old female with biliary atresia and numerous episodes of variceal bleeding. Access is obtained from a right hepatic vein to the right portal vein using an approach similar to that seen in Figure 34.5. The shunt (ends marked with black arrowheads) extends from the hepatic vein superiorly into the portal vein. Retrograde flow opacifies a hepatic vein branch (HV) and the superior mesenteric vein (SMV). Antegrade (hepatopedal) flow has been established through the shunt into the right atrium (RA).

(249) and reduction of intraoperative morbidity during liver transplant surgery (249–255). Candidates for TIPS should be evaluated with endoscopy to confirm the presence of varices and exclude other sources of hemorrhage for which alternative therapies would be more appropriate. Contraindications include heart failure, polycystic liver disease, infection, severe hepatic encephalopathy, and severe liver failure. TIPS is described as a safe and effective procedure for the reduction of PV pressure in children as young as 3 years of age, with a short-term success rate of 75–90% (247,256,257). Long-term complications arise due to intimal hyperplasia, stricture, and thrombus within the shunt lumen (258).

Minor procedural complications have occurred in approximately 10% of cases. Hepatic encephalopathy occurs in 5–35% of patients following TIPS (259). Other severe life-threatening complications occur less frequently being reported in ~5% of patients. These include haemoperitoneum, haemobilia and biliary fistula, acute hepatic ischemia, pulmonary hypertension, and pulmonary edema. Chronic complications include PV thrombosis, haemolysis, and shunt stenosis. Typed and crossed blood

should be available for emergent transfusion, and a PICU bed should be available for close monitoring, should the need arise.

Following the procedure, there is usually hepatopedal flow to the IVC with collapse of varices. Rarely, the varices persist despite a well functioning shunt, in which case embolization is performed. If the shunt is felt to be open but the portosystemic gradient remains above 12 mmHg, the stent is usually dilated to a diameter of 10 mm (260). If the pressure remains elevated, a second parallel stent can be placed (261).

8. VASCULAR INTERVENTIONS

Radiologists act to both diagnose and treat a large variety of congenital and acquired vascular abnormalities. Vascular anomalies of the liver will be used to illustrate the radiologist's role in this area.

8.1. Treatment of Vascular Malformations

The radiologist plays a key role in the management of these rare disorders as repeated radiological treatments and combined radiological and surgical treatments are sometimes required.

Managing these congenital anomalies demands a clear understanding of the lesions and their natural history. The congenital vascular anomalies include vascular tumors (i.e., hemangiomas and their variants) and vascular malformations. Hemangiomas are benign endothelial cell tumors, whilst vascular malformations are due to errors in vascular morphogenesis resulting in isolated channel anomalies (capillary/arterial/venous/lymphatic) or combined channel anomalies such as arteriovenous malformations and abnormal connections between vascular channels, for example, arterioportal fistulae (262). Vascular malformations include high flow (arteriovenous) and slow flow (venous) malformations.

Approximately 10% of hepatic hemangiomas require specific treatment in infancy as they endanger life from associated cardiac failure and thrombocytopenia. These are the most frequent lesions requiring arterial embolisation of the liver in early infancy. Lesions can be multifocal (which often have consistent imaging appearances) or focal (which have a more variable picture). Infants with high-output cardiac failure presenting at birth tend to have focal lesions and those with mutifocal lesions may present up to several months of age. The angiographic appearance, however, of all lesions is variable (263,264). Angiography is carried out in those infants with heart failure in whom embolization is being considered. It is important to fully evaluate the vascular geography of these lesions as they can be complex and derive an arterial supply from the internal mammary, the phrenic and the intercostal arteries, as well as the hepatic artery. Embolization/surgery should be considered in infants requiring mechanical ventilatory support or if infants remain in cardiac failure despite medical support and are failing to thrive (265,266). Embolization does not seem to help those with massive hemangioma who are not in cardiac failure. Embolization may need repeating if control of the cardiac failure is suboptimal. Surgical ligation of the hepatic artery may not provide control of cardiac failure as portohepatic shunts (between the portal and hepatic veins) as well as arteriovenous shunts (predominantly but not exclusively between the hepatic artery and hepatic veins) can occur (263, 264). Angiography is needed in these children before any intervention (however, it should be performed with a view to embolization during the same procedure).

Hepatic arteriovenous malformations (AVMs) are rare and can present with neonatal cardiac failure when it is probably best managed by embolization and surgical resection. In

this age group, an MR scan should be undertaken to distinguish a solitary hemangioma from an AVM. This will prevent the unecessary pharmacological treatment with angiogenesis inhibitors (including steroids) of AVMs.

Later in life, most hepatic AVMs are seen in patients with hereditary hemorrhagic telangiectasia (267). They can result in cardiac failure, hepatic ischemia, and portal hypertension. Embolization should be undertaken with caution as lesions are invariably diffuse and there is a significant risk of causing hepatic ischemia and necrosis (268).

Arterioportal fistula (APF) can be intra/extrahepatic and congenital/acquired. Most intrahepatic APF are isolated congenital anomalies and are seen in HHT and Ehlers–Danlos syndrome, whilst diffuse APFs are seen in EHBA and cirrhosis. Penetrating/blunt trauma can lead to APFs. The most important clinical manifestation of intrahepatic APF is portal hypertension. A small number of patients will present with high-output failure if the ductus venosus is patent. Acute closure of the DV has been implicated in fatal GI bleeding. Most APF have a large intrahepatic varix, which can be part of the draining portal vein or a segment of the umbilical vein. The fistula may be a single AV connection or multiple connections with the same varix. These lesions should be treated as soon as they are diagnosed because of potential damage to portal veins and bowel and most can be cured by embolization (269). Following embolization, heparinization should be considered to avoid portal vein thrombosis.

Acquired vascular abnormalities rarely present clinically and are often found on noninvasive imaging, for example, on a liver ultrasound scan performed after a percutaneous liver biopsy. Arterial pseudoaneurysms and arteriovenous fistulae can follow both penetrating and blunt liver trauma and most require no specific treatment; however, it is recommended to treat these on an individual basis. Follow-up is important and if lesions become life-threatening, such as with pseudoaneurysm rupture, then emergent catheter embolization should be undertaken.

In the Budd–Chiari syndrome, the obstruction to hepatic venous outflow (in native and grafted livers) usually involves the main hepatic veins, however, obstruction may involve the suprahepatic inferior vena cava. From our own experience we have found a variable angiographic appearance with coexistent critical stenoses and total occlusions of all hepatic veins. Catheter angiography is needed in these patients to demonstrate the stenoses and obstructions, measure pressure gradients, and, if suitable, permit vascular intervention. Balloon angioplasty is now recommended as the intital treatment of choice and other endovascular techniques such as thrombolysis/stenting could be considered in those with failed balloon angioplasty (270–273). Our experience using balloon angioplasty and other endovascular recannalizing techniques is encouraging with technical and early clinical successes seen in most patients.

9. FUTURE DIRECTIONS

Continuing advancements in medical imaging, such as interventional MRI, and minimally invasive procedure technologies, such as energy deposition methods, will likely change the face of medicine.

Interventional and intraoperative MRI promises to provide radiation free guidance for procedures. Advanced imaging techniques such as dynamic contrast enhancement and spectroscopy combined with superior soft tissue differentiation will allow MRI-guided procedures that are not yet possible.

Energy deposition technologies are another area currently being developed. Interstitial probes are inserted in order to deposit energy from various sources including

radiofrequency, laser, microwave, and ultrasound. The energy heats tissues and results in tissue coagulation and cellular death, allowing percutaneous treatment of unresectable tumors.

High intensity focused ultrasound (HIFU) is the only method of ablation that is currently available that can be performed through the intact skin and even the intact cranium (274,275). Accurate monitoring during ablation assures that only the tumor is treated and the surrounding structures are not damaged and tissues reach an appropriate level for cell necrosis. Ablation can be monitored by either ultrasound or MRI. MRI allows accurate, immediate, real-time monitoring of temperature and tissue destruction. This information can then be fed back to the ultrasound unit to further guide the ablation (274,275). Based on the positive results of the 1 year objective and subjective outcomes of MRI-guided laser ablation of fibroids (276), a pilot study of MRI-guided HIFU fibroid tumor ablation was performed in 17 women prior to hysterectomy (277). Decreased pain and improved menorrhagia was reported. Lesions on MRI correlated with those on pathologic examinations. In 165 ablations of various tumors, a group from China demonstrated the presence of vascular occlusion in addition to tissue coagulation (278).

Tissue heating below cell death threshold levels from HIFU can be used for focal gene or drug therapy. HIFU is currently thought to increase vascular and cell membrane permeability and disrupt tight junctions between cells. This causes drugs to leak into the tissues, between cells and into cells. Drugs and gene vectors can be used in natural form or in temperature-sensitive liposomes (279).

HIFU is still experimental and its use in children is extremely limited. To our knowledge, the only reported incidence in the pediatric population was included in a paper by Wu on pathologic changes related to HIFU ablation of malignant lesions (280). The stated range of patient age was 3–89 years. The number of pediatric patients and tumor types were not indicated. Potential uses of HIFU in the pediatric population include tumor ablation, local drug and gene therapy delivery, and treatment of vascular malformations.

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Future Directions

35
Ethical Issues in Pediatric Minimal Access Surgery

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1. INTRODUCTION

Case 1: JC is a 15-year-old boy, who suffers from pectus excavatum. While as a child, JC had never been self-conscious about his chest wall deformity. His mother reports that over the last two years he has become increasingly shy and will no longer swim or interact in team sports in order to avoid locker rooms. JC wants corrective surgery and has researched the Internet. He wants to have minimally invasive surgery and has chosen the Nuss procedure. The procedure involves the insertion of a convex steel bar underneath the sternum, for a period of two years to allow for chest wall remodeling. The bar is then removed as an outpatient procedure.

Case 2: GM is a 12-year-old girl suffering from ulcerative colitis since age eight who is deemed to have failed medical treatment. She is admitted for a lower GI bleed, requiring transfusion. She had previously received immunosuppressive medications. She has been

on prednisone and she has fallen off her growth curve. The gastroenterologist and you agree that GM should have a subtotal colectomy. There is a strong family history and the father has had a subtotal colectomy and J-pouch done when he was thirty. The family is quite well informed and is asking for their daughter to have a one-stage laparoscopic J-pouch. They are concerned about body image difficulties as GM is entering adolescence and wants her to avoid the stress of adapting to a stoma.

2. WHY IS THE ISSUE OF NEW TECHNOLOGIES AND SURGICAL INNOVATIONS IMPORTANT?

New technologies begin as innovative therapies, develop into cutting-edge technologies, challenge previously accepted paradigms on the choice of treatment, and then may become the practiced standard of care. Physicians and surgeons are faced with new ethical issues when a patient demands nonvalidated therapies or techniques not yet proven to be superior to the standard of care. The balancing of potential benefits and risks is part of everyday medical practice but is highlighted by the impact of new technologies. Technologies that promise less pain, trauma, and smaller scars, a speedier recovery, and comparable results are bound to find early enthusiasts in the medical field as well as in the public. The Internet, television, and other large-scale information systems make patients as well as physicians and surgeons more quickly aware of advances in medical technologies and surgical techniques, often encouraging patients to demand access to medical procedures before they are fully refined. New technologies are often viewed as new magic. Doctors and hospitals may be driven to new techniques by the demand of the public influenced more by the media than by solid clinical evidence. While the medical debate heats up over the proper role of many minimally invasive operations, many surgeons and their hospitals, fearful of being left behind, are not waiting for strong evidence of value before embarking on the technological bandwagon. As Rothschild points out (1), surgeons have to ask the question: for which surgical procedures has equipose been reached? When do these minimally invasive methods become the gold standard and when should traditional options be maintained?

There are several factors that influence the ethics of the decision to use the technology. The first factor is physician knowledge. The extent to which the physician understands the disease and how the device functions or a new procedure is performed are crucial components. This must include knowledge of the risks of the procedure and potential modes of device failure as well as appropriate actions for dealing with these problems. The second factor to be considered is the alternative: devices or procedures are safe when the risks of using them are less than of not using it. Other therapeutic options must be evaluated. The third factor is patient preference. Patient empowerment is one of the most powerful trends in health care today. What is the patient's willingness to take risks? What are the implications of a negative or positive outcome? Does the patient have the knowledge and mindset to participate in the decision?

3. ETHICAL ISSUES

3.1. Adaptation vs. Innovation and Experimentation

Surgeons perform the same surgery somewhat differently on each patient depending on anatomy and physiologic differences. If one patient has a better result, the surgeon may want to reproduce what was done with that patient when performing the next procedure.

Then what are the boundaries between innovation and experimentation? At some point these adaptations become innovation. How much change has to occur to constitute an innovative approach and call forth an open and unbiased effort to assess the effectiveness of a new treatment? (2) An incremental improvement in a conventional procedure in the course of a rational evolutionary process is typically within the bounds of acceptable clinical innovation, but modification of the fundamental concepts of the operation would not be. These are important distinctions both for ethical and medicolegal reasons. Surgeons must remain alert to the possibility of acceptable clinical innovation creeping inexorably toward reckless experimentation. The line is clearly crossed when surgeons pose clinical questions to which answers cannot be anticipated. This usually includes those occasions when surgeons attempt procedures that they personally have never performed before (3).

Surgical innovations must ultimately be put to the test with human subjects. After the introduction of the Nuremberg code, it was not until the mid-1960s that, after a publication from Beecher (4), the need for review of medical and surgical experimentation was brought to the public domain. Thus in 1966, the American Public Health Service required all research to be reviewed by an institutional committee to ensure ethical acceptability. A decade later the National Commission for the Protection of Human Subjects of Biomedical and Behavioral Research was established and the Belmont report was published, in 1979 (1), which remains in force today. Of special importance to the surgical community, the report noted that "new procedures, while not automatically research, in the sense that the innovator intends immediate patient benefit rather than the generation of new knowledge, should however, be made the object of formal research at an early stage in order to determine whether they are safe and effective" (5).

3.2. Informed Consent and Learning Curves

The surgical profession is currently faced with the ethical dilemma: how to reconcile the laudable quest for surgical innovations and the application of new technologies with the need to ensure patient safety and proper informed consent. The Belmont Report also affirmed the critical role that the informed consent of research subjects should play before investigations can proceed. The standards for research consents need to be higher than the standards for consent in medical practice. In research one cannot assume that participation is likely to benefit the subject. This emphasis in assuring freedom of choice or autonomy assumes paramount importance: there should be no immediate obligation to participate in any proposed investigation (1). The consent process in conventional treatment includes discussion of what is known, but in experimental or innovative treatment the discussion must concentrate on what is not known. Not only should the patients and their loved ones know that the proposed intervention constitutes a departure from what has usually been done in similar circumstances, they deserve information relevant to the fact that innovation may well add to the patient's burden, rather than relieve it. Information should be specific to the institution and surgeon. The patient should be informed of the number of procedures performed and the morbidity and mortality to date. It is not sufficient in these circumstances to cite statistics from other surgeons and institutions alone (2).

Patients' safety should always be the paramount concern in the introduction of a new minimal access procedure. New procedures and technologies require even the most technically experienced surgeons to submit to additional formal training. When new procedures and technologies are introduced, education is generally accomplished at short courses with little proficiency testing. Proctoring and apprenticeships are also employed, but there are few standards of performance (6). The shepherding of large numbers of

surgeons through the lengthy learning curve is posing a real challenge to the surgical community (7). The learning curve in such cases inevitably continues at the expense of early patients as the surgeons continue to refine newly acquired skills. A discussion analogous of what trainees should tell patients about their personal position on the learning curve should take place. The consensus seems to be that patients should be given that information, as a reasonable person would want to know it (8).

3.3. Conflict of Interest

With the introduction of new technologies in clinical practice there is the emergence of specific conflicts of interest. The treating physician or surgeon concomitantly becomes in many cases the scientific researcher. When this situation arises, it can no longer be assumed that the interest of the individual patient will prevail over scientific interests. The clinician/investigator's loyalty may become conflictual: is the patient or the research the primary concern? A useful test question should be whether the researcher would, under similar circumstances, refuse to enroll himself or herself or a relative. Patients need to be reassured that the patient–doctor relationship will in no way be altered if they choose not to enroll in the proposed study.

Conflict of interest may also arise when there is a very strong economical incentive for the surgeon to develop a reputation as a minimal access surgery expert and thus increase referrals. The interest of the individual patient must succeed over practice building strategies.

3.4. Evidence-Based Technology Assessment

In the era of evidence-based medicine, surgeons are similarly accountable when they employ procedures of uncertain worth or invent new operations. Surgeons will increasingly need to demonstrate that their preferred approach has solid justification through acceptable and appropriate research strategies and that the new techniques actually accomplish what they are designed to. What is new, especially when endorsed by influential members of the surgical community, stands a good chance of being adopted before its usefulness has been demonstrated adequately. In the past, the scientific basis for many such novelties has only been theoretical (2) before these novelties have been diffusely embraced.

New techniques have developed faster than the profession's ability to provide evidence-based data before widespread acceptance and application. It is necessary to speed up health technology assessment as new technologies may antiquate the results of well-designed and well-performed clinical trials, sometimes even before completion or diffusion of their results. The American College of Surgeons addressed the need to protect society while promoting scientific progress in its 1994 statement on Emerging Surgical Technologies. The College warned, "it is equally essential that the value and safety of a new procedure be established before it is widely used on patients" (9). New ideas come along at a furious pace and many obstacles exist, such as time, money, and bureaucracy, which inhibit or discourage the conversion of innovative ideas into formal research protocols. It is important when designing a trial for evaluating a new technique to ensure a fast enough accrual rate of subjects to complete the trial in a timely fashion, and thus avoid the trial being antiquated by the introduction of another refinement before completion.

Surgical procedures are frequently introduced into general practice on the basis of uncontrolled studies that are less rigorous than those required for the approval of medical intervention (10). The standard for the evaluation of surgical therapy is lower

because of the complexity of designing and conducting scientifically valid and ethically acceptable clinical trials for surgical procedures (11). There is a time-related phenomenon that cannot be ignored: the surgeon's experience with a procedure and the refining of the operative details make too much of a difference for surgery to be subjected to the same testing methods used in the rest of medicine. One needs to recognize the inadequacy of case studies and historical comparisons under most circumstances as those controls cannot account for a variety of confounding variables: selection of treated patients, temporal increase in total effectiveness of care, etc.

Randomized, double blind, placebo-controlled trials offer the most effective way to control the placebo effect of having surgery and investigator bias. Once the pilot study is concluded, based on well-thought-out criteria for prematurely stopping or completing the feasibility study, surgeons should undertake an appropriately designed controlled clinical trial with sufficient patients (i.e., statistical power) to reach generalizable results about the efficacy of the new treatment.

However, it is estimated that only 7% of surgical investigators use a randomized study design of any type (12). There is an under-usage of randomized control trial (RCT) in surgical research. The fundamental criticism is that RCTs deprive the surgeons and their patients of the opportunity to make choices based on the surgeon's clinical intuitions, skill, and experience and the patient's preferences, that is, RCTs undermine the fundamental obligation of the doctor to try to help the patient accept what the surgeon believes is best, under the circumstances. The question arises if individual freedom should ever trump valid attempts to gain the most knowledge, especially when we reflect on all the unnecessary disfiguring and disabling surgical treatments previously believed to offer great benefits?

The crucial issue is that surgeons as fiduciaries must balance technological advancements and ethical responsibilities, a subject rarely broached in our data-driven surgical publications. Ethical reasoning and scientific methodologies are different and equally important constructs. The ethics gap in surgery is becoming more apparent. As a task-oriented specialty, we must continually remind ourselves that positive scientific outcomes do not justify ethical breaches. Confusion can follow if one relies solely on surgical science as the measure of good practice (2).

3.5. Economical and Social Cost of New Technologies

The key to determining whether a new procedure is a true advance or a gimmick lies in the surgeons' ability not only to evaluate the outcomes, but also to observe the advantages of the procedure from the perspective of all the stakeholders in the medical system (patients, providers, payer, and industry) to determine its true value (13). Surgeons must recognize the fact that the high-tech arena is an expensive one in which to play. They must, therefore, balance the high costs against the scientifically proven advantages of each of the operations. Higher costs must be justified by superior results (14). How should society evaluate the costs and allocate additional resources among new technologies? With the present rate of growth in medical technology the gap is widening between what is medically possible for most given illnesses and what is affordable by society.

Clearly, technology is not an unequivocal savior. With it come difficult social, ethical, and economical choices. Some moral questions arise concerning the technology and whether it is justified in light of moral values. Technologies are expressions of fundamental values, such as the search for knowledge or the relief from suffering. However, these values can no longer be taken as implicitly given, but as the starting point of other motivating values in society (15). Some new technologies are advertised as solutions

for problems that did not previously exist; some are marketed without any identified need. Some technologies are looking for an application, creating their own market, inducing a particular need. The availability of technology may also induce for patients some alienation from their own subjective experience. For example, the possibility of kidney transplant in children and the advantages of living-related minimally invasive nephrectomy place family relations in a very different perspective. Technologies take over an increasing part of our lives and medical practice, and we need to reflect on their value and meaning before embracing them and incorporating them in our clinical armamentarium. Economical pressures to adopt new technologies in order to improve their share of the market, should not deter surgeons from basing their practice decisions on sound clinical and ethical evaluations of those technologies.

4. HOW SHOULD I DEAL WITH SURGICAL INNOVATIONS OR NEW TECHNOLOGIES IN MY PRACTICE?

Honesty and truly informed consent seem to be the two important concepts when dealing with new technologies and surgical innovations such as minimal access surgery. Patients should be well informed about the levels of evidence in the literature for a new technique or procedure. Each surgeon should divulge his level of training, expertise, and experience with a new procedure and be able to inform the patient of his/her true mortality and morbidity and how it compares to the gold standard of the institution. Surgeons should strive to formally evaluate new technologies and procedures with the ethics board-approved controlled trials before making them the standard of care and resist being pushed by the industry or the public in adopting nonvalidated therapies.

5. THE CASES

Case 1: You discuss with JC and his parents, the known risks and benefits of both the Nuss procedure and the Ravitch procedure (the present standard of care in your hospital). You also remind them that there are no long-term results available in his age group for the Nuss procedure. You reinforce that your experience with the Ravitch procedure is extensive, but that the Nuss procedure is new to your repertoire and your limited experience consists of two patients. There has been no mortality or morbidity so far for the Nuss procedure but the procedures were both done within the last 6 months. You have recently joined a multicenter study group comparing both procedures in a prospective trial. You explain to them the mechanism of the trial and what it entails for them. You explain that this trial is the only way to determine which of the two procedures is the best surgical option. They agree to participate in the trial.

Case 2: You review with GM and her parents the indications for subtotal colectomy. Her nutritional status is good despite her ulcerative colitis but she has been on recent immunosuppression and is still on prednisone even though on a low dose. You would recommend a two-stage approach for her subtotal colectomy, and J-pouch to minimize the chance of postoperative complications (16). Laparoscopic-assisted J-pouch has been reported in the pediatric literature (17). You explain to the parents that your institution has the experience of 20 laparoscopic-assisted subtotal colectomies, of which you have personally performed 10 but you have yet to perform a laparoscopic-assisted J-pouch. On reviewing your data, there has been no mortality and the morbidity is not statistically different in a retrospective comparison with open subtotal colectomy. Surprisingly you

have found that the length of stay, postoperatively, is the same for the two procedures. You offer them contact information for an adult colleague who you know has performed one-stage laparoscopic J-pouches, if they want to pursue a second opinion. The parents want to think about it some more and give you their decision at a later time.

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Education and Training for Pediatric Minimal Access Surgery

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1. INTRODUCTION

A review of the history of minimal access surgery (MAS) is notable in that pediatric surgeons played a substantial role in the early clinical application of the improving technology associated with this type of procedure (1,2). Yet, pediatric surgeons have been slow to adopt minimal access technology even when it was introduced by general surgeons (3). One possible explanation for this reticence is that "... many of the leaders in pediatric surgery were not skilled in this new technology ...." (4). Perhaps if there had been an

effective way to train pediatric surgeons in these new techniques, we would be leaders in MAS instead of followers.

2. A NEEDS ASSESSMENT FOR MAS TRAINING IN PEDIATRIC SURGERY

The limited information about MAS training needs for pediatric surgeons suggests that a significant number desire more of this type of instruction during their pediatric surgery training (5). The majority of pediatric surgeons who routinely perform MAS received their training in a postgraduate course (6). This is not an indictment of the residency program but speaks of the importance of postgraduate training in the career of a pediatric surgeon.

When contrasted to an adult surgery practice, pediatric surgery remains very broad in scope (7). The diversity of patient population is also quite wide now, ranging from the fetus to the adult (5). This diversity of practice creates substantial challenges in considering how MAS education can best be provided for pediatric surgeons and pediatric surgery residents. Possible solutions are suggested by first considering what is known of skills education for all surgeons.

3. EVIDENCE-BASED TECHNICAL SKILLS EDUCATION

With the advent of laparoscopic cholecystectomy, a huge demand for training in MAS developed among general surgeons. This demand was met through courses developed by surgeons who had already gained experience with these techniques. The workshop-style courses were typically fairly expensive, involved one to two days of instruction with the awarding of a certificate of attendance serving as the only measurable educational outcome. Concern about this type of training developed almost immediately and has persisted (8–15). The Society of American Gastrointestinal Surgeons (SAGES) moved quickly to develop guidelines for training (16) but the evidence is that a substantial proportion of surgeons did not follow these recommendations (17). Most participants felt that these types of courses did not adequately prepare them to perform the procedure (14). Additionally, it has been shown that the complication rate was higher for surgeons who began performing these procedures after having only taken one of these courses (18). Completely absent from the discussion about MAS training is the challenge of providing training for pediatric surgery residents.

We can conclude from this brief review that the workshop method of skills instruction is flawed. Therefore, there is an opportunity to consider other options in designing a more effective training program. To begin this process, we should consider what is known of surgical skills acquisition and retention. Further, we should consider the role of education in the process of surgical innovation.

4. PRINCIPLES FOR SKILLS CURRICULUM DESIGN

4.1. Psychomotor Skill Acquisition

The study of psychomotor acquisition begins with the introduction of telegraphy over a century ago (19). While there are a number of descriptions of how skill acquisition occurs (20), the most frequently cited model describes three stages of learning (21). The first stage is the cognitive stage during which the learner gains an understanding of the task. The second or associative stage follows where the learner begins to attempt to

perform the task. During this stage the surgeon practices the skill and eliminates error. Error identification and elimination occurs through both internal and external feedback. Internal feedback occurs as the surgeon makes his or her own comparisons between actual and ideal performance. External feedback occurs when an expert examines the performance, identifies errors, and prescribes a plan for eliminating them. Practice is recognized as a critical element in attaining expertise (22) and yet it is unlikely that a short course allows for an adequate period of practice. The result is that the course ends with the learner in the early aspect of this phase of learning and no opportunity for additional practice before they begin performing the procedure on patients. The final stage of psychomotor learning is the automatic phase where the task is performed without active conscious input.

4.2. The Learning Curve

When outcome measures of performance are plotted as a function of experience, the result is often called the learning curve. In fact, it is more accurately called a performance curve because it is performance that is being measured and learning should never stop (23,24). There are some generalizations about performance curves that have important implications for skills curricula design. The first is that there is substantial improvement in the early part of the experience (Fig. 36.1). This suggests that outcomes of an educational intervention designed to teach surgical skills could be significantly improved with little increase in resources. The second important generalization is that individual performance curves do not follow the pattern described for performance curves for groups. It is common to see plateaus in performance where no additional improvement is seen for several repetitions (Fig. 36.2). A course of study must then be designed so that all participants can attain an acceptable level of performance.

4.3. Skill Retention

The practice of pediatric surgery is unique in that it is common for surgeons to perform complex cases infrequently (7) and raises concerns about retention of skill in the absence of practice. Some preliminary evidence supports the idea that loss of skills is related to clinical volume (25). The area of MAS skill retention has not been investigated

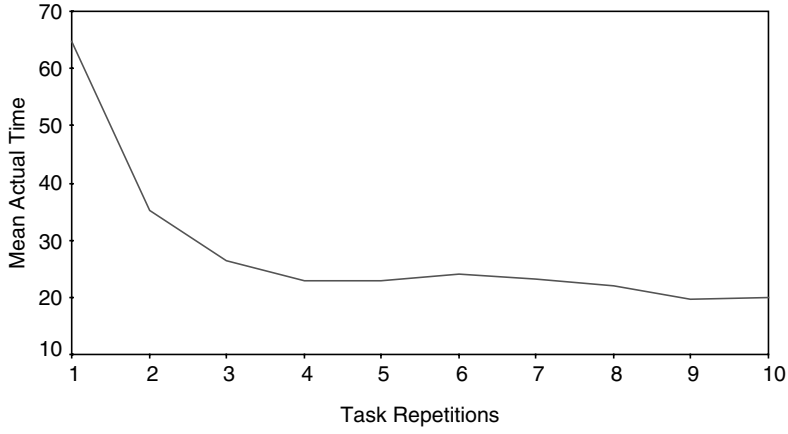


Figure 36.1 A performance curve that demonstrates the relationship between operative time and experience for a group of novice learners. (Courtesy of Dr. Jeannie MacDonald and the Southern Illinois University School of Medicine Department of Surgery Skills Laboratory.)

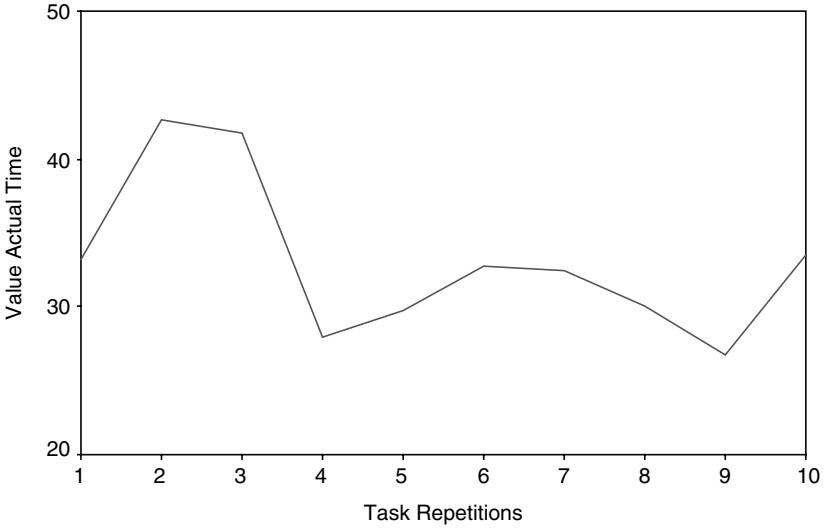


Figure 36.2 A performance curve that demonstrates the relationship between operative time and experience for a single novice learner. (Courtesy of Dr. Jeannie MacDonald and the Southern Illinois University School of Medicine Department of Surgery Skills Laboratory.)

but some general principles of retention do exist (19) and would seem to relate to surgical skill. For example, it has been shown that forgetting increases as a function of interval from learning and that relearning is more rapid than the original learning.

4.4. Surgical Innovation

MAS can properly be viewed as a substantial innovation in pediatric surgery. The process of the spread of diffusion has not been studied in surgery but some general principles of this process have been developed (26) that would seem to apply to the process of diffusion of surgical innovation. The first principle is that there are a number of factors that affect the rate at which an innovation is adopted. These factors include perceived attributes of the innovation (e.g., relative advantage), type of innovation-decision (e.g., optional), communication channels (e.g. media), nature of the social system (e.g., network interconnectedness) and extent of change agent's promotion efforts. An educational intervention would play several potential roles in the changing of perceptions about the innovations. First, information about the procedure could alter perceptions regarding the attributes of a new surgical procedure. Clear explanations might make it appear less complex. Further, educational sessions serve as one type of communication channel. In the case of laparoscopic cholecystectomy, this form of communication was much more rapid than communication that occurred through the traditional medical literature.

Another aspect of the innovation diffusion process that has application for MAS education is the description of how individuals adopt in an innovation. The first surgeons to adopt a new practice are the "Innovators." They are described as individuals who embrace new ideas and tolerate a high degree of uncertainty but who do not necessarily enjoy the respect of the remainder of the group. The second group to adopt the new practice is the "Early adopters" who do have the respect of the community. The third group to adopt the new practice is the "Early majority" and is described as being more deliberate in their decisions. The "Late majority" adopts ideas after the average period of adoption and

requires the pressure of peers to do so. Finally, the last group to adopt an innovation is the "Laggards."

A number of forces act to propel a surgical innovation. In the case of laparoscopic cholecystecomy in adults, a major force was demand by the patients and so surgeons and hospitals recognized that they must provide this procedure or lose a substantial share of their patient base. Given the frequency of this procedure, the costs of training and equipment were relatively small when compared to the loss of revenue gained from biliary surgery. In contrast, the number of Pediatric Surgery MAS at any given hospital is smaller and so the costs of initiating the program more substantial (3). Another powerful force is the perception by patients and their parents. This perception is now more often being influenced by information that they obtain via the Internet, much of which is inaccurate (27).

A review of the pediatric surgical literature provides an interesting contrast between two different MAS procedures in how they are accepted. In both cases, the external forces and individuals interact in the process of adoption or rejection of the innovation. The initial descriptions of laparoscopic cholecystecomy (28–30) in children were followed by very little opposing literature and this has rapidly become the standard approach to this procedure (6). In contrast, the initial description of the Nuss procedure (31) has generated a number of reports comparing the traditional Ravitch procedure that cast doubt that this will become the standard approach for the treatment of all children with pectus excavatum (32–34). Time will tell whether the Nuss procedure will become the standard approach to the correction for all chest wall deformities or for just a subset of these of patients. "Early adopters" serve as opinion leaders and play a major role in determining the outcome of an innovation. They, along with the "Innovators," are the logical choice for providing leadership in the education and training of the remaining pediatric surgeons.

The unique sociology of pediatric surgery provides an opportunity to create a model for other surgical disciplines in how education can provide for the most thoughtful and safest introduction of innovation. While the number of pediatric surgical training programs has increased (35,36) thus expanding the number of pediatric surgeons, the group remains quite small compared to other surgical subspecialties and behaves somewhat like an extended family (37). Bearing these unique attributes in mind and applying the principles of skills training above, it is possible to consider the development of an educational program for pediatric surgeons that will allow them to demonstrate that they are leaders in innovation.

5. PEDIATRIC SURGERY MAS TRAINING: AN OPPORTUNITY FOR LEADERSHIP

5.1. We Should be the Most Sophisticated Consumers of Skills Courses

Despite their shortcomings, skills workshops are utilized by a significant percentage of pediatric surgeons and it is anticipated that they will continue to meet a necessary educational need. Pediatric surgeons need to be sophisticated consumers of these types of courses and attend only those courses with specific attributes (Table 36.1).

5.2. We Should Create and Administer the Best Skills Courses

All pediatric surgery MAS courses should meet the requirements outlined by SAGES. A recent review describes various methods, including simulation, animate and inanimate models, that have been used in surgical skill instruction (39). Some specific work has

Table 36.1 Suggestions for Selecting an Optimal MAS Workshop

The course should have a set of formal educational objectives and a description of the assessment methodology
The faculty should be qualified to both perform and teach the procedure
The participants of a course should possess the appropriate prerequisite skills and knowledge
The facility should be adequate
The curriculum should be designed to minimize time required with the optimal outcome.

Source: Adapted from Rogers et al. (38).

been done for MAS (40,41) that validates the approach of using a series of basic structured tasks or drills to develop the skills necessary to do MAS procedures. Courses could be substantially improved by simply making them longer. A weeklong course would offer an opportunity for more practice, which is the element most lacking in the very short courses. It would be more expensive and less convenient but would accomplish some worthy educational goals that produce better patient outcomes. More opportunity for practice should position individuals further along their individual performance curves to assure that innovation is introduced with fewer errors.

5.3. We Should Create a Program Designed to Teach Pediatric Surgeons How to Teach Surgical Skills. The Program Should be Mandatory for All Those Instructing in Courses

Professional athletes gain a high level of expertise in their particular sport. They still require coaching and it is doubtful that their coaches are as proficient in the sport as are the athletes themselves. Further, not all professional athletes are excellent coaches. In the same way, gaining expertise in the performance of a surgical skill does not guarantee that one is expert at instructing others. The elements of effective skills instruction are fairly simple and educated instructors would enhance educational outcomes in a course.

5.4. We Should Develop Permanent Training Centers for Both Residents and Trained Surgeons

The historical evaluation of graduate and postgraduate training (42) has led to an unfortunate separation between education in these two continuous aspects in the pediatric surgeon's career. We should take the leadership in demonstrating that education is a constant feature in the life of a pediatric surgeon. Training programs should cooperate to develop courses where all trainees are exposed to existing or new surgical techniques. The precedent for this type of centralized training has already been established (43,44). These sessions could take place at some of the established pediatric surgery residency sites, a MAS center, or at one of the sites the commercial vendors of MAS equipment have erected. In addition to providing training in new techniques, an established center could allow a surgeon to return periodically to practice an established skill and thereby improve retention.

5.5. We Should Provide Leadership in Documenting the Improvement That Educational Courses Have in Practice

The effects of continuing medical education offerings have only been rarely followed into practice (45). What is needed is a network that would allow for the collection of

information about patient outcomes as pediatric surgeons begin to perform new procedures in patients. Such networks already exist for ECMO, childhood cancers, and trauma. It is time to apply the lessons learned from the creation and administration of these networks to all important clinical outcomes. This type of multi-institutional network would produce better early information about the value of a proposed innovation than is currently available from single institutional experiences. Further, it would also allow for an improved understanding of how an educational intervention affects physician learning and, ultimately, patient outcomes.

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37
Robotically Assisted Pediatric Surgery

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1. INTRODUCTION

Innovations in endoscopic technique and equipment continue to broaden the range of applications in minimal access surgery (MAS). However, in the adult population many of these procedures have yet to overtake the traditional open approach due to their technical difficulty. Difficulties remain in achieving dexterity and precision of instrument control within the confines of a limited operating space. These difficulties are further compounded by the need to operate from a video image. In the pediatric population, such challenges are amplified by the significantly smaller workspace, as well as the size and delicacy of tissues. The application of robotic technology has the potential to contribute significantly to the advancement of minimal access pediatric surgery.

2. LIMITATIONS OF MAS

While minimal access techniques have revolutionized many surgical procedures, the introduction of MAS also brought with it certain unique complexities that are not present with conventional open surgery.

2.1. Movement Limitation

First, MAS instruments work through cannulas, or ports in the body wall. These ports act as pivot points that consequently reverse the direction of motion of the instrument tip in relation to the motion of the instrument handle. For instance, in order to move the instrument tip to the left inside the body cavity, the surgeon is required to move his hands to the right outside the body, and so on. This reversal of movement requires nonintuitive instrument control that is mentally taxing, especially as the complexity of the surgical task increases.

Second, the majority of MAS instruments consist of an end-effector mounted to the tip of a long, rigid shaft. The endoscopic cannula allows these instruments to pivot around the fixed point within the body wall, but restricts motion laterally. The six degrees of freedom of position and orientation (defined as motion along the *X*, *Y*, *Z* axes, and rotation about each of these axes) of open instruments is therefore reduced to four degrees of motion (pitch, yaw, roll, and insertion) for MAS procedures (Fig. 37.1). An additional two degrees of freedom could be restored to MAS instruments by constructing articulations at the distal end, past the location of the cannula pivot point (Fig. 37.2). However, the precise and dynamic control of these distal articulations during an operative procedure would be difficult to coordinate without the assistance of computer control.

2.2. Haptic Limitations

The long shafts of MAS instruments force a separation of the surgeon's hands from the operative anatomy, which significantly decreases the amount of tactile sensation and force reflection available. The extended length of the instruments also significantly

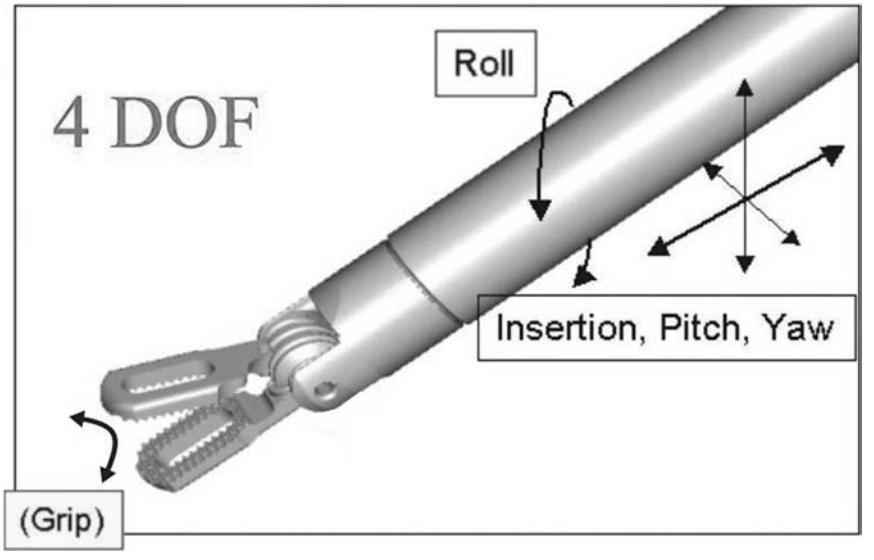


Figure 37.1 Traditional four degrees-of-freedom endoscopic instrument.

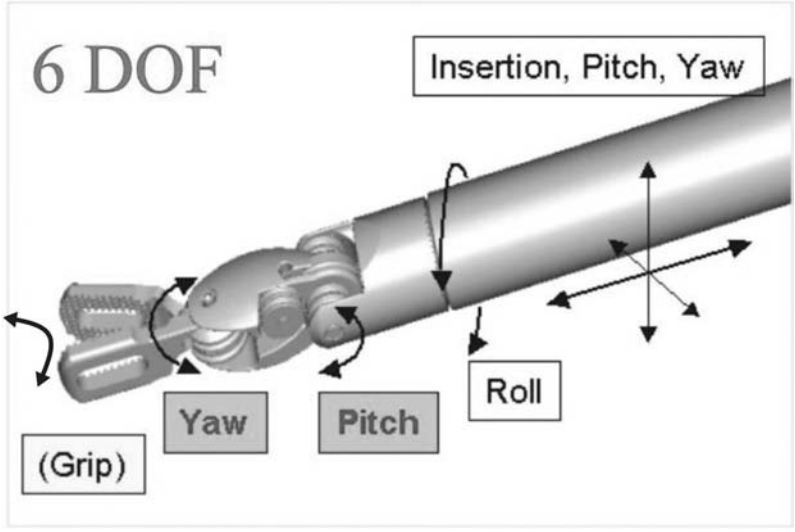


Figure 37.2 Fully articulated six degrees-of-freedom robotic instrument.

magnifies any existing hand tremor. Furthermore, the excursion of an instrument tip is highly dependent on its depth of insertion. For instance, an instrument that is shallowly inserted requires comparatively large hand movements to accomplish a given instrument movement inside the body, while a deeply inserted instrument requires much less hand movement to sweep the instrument tip around. Consequently, the dynamics of the instrument change constantly as it is inserted and retracted throughout a procedure. Overall, all these factors can lead to less precise and less predictable movements when compared to standard, open surgical techniques.

2.3. Visual Limitations

The introduction of an endoscope forces a surgeon to be guided by a video image instead of direct vision. The video monitor is often located on the far side of the patient, and the differences in orientation between the endoscope, instruments, and monitor requires the surgeon to perform a difficult mental transformation between the visual and motor coordinate frame (1). This problem is further exacerbated whenever an angled endoscope is used.

The majority of conventional endoscopes are built around a single lens train that is only capable of displaying images in a flat two-dimensional format. This removes many of the depth cues of normal binocular vision, complicating tasks such as dissection between tissue planes. Some stereoscopic vision systems exist, but their performance is limited by the resolution and contrast characteristics of the endoscopes themselves, as well as the display technologies. In addition to these limitations, conventional endoscopes often require a dedicated assistant to hold and manipulate them. The natural tremors and movements of the handling assistant are exacerbated by the magnified image.

3. ROBOTICS IN SURGERY

For several decades now, robots have served in a variety of applications such as manufacturing, deep-sea exploration, munitions detonation, military surveillance, and

entertainment. In contrast, the use of robotic technology in surgery is still a relatively young field. Improvements in mechanical design, kinematics, and control algorithms that were originally created for industrial robots are directly applicable to many surgical applications. The use of robotic technology is, in large part, aimed at addressing the numerous aforementioned limitations with conventional MAS.

The first recorded application of robotics in a surgical procedure was for CT-guided stereotactic brain biopsy in 1987 (2). Since then, technological advancements have led to the development of several different robotic systems. These systems vary significantly in complexity and function.

4. CLASSIFICATION OF ROBOTIC SURGERY SYSTEMS

Robots can interact with surgeons in many ways. One method of classifying robots is by their level of autonomy. Using this classification, there are currently three types of robots used in surgery: autonomous robots, surgical assist devices, and teleoperators (Table 37.1).

An autonomously operating robot carries out a preoperative plan without any immediate control from the surgeon. The tasks performed are typically focused or repetitive but require a degree of precision not attainable by human hands. An example is the ROBODOC® system used in orthopedic surgery to accurately mill out the femoral canal for hip implants (3). Another example is the CYBERKNIFE® system that consists of a linear accelerator mounted on a robotic arm that is used to precisely deliver radiotherapy to treat intracranial and spinal tumors (4,5).

The second class of robots is the surgical assist devices, where the surgeon and robot share control. The most well known example of this group is the AESOP® (Automatic Endoscopic System for Optimal Positioning; Computer Motion, Inc., Goleta, CA). This system allows a surgeon to attach an endoscope to a robotic arm that provides a steady image by eliminating the natural movements inherent in a live camera holder. The surgeon is then able to reposition the camera by voice commands. Today, the AESOP is used in many different surgical disciplines including general surgery (6,7), gynecologic surgery (8), cardiothoracic surgery (9), and urology (10).

The final class consists of robots whose every function is explicitly controlled by the surgeon. The hand motions of the surgeon at a control console are tracked by the electronic controller and then relayed to the slave robot in such a manner that the instrument tips perfectly mirror every movement of the surgeon. Because the control console is physically separated from the slave robot, these systems are referred to as teleoperators. All the recent advances in robotically assisted procedures in pediatric surgery have involved this class of machines.

Table 37.1 Classification of Robotic Surgical Systems

Type of system	Definition	Example
Autonomous	System carries out treatment without immediate input from the surgeon	Cyberknife Robodoc
Surgical assist	Surgeon and robot share control	Aesop
Teleoperators	Input from the surgeon directs movement of instruments	Intuitive Surgical da Vinci System Computer Motion Zeus System

5. TELEOPERATORS

In the realm of true operative procedures, previously there were two systems commercially available—the da Vinci Surgical System by Intuitive Surgical, Inc. (Sunnyvale, CA) and the Zeus system by Computer Motion (Goleta, CA). Recently, the two companies have merged, and it is expected that the da Vinci system will become the predominant robotic operative platform.

Though these systems are popularly referred to as surgical robots, this is a misnomer as "robot" implies autonomous movement. In neither the da Vinci nor the Zeus does the system operate without the immediate control of a surgeon. A better term may be "computer-enhanced telemanipulators." However, for the sake of consistency with published literature, this chapter will continue to refer to such systems as robots.

The integration of computer technology into both the da Vinci and Zeus helps to resolve many of the limitations of MAS surgery. By scanning the surgeon's hand motions, information is relayed to the instruments in order to move them in the corresponding direction and orientation. Intuitive nonreversed instrument control is therefore restored, while preserving the minimal access nature of the approach.

The presence of a computer control system allows one to filter out inherent hand tremor, thus making the motion of the endoscope and the instrument tips steadier than with the unassisted hand. In addition, the system allows for variable motion scaling from the surgeon's hand to the instrument tips. For instance, a 3 : 1 scale factor maps 3 cm of movement of the surgeon's hand into 1 cm of motion at the instrument tip. In combination with image magnification from the video endoscope, motion scaling makes delicate motions easier and more precise to perform (11).

In both systems, the instruments are also engineered with articulations at the "wrist" distally that increases their dexterity compared to simpler MAS tools. The da Vinci system alone possesses instruments capable of the full six degrees of freedom of the human wrist.

5.1. The Zeus System

The Zeus system (Computer Motion, Goleta, CA) is a telemanipulator system that consists of a surgeon's console and three robotic arms (Figs. 37.3–37.5). The surgeon operates from a console several feet away from the operating table. There he uses handheld manipulators to control the two robotic arms and surgical instruments, a foot pedal to activate the computer driven system, and voice commands to direct a camera controlled by an AESOP arm (12). Like the da Vinci system, the Zeus system offers tremor reduction and motion scaling.

The Zeus system consists of three modular, freestanding robotic arms that are attached to the operating table. This design allows the system to be oriented to many different configurations. The Zeus system also features 3.5–5 mm instruments, several of which are capable of increased articulation through the Zeus Microwrist. This joint provides the instrument with an additional degree of freedom at the wrist, giving a total of five degrees of freedom. The Zeus system also features the ability to accommodate a variety of visualization options (3D and 2D) and telescope sizes.

The Zeus system has received generalized clearance for surgery under Conformité Européenne (CE) guidelines. In the United States, the Zeus system received Food and Drug Administration (FDA) clearance for general laparoscopy and has been used for thoracic and cardiac procedures. To date, the Zeus system has been used to perform multiple operations in adults in different surgical specialties throughout the world (13–18).



Figure 37.3 The Computer Motion Zeus robotic surgical system. (Courtesy of Intuitive Surgical, Sunnyvale, CA.)

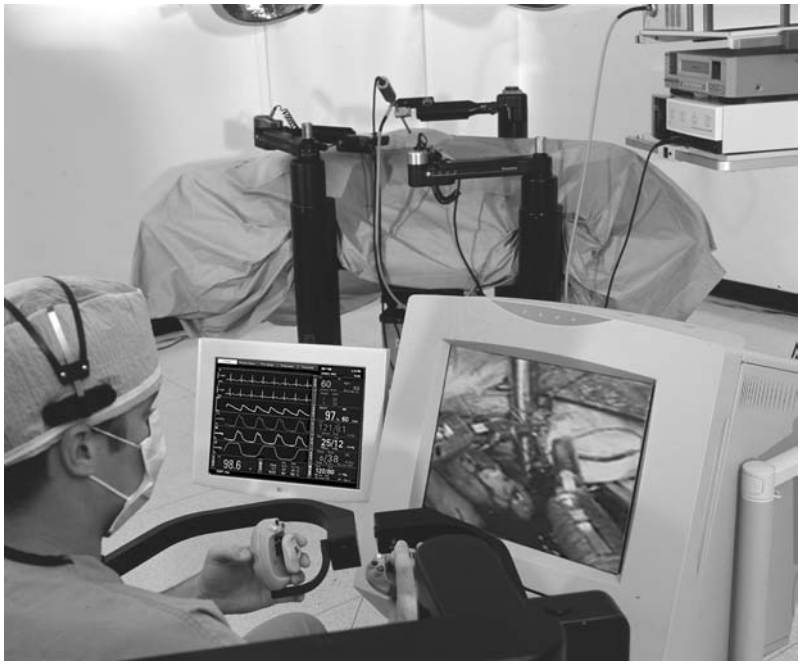


Figure 37.4 The Zeus surgeon's console with its video display and master controls. (Courtesy of Intuitive Surgical, Sunnyvale, CA.)



Figure 37.5 The Zeus system as arranged in an operating room. (Courtesy of Computer Motion, Goleta, CA.)

5.2. The da Vinci System

The da Vinci system is made up of two major components (19) (Figs. 37.6–37.8). The first is the surgeon's console, which houses the visual display system, the surgeon's control handles, the user interface buttons, and the electronic controller. The second component is the patient side cart, which consists of two arms that control the operative instruments and a third arm that controls the video endoscope.

The operative surgeon is seated at the surgeon's console, which can be located up to 10 m away from the operating table. Within the console are located the surgeon's control handles, or masters, which act as high resolution input devices that read the position, orientation, and grip commands from the surgeon's finger tips. They also act as haptic displays that transmit forces and torques back to the surgeon's hand in response to various measured and synthetic force cues. This control system also allows for computer enhancement of functions such as motion scaling and tremor reduction.

The image of the operative site is projected to the surgeon through a high-resolution stereo display system that uses two medical grade cathode ray tube (CRT) monitors to display a separate image to each of the surgeon's eyes. The surgeon's brain then fuses the two separate images into a virtual three-dimensional construct. The image plane of the stereo viewer is superimposed over the range of motion of the masters, which restores visual alignment and hand–eye coordination. In addition, because the image of the endoscopic instrument tips is overlaid on top of where the surgeon senses his hands, the end effect is that the surgeons feels that his hands are virtually inside the patient's body.

Since its inception in 1995, the da Vinci system has received generalized clearance under European CE guidelines for all surgical procedures. In the United States it has received clearance for general, thoracic surgery, and urologic procedures. In addition, the da Vinci system recently received FDA clearance for cardiac procedures involving

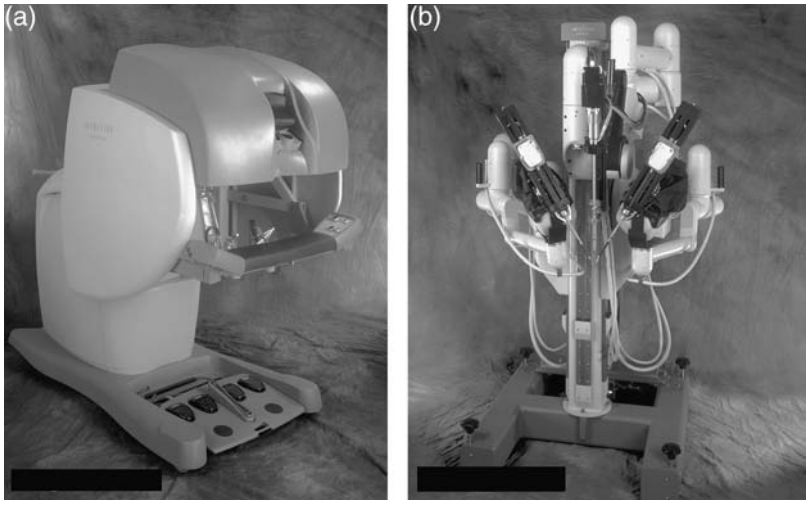


Figure 37.6 (Courtesy of Computer Motion, Goleta, CA.)

a cardiotomy. To date, thousands of surgical procedures in multiple disciplines have been performed using the da Vinci system (20–27).

6. APPLICATION OF ROBOTIC TECHNOLOGY TO PEDIATRIC SURGERY

To date, there is only a small body of literature regarding the application of robotic technology for pediatric surgical procedures. Hollands (12, 28, 29) and Lorincz (30) have both described the application of the Zeus robotic system in a porcine model. Technically challenging procedures such as entero-enterostomy, hepaticojejunostomy, portoenterostomy, and esophago-esophagostomy were all demonstrated to be technically feasible

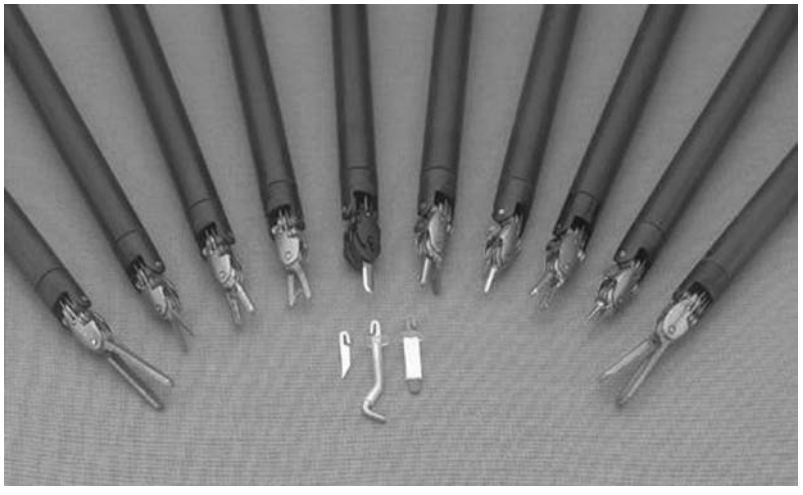


Figure 37.7 An array of fully articulated, 6 degrees-of-freedom robotic endoscopic instruments. (Courtesy of Intuitive Surgical, Sunnyvale, CA.)



Figure 37.8 At the surgeon's console, alignment of the visual axis to the master controls creates the illusion that the surgeon's hands are operating virtually within the patient. (Courtesy of Intuitive Surgical, Sunnyvale, CA.)

(Table 37.2). Similarly, Malhotra (31), Aaronson (32), and Olsen (33) have described the application of the da Vinci system in animal models to perform complex pediatric cardiovascular, neurosurgical, and urologic procedures.

Several authors have reported a small but assorted variety of human pediatric cases performed with both the Zeus and da Vinci systems (Table 37.3). On average, setup and operative times were longer with the robotic cases when compared to standard laparoscopy. The rate of complications or conversion to open surgery has been low. Significant long-term follow-up for any differences in clinical outcome has yet to be reported. Specialized training of the surgical teams and the expense of the equipment remain substantial obstacles to widespread adoption. However, the authors in common have applauded the introduction of high-quality three-dimensional vision, articulated instrument tips, and intuitive instrument control, all of which seem to enhance surgical precision. To fully evaluate the potential benefits and application to pediatric surgery, further studies are warranted.

Procedures such as repair of esophageal atresia, portoenterostomy, and ureteral reimplantation can all be performed using existing endoscopic equipment. However,

Table 37.2 Robotic Procedures in Porcine Models Using the Zeus Surgical System

Author	Operative procedure	Robotic system	Procedure times, laparoscopic (mean)	Procedure times, robotic (mean)
Hollands et al. (12,28,29)	Porcine Enteroenterostomy (28) (5 laparoscopic, 5 robotic)	Zeus	Anesthesia = 176 min Operative time = 143 min Anastomotic time = 109 min	Anesthesia = 154 min Operative time = 139 min Anastomotic time = 93 min
	Porcine Hepaticojejunostomy (12) (5 laparoscopic, 5 robotic)	Zeus	Anastomotic time = 66 min	Anastomotic time = 93 min
	Porcine Esophagoesophagostomy (29) (5 laparoscopic, 5 robotic)	Zeus	Anesthesia time = 124 min Operative time = 97 min Anastomotic time = 89 min	Anesthesia time = 151 min Operative time = 131 min Anastomotic time = 125 min
	Porcine Portoenterostomy (12) (10 laparoscopic, 10 robotic)	Zeus	Anesthesia time = 125 min Operative time = 98 min Anastomotic time = 60 min	Anesthesia time = 164 min Operative time = 137 min Anastomotic time = 94 min
Lorincz et al. (30)	Porcine Portoenterostomy (8 robotic)	Zeus	N/A	Mean operative time 330 min
	Porcine Esophagoesophagostomy (7 robotic)	Zeus	N/A	Mean operative time 120 min
Malhotra et al. (31)	Juvenile ovine thoracic aortic anastomosis (4 robotic); juvenile ovine thoracic longitudinal aortotomy with patch aortoplasty (1 robotic)	da Vinci	N/A	Aortic clamp time = 47 min Anastomotic time = 26 min
Aaronson et al. (32)	Intrauterine fetal ovine simulated myelomeningocele repair—creation and repair of full-thickness skin lesions (6 robotic)	da Vinci	N/A	In utero procedure time = Steep learning curve from just under 120 min to just over 30 min within 6 cases
Olsen et al. (33)	Porcine Cohen cross-trigonal ureter reimplantations (14 robotic reimplantations)	da Vinci	N/A	Mean operative time = 68 min

Table 37.3 Pediatric Clinical Experience with the Zeus and da Vinci Robotic Systems

Author	Robotic system	Patient (*N*)	Operative procedure	Results	Comments
Le Bret et al. (13)	Zeus	56	Patent ductus arteriosus ligation (28 thoracoscopic, 28 robotic)	Operating room time thoracoscopic = 83 min; robotic = 162 min Surgical procedure time thoracoscopic = 24 min; robotic = 50 min	Longer operative time for robotic group; 1 conversion to videothoracoscopic; no significant difference in complications or outcome
Lorincz et al. (30)	Zeus	7	5 Nissen fundoplication; 1 cholecystectomy; 1 Heller myotomy	Operative time for fundoplication reduced from 4.5 h to 1.5 h	Rapid improvement in case times as team progressed along learning curve
Gutt et al. (39) and Heller et al. (40)	da Vinci	14	11 fundoplication; 2 cholecystectomy; 1 bilateral salpingo-oophorectomy	Operative times: fundoplication = 146 min (mean); Cholecystectomy 105, 150 min; salpingo-oophorectomy 95 min	No complications or conversion to laparotomy
Luebbe et al. (41)	da Vinci	20	10 fundoplication; 3 cholecystectomy; 2 splenectomy; 1 urachus resection; 1 Morgagni diaphragmatic hernia; 3 biopsies; 1 lymphadenectomy	Mean times: OR setup = 45 min; patient preparation = 17 min; console operating time = 93 min (range 10–299 min)	15% complications (2 conversions to laparotomy for bleeding, 1 pneumothorax)
Mijalijavic et al. (42)	da Vinci	2	Vascular ring dissection	Total operative times: 172.5 min (mean) Robotic procedure times: 106.5 min (mean)	Total operative time longer than usually required for standard thoracoscopic procedure due to setup; dissection time slightly shorter in robotic cases

mastery of these complex techniques in an endoscopic environment is challenging. Compared to open surgery, few would argue that there are significant sacrifices made in terms of the maneuverability and dexterity of the instrument tips, the precision and delicacy of dissection, and the sheer ease with which procedures may be accomplished.

There are a number of distinct and compelling advantages associated with the use of the surgical robot that may enhance current operative technique. Unlike conventional laparoscopic instrumentation, which requires manipulation in reverse, the proportional movement of the robotic device allows the instruments to follow directly the movement of the surgeon's hands. The intuitive control of the instruments is particularly advantageous for the novice endoscopist. In addition to mimicking the surgeon's movements in an intuitive manner, the robotic instruments also offer six degrees of freedom and grip, two more than conventional instruments. This technology permits a large range of motion and rotation that follows the natural range of articulation of the human wrist, and may be particularly helpful when working space is limited. The electronic control system is capable of filtering out hand tremors as well as motion scaling, whereby gross hand movements at the surgeon's console may be translated into much finer movement of the instrument tips at the operative site.

The three-dimensional vision system adds an additional measure of safety and surgical control beyond what is available with the traditional endoscope. The three-dimensional display improves depth perception, and the ability to magnify images by a factor of ten, and allows extremely sensitive and accurate surgical manipulation. The alignment of the visual axis with the surgeon's hands in the console further enhances hand–eye coordination to a degree uncommon in traditional endoscopic surgery.

Despite these improvements, there remain significant obstacles to the widespread adoption of these robotic systems. Chief among these is the cost for both the robotic systems and their array of instruments. Robotic procedures, times are predictably longer when compared to the conventional laparoscopic approach, at least for the initial series of cases until the surgical team becomes facile with the use of the new technology. The robotic systems themselves are somewhat large and obtrusive, at times impeding access by the anesthesiologist or patient side surgeon. Until recently, the robotic instruments were significantly larger than their laparoscopic equivalents. For instance, the da Vinci instruments required 8-mm ports and the 3D endoscope required a 12-mm port. Finally, the lack of significant haptic feedback continues to be a major drawback to precise surgical dissection. Undoubtedly, many of these issues will be remedied in the next generation of equipment as the technology continues to improve.

7. FUTURE DIRECTIONS

While the current robotic systems represent great strides in technology, the possibilities for innovation are virtually endless. The use of a video image that is processed through a computer system, rather than direct vision, allows for the overlay of any number of images or information. For instance, vital signs and other patient data may be projected directly in front of the surgeon's eyes while he is operating. A three-dimensional image of a tumor may be directly overlaid on top of the operative field as the dissection is carried out. Virtual models of heart valves, orthopedic implants, or vascular conduits may be test-fitted before the costly objects are requisitioned.

Because the computer systems may be made aware of both the patient's anatomy as well as the position of the operative instruments, a virtual "safety envelope" may be

defined. The system can then track the surgeon's hand movements and prevent inadvertent damage to collateral tissue.

One of the "holy grails" of robotic surgery is to endow the systems with true force reflection and haptic feedback. However, the presence of the numerous mechanical joints inherently imparts additional friction to the entire kinematic chain. It is therefore difficult to distinguish friction that originates from the robotic system versus forces from living tissue. This limitation will be overcome with the development of newer computer algorithms and microsensors that can be positioned at the tips of the instruments.

The control handles of a system, such as the da Vinci, not only sense a surgeon's hand movements, but are also electronically powered and can relay forth information back to the surgeon. Not only can tissue tension be delivered as is familiar in conventional surgery, but also any range of biological data. For instance, the pulsations of a diminutive artery can be enhanced and magnified such that it is palpable to the surgeon at the console. Other variables that are not in the average realm of human perception such as oxygen tension, temperature, and tissue density may also be conveyed, as demonstrated by the NASA Smart Probe project (34).

Computer systems are also much more facile than the human mind at processing complex coordinate frames of reference. For instance, the operative instruments can be programmed to always align with the axis of view of the endoscope. Thus wherever the endoscope is angled, it would appear to the surgeon that he is positioned at the end of the endoscope. For example, an angled endoscope inserted into the mouth and directed back towards the nasopharynx could establish a vantage point for the operative instruments such that one would seem to operate through the back of the patient's head.

The fact that robotic systems can track a surgeon's hand movements brings with it the ability to record a wealth of data. Thus every nuance of a master surgeon's performance, as well as the visual information from the operation, may be preserved. All that information may then be replayed in its entirety for those in training. Rather than stumble through an operation step by step, a novice may be able to first mimic and then perform an operation as it was meant to be. This "player piano" model may be invaluable in surgical education and could change the manner in which future generations learn to operate.

A much-popularized idea is the concept of telesurgery, whereby a surgeon can perform an operation from a distance by means of a remote interface. This concept, first conceived for military applications, would allow for the delivery of surgical care to remote or inhospitable areas. It also allows a surgical authority to perform operations far beyond his immediate geographical vicinity. Recently, the world's fist trans-Atlantic laparoscopic cholecystectomy was performed remotely, in which a surgeon located in New York operated on a patient in Strasbourg, France (35). However, this concept is still severely restricted by the cost and capacity of current bandwidth technology, as well as the inviolate limit of the speed of light. A less ambitious application is telementoring, whereby an experienced specialist can observe and advise a surgical team operating in a remote location. Already, a growing number of procedures have been accomplished using this technology (36–38).

8. CONCLUSION

The advent of minimal access surgery has brought with it a wealth of potential benefits for both the patient and our health care system. However the inherent limitations of operating in an endoscopic setting pose significant challenges for the surgeon, and this is only magnified as procedures become more complex such as those encountered in pediatric surgery.

The incorporation of robotic and computer technology has the potential of contributing significantly to the advancement of this area. As the technology continues to be refined, its ultimate acceptance will demand that the issues of cost, training, safety, efficacy, and clinical utility, will all have to be addressed.

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about the book...

This authoritative guide reviews the evoloving evidence-based literature for a wide variety of procedures and techniques in pediatric minimal access surgery (MAS)—providing expertly written chapters that cover the techniques specific to a broad range of diseases, as well as current advances in urology, neurosurgery, and spinal and cardiac surgery.

Unlike any other source on the topic, this book focuses on key theories and applications rather than technical details...emphasizes the principles of practice for this technology and the most recent evidence to support its use...covers individual diseases and procedures within pediatric general surgery and its subspecialties...describes applications of minimal access surgery within other pediatric surgical specialties and considers emerging technologies in the field...emphasizes evidence-based approaches...and offers an international perspective on the topic.

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